ANNUAL REPORT

2006

with data for 2004

INTERNATIONAL CLEARINGHOUSE FOR BIRTH DEFECTS SURVEILLANCE AND RESEARCH

Published by

INTERNATIONAL CLEARINGHOUSE FOR BIRTH DEFECTS SURVEILLANCE AND RESEARCH

Via Pilo Albertelli, 9 - 00195 Roma - Italy

INTERNATIONAL CLEARINGHOUSE FOR BIRTH DEFECTS SURVEILLANCE AND RESEARCH (ICBDSR)

A non-governmental organisation in official relations
with the World Health Organization

ANNUAL REPORT

2006

with data for 2004

Officers 2005/2006

Chairperson
John Harris (California, USA)

Vice-Chairperson
Carol Bower (Australia)

Secretary
Michiko Yamanaka (Japan)

Treasurer
Barry Borman (New Zealand)

Published by
THE INTERNATIONAL CENTRE ON BIRTH DEFECTS

Via Carlo Mirabello 19,
00195 Roma, Italy
phone: 39-06-3701905
Fax: +39-06-3701904
e-mail: icbd@icbd.org
website: http://www.icbd.org/

ISSN 0743-5703
The International Centre on Birth Defects
acknowledges the financial support from the Centers for Diseases Control and Prevention, Atlanta, USA
(CDC Grant no. U50/CCU207141 - 14).

INTERNATIONAL CLEARINGHOUSE FOR BIRTH DEFECTS SURVEILLANCE AND RESEARCH ANNUAL REPORT 2006 (WITH DATA FOR 2004)

Contents

Introduction

A word on the structure of the report

Because of collaborative research is the most important functions of the International Clearinghouse for Birth Defects Surveillance and Research (ICBDSR), summaries of these activities open this report.

Descriptions of the individual Programmes and tabulations of their annual data follow.

Each monitoring system monitors all birth defects. However, the tables and graphs present data for selected defects. The selection is made at the Annual Meeting and is quite arbitrary, moreover it may change year by year.

The main aim of the tables and graphs is to show some basic data and the rates of some specific defects in each surveillance system. Figures are presented in:

(a) a graph showing the total number of births per year
(b) two graphs showing the percentage of births per year and maternal age classes (<20, 20-24, 25-29; 30-34, 35-39, 40+)
(c) a table showing data for 2004
(d) a table showing data for the previous longest available period of each register and for each malformation up to 2004
(e) some graphs showing the yearly rates represented in the following style:
  a. bars represent real patterns of prevalence
  b. blue bars stand for live+still births rates
  c. grey bars stand for termination of pregnancy rates.

The tables have a standard format for all the registries. If a malformation is not reported the row says "not reported".
The prevalence rates graphs are presented only for those malformations which have:
(a) figures at least for 8 years,
(b) number of cases per year different from zero in at least half the available years.

This way of presenting data underlines the recommendation to avoid the comparison of rates of a birth defect among Programmes, as there are important differences in the methodology of registration, in defining live births, still births and abortions, including birth defects observed in pregnancy terminations, and last but not least, in defining every single birth defect. Some of the differences in birth defects definitions are highlighted in the description of each surveillance system and in the table "Synopsis of Surveillance Systems" and in the table "Deviations from the ICBDSR definitions by Registry".

Birth defect rates are calculated by including all cases of each defect, whether isolated or associated to other defects. In some instances, therefore, the same baby may be counted more than once in the tables (i.e.: a baby with cleft lip and limb deficiency is counted twice). In the data from Hungary, however, only isolated defects are reported.

Not all Registries report pregnancy terminations either because the data are not available to the Registry or pregnancy termination is not legal in that country. The inclusion of pregnancy terminations is noted in the tables.

Note

Rates

When calculating rates among live born infants and stillbirths, the denominator used is total births. When terminations are included, the total number of terminations for birth defects is added to the denominator. The denominator used for age-specific rates for Down syndrome consists of the total number of live born infants, stillbirths and, if appropriate, terminations for Down syndrome, whose mothers are in that age group.

Collaborative Research Projects

Multiple Congenital Anomalies (MCA), 2004

Jorge Lopez Camelo (South America, ECLAMC)
Monica Rittler (South America, ECLAMC)

Introduction

For the year 2004, we received data from 7 programmes, for a total of 1091 reported cases, among 668,022 births (Table 1). Of these, 175 were reported as syndromes and 726 had at least two major congenital anomalies, which is our current case definition of multiple congenital anomaly (MCA). Coding and interpretation of the results were done by Monica Rittler, statistical analyses, review and report writing by Jorge Lopez-Camelo.

Main findings and comments

This year, among the 48 defect groups, 28 (58.3%) were associated with an O/E ratio greater than 1. One of them (severe ear defects) reached statistical significance (Obs=15, Exp=4.8). Two defects showed a statistically significant decrease: spina bifida (Obs=22, Exp=30.2) and syndactyly (Obs=11, Exp=23.8).
A significant excess was found for 8 two-defects (Table 2) and for 3 three-defects combinations (Table 3).
For all comparisons, the data reported from 1992 through 2000, over 2,828,315 births were used as baseline.

Table 1: Cases of multiple congenital anomalies, by programme and number of defects (2004).

PROGRAMME	Births	Total cases Reported	known etiology (syndromes)	Major defects Unrelated<2	2 or +	Rate
Canada British Columbia	40878	87	13	44	30	7.3
Finland	57945	119	43	10	66	11.4
France Central East	105731	228	35	40	153	14.5
Israel IBDSP	37489	35	0	6	29	7.7
Mexico RYVEMCE	32769	47	7	7	33	10.0
South America ECLAMC	191201	541	66	83	392	20.5
USA Atlanta	52356	34	11	0	23	4.4
TOTAL	**668022**	**1091**	**175**	**190**	**726**	**14.0**

Table 2: Two-defect combinations.

Defect Combination	Observed	Expected	O/E
Hydrocephaly + Craniosynostosis	2	0	
Holoprosencephaly + Anorectal atresia	4	0.6	6.7
Other specified CNS defects + Diaphragmatic hernia	6	1.3	4.6
Other eye malformations + Severe ear malformations	5	0.6	8.3
Severe ear malformations + Choanal atresia	3	0.2	15
Cleft lip with/without palate + Cystic Kydney	6	0.9	6.7
Cleft palate + Anorectal atresia	5	1.3	3.8
Choanal atresia + Hypospadias	3	0.2	15

Table 3: Three-defects combinations.

Defect Combination	Observed	Expected	O/E
Holoprosencephaly + ear canal atresia + Anorectal atresia	3	0.0	
Anorectal atresia + Congenital Heart Defects + Preaxial limb reduction defects	6	0.6	10
Congenital Heart Defects + Cystic Kidney + Polydactyly	3	0.0	

Some of the observed significant associations, such as craniostenosis + hydrocephalus, and severe ear malformations + choanal atresia, could be due to methodological factors. Both defects could have been interpreted as pathogenetically related, leading to an underreporting or -coding of one of them, which would explain the low expected values.
Noticeably all cases with choanal atresia showing this and other significant associations that included choanal atresia, were reported by the same programme (France CE).
Other specified CNS defects + diaphragmatic hernia, as well as other eye malformations + severe ear defects: Categories of "other" defects include a heterogeneous set of anomalies, with a low number of cases for each particular type.
No methodological causes could be identified for the remaining significant associations.
Anorectal atresia + congenital heart defect + preaxial limb reduction defect: four of the six cases with this combination had other defects belonging to the Vater association.
Cleft lip with or without cleft palate + cystic kidneys: at least some of the cases could represent undiagnosed syndromes, such as Meckel.
Congenital heart defect + cystic kidney + polydactyly: all three cases had a type of heart or spleen anomaly suggesting a laterality defect.
An eventual heterogeneity across programmes for the observed decrease of spina bifida and syndactyly remains to be established.

Prenatal Diagnosis and Down Syndrome, 2004

Guido Cocchi (Italy: IMER)
Silvia Gualdi (Italy: IMER)

Introduction

The aim of the survey was to assess the variability in time and in the Monitoring System of the use and the spread of prenatal diagnostic techniques and to analyse the impact of elective termination on prevalence rates at birth of Down Syndrome (DS), in countries where elective abortions are performed.

Participation in the Clearinghouse Monitoring Systems worldwide provides a unique opportunity to analyse international variations on the use of prenatal diagnosis (Chorion Villus Sampling = CVS, Amniocentesis = AC, Cordocentesis= CC), and access to screening, as well as differences in advice and abortion legislation. In addition, repeating this study over time has made it possible to follow the evolution of these techniques and to evaluate the impact of each practice on the prevalence of DS.

2004 Data

During 2004, 21 Programs provided data on 2892 DS cases. Three Programs joined the survey for the first time and sent data: Slovak Republic, USA:Utah and Wales, two others sent data again: England-Wales and Italy: North-East after a year's lack of data). 1,541 out of 2,892 cases (53.3%) were prenatally diagnosed and terminated (Table 1).

The total number of births under surveillance in the 21 Programs was 1,652,215.

The percentage of terminations of pregnancy (ToP) ranged from the lowest values in USA:Utah (6.6%), RUSSIA:Moscow Region (7.6%), SLOVAK (10%), CANADA:Alberta (17.3%) and USA:Atlanta (19%), to the highest in the registries of French and Italian regions: FRANCE:Paris (87%), ITALY:Tuscany (78.9%), FRANCE:Central-East (74.4%), ITALY:IMER (72.9%) and FRANCE:Strasbourg (70.3%). Other Programs show percentages of terminations over 60%: AUSTRALIA:Victoria (62.5%), ITALY:BDRCam (68.8%) and CZECH Republic (69.3%).

In the European programs that provided a data set of 12 years (1993-2004), a regular increase in the percentage of ToP has been observed: 41.5% in 1993, 45.9% in 1994, 48.5% in 1995, 50.9% in 1996, 52.2% in 1997, 53.8% in 1998, 55.2% in 1999, 57.8% in 2000, 57.1% in 2001, 58.7% in 2002, 68.9% in 2003 and 69.7% in 2004. The increased rate for 2003 and 2004 is related to the lack of data of England and Wales, in fact if we add the data for 2004 of E-W the rate decreases to 59.9%.

The comparison of the rate of ToPs in 2004, between European Countries and non-European Countries (i.e. Australia:Victoria, Canada:Alberta and USA:Atlanta) is significantly different (59.9% vs 36% , ¯2 = 38.04 p< 0.001).

The terminations are related to the maternal age as shown in Table 2: the percentage of ToPs is lower in the lowest maternal age class (≤29 years). On the contrary in the higher maternal age classes: i.e. over 35 years (38-39 and ? 40) the percentage of terminations is higher.

The percentage of mothers aged over 35 years, has increased year by year and in many registries is over 20%: Sweden: 20.2%; Northern-Netherlands: 21.6%; Australia:Victoria: 22.4%; Italy:IMER: 26.4%; Italy:Tuscany: 28.1%; France:Paris: 28.7%.

The greater percentages of termination are typically detected in the Programs that show the higher values of higher aged mothers (Table3). In fact overall, the proportion of DS pregnancies which were terminated among women at higher risk (?35 years old), was over 80% in five Monitoring Systems: France:Paris and France:Central-East (92% and 85.3% respectively); in France:Strasbourg, Czech Republic and Italy:Tuscany the percentage of terminations was very similar 81%, 81.2 and 81.3 respectively. Percentages of ToPs less than 40% were observed in many programs: USA:Utah (13.8%), Russia:Moscow Region (16.7%); Canada:Alberta (20%), USA:Atlanta (25%); Italy:BDRCam (30%) and Israel:IBDSP (31.6%) (Table 3).

The most common technique of prenatal diagnosis remained AC in 2004 (Table 4), with a mean value of 67.9%. CVS, with a mean value of 30%, shows a progressive increase year by year: 18.3% in 1995, 19.0% in 1996, 19.3% in 1997, 18.2% in 1998, 20.2% in 1999, 21.8% in 2000, 22.9% in 2001, 28.6 in 2002 and in 2003.

In Australia:Victoria CVS is the first technique of prenatal detection used with a rate of 58.9%, while AC is less used. CVS has been used mainly in France:Strasbourg (50%); Finland (48.4%), England and Wales (45.5%). In the Registries of France the mean percentage is 23.6% (vs 16% of the previous year) while the mean value in Italy is 16% (vs 9.4% of the previous year). If we analyse the use of CVS in Italy we can observe a positive increase from south to north: Campania: 3.3%, Tuscany: 18.2%, Emilia-Romagna: 25%, Nord East: 29.2% (Table 4).

The Programs where CVS is most frequently used show the lowest mean gestational ages at pregnancy termination in the older maternal age group (>35) as in Australia:Victoria (13.3±1.7 wks)

(Table 5). The mean age (wks) of terminations after CVS diagnosis is heterogeneous and significantly different in the programs in both maternal age groups. In the younger group (<35 years) there is a lower limit of around 13 wks in many Registries such as: Australia:Victoria, France:Paris, Italy:IMER and Italy:Tuscany and Wales to an upper limit of 21 wks in USA.Atlanta. (Table 5).

The prevalence at birth of DS has decreased in the majority of the 14 Programs that can provide the rates for all the 12 year period. A significant negative temporal trend was observed only in the registries that showed, as expected, an increase in the termination of pregnancies: the Czech Republic, the three Monitoring Systems of France (Central-East, Paris and Strasbourg) and the three Italian Monitoring Systems (BDRCam, IMER and Tuscany) (Table 6) . These Programs are the same ones that showed the highest rates of ToPs and an increase in the terminations year by year. In the same way, the highest rates of prevalence at birth were observed in the Programs where ToPs were lowest i.e Canada:Alberta and USA:Atlanta. The Canada:Alberta registry shows a significant increase in the years. Controversial data are shown by Finland, Germany:Saxony-Anhalt registries where despite quite high rates of ToP (56.4% and 46.7% respectively see Table 2) they show a high rate of prevalence at birth i.e 12.25 per 10.000 in Finland and 9.18 in Germany:Saxony-Anhalt .

Table 1. List of the programs participating in the Prenatal Diagnosis Study in the years.

	1993	1994	1995	1996	1997	1998	1999	2000	2001	2002	2003	2004
Australia	X	X	X	X	X	X				* X	*X	*X
Canada: Alberta					X	X	X	X	X	X	X	X
Czech Republic	X	X	X	X	X	X	X	X	X	X	X	X
England & Wales	X	X	X	X	X	X	X	X	X	X		X
Finland	X	X	X	X	X	X	X	X	X	X	X	X
France:Central-East	X	X	X	X	X	X	X	X	X	X	X	X
France: Paris	X	X	X	X	X	X	X	X	X	X	X	X
France: Strasbourg	X	X	X	X	X	X	X	X	X	X	X	X
Germany: Saxony-Anhalt								X	X	X	X	X
Israel: IBDSP	X	X	X	X	X	X	X	X	X	X	X	X
Italy: BDRCam	X	X	X	X	X	X	X	X	X	X	X	X
Italy: IMER	X	X	X	X	X	X	X	X	X	X	X	X
Italy: North-East	X	X	X	X	X	X	X	X	X	X		X
Italy: Tuscany	X	X	X	X	X	X	X	X	X	X	X	X
Russia: Moscow Region											X	X
Northern Netherlands	X	X	X	X	X	X	X	X	X	X	X	X
Slovak Republic												X
Sweden								X	X	X	X	X
Usa: Atlanta	X	X	X	X	X	X	X	X	X	X	X	X
Usa: Utah UBDN												X
Wales: CARIS												X

** Australia: Victoria*

Table 2. Percentage (%) of terminations (TOP) among the total number of cases recorded in 2004

Monitoring Program	Maternal Age (years)					
	<= 29	30 – 34	35 – 37	38 – 39	>= 40	Total
Australia: Victoria	30.4	43.8	77.3	55.0	70.6	62.5
Canada: Alberta	5.6	22.2	20.0	7.1	36.4	17.3
Czech Republic	54.8	71.1	84.6	65.2	95	69.3
England and Wales	31.1	48.2	54.7		58.9	48.0
Finland	62.5	48.0	55.8	62.8	55.3	56.4
France: Central East	64.5	71.7	82.2	88.9	84.9	74.4
France: Paris	80.8	77.8	94.7	88.0	92.9	87.0
France: Strasbourg	50.0	58.3	100	100	50.0	71.8
Germany: Saxony-Anhalt	22.2	60.0	50.0	50.0	71.4	46.7
Israel: IBDSP	45.5	33.3	60.0	50.0	10.0	35.9
Italy: BDRCam	50.0	50.0	82.6	70.0	81.1	68.8
Italy: IMER	33.3	88.9	80.0	60.0	83.3	72.9
Italy: North-East	20.0	41.7	52.4	55.6	82.4	50.0
Italy: Tuscany	0	75.0	85.7	84.6	76.2	78.9
Northern Netherlands	12.5	28.6	50.0	66.7	57.1	37.9
Russia: Moscow Region	3.6	5.0	0	0	30.0	7.6
Slovak Republic	5.0	0	23.1		12.5	10.0
Sweden	45.7	38.2	47.5	80.0	72.6	56.3
USA: Atlanta	6.3	15.0	46.7	18.2	13.6	19.0
USA : Utah UBDN	0	7.7	10.0	20.0	14.3	6.6
Wales: CARIS	15.0	62.5	44.7	75.0	66.7	48.6

Table 3. Percentage of mothers aged 35 and over in the monitoring programs participating in the study and percentage of terminations (ToP) in the same group of mothers. Prevalence rate in live and stillbirths (per 10,000) and comparison with the rate after inclusion of ToP

Monitoring Program	% of mothers	% of ToP in mothers	Prevalence rate (* 10,000)	
	aged >=35	aged >=35	L+S	L+S+ToP
Australia: Victoria	22.4	70.4	24.3	82.2
Canada: Alberta	14.8	20.0	53.4	75.1
Czech Republic	8.1	81.2	16.3	86.6
England and Wales	19.2	56.2	17.5	40.0
Finland	19.5	56.6	40.6	93.7
France: Central East	18.6	85.3	10.7	72.6
France: Paris	28.7	92.0	7.0	88.0
France: Strasbourg	15.1	81.0	19.7	103.3
Germany: Saxony-Anhalt	11.7	60.0	29.4	73.5
Israel: IBDSP	16.9	31.6	20.5	29.9
Italy: BDRCam	17.6	30.0	10.3	51.3
Italy: IMER	26.4	73.8	14.9	56.9
Italy: Tuscany	28.1	81.3	11.4	60.7
Northern Netherlands	21.6	57.1	14.5	33.9
Russia: Moscow region	7.0	16.7	37.3	44.8
Sweden	20.2	65.2	26.4	75.9
USA: Atlanta	16.5	25.0	41.7	55.7
USA: Utah UBDN	9.0	13.8	54.9	63.7
Wales: CARIS	15.6	60.5	29.7	75.1

Italy: North East and Slovak Republic not included for lack of data

Table 4 . Down Syndrome techniques of prenatal diagnosis (number of cases) registered in 2003 by maternal age classe.

Monitoring Program	<35				35-39				>39				Tot			
	CVS	AC	CC	UK	CVS	AC	CC	UK	CVS	AC	CC	UK	CVS	AC	CC	UK
Australia: Victoria	10	10	0	1	26	19	0	0	30	17	0	2	66	46	0	3
Canada: Alberta	0	5	0	0	0	5	0	0	0	3	0	1	0	13	0	1
Czech Republic	2	64	0	0	1	36	0	0	1	18	0	0	4	118	0	0
England and Wales	52	70	nr	nr	77	79	nr	nr	53	39	nr	nr	182	188	30	19
Finland	12	20	0	0	16	17	0	1	16	10	0	0	44	47	0	1
France: CentralEast	5	40	0	8	15	58	0	4	6	32	0	7	26	134	0	20
France: Paris	14	35	0	0	11	29	0	0	13	38	0	1	38	102	0	1
France: Strasbourg	5	4	0	0	6	7	0	0	2	2	0	0	13	13	0	0
Germany: Saxony-Anhalt	0	5	0	0	0	4	0	0	0	5	0	0	0	14	0	0
Israel: IBDSP	0	8	0	0	0	5	0	0	0	1	0	0	0	14	0	0
Italy: BDRCam	0	15	0	0	0	26	0	0	0	18	0	0	2*	59	0	0
Italy: IMER	1	9	0	0	6	15	0	0	3	6	0	1	10	30	0	1
Italy: North-East	1	4	0	1	3	9	0	4	3	4	0	7	7	17	0	12
Italy: Tuscany	3	3	0	0	1	21	0	0	4	12	0	0	8	36	0	0
Northern Netherlands.	1	2	0	0	2	1	0	1	1	3	0	0	4	6	0	1
Sweden	6	30	0	1	12	50	0	2	5	32	0	0	23	112	0	3
USA: Atlanta	1	3	0	0	2	7	0	0	1	2	0	0	4	12	0	0
USA: Utah UBDN	0	1	0	0	0	2	0	0	0	2	0	0	0	5	0	0
Wales: CARIS	1	12	0	0	4	9	0	0	3	7	0	0	8	28	0	0
Total	**104**	**340**	**0**	**11**	**182**	**399**	**0**	**12**	**141**	**251**	**0**	**19**	**439**	**994**	**30**	**62**

CVS = Chorion Villus sampling
AC = Amniocentesis
CC = Chordocentesis
UK = Unknown
* maternal age unknown

Table 5. Mean gestational age (weeks) and Standard Deviation of induced abortions by maternal age group and by type of prenatal diagnosis.

Monitoring Program	<=34			>=35		
	CVS	AC	Total	CVS	AC	Total
Australia: Victoria	13.2±0.8	17.3±1.3	15.4±2.4	13.3±1.7	16.9±1.7	14.9±2.5
Canada: Alberta	-	17.8±3.4		-	18.6±2.3	
Czech Republic	15.5±0.7	19.3±2.1	19.1±2.1	14.5±2.1	19.4±1.9	19.2±2.0
Finland	15.1±1.8	18.0±2.3	16.9±2.3	14.6±1.3	18.0±1.3	16.1±2.2
France: Central East	14.4±0.9	22.0±5.1	20.8±5.5	14.5±1.8	19.7±3.0	18.6±3.5
France: Paris	13.5±0.7	21.0±4.7	18.9±5.6	14.4±1.8	21.3±4.8	19.5±5.2
France: Strasbourg	16.2±3.4	23.2±2.5	19.3±4.7	14.6±1.7	22.7±5.4	18.9±5.7
Germany: Saxony-Anhalt	-	20.2±3.7		-	17.9±2.2	
Israel: IBDSP	-	22.8±1.3		-	21.5±1.4	
Italy: BDRCam	-	21.1±2.1		-	20.4±2.3	
Italy: IMER	13.0±0.0	20.7±1.0	19.6±3.1	14.0±0.9	21.4±1.3	19.2±3.7
Italy: North-East	15.0±0.0	18.8±1.5	18.0±2.1	14.5 ±1.4	19.3±1.6	17.8±2.7
Italy: Tuscany	13.0±1.4	16.0±2.7	14.8±2.6	13.8±4.6	18.9±1.1	18.2±2.6
Northern Netherlands	15.0±0.0	18.0±1.4	17.0±2.0	14.0±2.0	18.3 ±1.0	16.4±2.6
Sweden	14.7±1.5	17.4±2.1	17.1±2.2	13.8±1.3	17.2±1.5	17.0±1.7
USA: Atlanta	21.0±0.0	23.0±0.0	22.0±1.4	19.7±5.5	20.0±3.6	19.8±4.2
USA: Utah UBDN	-	22.0±0.0		-	18.8±1.0	
Wales: CARIS	13.0±.0	18.7±3.1	18.2±3.1	14.0±1.3	18.5±2.2	17.1±2.9

England and Wales and Russia: Moscow Region not included for lack of data

Table 6. Prevalence at birth (x 10,000) in the years of DS in the 14 programs participating in the survey for all the years.

Programme	1993	1994	1995	1996	1997	1998	1999	2000	2001	2002	2003	2004	P
Canada Alberta	11.4	11.1	13.2	8.5	11.1	14.0	11.6	14.7	15.2	12.7	19.2	0.7	<0.01*
Czech Republic	7.5	7.7	7.3	5.5	5.1	6.7	6.6	5.4	5.5	5.4	6.4	5.5	<0.05**
Finland	13.2	12.8	12.9	10.3	10.1	1.3	10.0	11.8	14.2	14.2	12.3	12.3	ns
France Central East	11.0	10.4	8.9	9.5	9.0	6.8	4.9	5.8	5.9	5.5	4.9	5.9	<0.0001**
France Paris	10.6	9.2	7.1	9.7	7.8	10.5	5.2	7.9	7.8	6.2	4.7	5.3	<0.01**
France Strasbourg	16.8	17.9	24.0	17.4	28.0	2.2	4.3	5.6	2.2	3.0	5.2	8.2	<0.01**
Germany Saxony Anhalt	5.8	6.3	7.4	7.9	8.3	13.7	6.1	6.4	8.3	9.1	5.3	9.2	ns
Israel IBDSP	5.1	5.0	6.3	4.9	9.1	3.3	6.0	5.7	6.2	4.8	6.5	6.7	ns
Italy BDRCam	19.9	7.6	10.0	9.2	6.7	8.7	6.3	3.0	6.8	5.4	5.2	4,8	<0.005**
Italy IMER	9.0	9.3	10.2	8.0	7.3	9.4	9.6	6.5	6.3	6.2	8.1	5.7	<0.05**
Italy Tuscany	11.8	9.8	11.4	6.9	7.3	6.3	6.1	4.9	5.7	3.8	4.0	4.1	<0.0001**
Northern Netherlands	9.9	5.7	9.4	13.7	11.9	10.0	8.4	6.4	9.3	13.3	6.0	9.4	ns
Sweden	12.3	15.3	12.3	13.7	12.4	11.9	14.0	11.0	14.6	13.3	15.5	10.6	ns
USA Atlanta	12.0	13.8	10.9	12.0	10.5	11.5	12.0	11.1	13-3	0.5	13.0	13.0	ns

** Increasing Trend*

*** Decreasing Trend*

Monitoring Program	Coverage	Year Joined ICBDSR	Maximum age at diagnosis	Criteria defining stillbirths
Australia: National	Population-based National	1981	1 year	20 weeks or 400 grams
Australia:VBDR	Population-based Statewide	2002	Up to 15 years	20 weeks or 400 grams
Australia: WABDR	Population-based, Statewide	2002	Up to 6 years	20 weeks or 400 grams
Canada: Alberta	Population-based Provincial	1996	1 year	20 weeks or 500 grams
Canada: British Columbia	Population-based Provincial	2001	No limit	At least 20 weeks or 500 grams
Canada: National	Population-based National	1996	1 year	20 weeks or 500 grams
Chile-Maule	Hospital-based Regional	2003	Hospital discharge	500 grams
China: Beijing	Population-based Four Provinces	1997	6 weeks	20 weeks
China: CBDMN	Hospital-based	1985	7 days	28 weeks
Costa Rica: CREC	Population-based National	2003	3 days	22 weeks or 500 grams
Cuba	Hospital based National	2003	Hospital discharge	500 grams
Czech Republic	Population-based National	1974	Up to 15 years	non-viable fetuses, 28 weeks or >1000 grams
England and Wales	Population-based National	1974	1995 onwards no limit	24 weeks
Finland	Population-based National	1974	1 year	22 weeks or 500 grams
France: Central-East	Population-based Regional	1974	1 year	22 weeks
France: Paris	Population-based Regional	1982	Hospital discharge	22 weeks
France: Strasbourg	Population-based Regional	1982	2 years	22 weeks or 500 grams
Germany: Saxony-Anhalt	Population-based (Federal State)	2001	Hospital discharge (almost first week of life) – up to 1 year	>/= 500 grams
Hungary	Population-based National	1974	1 year	24 weeks or 500 grams
Ireland: Dublin	Population-based Regional	1997	5 years	24 weeks or 500 grams
Iran: Tabriz	Hospital-based Regional	2006	1 year	20 weeks or 400 grams
Israel: IBDSP	Hospital-based Regional	1974	Hospital discharge 2-5 days	20weeks or 500 grams
Italy: BDRCam	Population-based Regional	1996	7 days	180 days (25 weeks + 5 days)
Italy: IMER	Population-based Regional	1985	7 days	180 days (25 weeks + 5 days)
Italy: ISMAC	Hospital-based Regional	1991	1 year	180 days (25 weeks + 5 days)
Italy: North East	Population-based Regional	1997	7 days	180 days (25 weeks + 5 days)
Italy: Tuscany	Population-based Regional	1998	1 year	180 days (25 weeks + 5 days)
Japan: JAOG	Hospital-based National	1988	7 days	22 weeks
Malta	Population-based National	2000	1 year	20 weeks
Mexico: RYVEMCE	Hospital based-National	1980	72 hours	20 weeks or 500 grams
New Zealand	Population-based National	1979	No limit	20 weeks or 400 grams
Northern Netherlands	Population-based Regional	1993	Up to 15 years	24 weeks
Norway	Population-based National	1974	Hospital discharge Lifelong for mortality (from 2002 1 year)	16 weeks (12 weeks from 1999)
Russia Moscow Region	Population-based Regional	2001	1 year	28 weeks
Slovak Republic	Population-based Regional	2003	1 year	Non-viable fetuses, 28 weeks or >1000 grams
South Africa: SABDSS	Hospital-based	1992	Hospital discharge (usually 4 days)	Stillbirths not recorded
South America: ECLAMC	Hospital-based Multinational	1977	3 days	500 grams
Spain: ECEMC	Hospital-based National	1979	3 days	24 weeks or 500 grams
Sweden	Population-based National	1974	28 days	22 weeks
Ukraine	Population-based Regional	2001	28 days	500 grams
United Arab Emirates	Hospital-based Regional	1995	7 days	23 weeks
USA: Atlanta	Population-based Regional	1974	6 years	20 weeks
USA: Texas	Population-based Regional	2004	1 year	Before 1999: 20 weeks. Since 1999: All stillbirths with documented birth defects included
USA: Utah UBDN	Population-based Regional	2005	2 years	20 weeks
Wales: CARIS	Population-based Regional	2005	1 year	24 weeks

The following definitions have been adopted by all monitoring systems except when indicated in the Table 7.1

1. Anencephaly: a congenital malformation characterized by the total or partial absence of the cranial vault, the covering skin, and the brain missing or reduced to small mass. Incl. craniorachischisis. Incl. infants with iniencephaly and other neural tube defects as encephalocele or open spina bifida, when associated with anencephaly. Excl. acephaly, that is, absence of head observed in amorphous acardiac twins.

2. Spina bifida: a family of congenital malformation defects in the closure of the spinal column characterized by herniation or exposure of the spinal cord and/or meninges through an incompletely closed spine. Incl. meningocele, meningomyelocele, myelocele, myelomeningocele, rachischisis. Spina bifida is not counted when present with anencephaly. Excl. spina bifida occulta, sacrococcygeal teratoma without dysraphism .

3. Encephalocele: a congenital malformation characterized by herniation of the brain and/or meninges through a defect in the skull. Encephalocele is not counted when present with spina bifida.

4. Microcephaly: a congenitally small cranium, defined by an occipito frontal circumference (OFC) 3 standard deviation below the age and sex appropriate distribution curves. [If using a different definition or cut off point (e.g., 2 standard deviations), report but specify criteria]. Excl. microcephaly associated with anencephaly or encephalocele.

5. Arhinencephaly/holoprosencephaly: a congenital malformation of the brain, characterized by various degrees of incomplete lobation of the brain hemispheres. Olfactory nerve tract may be absent. Holoprosencephaly includes cyclopia, ethmocephaly, cebocephaly, and premaxillary agenesis.

6. Hydrocephaly: a congenital malformation characterized by dilatation of the cerebral ventricles, not associated with a primary brain atrophy, with or without enlargement of the head, and diagnosed at birth. Not counted when present with encephalocele or spina bifida. Excl. macrocephaly without dilatation of ventricular system, skull of macerated fetus, hydranencephaly, holoprosencephaly, and postnatally acquired hydrocephalus.

7. Anophthalmos/microphthalmos: apparently absent or small eyes. Some normal adnexal elements and eyelids are usually present. In microphthalmia, the corneal diameter is usually less than 10 mm. and the antero posterior diameter of the globe is less than 20 mm.

8. Anotia/microtia: a congenital malformation characterized by absent parts of the pinna (with or without atresia of the ear canal) commonly expressed in grades (I IV) of which the extreme form (grade IV) is anotia, absence of pinna.
Excl. small, normally shaped ears, imperforate auditory meatus with a normal pinna, dysplastic and low set ears.

9. Transposition of great vessels: a cardiac defect where the aorta exits from the right ventricle and the pulmonary artery from the left ventricle, with or without other cardiac defects. Incl. double outlet ventricle so called corrected transposition.

10. Tetralogy of Fallot: a condition characterized by ventricular septal defect, overriding aorta, infundibular pulmonary stenosis, and often right ventricular hypertrophy.

11. Hypoplastic left heart syndrome: a cardiac defect with a hypoplastic left ventricle, associated with aortic and/or mitral valve atresia, with or without other cardiac defect.

12. Coarctation of the aorta: an obstruction in the descending aorta, almost invariably at the insertion of the ductus arteriosus

13. Choanal atresia, bilateral: congenital obstruction (membraneous or osseous) of the posterior choana or choanae. Excl. choanal stenosis and congestion of nasal mucosa.

14. Cleft palate without cleft lip: a congenital malformation characterized by a closure defect of the hard and/or soft palate behind the foramen incisivum without cleft lip. Incl. submucous cleft palate. Excl. cleft palate with cleft lip, cleft uvula, functional short palate, and high narrow palate.

15. Cleft lip with or without cleft palate: a congenital malformation characterized by partial or complete clefting of the upper lip, with or without clefting of the alveolar ridge or the hard palate. Excl. midline cleft of upper or lower lip and oblique facial fissure (going towards the eye).

16. Oesophageal atresia/stenosis: a congenital malformation characterized by absence of continuity or narrowing of the esophagus, with or without tracheal fistula. Incl. tracheoesophageal fistula with or without mention of atresia or stenosis of oesophagus.

17. Small intestine atresia/stenosis: complete or partial occlusion of the lumen of a segment of the small intestine. It can involve a single area or multiples areas of the jejunum or ileum. Excl. duodenal atresia.

18. Anorectal atresia/stenosis: a congenital malformation characterized by absence of continuity of the anorectal canal or of communication between rectum and anus, or narrowing of anal canal, with or without fistula to neighboring organs. Excl. mild stenosis which does not need correction, and ectopic anus.

19. Undescended testis: bilateral undescended testes in at term newborn or at least unilateral undescended testis in males more than 1 year of age. Excl. retractrile testis.

20. Hypospadias: a congenital malformation characterized by the opening of the urethra on the ventral side of the penis, distally to the sulcus.
Incl. penile, scrotal, and perineal hypospadias. Excl. glandular or first degree hypospadias and ambiguous genitalia (intersex or pseudohermaphroditism).

21. Epispadias: a congenital malformation characterized by the opening of the urethra on the dorsal surface of the penis. Not counted when part of exstrophy of the bladder.

22. Indeterminate sex: genital ambiguity at birth that does not readily allow for phenotypic sex determination. Incl. male or female true or pseudohermaphroditism.

23. Renal agenesis: a congenital malformation characterized by complete absence of kidneys bilaterally or severely dysplastic kidneys.

24. Cystic kidney: a congenital malformation characterized by multiple cysts in the kidney. Incl. infantile polycystic kidney, multicystic kidney, other forms of cystic kidney and unspecified cystic kidney. Excl. single kidney cyst.

25. Bladder exstrophy: complex malformation characterized by a defect in the closure of the lower abdominal wall and bladder. Bladder opens in the ventral wall of the abdomen between the umbilicus and the symphysis pubis. It is often associated with epispadias and structural anomalies of the pubic bones.

26. Polydactyly, preaxial: extra digit(s) on the radial side of the upper limb or the tibial side of the lower limb. It can affect the hand, the foot, or both.

27. Limb reduction defects: a congenital malformation characterized by total or partial absence or severe hypoplasia of skeletal structures of the limbs.
Incl. femoral hypoplasia. Excl. mild hypoplasia with normal shape of skeletal parts, brachydactyly, finger or toe reduction directly associated with syndactyly, general skeletal dysplasia and sirenomelia.

28. Diaphragmatic hernia: a congenital malformation characterized by herniation into the thorax of abdominal contents through a defect of the diaphragm. Incl. total absence of the diaphragm. Excl. hiatus hernia, eventration and phrenic palsy.

29. Abdominal wall defects: cases specified as omphalocele and/or gastroschisis plus unspecified cases.

30. Omphalocele: a congenital malformation characterized by herniation of abdominal contents through the umbilical insertion and covered by a membrane which may or may not be intact. Excl. gastroschisis (para umbilical hernia), a or hypoplasia of abdominal muscles, skin covered umbilical hernia.

31. Gastroschisis: a congenital malformation characterized by visceral herniation through a right side abdominal wall defect to an intact umbilical cord and not covered by a membrane. Excl. a or hypoplasia of abdominal muscles, skin covered umbilical hernia, omphalocele.

32. Prune belly sequence: a complex congenital malformation characterized by deficient abdominal muscle and urinary obstruction/distension. It can be caused by urethral obstruction secondary to posterior urethral valves or urethral atresia. In the affected fetus the deficiency of the abdominal muscle may not be evident. It can be associated with undescended testes, clubfoot, and limb deficiencies.

33. Trisomy 13: a congenital chromosomal malformation syndrome associated with extra chromosome 13 material. Incl. translocation and mosaic trisomy 13.

34. Trisomy 18: a congenital chromosomal malformation syndrome associated with extra chromosome 18 material. Incl. translocation and mosaic trisomy 18

35. Down syndrome: a congenital chromosomal malformation syndrome characterized by a well known pattern of minor and major anomalies and associated with excess chromosomal 21 material. Incl. trisomy mosaicism and translocations of chromosome 21.

Deviations from the ICBDSR Definitions by Registry

	Encephalocele	Microcephaly	Arhinencephaly / Holoprosencephaly	Hydrocephaly	Anophthalmos / Microphthalmos	Anotia	Transposition of great vessels	Tetralogy of Fallot	Choanal atresia, bilateral	Cleft palate without cleft lip	Cleft lip with or without cleft palate	Oesophageal atresia / stenosis	Small intestine atresia / stenosis	Anorectal atresia / stenosis	Undescended testis	Hypospadias	Epispadias	Indeterminate sex	Renal agenesis	Cystic kidney	Polydactyly, preaxial	Limb reduction defects	Prune belly sequenze	Trisomy 13	Trisomy 18	Down syndrome
Australia: VBDR									11							25					35					
Australia: WABDR									11							25			28		35					
Canada: Alberta		2			2	7	8		11\12							25					35					2
Canada: British Columbia	1	2	4	6	2	7	8	10	11\12	13	15		18\19			25	25\26	27	28	30	35	37		2	2	2
Canada: National	1	1\2		6	2				11\12	14			18	21	23	25	26		28	31	35		40	2	2	2
China: Beijing																					35					
China: CBDMN	1	2		6	2	7	9		12				18			25		27	28	31	35	37		2	2	2
Costa Rica: CREC				6			9		11\12								26	27	28	31	35			2	2	2
Cuba: RECUMAC	1	2		6	2	7			11	14	15		18			25	26	27	28	32	35	37		2	2	2
Czech Republic																25					35					
England and Wales																										
Finland	1	2			2		8		11							25		27		32		37		2	2	2
France: Central East																25										2
France: Paris																25										
France: Strasbourg		2			2		9						18\19						28\29	30\33						
Germany: Saxony-Anhalt																										
Hungary	1	2			2		9									25	26				35	38\39		2	2	2
Ireland: Dublin		2			2				11				18\19			25	26				35			2	2	2
Israel: IBDMS							8									25				33						
Italy: BDRCAM																								2	2	2
Italy: IMER																25					35					
Italy: ISMAC																25										
Italy: North East			5		2					13	15	17	18\20	22					29		35					2
Italy: Tuscany							8																			
Japan: JAOG		2			2															31						
Malta		2			2		9		11									27		31	35	37		2	2	2
Mexico: RYVEMCE	1	2			2				11\12				18					27	28	30	35	37		2	2	2
New Zealand					2											25	26							2	2	2
Northern Netherlands															24	25					35					
Norway																										
Russia: Moscow region		2					9			13	15		18			25				31	36					2
South Africa: SABDSS	1	2			2				11\12							25		27		31	35	37		2	2	2
South America: ECLAMC																25										
Spain: ECEMC		3			2													27				37				2
Sweden		2			2				11							25			28	32						2
Ukraine		2\3		6	2	7	9				16							27						2	2	2
United Arab Emirates		2			2	7	8	10	11				18						28/29	31						
USA: Atlanta									12		16															
Wales: CARIS	1	2			2	7									24	25								2	2	2

ICBDSR Definitions of the Reported Malformations

1 = when present with spina bifida counted
2 = clinical diagnosis included
3 = OCF below 3rd percentile
4 = there may be other defects with the same code
5 = only cyclopia included
6 = hydranencephaly included
7 = absence of auricle
8 = double outlet right ventricle excluded
9 = all kind of transposition included
10 = Trilogy of Fallot included
11 = unilateral cases included
12 = stenosis included
13 = submucous cleft palate excluded
14 = Cleft uvula included
15 = midline and oblique facial clefts included
16 = clefts of the alveolar ridge without cleft lip included
17 = stenosis excluded
18 = doudenal atresia included
19 = doudenal stenosis excluded
20 = intestinal stenosis excluded
21 = Large intestine atresia/stenosis included
22 = stenosis excluded
23 = No gestational age information
24 = Registered when it is combined with other defects
25 = all types included
26 = epispadias counted with hypospadias
27 = genital ambiguity and absent genitalia included
28 = unilateral defects included
29 = severely dysplastic kidneys excluded
30 = single cyst included
31 = all kind of cystic kidney included
32 = all cystic kidneys are included except for single renal cysts
33 = AR polycystic kidney excluded
34 = some autosomalrecessive polycystyc kidneys are not excluded
35 = any type of polydactyly included
36 = polysyndactyly preaxial excluded
37 = any hypoplasia of skeletal limb structures included except branchydactyly and hypoplasia as part of skeletal dysplasia
38 = any hypoplasia of skeletal structures included
39 = sirenomelia included
40 = Prune belly sequence counted with Total abdominal wall defects

Australia: VBDR
Victoria Birth Defects Registry

History:
In 1979 the Commonwealth Government agreed in principle to collect more information about births and birth defects. It was decided that the States would be responsible for setting up their own systems and the Commonwealth would establish a National Perinatal Statistics Unit, to collate information from all the states and provide an overall picture. The Victorian Perinatal Data Collection Unit (PDCU), established under the Health Act of 1958, operates under the aegis of the Consultative Council on Obstetric and Paediatric Mortality and Morbidity (the Council). One of the fundamental purposes of the PDCU was the establishment and maintenance of a Birth Defects/Congenital Malformations Register (BDR). The PDCU and BDR were established in 1982.

Size and coverage:
The BDR collects information on all birth defects for livebirths, stillbirths and terminations of pregnancy pre 20 wks gestation and children up to 15 yrs of age (irrespective of the age at diagnosis). Approximately 3.6% of babies are born with a birth defect at or after 20 weeks gestation. We also follow up terminations for malformations before 20 weeks, once these are included the overall prevalence is approximately 4%. Birth defects are notified to the register for those babies/fetus' who were born in Victoria

Legislation and funding:
The ongoing maintenance of the BDR is enshrined in the legislation pertaining to the PDCU (Health Act 1958) and is an ongoing function of the PDCU, however notification of birth defects outside the reporting period on the Perinatal Morbidity Statistics form (28 days) is a voluntary process. There is a section for reporting of birth defects on the Perinatal form which is completed at the time of birth. Several measures are taken to ensure the ascertainment of birth defects outside this reporting period which will be specified in 'sources of ascertainment'. The PDCU & BDR are funded by the Department of Human Services (State Government)

Sources of ascertainment:
Perinatal forms (approx 52.5%)
Hospital listings* (aprox 27.1%)
Perinatal Death Certificates (approx 5.2%)
Autopsy Reports (approx 2.9%)
Cytogenetic Reports (approx 6.7%)
Maternal & Child Health Nurse (approx 4.7%)
Other professionals/parents (approx0.1%)

* these include obtaining inpatient listings from the Royal Children's Hospital (RCH) detailing all children born since 1982 who have been subsequently admitted to the RCH with a birth defect. We also obtain listings of all children born since 1982 who have visited the RCH Cardiology Unit and Metabolic Clinic, either as an inpatient or an outpatient. This procedure has also been adopted for Monash Medical Centre. Other listing received include cystic fibrosis, hypothyroidism, cerebral palsy.

Exposure information:
No exposure information is available

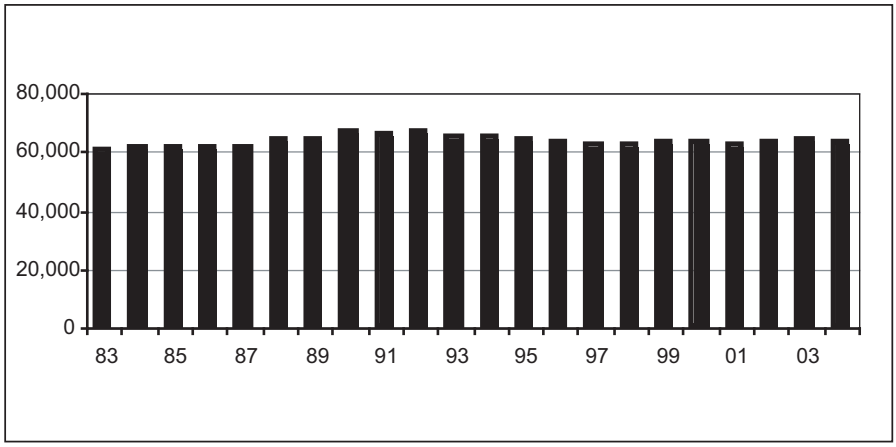
Addresses and Staff:
Jane Halliday PhD
Victorian Birth Defects Registry:
Epidemiologist, Birth Defects Register
Perinatal Data Collection Unit
7/589 Collins St, Melbourne 3000

Phone: 96162729

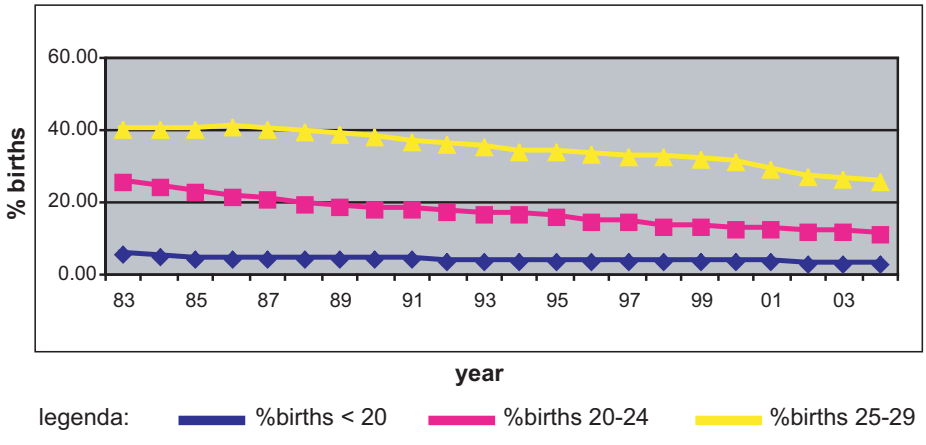
E-mail: Jane.Halliday@dhs.vic.gov.au

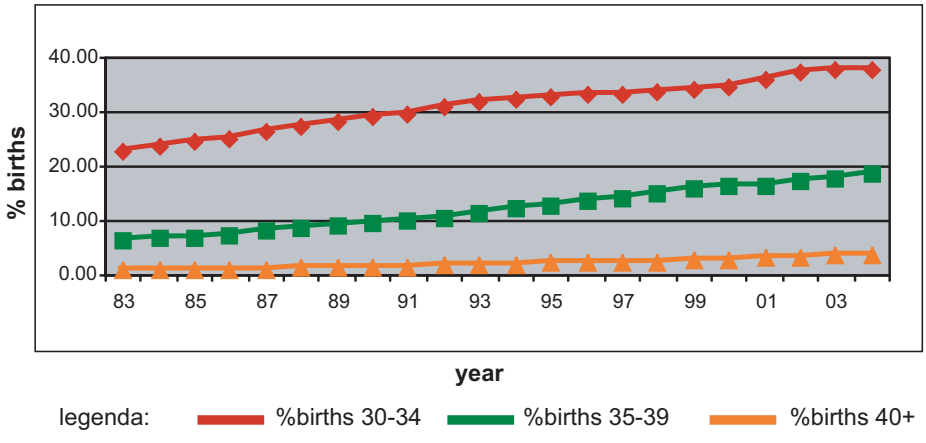
Australia: VBDR

Total births by year



Percentage of births by maternal age





Australia: VBDR, 2004

Live births (LB)	63,082
Stillbirths (SB)	618
Total births	63,700
Number of terminations of pregnancy (ToP) for birth defects	331

Birth Defects	Number of cases			Rates*10,000
	LB	SB	ToP	Total rate
Anencephaly	5	4	32	6.44
Spina bifida	9	14	15	5.97
Encephalocele	3	3	4	1.57
Microcephaly	18	0	2	3.14
Arhinencephaly / Holoprosencephaly	2	1	4	1.10
Hydrocephaly	38	9	12	9.26
Anophthalmos	1	1	0	0.31
Microphthalmos	3	0	0	0.47
Unspecified Anophthalmos/ Microphthalmos	0	0	0	---
Anotia	4	1	0	0.78
Microtia	2	0	0	0.31
Unspecified Anotia/Microtia	0	0	0	---
Transposition of great vessels	30	5	3	5.97
Tetralogy of Fallot	18	3	3	3.77
Hypoplastic left heart syndrome	15	9	4	4.40
Coarctation of aorta	18	2	0	3.14
Choanal atresia, bilateral	10	0	0	1.57
Cleft palate without cleft lip	52	3	3	9.11
Cleft lip with or without cleft palate	59	3	3	10.20
Oesophageal atresia / stenosis with or without fistula	13	2	0	2.35
Small intestine atresia / stenosis	15	1	0	2.51
Anorectal atresia / stenosis	12	3	2	2.67
Undescended testis (36 weeks of gestation or later)	309	0	0	48.51
Hypospadias	191	0	0	29.98
Epispadias	3	0	0	0.47
Indeterminate sex	8	0	3	1.73
Renal agenesis	32	7	4	6.75
Cystic kidney	36	3	2	6.44
Bladder exstrophy	2	0	0	0.31
Polydactyly, preaxial	74	2	5	12.72
Total Limb reduction defects (include unspecified)	32	10	1	6.75
Transverse	nr	nr	nr	nr
Preaxial	nr	nr	nr	nr
Postaxial	nr	nr	nr	nr
Intercalary	nr	nr	nr	nr
Mixed	nr	nr	nr	nr
Unspecified	nr	nr	nr	nr
Diaphragmatic hernia	16	3	3	3.45
Omphalocele	5	5	6	2.51
Gastroschisis	9	0	1	1.57
Unspecified Omphalocele/Gastroschisis	1	0	0	---
Prune belly sequence	1	0	0	0.16
Trisomy 13	3	1	16	3.14
Trisomy 18	4	11	50	10.20
Down syndrome, all ages (include age unknown)	59	10	115	28.89
<20	0	0	0	0.00
20-24	5	0	2	9.85
25-29	9	2	5	9.93
30-34	13	5	14	13.62
35-39	19	0	45	54.42
40-44	12	2	33	212.29
45+	1	0	3	449.44
unknown	0	0	14	---

nr= not reported

Australia: VBDR, Previous years rates 1983- 2004

Birth prevalence rates: (LB+SB+TOP) * 10,000

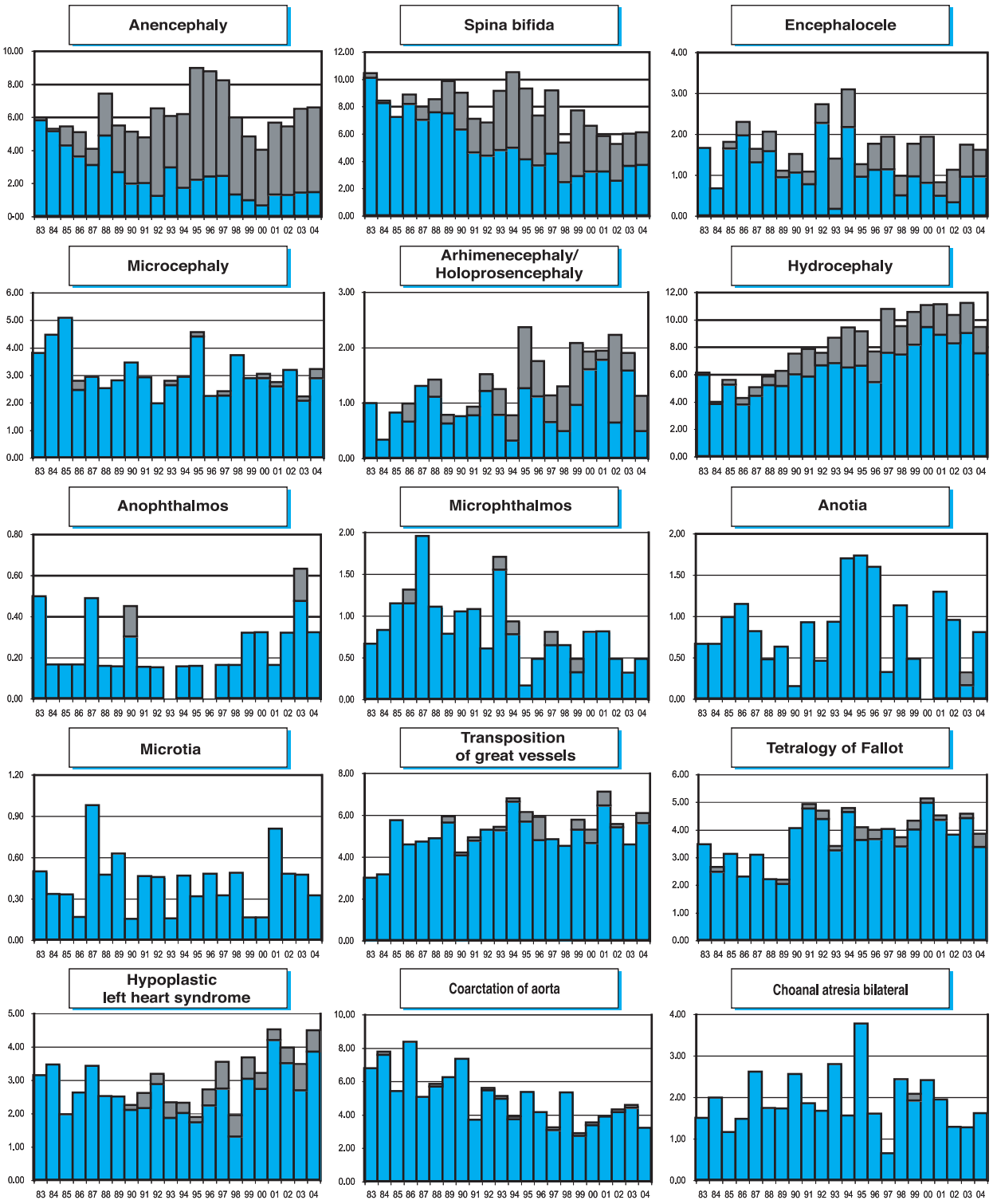
	1974-79	1980-84*	1985-89	1990-94	1995-99	2000-04
Total births		**121,365**	**311,930**	**328,100**	**313,756**	**313,876**
Anencephaly		5.60	5.48	5.70	7.33	5.61
Spina bifida		9.39	8.46	8.47	7.74	5.93
Encephalocele		1.15	1.76	1.95	1.53	1.43
Microcephaly		4.12	3.21	2.80	3.16	2.87
Arhinencephaly / Holoprosencephaly		0.66	1.06	1.04	1.72	1.82
Hydrocephaly		5.03	5.39	8.17	9.50	10.61
Anophthalmos		0.33	0.22	0.18	0.16	0.35
Microphthalmos		0.74	1.25	1.07	0.51	0.57
Unspecified Anophthalmos / Microphthalmos		---	---	---	---	---
Anotia		0.66	0.80	0.82	1.05	0.67
Microtia		0.41	0.51	0.34	0.35	0.45
Unspecified Anotia / Microtia		---	---	---	---	---
Transposition of great vessels		3.05	5.16	5.30	5.42	5.70
Tetralogy of Fallot		3.05	2.56	4.36	4.02	4.36
Hypoplastic left heart syndrome		3.30	2.60	2.53	2.74	3.92
Coarctation of aorta		7.25	6.16	5.12	4.18	3.89
Choanal atresia, bilateral		1.73	1.73	2.07	2.10	1.69
Cleft palate without cleft lip		9.06	7.08	6.92	8.32	9.69
Cleft lip with or without cleft palate		10.88	10.26	10.64	10.14	10.58
Oesophageal atresia / stenosis with or without fistula		3.87	3.33	3.78	3.86	2.71
Small intestine atresia / stenosis		1.73	2.63	2.56	2.84	3.35
Anorectal atresia / stenosis		3.63	3.40	4.36	5.10	3.73
Undescended testis (36 weeks of gestation or later)		6.84	20.93	42.03	48.64	46.58
Hypospadias		17.14	21.03	31.03	35.06	34.15
Epispadias		0.08	0.42	0.30	0.54	0.57
Indeterminate sex		1.24	2.31	2.74	2.01	1.56
Renal agenesis		4.28	5.23	4.69	7.08	6.85
Cystic kidney		2.47	3.72	4.97	6.98	6.66
Bladder exstrophy		0.41	0.42	0.24	0.54	0.51
Polydactyly, preaxial		7.09	8.05	9.75	10.65	10.80
Total Limb reduction defects (include unspecified)		6.10	5.87	6.22	7.46	5.48
Transverse		nr	nr	nr	nr	nr
Preaxial		nr	nr	nr	nr	nr
Postaxial		nr	nr	nr	nr	nr
Intercalary		nr	nr	nr	nr	nr
Mixed		nr	nr	nr	nr	nr
Unspecified		nr	nr	nr	nr	nr
Diaphragmatic hernia		3.13	2.60	4.08	4.21	3.06
Omphalocele		1.90	3.11	3.05	3.86	3.15
Gastroschisis		0.49	1.25	1.68	2.68	2.36
Unspecified Omphalocele / Gastroschisis		---	---	---	---	---
Prune belly sequence		0.33	0.32	0.27	0.41	0.06
Trisomy 13		0.82	1.25	1.80	2.61	2.99
Trisomy 18		1.65	2.98	4.54	6.31	7.87
Down syndrome, all ages (include age unknown)		11.21	15.48	17.40	21.99	28.36
<20		11.76	5.87	7.18	8.55	3.12
20-24		7.62	6.85	8.95	7.24	9.06
25-29		7.26	9.22	8.54	8.30	10.21
30-34		12.13	16.96	15.46	16.66	17.84
35-39		30.50	49.59	45.90	50.02	57.93
40-44		106.08	116.28	135.11	195.12	185.47
45+		153.85	72.46	368.42	438.25	318.30
unknown		---	---	---	---	---

* data include less than 5 years

nr = not reported

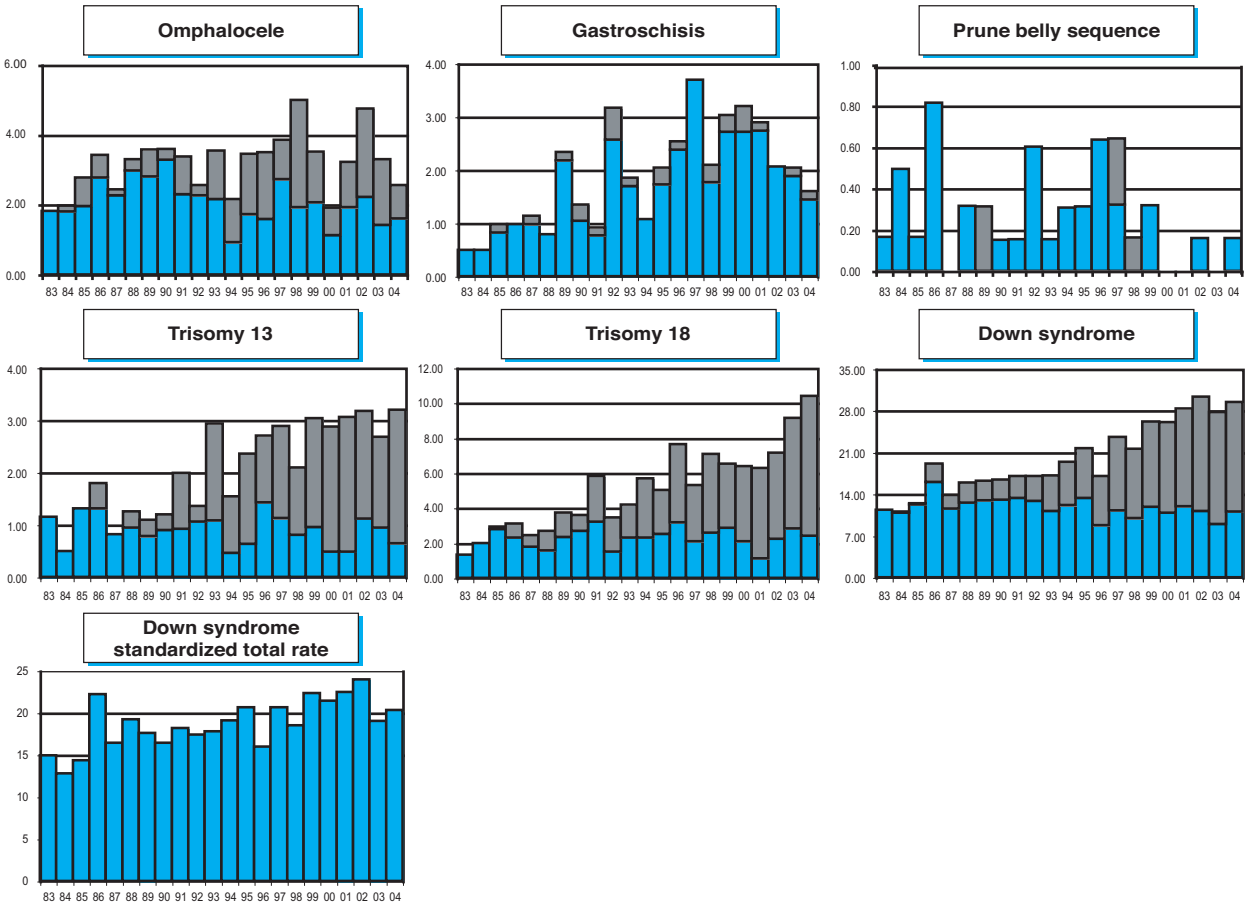
Australia: VBDR

'Time trends 1983-2004 (Birth prevalence rates per 10,000)



Note: L+S rates, ToP rates

Note: L+S rates, ToP rates



Note: L+S rates, ToP rates

Australia: WABDR
Western Australian Birth Defects Registry

History:
The Registry was established in 1980, and is currently located in a teaching obstetric hospital. The objectives of the Registry have always been to establish how often birth defects occur, to conduct research into causes and prevention of birth defects, to provide health professionals and the public with information about birth defects, and to monitor and evaluate screening, treatment and prevention programs.

Size and coverage
Population-based in the state of Western Australia. 25,000 birth a year, ~6% reported with a birth defect. Birth defects diagnosed prenatally and up to the age of 6 years, in stillbirths, terminations of pregnancy and livebirths are included.

Legislation and funding
Following a period of short term funding from both Federal and State sources, the Registry is now wholly funded by the Western Australian Department of Health. There are several statutory sources of information (birth, death and hospital data collections), and a large number of voluntary sources. Statutory notification is being considered by the Department of Health.

Sources of ascertainment
Statutory sources:
Midwives' Notification of Birth Forms (all births over 20 weeks gestation), Death Certificates (perinatal, infant and childhood); Hospital Morbidity (all hospital discharges in Western Australia).
Voluntary sources: Maternity and paediatric hospitals, Obstetricians, paediatricians, orthopaedic surgeons, Community and Child Health Nurses, Cytogenetic laboratories, Pathology services (including prenatal screening services), Ultrasound practices Genetic Services, Disability services

Exposure information
No exposure information is routinely collected

Background information
The data on the Registry are routinely linked to the linked dataset of all births, deaths and hospital admissions for Western Australia. This linkage provides information on variables such as maternal and paternal age, labour and delivery data, and maternal illnesses, for both cases of birth defects (numerators) and all births in Western Australia (denominators).
Data from the Registry are provided to the National Perinatal Statistics Unit for monitoring birth defects in Australia as a whole.

Addresses and Staff:
Clinical Professor Carol Bower
Programme Director Western Australia Birth Defects Registry (WABDR)
King Edward Memorial Hospital PO Box 134 SUBIACO 6904 Western Australia

Phone: 618 9340 2721

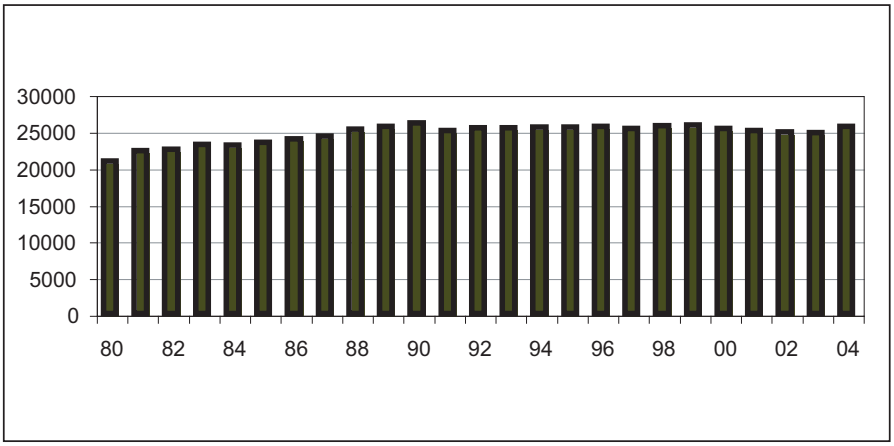
Fax: 618 9340 2636

Email: caroline.bower@health.wa.gov.au

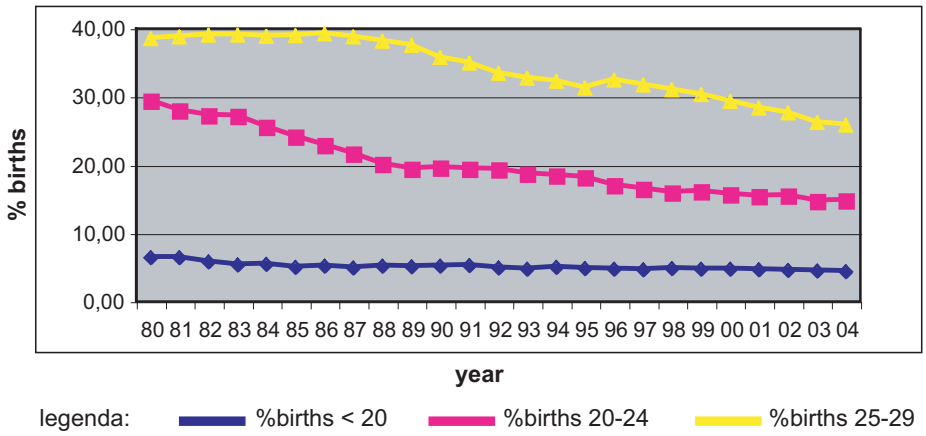
Web site:
http://www.kemh.health.wa.gov.au/services/birth_defects/index.htm

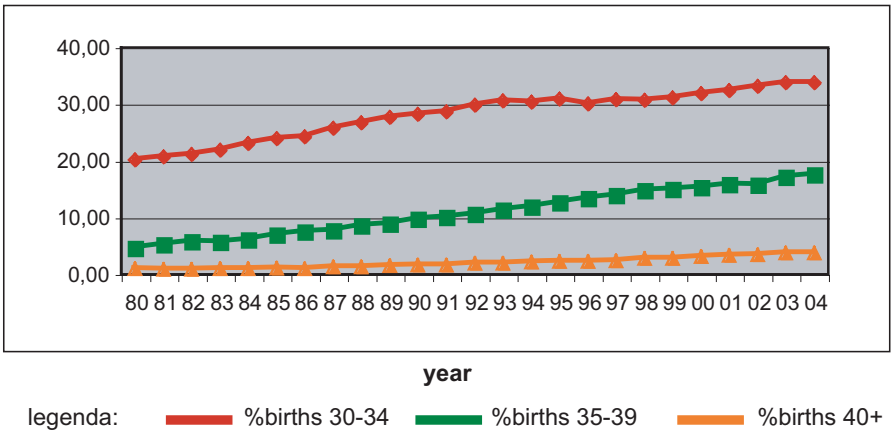
Australia: WABDR

Total births by year



Percentage of births by maternal age





Australia: WABDR, 2004

Live births (LB)	25,340
Stillbirths (SB)	188
Total births	25,528
Number of terminations of pregnancy (ToP) for birth defects	153

Birth Defects	Number of cases			Rates*10,000
	LB	SB	ToP	Total rate
Anencephaly	0	0	15	5.84
Spina bifida	10	1	15	10.12
Encephalocele	1	0	2	1.17
Microcephaly	7	0	1	3.12
Arhinencephaly / Holoprosencephaly	2	0	4	2.34
Hydrocephaly	8	1	5	5.45
Anophthalmos	1	0	1	0.78
Microphthalmos	2	0	2	1.56
Unspecified Anophthalmos/ Microphthalmos	0	0	0	---
Anotia	3	1	0	1.56
Microtia	2	1	0	1.17
Unspecified Anotia/Microtia	0	0	0	---
Transposition of great vessels	10	0	4	5.45
Tetralogy of Fallot	6	0	0	2.34
Hypoplastic left heart syndrome	1	0	4	1.95
Coarctation of aorta	10	0	6	6.23
Choanal atresia, bilateral	0	0	0	0.00
Cleft palate without cleft lip	20	1	0	8.18
Cleft lip with or without cleft palate	32	3	5	15.58
Oesophageal atresia / stenosis with or without fistula	9	1	2	4.67
Small intestine atresia / stenosis	4	1	0	1.95
Anorectal atresia / stenosis	14	0	3	6.62
Undescended testis (36 weeks of gestation or later)	58	0	0	22.58
Hypospadias	69	0	1	27.26
Epispadias	0	0	1	0.39
Indeterminate sex	0	0	1	0.39
Renal agenesis	5	1	10	6.23
Cystic kidney	15	1	6	8.57
Bladder exstrophy	0	0	1	0.39
Polydactyly, preaxial	17	0	5	8.57
Total Limb reduction defects (include unspecified)	8	1	3	4.67
Transverse	nr	nr	nr	nr
Preaxial	nr	nr	nr	nr
Postaxial	nr	nr	nr	nr
Intercalary	nr	nr	nr	nr
Mixed	nr	nr	nr	nr
Unspecified	nr	nr	nr	---
Diaphragmatic hernia	5	0	2	2.73
Omphalocele	3	2	7	4.67
Gastroschisis	7	0	0	2.73
Unspecified Omphalocele/Gastroschisis	0	0	0	---
Prune belly sequence	0	0	0	0.00
Trisomy 13	3	1	6	3.89
Trisomy 18	3	0	9	4.67
Down syndrome, all ages (include age unknown)	28	3	44	29.20
<20	0	0	0	0.00
20-24	1	0	2	7.94
25-29	6	0	5	16.68
30-34	6	1	8	17.39
35-39	6	2	21	64.49
40-44	8	0	7	163.76
45+	1	0	1	444.44
unknown	1	0	0	---

nr = not reported

Australia: WABDR, Previous years rates 1980 - 2004

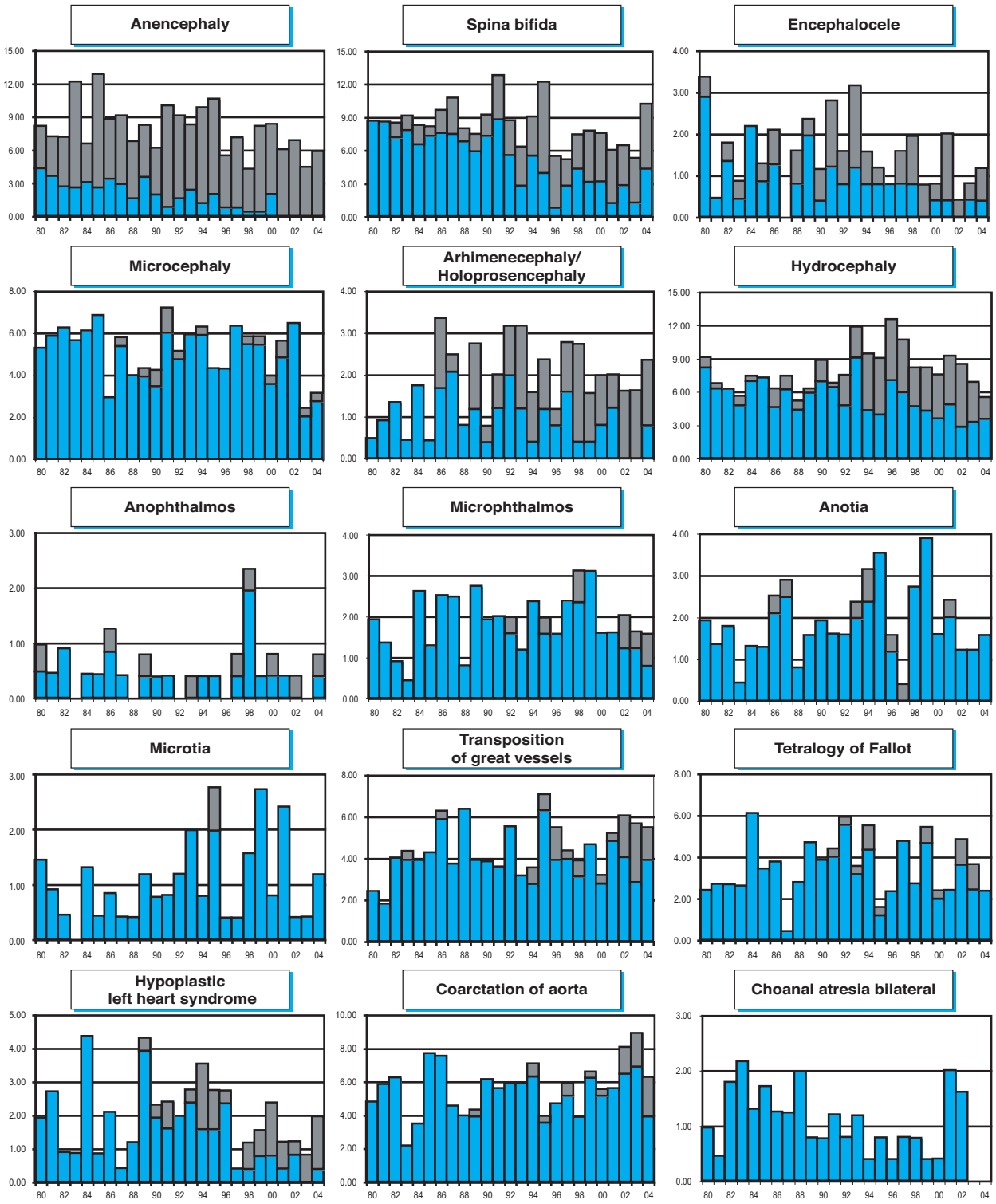
Birth prevalence rates: (LB+SB+TOP) * 10,000

	1974-79	1980-84	1985-89	1990-94	1995-99	2000-04
Births		**88,499**	**119,571**	**127,229**	**127,395**	**125,410**
Anencephaly		8.25	9.09	8.65	7.12	6.31
Spina bifida		8.61	8.76	9.20	7.59	7.11
Encephalocele		1.70	1.47	2.05	1.25	1.04
Microcephaly		5.83	4.75	5.74	5.32	4.31
Arhinencephaly / Holoprosencephaly		0.99	1.96	2.12	2.11	1.92
Hydrocephaly		7.00	6.47	8.89	9.71	7.51
Anophthalmos		0.54	0.57	0.31	0.78	0.48
Microphthalmos		1.44	1.96	1.89	2.43	1.68
Unspecified Anophthalmos / Microphthalmos		---	---	---	---	---
Anotia		1.35	1.80	2.12	2.43	1.60
Microtia		0.81	0.65	1.10	1.57	1.04
Unspecified Anotia / Microtia		---	---	---	---	---
Transposition of great vessels		3.32	4.91	3.93	5.09	5.11
Tetralogy of Fallot		3.32	3.03	4.64	3.37	3.12
Hypoplastic left heart syndrome		2.15	1.80	2.60	1.72	1.52
Coarctation of aorta		4.49	5.57	6.14	5.01	6.87
Choanal atresia, bilateral		1.35	1.39	0.87	0.63	0.80
Cleft palate without cleft lip		8.52	8.51	10.86	11.74	11.66
Cleft lip with or without cleft palate		11.93	14.08	9.99	12.37	13.42
Oesophageal atresia / stenosis with or without fistula		3.05	3.52	2.75	3.29	3.91
Small intestine atresia / stenosis		3.05	2.54	2.52	2.82	2.96
Anorectal atresia / stenosis		5.83	4.91	6.69	5.79	6.71
Undescended testis (36 weeks of gestation or later)		65.13	66.80	66.79	56.99	37.07
Hypospadias		27.09	29.88	34.93	36.09	33.39
Epispadias		0.27	0.41	0.16	0.16	0.16
Indeterminate sex		0.18	0.41	0.31	0.16	0.32
Renal agenesis		4.13	3.27	4.48	5.40	4.87
Cystic kidney		2.42	4.18	7.08	7.52	9.99
Bladder exstrophy		0.18	0.16	0.31	0.70	0.32
Polydactyly, preaxial		9.42	10.64	10.07	12.06	10.07
Total Limb reduction defects (include unspecified)		3.77	4.75	6.06	7.91	6.47
Transverse		nr	nr	nr	nr	nr
Preaxial		nr	nr	nr	nr	nr
Postaxial		nr	nr	nr	nr	nr
Intercalary		nr	nr	nr	nr	nr
Mixed		nr	nr	nr	nr	nr
Unspecified		---	---	---	---	---
Diaphragmatic hernia		3.23	2.70	2.83	4.31	2.80
Omphalocele		1.79	3.11	3.70	3.21	4.79
Gastroschisis		1.35	1.80	2.52	4.15	3.52
Unspecified Omphalocele / Gastroschisis		---	---	---	---	---
Prune belly sequence		0.54	0.74	0.55	0.31	0.00
Trisomy 13		0.72	1.15	1.42	1.72	3.20
Trisomy 18		1.70	1.72	3.85	5.17	7.35
Down syndrome, all ages (include age unknown)		11.57	15.23	17.23	19.65	26.28
<20		4.59	6.47	6.29	6.61	14.10
20-24		5.23	5.69	7.83	6.55	6.83
25-29		9.01	7.87	8.38	9.24	13.39
30-34		11.72	14.35	18.93	16.07	19.37
35-39		47.15	47.27	38.06	43.67	51.37
40-44		104.82	257.51	146.54	173.83	154.22
45+		615.38	461.54	512.82	136.99	631.07
unknown		---	---	---	---	---

nr= not reported

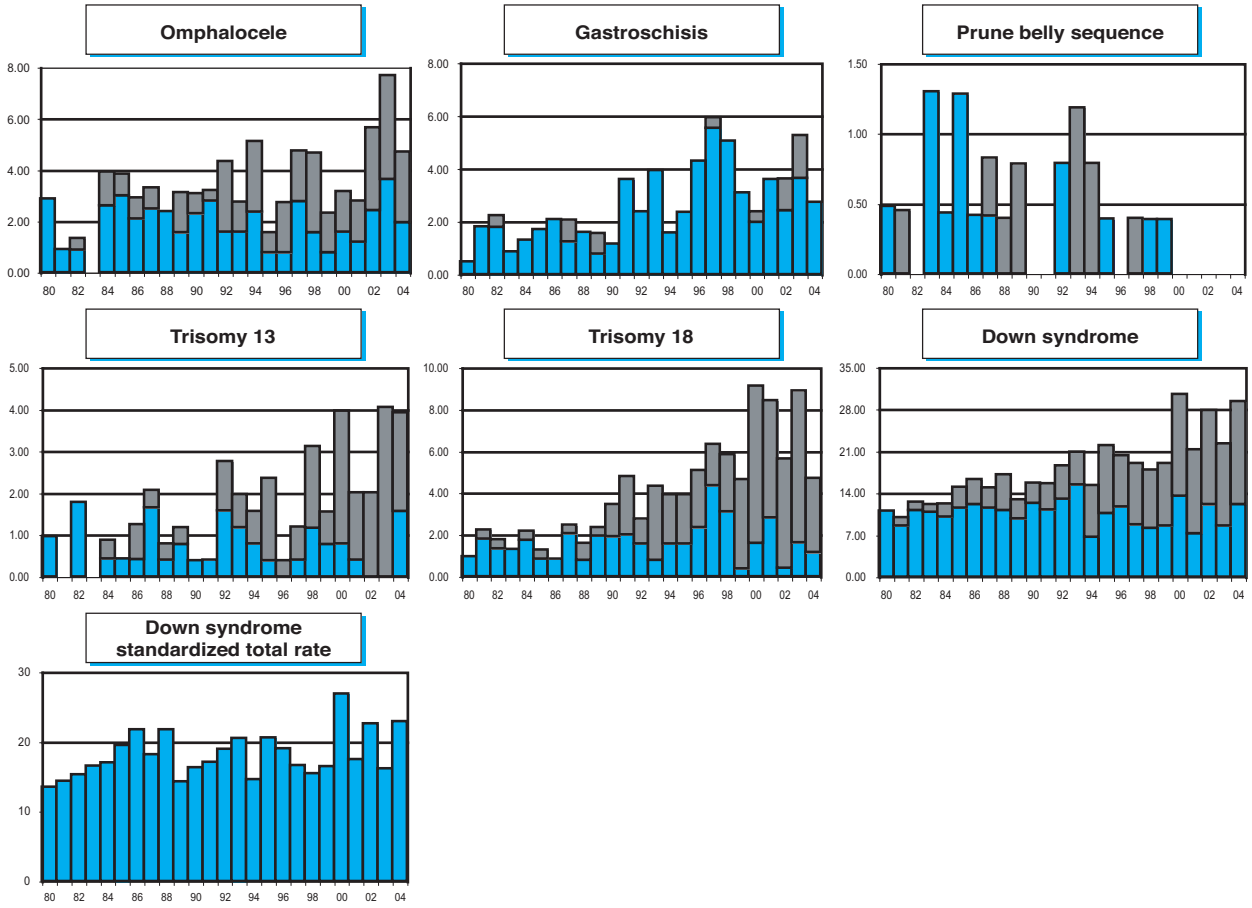
Australia: WABDR

'Time trends 1980-2004 (Birth prevalence rates per 10,000)



Note: L+S rates, ToP rates

Note: L+S rates, ToP rates



Note: L+S rates, ToP rates

Canada: Alberta

Alberta Congenital Anomalies Surveillance System

History:
The Programme began in 1966 as a general Registry for Handicapped Children. This was disbanded in 1980 and continued as a surveillance Programme for live and stillborn infants with congenital anomalies who were born in the Province of Alberta.

Size and coverage:
All live and stillbirths in the province are covered which at present comprises about 40,000 births per year. The definition of stillbirth is 20 weeks or more or 500 grams or more. The vast majority of births occur in hospital (approximately 97%). In 1997 a special fetal congenital anomalies surveillance system was started to include those fetuses with congenital anomalies who were either spontaneously lost prior to 20 weeks or where there was termination as a result of prenatal diagnosis.

Legislation and funding:
Reporting is voluntary. The system is run by members of the Department of Medical Genetics, Alberta Children's Hospital/University of Calgary reporting to Alberta Vital Statistics and Alberta Health. Funding is from Alberta Ministry of Health.

Sources of ascertainment:
Reports are obtained from physician's notice of birth, live birth and stillbirth registrations, death registrations and a special congenital anomalies reporting form (CARF) from hospitals. This is based on discharge diagnosis, including readmissions for any reason up to one year of age. Additional sources are speciality clinics, such as medical genetics and cytogenetics laboratories.

Exposure information:
None is routine.

Background information:
Linkage studies are possible with other statistical data from Alberta Health.

Addresses and Staff:
R. Brian Lowry, MD, Programme Director
ACASS / Clinical Genetics
Alberta Children's Hospital
2888, Shaganappi Trail NW,
Calgary, AB, Canada. T3B 6A8

Phone: 403-955-7370

Fax: 403-955-2870

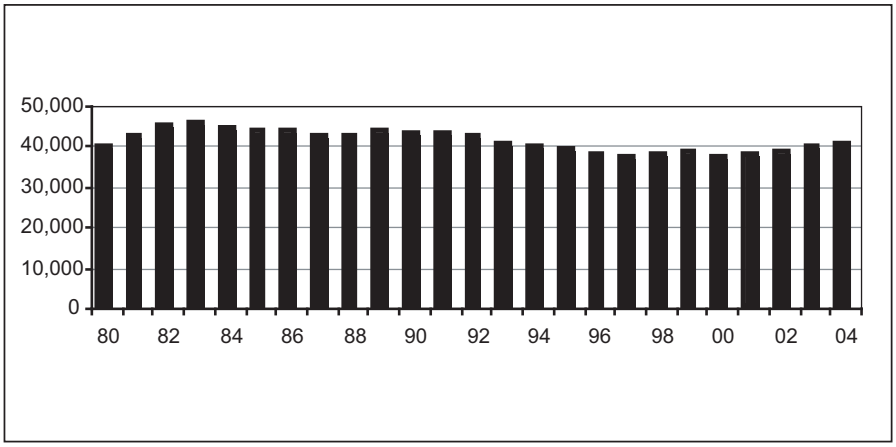
E-mail: brian.lowry@calgaryhealthregion.ca

Barbara Sibbald - Manager

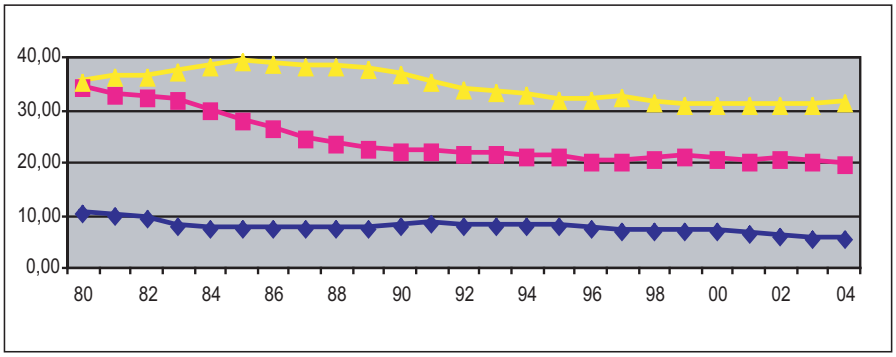
E-mail: barbara.sibbald@calgaryhealtregion.ca

Canada: Alberta

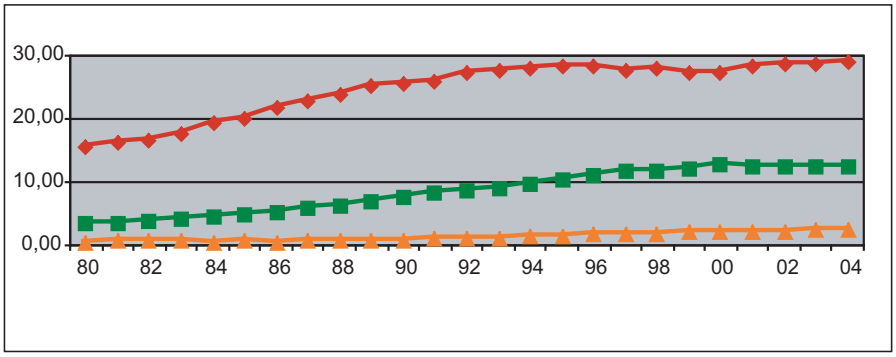
Total births by year



Percentage of births by maternal age



legenda: %births < 20 %births 20-24 %births 25-29



legenda: %births 30-34 %births 35-39 %births 40+

Canada: Alberta, 2004

Live births (LB)	40,267
Stillbirths (SB)	285
Total births	40,552
Number of terminations of pregnancy (ToP) for birth defects	83

Birth Defects	Number of cases			Rates*10,000
	LB	SB	ToP	Total rate
Anencephaly	2	1	7	2.46
Spina bifida	9	4	2	3.69
Encephalocele	2	1	0	0.74
Microcephaly	7	3	0	2.46
Arhinencephaly / Holoprosencephaly	3	2	1	1.48
Hydrocephaly	15	7	0	5.41
Anophthalmos	0	0	0	0.00
Microphthalmos	6	2	0	1.97
Unspecified Anophthalmos/ Microphthalmos	0	0	0	---
Anotia	1	0	0	0.25
Microtia	11	1	0	2.95
Unspecified Anotia/Microtia	0	0	0	---
Transposition of great vessels	19	1	1	5.17
Tetralogy of Fallot	8	0	0	1.97
Hypoplastic left heart syndrome	6	2	0	1.97
Coarctation of aorta	6	0	0	1.48
Choanal atresia, bilateral	5	0	0	1.23
Cleft palate without cleft lip	33	1	1	8.61
Cleft lip with or without cleft palate	42	4	3	12.06
Oesophageal atresia / stenosis with or without fistula	11	0	0	2.71
Small intestine atresia / stenosis	10	3	0	3.20
Anorectal atresia / stenosis	15	7	6	6.89
Undescended testis (36 weeks of gestation or later)	111	0	0	27.32
Hypospadias	91	1	0	22.64
Epispadias	2	0	0	0.49
Indeterminate sex	8	1	0	2.21
Renal agenesis	16	4	3	5.66
Cystic kidney	21	5	4	7.38
Bladder exstrophy	0	0	0	0.00
Polydactyly, preaxial	82	3	3	21.66
Total Limb reduction defects (include unspecified)	23	13	9	11.07
Transverse	nr	nr	nr	nr
Preaxial	nr	nr	nr	nr
Postaxial	nr	nr	nr	nr
Intercalary	nr	nr	nr	nr
Mixed	nr	nr	nr	nr
Unspecified	nr	nr	nr	---
Diaphragmatic hernia	5	0	2	1.72
Omphalocele	2	6	0	1.97
Gastroschisis	12	1	0	3.20
Unspecified Omphalocele/Gastroschisis	nr	nr	nr	---
Prune belly sequence	2	1	0	0.74
Trisomy 13	4	3	3	2.46
Trisomy 18	7	5	6	4.43
Down syndrome, all ages (include age unknown)	56	11	14	19.93
<20	1	0	0	4.73
20-24	5	2	0	8.85
25-29	7	2	1	7.84
30-34	11	3	4	15.29
35-39	26	3	5	68.03
40-44	6	1	4	114.11
45+	0	0	0	0.00
unknown	0	0	0	---

Canada: Alberta, Previous years rates 1980 - 2004

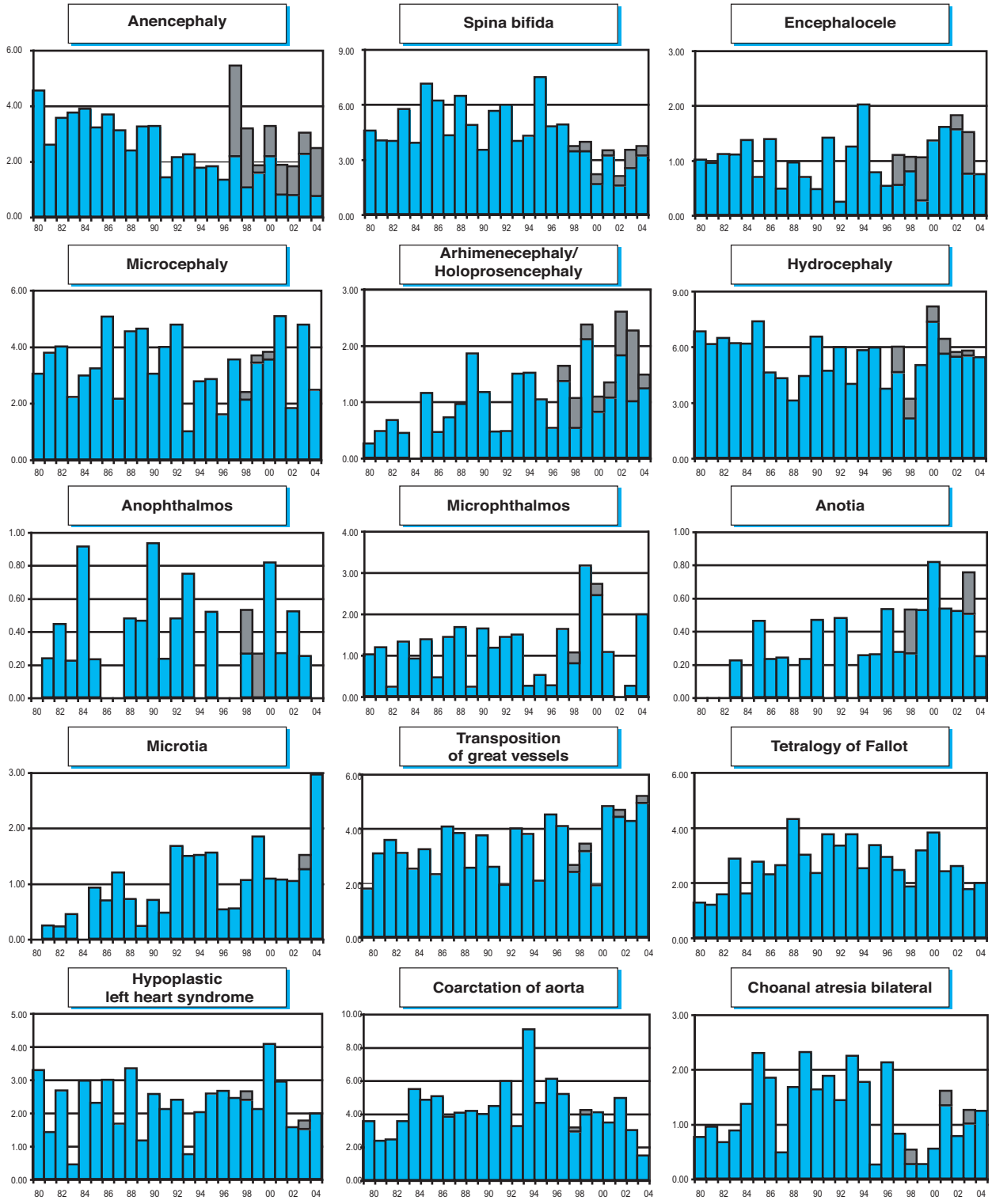
Birth prevalence rates: (LB+SB) * 10,000 until 1996
Birth prevalence rates: (LB+SB+TOP) * 10,000 since 1997

	1974-79	1980-84	1985-89	1990-94	1995-99	2000-04
Births		**216,350**	**214,267**	**207,397**	**189,126**	**193,241**
Anencephaly		3.65	3.13	2.17	2.70	2.48
Spina bifida		4.44	5.79	4.68	4.97	3.00
Encephalocele		1.11	0.84	1.06	0.90	1.35
Microcephaly		3.19	3.92	3.13	2.80	3.52
Arhinencephaly / Holoprosencephaly		0.37	1.03	1.01	1.32	1.66
Hydrocephaly		6.33	4.76	5.40	4.76	6.21
Anophthalmos		0.37	0.23	0.48	0.26	0.36
Microphthalmos		0.92	1.03	1.21	1.32	1.19
Unspecified Anophthalmos / Microphthalmos		---	---	---	---	---
Anotia		0.05	0.23	0.24	0.42	0.57
Microtia		0.18	0.75	1.16	1.11	1.45
Unspecified Anotia / Microtia		---	---	---	---	---
Transposition of great vessels		2.82	3.17	3.18	3.33	4.19
Tetralogy of Fallot		1.71	2.99	3.13	2.75	2.54
Hypoplastic left heart syndrome		2.13	2.29	1.98	2.49	2.43
Coarctation of aorta		3.47	4.39	5.30	4.65	3.36
Choanal atresia, bilateral		0.92	1.73	1.78	0.79	0.98
Cleft palate without cleft lip		6.42	7.05	8.05	8.57	8.12
Cleft lip with or without cleft palate		10.21	11.99	11.52	12.11	12.42
Oesophageal atresia / stenosis with or without fistula		2.77	3.17	2.17	2.64	1.97
Small intestine atresia / stenosis		0.79	1.03	1.40	1.80	1.97
Anorectal atresia / stenosis		3.24	5.18	5.11	5.02	6.93
Undescended testis (36 weeks of gestation or later)		26.44	28.24	28.50	22.15	25.82
Hypospadias		17.15	24.27	24.30	17.55	20.80
Epispadias		0.55	0.19	0.53	0.32	0.67
Indeterminate sex		0.37	0.65	1.16	0.85	1.40
Renal agenesis		3.10	4.62	5.45	4.39	6.16
Cystic kidney		2.31	3.78	4.82	5.29	7.81
Bladder exstrophy		0.37	0.19	0.39	0.16	0.41
Polydactyly, preaxial		9.80	13.95	16.49	11.84	16.30
Total Limb reduction defects (include unspecified)		6.33	9.01	10.51	9.89	11.44
Transverse		nr	nr	nr	nr	nr
Preaxial		nr	nr	nr	nr	nr
Postaxial		nr	nr	nr	nr	nr
Intercalary		nr	nr	nr	nr	nr
Mixed		nr	nr	nr	nr	nr
Unspecified		---	---	---	---	---
Diaphragmatic hernia		3.37	3.31	2.60	2.75	3.98
Omphalocele		1.62	2.29	1.88	2.22	2.43
Gastroschisis		1.34	1.49	1.69	2.49	3.21
Unspecified Omphalocele / Gastroschisis		---	---	---	---	---
Prune belly sequence		0.51	0.28	0.14	0.42	0.36
Trisomy 13		0.83	0.65	1.21	1.53	1.81
Trisomy 18		1.66	1.68	2.03	3.81	4.35
Down syndrome, all ages (include age unknown)		8.92	9.80	11.19	14.54	19.35
<20		nr	5.63*	4.33	7.34	7.99
20-24		nr	4.38*	8.05	4.64	5.92
25-29		nr	6.46*	7.61	6.89	10.70
30-34		nr	12.26*	13.33	13.52	16.23
35-39		nr	34.14*	22.44	41.48	52.52
40-44		nr	107.05*	86.25	135.09	144.29
45+		nr	0.00*	405.41	400.00	62.11
unknown		---	---	---	---	---

nr= not reported

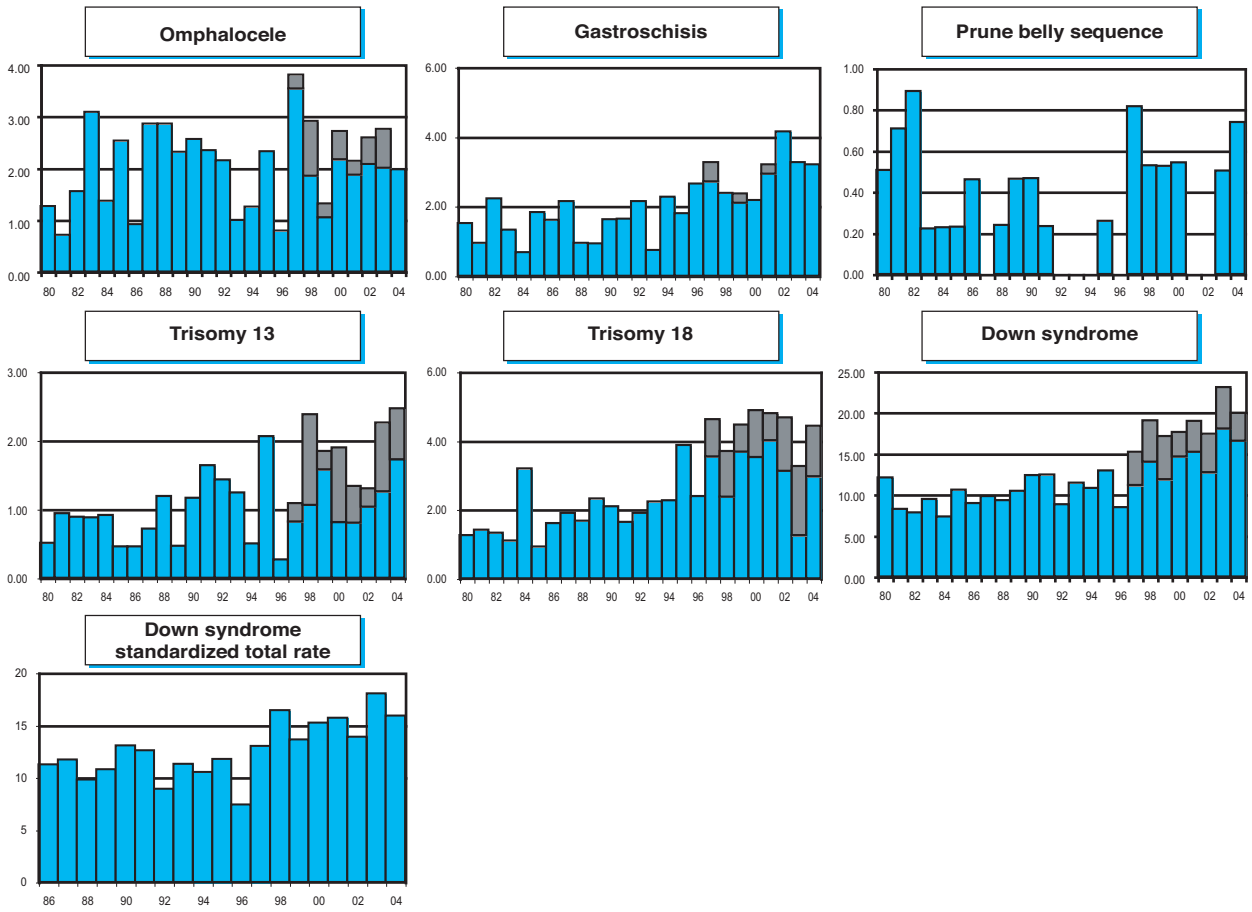
Canada: Alberta

'Time trends 1980-2004 (Birth prevalence rates per 10,000)



Note: L+S rates, ToP rates

Note: L+S rates, ToP rates



Note: L+S rates, ToP rates

Canada: British Columbia

British Columbia Health Status Registry (BCHSR)
Congenital Anomalies Surveillance Programme

History:
The Programme was established in 1952 as the Crippled Children's Registry. Until 1959 the Programme had an age limit of 21, but this was removed in 1960 and the name was change to the Registry for Handicapped Children and Adults and included all familial conditions and congenital malformations. In 1975, the Registry's name was changed to the Health Surveillance Registry as risk registers for amniocentesis, rubella, hyaline membrane disease, and fetal alcohol syndrome were added. In 1991, the Royal Commission Report on Health Care and Costs contained a recommendation that Vital Statistics should develop and maintain a registry of individuals with disabilities to assist in the development of long-range plans and to monitor the changing needs of the population. Subsequently, in September 1992, amendments to the Health Act established the legislative mandate and responsibilities for the HSR. The Registry's current name, Health Status Registry, was acquired in 1992. In order to refocus the Registry's emphasis on children, the criteria for registration of individuals with long-term physical, mental and/or emotional problems was restricted to persons under the age of 20 years old, however registration of persons with genetic conditions was not age limited. By 2000 there were approximately 215,000 records in the Registry.

Size and coverage
The registry covers all births in the province approximately 45,000 births annually including stillbirths with at least 20 weeks gestation or birth weight 500 grams or more.

Legislation and funding
In 1992, amendments to the Health Act established the legislative mandate and responsibilities for the BC HSR. Funding comes from the British Columbia Vital Statistics Agency.

Sources of ascertainment
Sources include: Notice of Live and Stillbirth, Death registrations, Hospital Admission/Discharge Abstracts, Children's Hospital, Sunnyhill Hospital, UBC and Victoria General Medical Genetics Clinics, Child Development Centres, Health Regions, the Asante Centre for Fetal Alcohol Syndrome.

Exposure information
Information on complications of pregnancy, labour or delivery is available on Vital Statistics birth registrations and environmental/occupational and drug/alcohol/smoking lifestyle related information can be obtained from the death registrations for the deceased.

Background information
The registry data are regularly matched to Vital Statistics birth registrations to obtain birth particulars of the registrants and maternal/paternal information, and also matched to death registrations to get the date of death and causes of death if the registered person was deceased. The registry is also working on the collection of the medically terminated pregnancies due to congenital anomalies.

Addresses and Staff:
Ron Danderfer, Programme Director
Manager, BC Vital Statistics Agency
818 Fort Street, Victoria,
British Columbia, CANADA, V8W 1H8

Phone: 1-250-952-2567

Fax: 1-250-952-2587

E-mail: ron.danderfer@gems1.gov.bc.ca

Shelly Wells - Medical Advisor
Terry McMahon - Medical Classification Officer
Debbie Hall - Medical Coding Clerk

Soo-Hong Uh, Consultant, BC Health Status Registry, KMT Division, BC Ministry of Health, 7-1, 1515 Blanshard Street, Victoria, BC, V8W3C8, Canada

Phone: 1-250-217-1346

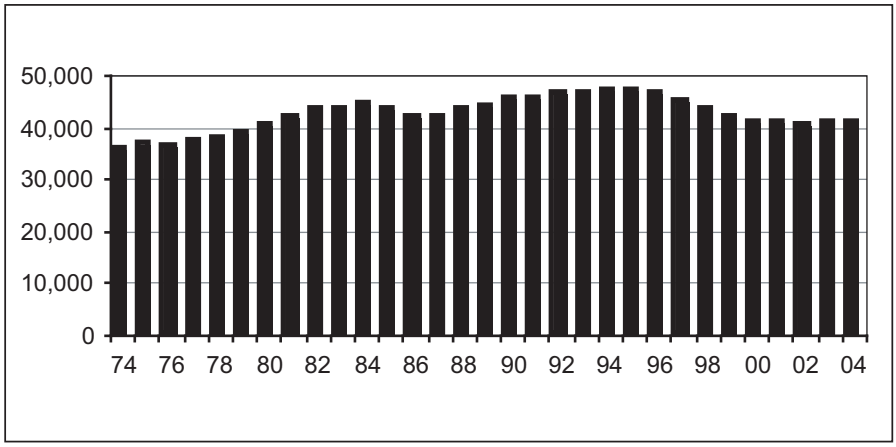
Fax: 1-250-952-1827

E-mail: soohong.uh@gov.bc.ca

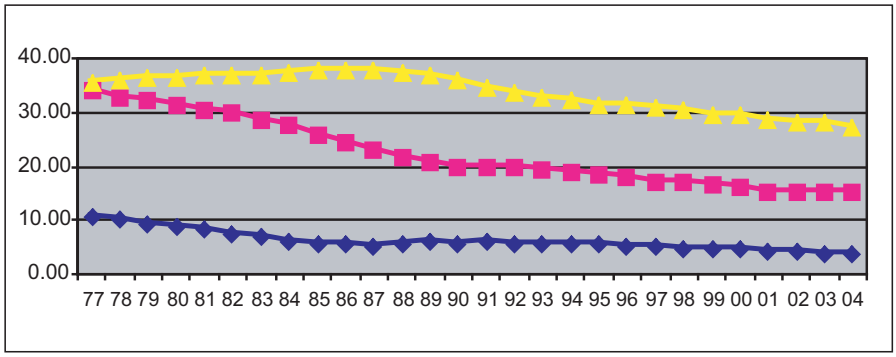
Web site:
http://www.vs.gov.bc.ca/stats/hsr/index.html

Canada: British Columbia

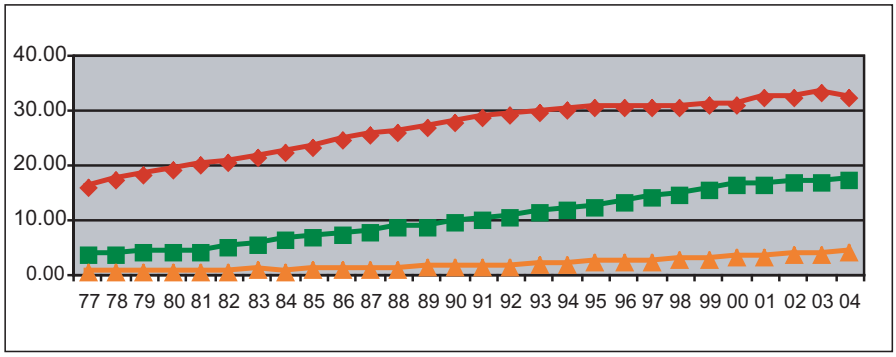
Total births by year



Percentage of births by maternal age



legenda: %births < 20 %births 20-24 %births 25-29



legenda: %births 30-34 %births 35-39 %births 40+

Canada: British Columbia, 2004

Live births (LB)	40,598
Stillbirths (SB)	280
Total births	40,878
Number of terminations of pregnancy (ToP) for birth defects	nr

Birth Defects	Number of cases			Rates*10,000
	LB	SB	ToP	Total rate
Anencephaly	4	6	nr	2.45
Spina bifida	10	4	nr	3.42
Encephalocele	1	0	nr	0.24
Microcephaly	14	1	nr	3.67
Arhinencephaly / Holoprosencephaly	27	9	nr	8.81
Hydrocephaly	7	2	nr	2.20
Anophthalmos	2	0	nr	0.49
Microphthalmos	2	0	nr	0.49
Unspecified Anophthalmos/ Microphthalmos	0	0	nr	---
Anotia	1	0	nr	0.24
Microtia	1	0	nr	0.24
Unspecified Anotia/Microtia	3	0	nr	---
Transposition of great vessels	9	0	nr	2.20
Tetralogy of Fallot	16	1	nr	4.16
Hypoplastic left heart syndrome	6	2	nr	1.96
Coarctation of aorta	19	1	nr	4.89
Choanal atresia, bilateral	4	0	nr	0.98
Cleft palate without cleft lip	22	1	nr	5.63
Cleft lip with or without cleft palate	40	4	nr	10.76
Oesophageal atresia / stenosis with or without fistula	10	0	nr	2.45
Small intestine atresia / stenosis	22	0	nr	5.38
Anorectal atresia / stenosis	18	0	nr	4.40
Undescended testis (36 weeks of gestation or later)	37	0	nr	9.05
Hypospadias	54	0	nr	13.21
Epispadias	4	0	nr	0.98
Indeterminate sex	0	0	nr	0.00
Renal agenesis	0	1	nr	0.24
Cystic kidney	1	0	nr	0.24
Bladder exstrophy	4	0	nr	0.98
Polydactyly, preaxial	25	2	nr	6.61
Total Limb reduction defects (include unspecified)	10	1	nr	2.69
Transverse	nr	nr	nr	nr
Preaxial	nr	nr	nr	nr
Postaxial	nr	nr	nr	nr
Intercalary	nr	nr	nr	nr
Mixed	nr	nr	nr	nr
Unspecified	nr	nr	nr	---
Diaphragmatic hernia	13	2	nr	3.67
Omphalocele	8	2	nr	2.45
Gastroschisis	22	4	nr	6.36
Unspecified Omphalocele/Gastroschisis	1	1	nr	---
Prune belly sequence	2	0	nr	0.49
Trisomy 13	4	2	nr	1.47
Trisomy 18	3	9	nr	2.94
Down syndrome, all ages (include age unknown)	42	16	nr	14.19
<20	1	0	nr	6.95
20-24	3	1	nr	6.50
25-29	5	0	nr	4.51
30-34	12	7	nr	14.39
35-39	15	5	nr	28.09
40-44	5	3	nr	53.26
45+	1	0	nr	116.28
unknown	0	0	nr	---

nr = not reported

Canada: British Columbia, Previous years rates 1974- 2004

Birth prevalence rates: (LB+SB) * 10,000

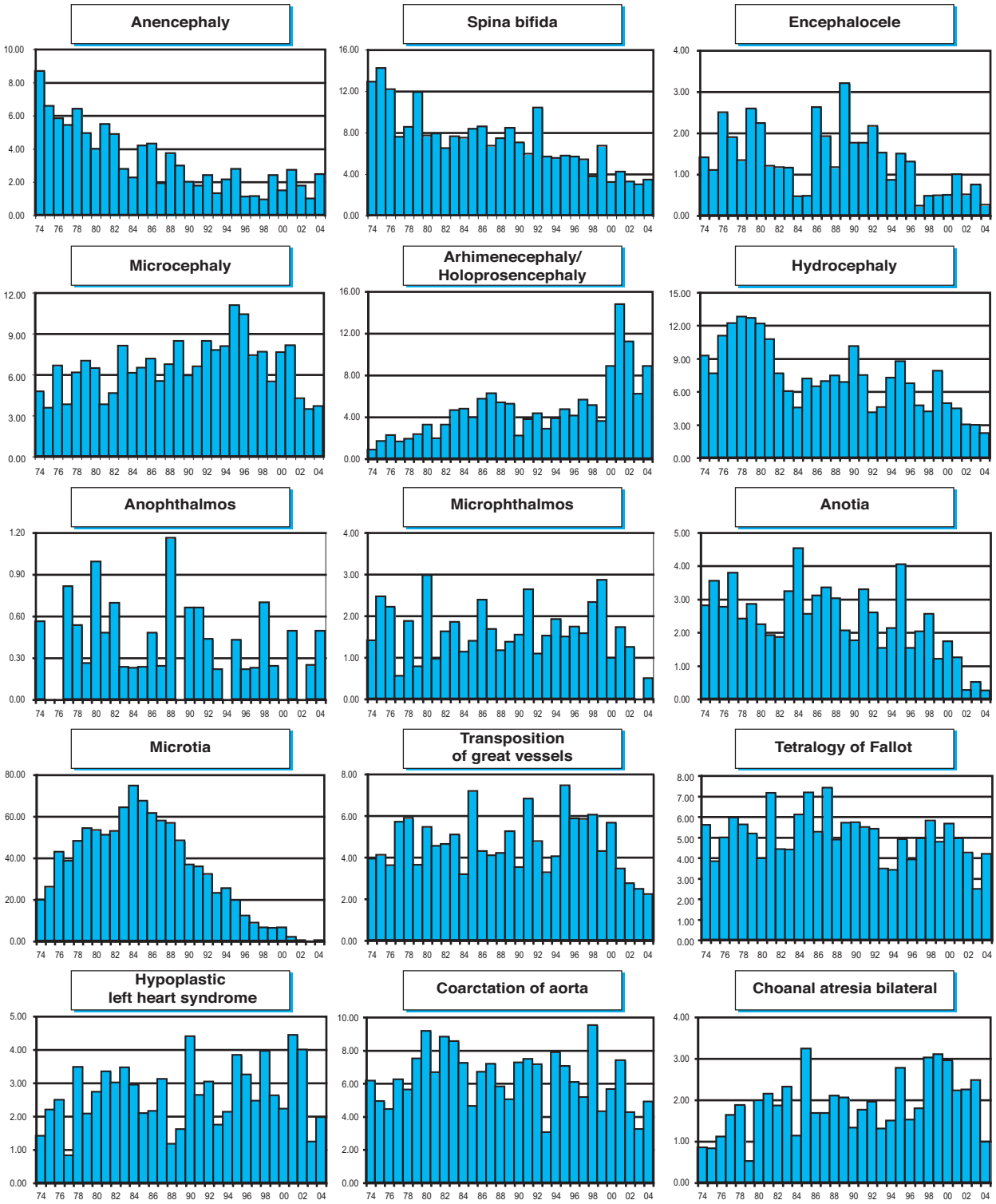
	1974-79	1980-84	1985-89	1990-94	1995-99	2000-04
Total births	**222,046**	**252,174**	**258,649**	**274,923**	**270,353**	**245,134**
Anencephaly	6.26	3.84	3.41	1.90	1.66	1.87
Spina bifida	11.17	7.40	7.89	6.88	5.42	3.40
Encephalocele	1.80	1.22	1.87	1.60	0.81	0.59
Microcephaly	5.31	5.81	6.86	7.36	8.47	5.42
Arhinencephaly / Holoprosencephaly	1.76	3.56	5.27	3.38	4.61	9.95
Hydrocephaly	10.94	8.11	6.95	6.67	6.45	3.50
Anophthalmos	0.36	0.52	0.42	0.39	0.36	0.25
Microphthalmos	1.53	1.69	1.59	1.73	1.97	0.89
Unspecified Anophthalmos / Microphthalmos	---	---	---	---	---	---
Anotia	3.02	2.76	2.80	2.25	2.28	0.79
Microtia	38.42	59.32	58.19	30.43	10.75	1.72
Unspecified Anotia / Microtia	---	---	---	---	---	---
Transposition of great vessels	4.46	4.54	4.99	4.46	5.91	3.30
Tetralogy of Fallot	5.18	5.20	6.07	4.68	4.84	4.28
Hypoplastic left heart syndrome	2.07	3.09	2.01	2.77	3.23	2.76
Coarctation of aorta	5.81	8.06	5.83	6.54	6.41	5.07
Choanal atresia, bilateral	1.13	1.87	2.15	1.56	2.42	2.17
Cleft palate without cleft lip	10.36	11.85	12.04	13.59	11.47	7.98
Cleft lip with or without cleft palate	14.01	15.88	13.67	15.11	12.95	10.14
Oesophageal atresia / stenosis with or without fistula	3.06	4.26	3.13	3.38	3.40	2.91
Small intestine atresia / stenosis	2.21	3.42	3.59	3.16	4.52	4.09
Anorectal atresia / stenosis	5.00	4.54	4.53	5.50	4.84	5.17
Undescended testis (36 weeks of gestation or later)	73.41	74.40	70.69	68.96	55.64	30.58
Hypospadias	27.65	32.99	32.52	37.53	35.44	20.78
Epispadias	0.00	0.05	0.00	0.00	0.00	0.49
Indeterminate sex	1.04	1.59	0.70	1.17	1.43	0.10
Renal agenesis	5.00	6.14	6.81	7.06	5.91	1.67
Cystic kidney	3.56	4.40	5.55	6.19	6.99	1.82
Bladder exstrophy	0.36	0.47	0.61	0.39	0.40	0.54
Polydactyly, preaxial	23.10	21.60	21.18	22.64	21.59	11.96
Total Limb reduction defects (include unspecified)	10.40	8.15	7.98	6.41	6.72	3.20
Transverse	nr	nr	nr	nr	nr	nr
Preaxial	nr	nr	nr	nr	nr	nr
Postaxial	nr	nr	nr	nr	nr	nr
Intercalary	nr	nr	nr	nr	nr	nr
Mixed	nr	nr	nr	nr	nr	nr
Unspecified	---	---	---	---	---	---
Diaphragmatic hernia	4.41	4.50	3.31	3.85	4.75	3.05
Omphalocele	0.00	0.05	0.00	0.04	0.13	2.17
Gastroschisis	0.00	0.00	0.05	0.04	0.18	4.68
Unspecified Omphalocele / Gastroschisis	---	---	---	---	---	---
Prune belly sequence	0.00	0.00	0.05	0.00	0.04	0.20
Trisomy 13	0.68	0.56	1.26	1.08	1.34	1.43
Trisomy 18	1.40	1.97	2.01	2.12	3.58	3.79
Down syndrome, all ages (include age unknown)	13.02	13.59	14.19	15.80	17.16	17.28
<20	3.54*	8.82	8.48	11.73	10.83	6.16
20-24	6.19*	5.41	7.08	9.27	7.93	7.72
25-29	7.87*	8.01	5.99	7.18	11.24	9.08
30-34	20.11*	12.04	14.85	15.26	14.37	13.50
35-39	38.08*	32.54	17.73	23.45	26.85	32.03
40-44	136.52*	88.78	76.65	62.68	72.14	66.53
45+	232.56*	196.08	131.58	307.69	494.30	93.17
unknown	---	---	---	---	---	---

* data include less than 5 years

nr = not reported

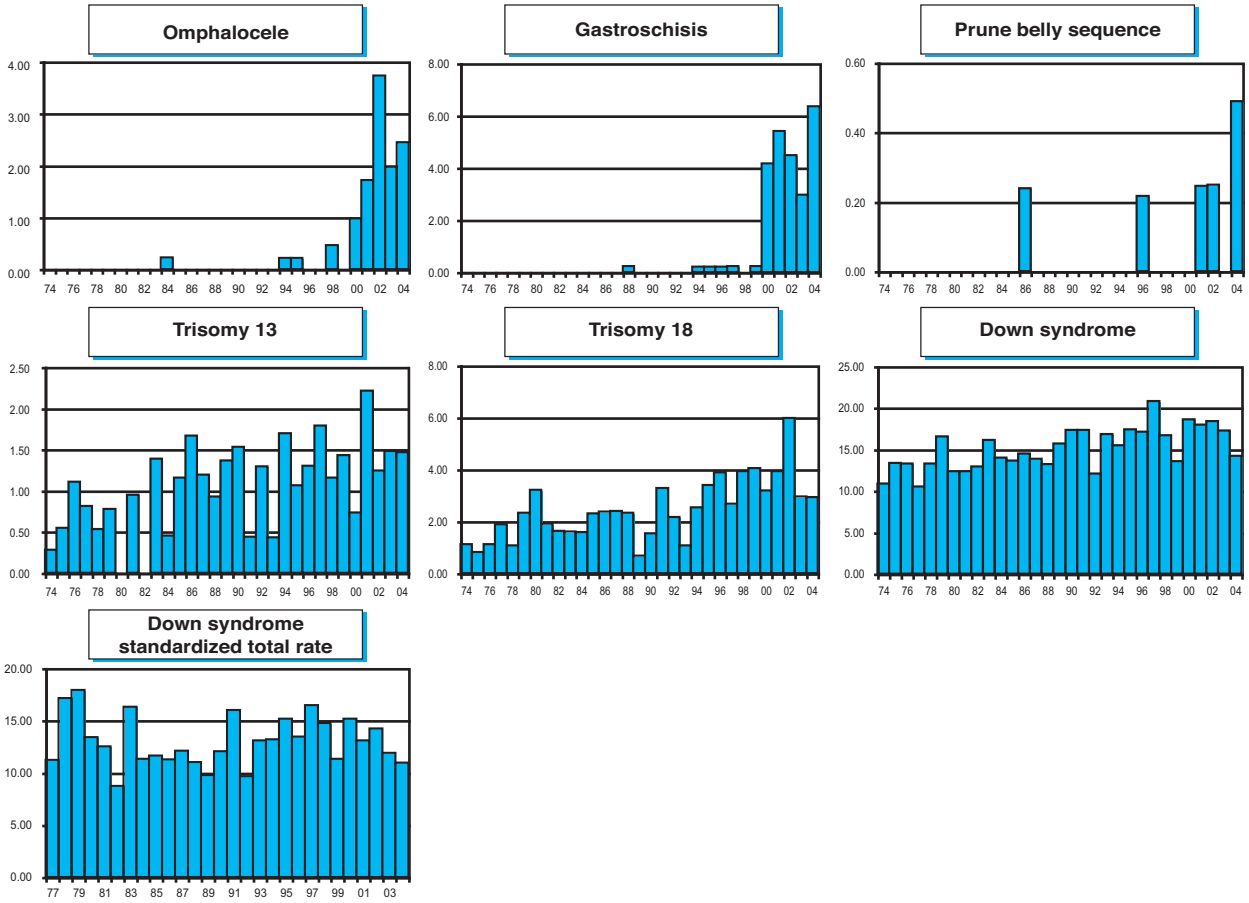
Canada: British Columbia

Time trends 1974-2004 (Birth prevalence rates per 10,000)



Note: L+S rates

Note: L+S rates



Note: L+S rates

Canada: National

Canadian Congenital Anomalies Surveillance Network (CCASN)

History:
The Programme was started in 1966. The Programme was a full member until 1987, when it became an associate member. The Programme was discontinued as an associate member of the ICBDSR in the early 1990s, and reinstated its associate member status in 1996.

Size and coverage
This system presently monitors about 330,000 births annually, which captures virtually all births in the 10 provinces and 3 territories of Canada. Data from Nova Scotia have not been included in the national statistics provided to the ICBDSR, however, efforts are being made to include this province in future submissions. Live births to 1 year of age and registered stillbirths (a birth weight of greater or equal to 500 grams, or greater than or equal to 20 weeks in pregnancy) were captured until 2000. Since 2001, all data provided by Canadian Institute for Health Information (CIHI) only include a 30 days followup period.

Legislation and funding
Reporting is based on an agreement between the Canadian Institute for Health Information (CIHI), a non-profit organization, which collects and disseminates data on hospital admission/separation in Canada, and the central registry, which is run and funded by the Public Health Agency of Canada. The Alberta Congenital Anomalies Surveillance System and Med-Echo (Système de maintenance et d'exploitation des données pour l'étude de la clientèle hospitalière) for the province of Québec provide their data separately.

Sources of ascertainment
Cases from most provinces and territories are ascertained from hospital admission/separation summary records collected by the Canadian Institute for Health Information (CIHI) and Med-Echo. The Alberta Congenital Anomalies Surveillance System provides its own separate provincial data. All data sources had a one year follow-up period until 2000. Since 2001, all data provided by Canadian Institute for Health Information (CIHI) only include a 30 days followup period.

Exposure information
No exposure information is routinely collected in the central registry.

Background information
Background information is based on hospital admission/separation summary records from the Canadian Institute for Health Information (CIHI) and Med-Echo. Alberta Congenital Anomalies Surveillance provides its own background information. Interpretation of trends should be done cautiously, since 2001 an increasing percentage of records are being coded using ICD-10 CA and may cause discrepancies from previously used ICD-9 coding. Also, as mentioned previously the variation in the follow-up period is another factor which may alter reporting of trends.

Addresses and Staff:
Hajnal Molnar-Szakacs, MD, MSc
Chief, Maternal and Infant Health
Public Health Agency of Canada
10th Floor, Jeanne Mance Bldg, A.L. 1910C,
Tunney's Pasture
Ottawa, Ontario Canada K1A OK9

Phone: 1-613-952-9855

Fax: 1-613-941-9927

E-mail: hajnal_molnar-szakacs@phac-aspc.gc.ca

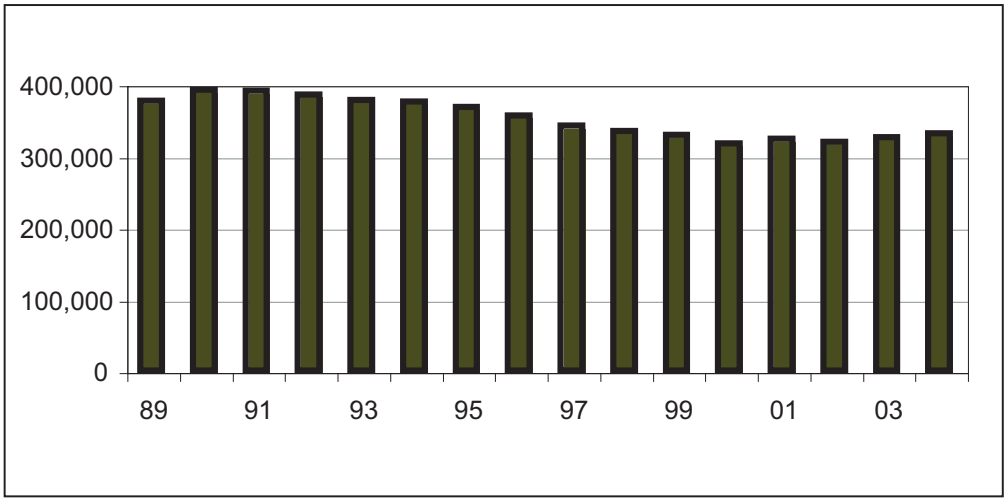
Jocelyn Rouleau, Senior Research Assistant

Phone: 1-613-954-4316

E-mail: jocelyn_rouleau@phac-aspc.gc.ca

Canada: National

Total births by year



Canada: National, 2004

Live births (LB)	328,972
Stillbirths (SB)	2,198
Total births	331,170
Number of terminations of pregnancy (ToP) for birth defects	nr

Birth Defects	Number of cases			Rates*10,000
	LB	SB	ToP	Total rate
Anencephaly	14	17	nr	0.94
Spina bifida	75	11	nr	2.60
Encephalocele	10	1	nr	0.33
Microcephaly	127	2	nr	3.90
Arhinencephaly / Holoprosencephaly	nr	nr	nr	nr
Hydrocephaly	128	16	nr	4.35
Anophthalmos	36	0	nr	1.09
Microphthalmos	6	0	nr	0.18
Unspecified Anophthalmos / Microphthalmos	0	0	nr	0.00
Anotia nr	nr	nr	nr	
Microtia	nr	nr	nr	nr
Unspecified Anotia / Microtia	nr	nr	nr	nr
Transposition of great vessels	159	1	nr	4.83
Tetralogy of Fallot	96	7	nr	3.11
Hypoplastic left heart syndrome	69	9	nr	2.36
Coarctation of aorta	150	0	nr	4.53
Choanal atresia, bilateral	90	0	nr	2.72
Cleft palate without cleft lip	210	2	nr	6.40
Cleft lip with or without cleft palate	308	11	nr	9.63
Oesophageal atresia / stenosis with or without fistula	103	1	nr	3.14
Small intestine atresia / stenosis	120	0	nr	3.62
Anorectal atresia / stenosis	134	0	nr	4.05
Undescended testis (36 weeks of gestation or later)	1227	0	nr	37.05
Hypospadias**	954	0	nr	28.81
Epispadias	nr	nr	nr	nr
Indeterminate sex	33	0	nr	1.00
Renal agenesis	144	9	nr	4.62
Cystic kidney	231	3	nr	7.07
Bladder exstrophy	19	0	nr	0.57
Polydactyly, preaxial	473	3	nr	14.37
Total Limb reduction defects (include unspecified)	109	4	nr	3.41
Transverse	nr	nr	nr	nr
Preaxial	nr	nr	nr	nr
Postaxial	nr	nr	nr	nr
Intercalary	nr	nr	nr	nr
Mixed	nr	nr	nr	nr
Unspecified	nr	nr	nr	nr
Diaphragmatic hernia	94	4	nr	2.96
Omphalocele***	172	19	nr	5.77
Gastroschisis	nr	nr	nr	nr
Unspecified Omphalocele/Gastroschisis	nr	nr	nr	nr
Prune belly sequence	nr	nr	nr	nr
Trisomy 13	23	11	nr	1.03
Trisomy 18	29	30	nr	1.78
Down syndrome, all ages (include age unknown)	403	37	nr	13.29
<20	nr	nr	nr	nr
20-24	nr	nr	nr	nr
25-29	nr	nr	nr	nr
30-34	nr	nr	nr	nr
35-39	nr	nr	nr	nr
40-44	nr	nr	nr	nr
45+	nr	nr	nr	nr
unknown	nr	nr	nr	nr

* All provinces and territories except Nova Scotia
**= include Epispadias
*** = include Gastroschisis and Unspecified Omphalocele/Gastroschisis
nr = not reported

Canada: National, Previous years rates 1989 - 2004

Prevalence rates: (LB+SB) * 10,000

	1974-79	1980-84	1985-89*	1990-94	1995-99	2000-04
Births			**375,840**	**1,918,123**	**1,728,310**	**1,615,779**
Anencephaly			2.16	1.90	1.26	1.01
Spina bifida			7.96	6.88	5.07	3.13
Encephalocele			1.46	1.36	0.93	0.66
Microcephaly			5.40	5.49	5.29	5.04
Arhinencephaly / Holoprosencephaly			nr	nr	nr	nr
Hydrocephaly			7.21	7.05	6.79	5.42
Anophthalmos			0.19	0.35	0.25	0.54
Microphthalmos			0.82	1.07	1.06	0.59
Unspecified Anophthalmos / Microphthalmos			0.00	0.00	0.00	0.00
Anotia			nr	nr	nr	nr
Microtia			nr	nr	nr	nr
Unspecified Anotia / Microtia			nr	nr	nr	nr
Transposition of great vessels			3.86	4.57	5.54	4.73
Tetralogy of Fallot			4.98	4.48	5.11	4.16
Hypoplastic left heart syndrome			2.87	2.86	2.80	2.69
Coarctation of aorta			5.51	5.34	5.95	5.19
Choanal atresia, bilateral			2.66	2.03	2.46	2.73
Cleft palate without cleft lip			6.70	7.17	7.38	6.84
Cleft lip with or without cleft palate			10.70	11.37	10.90	9.69
Oesophageal atresia / stenosis with or without fistula			3.35	3.47	3.36	3.00
Small intestine atresia / stenosis			3.49	3.47	3.55	3.92
Anorectal atresia / stenosis			5.75	5.13	4.91	4.41
Undescended testis (36 weeks of gestation or later)			36.03	34.29	32.74	35.48
Hypospadias**			27.11	26.78	26.93	29.57
Epispadias			nr	nr	nr	nr
Indeterminate sex			0.88	0.67	0.69	1.01
Renal agenesis			5.27	4.97	5.05	4.84
Cystic kidney			4.47	4.96	5.99	6.99
Bladder exstrophy			0.35	0.45	0.36	0.40
Polydactyly, preaxial			12.21	11.88	11.95	14.41
Total Limb reduction defects (include unspecified)			4.76	4.65	4.14	3.82
Transverse			nr	nr	nr	nr
Preaxial			nr	nr	nr	nr
Postaxial			nr	nr	nr	nr
Intercalary			nr	nr	nr	nr
Mixed			nr	nr	nr	nr
Unspecified			nr	nr	nr	nr
Diaphragmatic hernia			3.59	3.77	3.60	3.33
Omphalocele***			3.72	5.67	5.99	6.57
Gastroschisis			nr	nr	nr	nr
Unspecified Omphalocele/Gastroschisis			nr	nr	nr	nr
Prune belly sequence			nr	nr	nr	nr
Trisomy 13			1.41	1.13	1.11	1.14
Trisomy 18			2.02	2.18	2.37	2.22
Down syndrome, all ages (include age unknown)			12.13	13.21	13.48	14.63
<20			nr	nr	nr	nr
20-24			nr	nr	nr	nr
25-29			nr	nr	nr	nr
30-34			nr	nr	nr	nr
35-39			nr	nr	nr	nr
40-44			nr	nr	nr	nr
45+			nr	nr	nr	nr
unknown			nr	nr	nr	nr

*data include less than 5 years

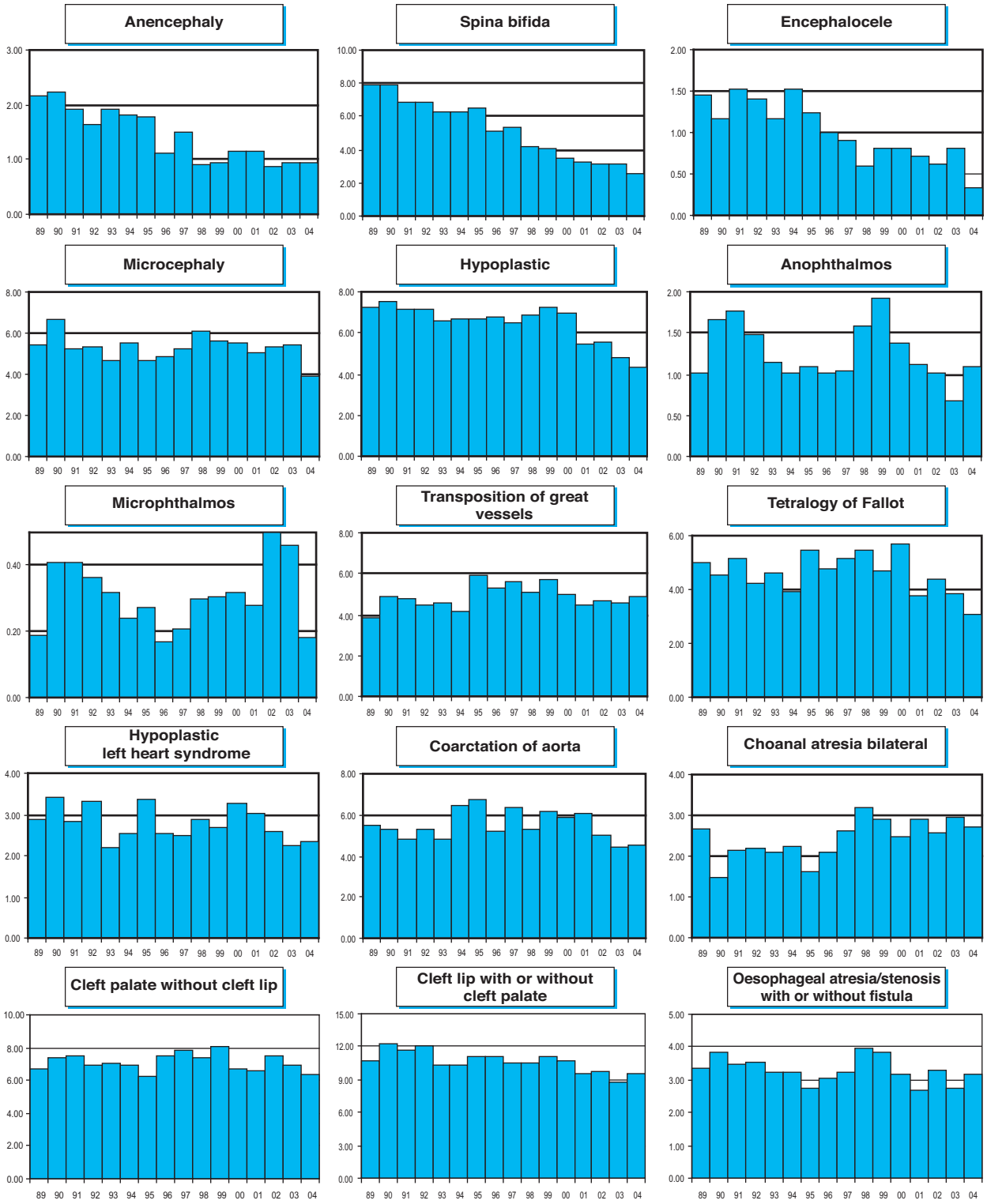
**= include Epispadias

*** = include Gastroschisis and Unspecified Omphalocele/Gastroschisis

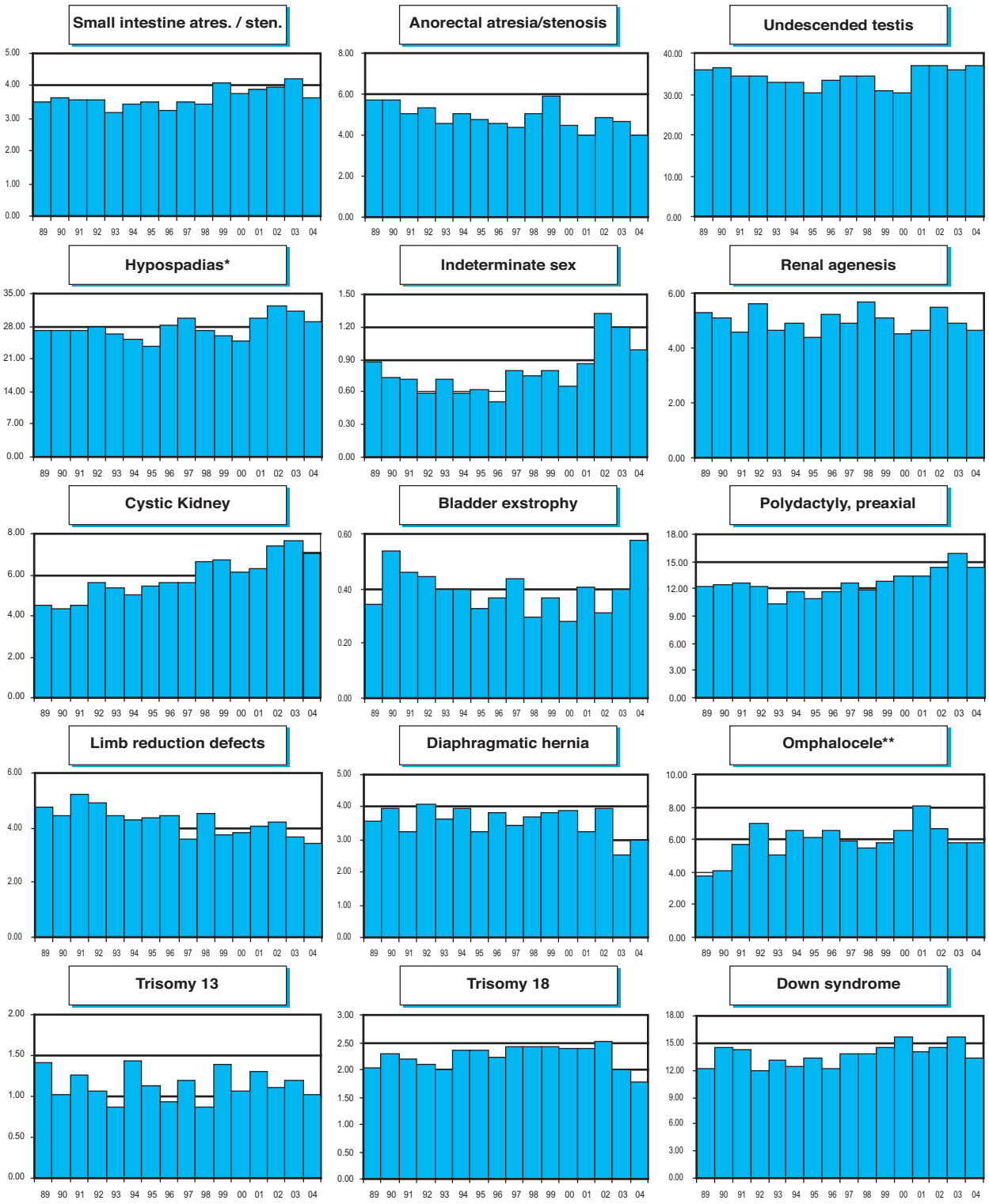
nr = not reported

Canada: National

Time trends 1989-2004 (Birth prevalence rates per 10,000)



Note: L+S rates



Note: L+S rates

Chile, Maule: RRMC-SSM

Regional Register Congenital Malformational Maule Health Service

History:
The register started in 2001 defined by order of Director Maule Health Service and assesored for South America.ECLAMC, Eduardo Castilla.
RRMC-SSM became a associated member of ICBDSR in 2003.

Size and coverage:
RRMC-SSM is located in a Region in the center of Chile, in Talca Maule Region.
Maule Region is situated between 34° 41' & 36° 33' S and 70° 20' & 72° 44' W. The surface is 30.535 kms2 (4 % of Chile). 930,306 habitants. 37,4% rurality.
Cellulosa producer and agricultural products.
The number of participating are 13 public hospitals from 2001 and since 2004 will included the unique private maternity of the region. There are around 13.500 births annually (2002).
The information about livebirths and stillbirths are collected from 13 maternity hospitals in the region for pediatricians and midwives.
Stillbirths of at least 500g birthweight have been included since 2001.

Legislation and funding:
The registry is based on the information of births and notification of congenital malformation ECLAMC from 2001 and funded by the Maule health Service.

Sources of ascertainment:
Reporting is made by collaborating pediatricians and midwives at the delivery units of partcipating hospitals.

Exposure information:
Detailed information on various risk factor exposures, maternal and paternal occupation, diseases and other information available.

Background information:
Epidemiological information on all births is available from participating hospitals and statisticals units.

Addresses and Staff:
M.Aurora Canessa, LinaresHospital, Maule Region-Chile. Av. Brazil 753, Linares, Chile.

Phone: 56-73-563276, 56-73-219879.

Fax: 56-73-219111, 56-73-219879.

E-mail: rrmc@ssmaule.cl

Chile Maule: RRMC-SSM, 2004

Live births (LB)	13,050
Stillbirths (SB)	57
Total births	13,107
Number of terminations of pregnancy (ToP) for birth defects	not permitted

Birth Defects	Number of cases			Rates*10,000
	LB	SB	ToP	Total rate
Anencephaly	2	2		3.05
Spina bifida	4	0		3.05
Encephalocele	2	0		1.53
Microcephaly	4	0		3.05
Arhinencephaly / Holoprosencephaly	1	0		0.76
Hydrocephaly	3	0		2.29
Anophthalmos	0	0		0.00
Microphthalmos	1	0		0.76
Unspecified Anophthalmos/ Microphthalmos	0	0		---
Anotia	0	0		0.00
Microtia	1	0		0.76
Unspecified Anotia/Microtia	0	0		---
Transposition of great vessels	2	0		1.53
Tetralogy of Fallot	1	0		0.76
Hypoplastic left heart syndrome	0	0		0.00
Coarctation of aorta	0	0		0.00
Choanal atresia, bilateral	1	0		0.76
Cleft palate without cleft lip	4	0		3.05
Cleft lip with or without cleft palate	11	0		8.39
Oesophageal atresia / stenosis with or without fistula	0	0		0.00
Small intestine atresia / stenosis	0	0		0.00
Anorectal atresia / stenosis	3	0		2.29
Undescended testis (36 weeks of gestation or later)	6	0		4.58
Hypospadias	10	0		7.63
Epispadias	0	0		0.00
Indeterminate sex	0	0		0.00
Renal agenesis	1	0		0.76
Cystic kidney	1	0		0.76
Bladder exstrophy	1	0		0.76
Polydactyly, preaxial	0	0		0.00
Total Limb reduction defects (include unspecified)	7	0		5.34
Transverse	7	0		5.34
Preaxial	0	0		0.00
Postaxial	0	0		0.00
Intercalary	0	0		0.00
Mixed	0	0		0.00
Unspecified	0	0		---
Diaphragmatic hernia	0	0		0.00
Omphalocele	1	1		1.53
Gastroschisis	2	1		2.29
Unspecified Omphalocele/Gastroschisis	0	0		---
Prune belly sequence	0	0		0.00
Trisomy 13	1	0		0.76
Trisomy 18	0	0		0.00
Down syndrome, all ages (include age unknown)	38	3		31.28
<20	0	0		0.00
20-24	4	0		12.49
25-29	3	0		9.83
30-34	8	0		30.65
35-39	9	3		76.97
40-44	13	0		304.45
45+	1	0		1000.00
unknown	0	0		---

China: BDSS-Beijing

Birth Defect Surveillance System in Thirty Counties of Four Provinces, People's Republic of China

History:
The Programme began in 1992. It became a full member of the ICBDSR in 1997.

Size and coverage:
This is a population based monitoring system. Reports were obtained from all hospitals and village health stations, which together cover all geographically defined population. Total number of population in these areas is around 17 millions and total number of births per year is around 150,000.

Legislation and funding:
Funding is from China Ministry of Health and local health authorities.

Sources of ascertainment:
Reports are obtained from delivery units, paediatric clinics, ultrasound departments, pathology departments and perinatal health care departments of different level hospitals, MCH institutes and village health stations in the participating counties and cities.

Exposure information:
Exposure information is obtained from the perinatal health care surveillance system (PHCSS) in the same areas for all women and their babies from pre-marital examination till six weeks after birth. BDSS data is linked with PHCSS data by using an ID number assigned to each woman.

Background information:
Background information is also obtained from PHCSS data.

Addresses and Staff:
Zhu Li, MD, Programme Director, National Centre for Maternal and Infant Health, Beijing Medical University Rm 115 Research Centre, 38 College Rd. Beijing 100083 PR China

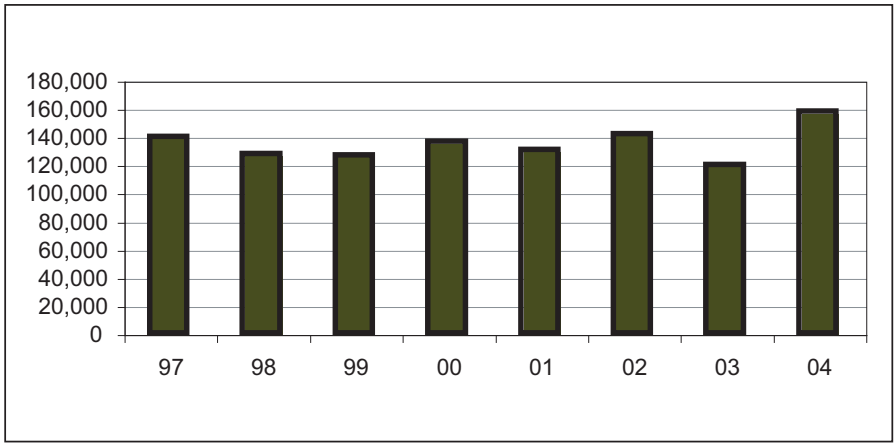
Phone: 86-10-62091138

Fax: 86-10-62091141

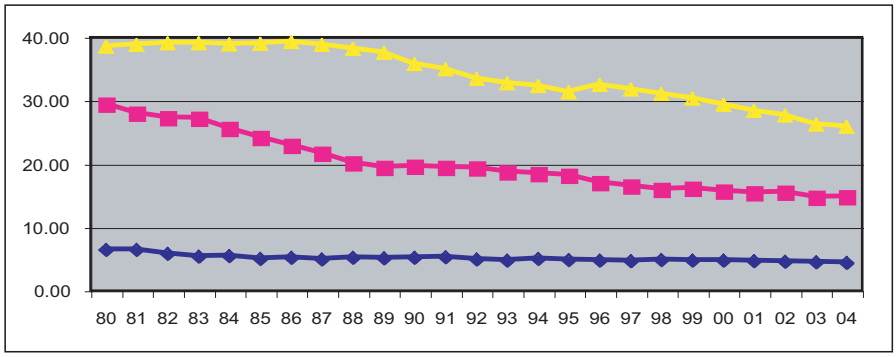
E-mail: lzh@public.bta.net.cn

China: BDSS-Beijing

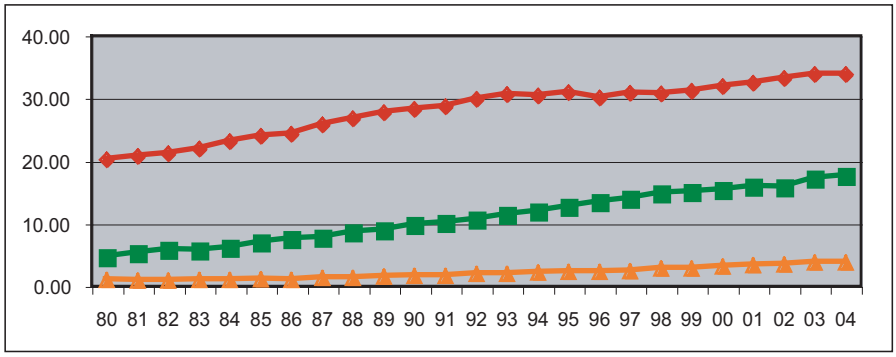
Total births by year



Percentage of births by maternal age



legenda: %births < 20 %births 20-24 %births 25-29



legenda: %births 30-34 %births 35-39 %births 40+

China: BDSS-Beijing, 2004

Live births (LB)	157,188
Stillbirths (SB)	527
Total births	157,715
Number of terminations of pregnancy (ToP) for birth defects	nr

Birth Defects	Number of cases			Rates*10,000
	LB	SB	ToP	Total rate
Anencephaly	0	41	nr	2.60
Spina bifida	8	17	nr	1.59
Encephalocele	4	11	nr	0.95
Microcephaly	2	0	nr	0.13
Arhinencephaly / Holoprosencephaly	1	3	nr	0.25
Hydrocephaly	8	58	nr	4.18
Anophthalmos	1	0	nr	0.06
Microphthalmos	2	0	nr	0.13
Unspecified Anophthalmos/ Microphthalmos	0	0	nr	0.00
Anotia	5	0	nr	0.32
Microtia	38	1	nr	2.47
Unspecified Anotia/Microtia	0	0	nr	0.00
Transposition of great vessels	nr	nr	nr	nr
Tetralogy of Fallot	nr	nr	nr	nr
Hypoplastic left heart syndrome	nr	nr	nr	nr
Coarctation of aorta	nr	nr	nr	nr
Choanal atresia, bilateral	nr	nr	nr	nr
Cleft palate without cleft lip	28	2	nr	1.90
Cleft lip with or without cleft palate	103	44	nr	9.32
Oesophageal atresia / stenosis with or without fistula	nr	nr	nr	nr
Small intestine atresia / stenosis	nr	nr	nr	nr
Anorectal atresia / stenosis	16	4	nr	1.27
Undescended testis (36 weeks of gestation or later)	4	0	nr	0.25
Hypospadias	8	0	nr	0.51
Epispadias	1	0	nr	0.06
Indeterminate sex	9	5	nr	0.89
Renal agenesis	nr	nr	nr	nr
Cystic kidney	nr	nr	nr	nr
Bladder exstrophy	1	0	nr	0.06
Polydactyly, preaxial	88	4	nr	5.83
Total Limb reduction defects (include unspecified)	27	17	nr	2.79
Transverse	24	11	nr	2.22
Preaxial	3	8	nr	0.70
Postaxial	0	0	nr	0.00
Intercalary	1	0	nr	0.06
Mixed	1	2	nr	0.19
Unspecified	0	0	nr	0.00
Diaphragmatic hernia	nr	nr	nr	nr
Omphalocele	6	9	nr	0.95
Gastroschisis	7	23	nr	1.90
Unspecified Omphalocele/Gastroschisis	0	0	nr	0.00
Prune belly sequence	2	10	nr	0.76
Trisomy 13	nr	nr	nr	nr
Trisomy 18	nr	nr	nr	nr
Down syndrome, all ages (include age unknown)	nr	nr	nr	nr
<20	nr	nr	nr	nr
20-24	nr	nr	nr	nr
25-29	nr	nr	nr	nr
30-34	nr	nr	nr	nr
35-39	nr	nr	nr	nr
40-44	nr	nr	nr	nr
45+	nr	nr	nr	nr
unknown	nr	nr	nr	nr

nr = not reported

China: BDSS-Beijing, Previous years rates 1997 - 2004

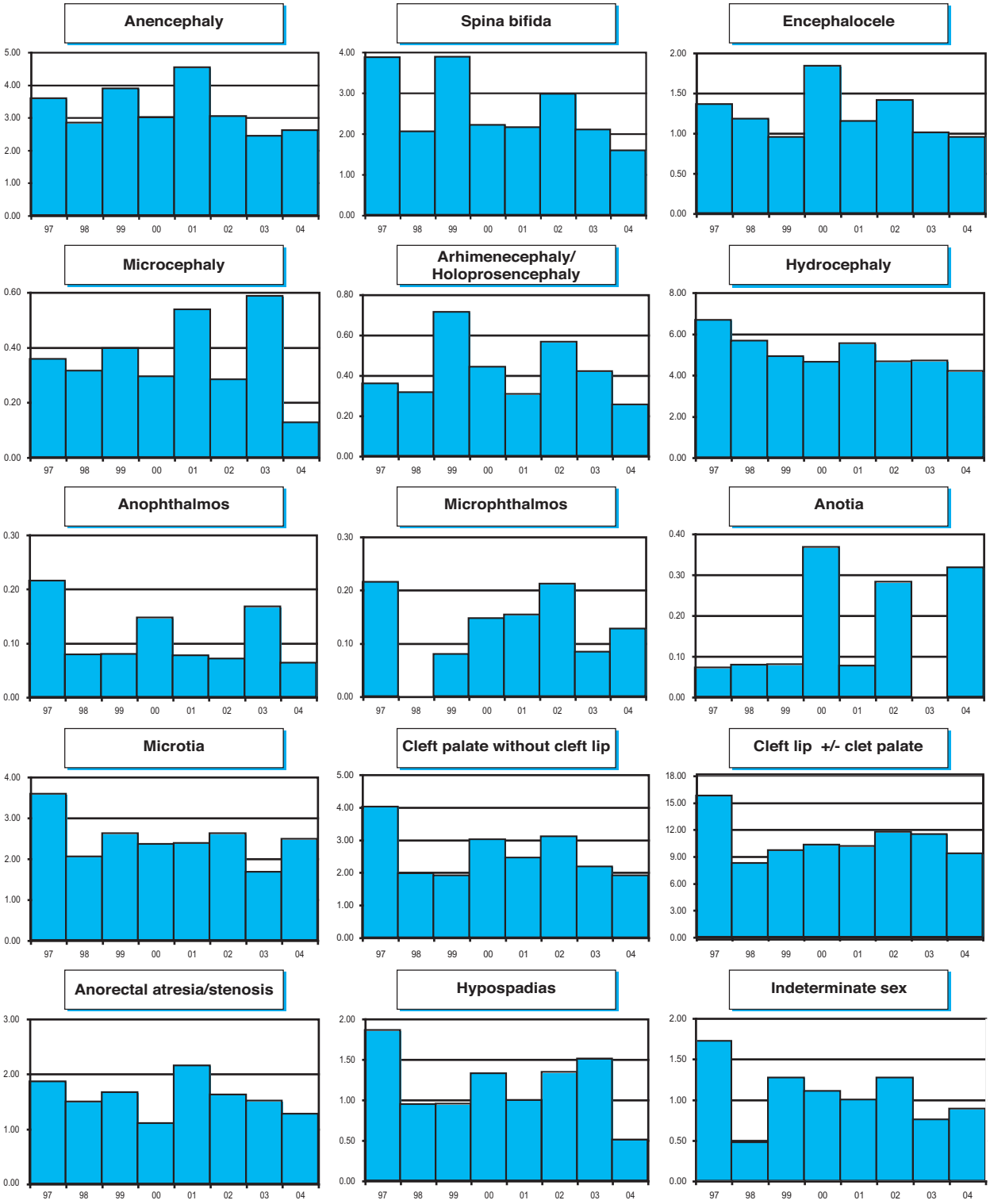
Birth prevalence rates: (LB+SB) * 10,000

	1974-79	1980-84	1985-89	1990-94	1995-99*	2000-04
Births					**393,354**	**685,662**
Anencephaly					3.43	3.11
Spina bifida					3.28	2.19
Encephalocele					1.17	1.27
Microcephaly					0.36	0.35
Arhinencephaly / Holoprosencephaly					0.46	0.39
Hydrocephaly					5.77	4.71
Anophthalmos					0.13	0.10
Microphthalmos					0.10	0.15
Unspecified Anophthalmos / Microphthalmos					---	---
Anotia					0.08	0.22
Microtia					2.77	2.32
Unspecified Anotia / Microtia					---	---
Transposition of great vessels					nr	nr
Tetralogy of Fallot					nr	nr
Hypoplastic left heart syndrome					nr	nr
Coarctation of aorta					nr	nr
Choanal atresia, bilateral					nr	nr
Cleft palate without cleft lip					2.67	2.52
Cleft lip with or without cleft palate					11.36	10.53
Oesophageal atresia / stenosis with or without fistula					nr	nr
Small intestine atresia / stenosis					nr	nr
Anorectal atresia / stenosis					1.68	1.52
Undescended testis (36 weeks of gestation or later)					0.23	0.25
Hypospadias					1.27	1.11
Epispadias					0.00	0.03
Indeterminate sex					1.17	1.01
Renal agenesis					nr	nr
Cystic kidney					nr	nr
Bladder exstrophy					0.05	0.03
Polydactyly, preaxial					7.14	5.85
Total Limb reduction defects (include unspecified)					2.39	2.57
Transverse					1.50	1.93
Preaxial					0.32	0.42
Postaxial					0.00	0.00
Intercalary					0.04	0.04
Mixed					0.00	0.07
Unspecified					---	---
Diaphragmatic hernia					nr	nr
Omphalocele					1.25	0.80
Gastroschisis					1.68	1.91
Unspecified Omphalocele / Gastroschisis					---	---
Prune belly sequence					1.65	0.79
Trisomy 13					nr	nr
Trisomy 18					nr	nr
Down syndrome, all ages (include age unknown)					nr	nr
<20					nr	nr
20-24					nr	nr
25-29					nr	nr
30-34					nr	nr
35-39					nr	nr
40-44					nr	nr
45+					nr	nr
unknown					---	---

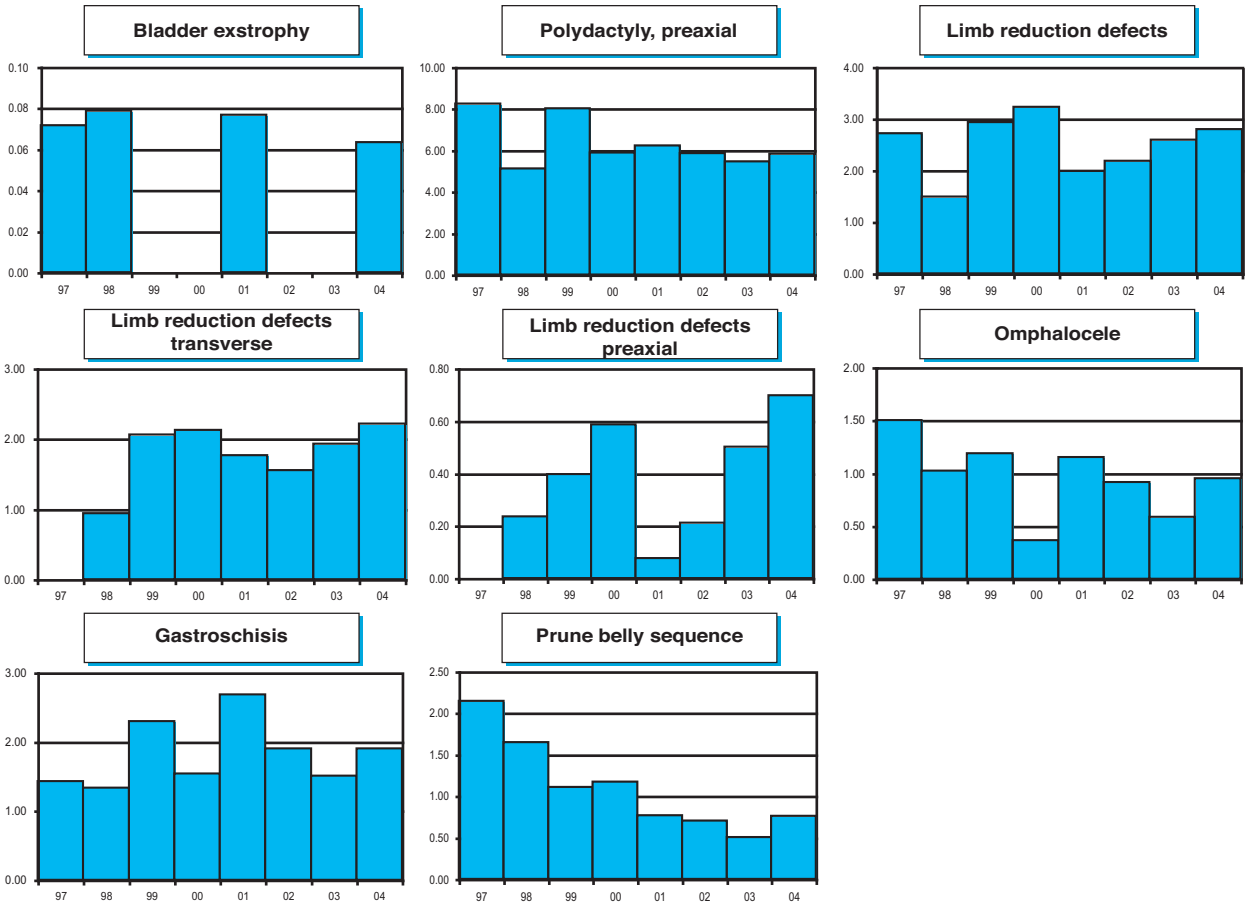
nr= not reported

China: BDSS-Beijing

Time trends 1997-2004 (Birth prevalence rates per 10,000)



Note: L+S rates



Note: L+S rates

China: CBDMN

Chinese Birth Defects Program of Sichuan Province, China (until 1994)
Chinese Birth Defects Monitoring Network

History:
The Programme began in 1984. It became an associate member of the ICBDSR in 1985 and a full member in 1987.

Size and coverage:
In 1984, reports were obtained from 100 hospitals but participation has increased. In 1985,205 hospitals participated. At present, the Programme covers approximately 260,000 births annually in 31 provinces.

Since we resumed reporting data, only one part of data (20 provinces, I remember apprising you by E-mail several years ago) is sent to ICBD. The nationwide programme covers approximately 450,000~500,000 births annually in provinces.

Legislation and funding:
Participation is voluntary. Funding is mainly from local health authorities, also supported by Ministry of health.

Sources of ascertainment:
Reports are obtained from delivery units, paediatric clinics, and pathology departments of the participating hospitals.

Exposure information:
Exposure information is obtained by interviews of mothers of the reported malformed infants. No information is available on exposures in controls.

Background information:
Total number of births from each participating hospital is known.

Addresses and Staff:
Zhu Jun, MD, Programme Director National Centre Birth Defects Monitoring (CNBDM) West China University of Medical Sciences No. 17 Section 3-Ren Min Nam Lu Chengdu - PRC - 610041, China

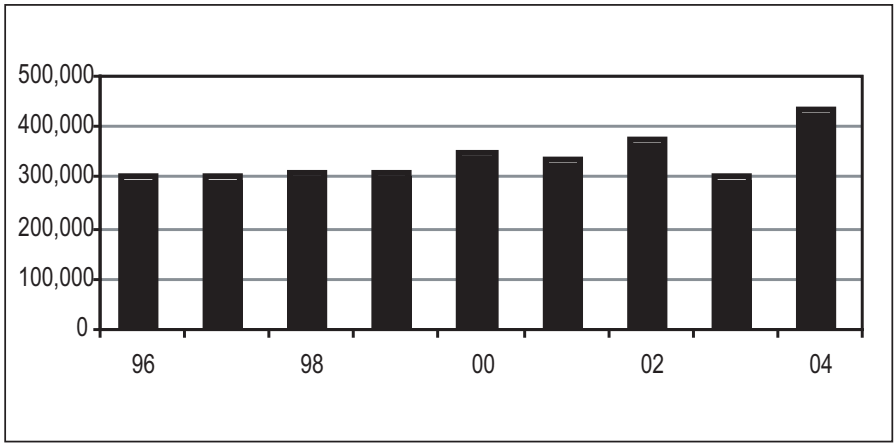
Phone: 86-28-5501363

Fax: 86-28-5501363

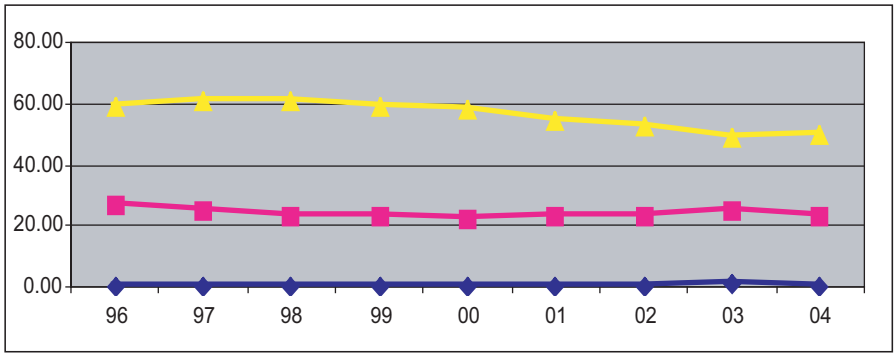
E-mail: cnbdms@mail.sc.cninfo.net

China: CBDMN

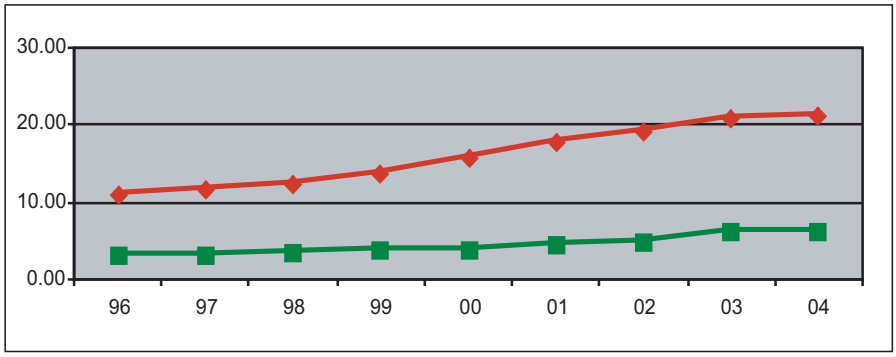
Total births by year



Percentage of births by maternal age



legenda: %births < 20 %births 20-24 %births 25-29



legenda: %births 30-34 %births 35-39 %births 40+

China: CBDMN, 2004

Live births (LB)	423,974
Stillbirths (SB)	3,729
Total births	427,703
Number of terminations of pregnancy (ToP) for birth defects	nr

Birth Defects	Number of cases			Rates*10,000
	LB	SB	ToP	Total rate
Anencephaly	15	107	nr	2.85
Spina bifida	112	147	nr	6.06
Encephalocele	29	34	nr	1.47
Microcephaly	4	2	nr	0.14
Arhinencephaly / Holoprosencephaly	nr	nr	nr	nr
Hydrocephaly	46	286	nr	7.76
Anophthalmos*	8	3	nr	0.26
Microphthalmos	nr	nr	nr	nr
Unspecified Anophthalmos/ Microphthalmos	nr	nr	nr	---
Anotia**	115	11	nr	2.95
Microtia	nr	nr	nr	nr
Unspecified Anotia/Microtia	nr	nr	nr	---
Transposition of great vessels	nr	nr	nr	nr
Tetralogy of Fallot	nr	nr	nr	nr
Hypoplastic left heart syndrome	nr	nr	nr	nr
Coarctation of aorta	nr	nr	nr	nr
Choanal atresia, bilateral	nr	nr	nr	nr
Cleft palate without cleft lip	138	4	nr	3.32
Cleft lip with or without cleft palate	470	145	nr	14.38
Oesophageal atresia / stenosis with or without fistula	34	14	nr	1.12
Small intestine atresia / stenosis	nr	nr	nr	nr
Anorectal atresia / stenosis	104	15	nr	2.78
Undescended testis (36 weeks of gestation or later)	47	1	nr	1.12
Hypospadias	214	7	nr	5.17
Epispadias	nr	nr	nr	nr
Indeterminate sex	32	25	nr	1.33
Renal agenesis	7	18	nr	0.58
Cystic kidney	15	46	nr	1.43
Bladder exstrophy	1	0	nr	0.02
Polydactyly, unspecified	580	24	nr	14.12
Total Limb reduction defects (include unspecified)	125	75	nr	4.68
Transverse	nr	nr	nr	nr
Preaxial	nr	nr	nr	nr
Postaxial	nr	nr	nr	nr
Intercalary	nr	nr	nr	nr
Mixed	nr	nr	nr	nr
Unspecified	nr	nr	nr	---
Diaphragmatic hernia	14	18	nr	0.75
Omphalocele	41	32	nr	1.71
Gastroschisis	60	49	nr	2.55
Unspecified Omphalocele/Gastroschisis	0	0	nr	---
Prune belly sequence	nr	nr	nr	nr
Trisomy 13	0	2	nr	0.05
Trisomy 18	2	2	nr	0.09
Down syndrome, all ages (include age unknown)	101	10	nr	2.60
<20	2	0	nr	10.74
20-24	11	2	nr	1.33
25-29	49	3	nr	2.45
30-34	25	3	nr	3.12
35+	14	2	nr	6.21
unknown	0	0	nr	---

* = include Microphthalmos and Unspecified Anophthalmos/ Microphthalmos
** = include Microtia and Unspecified Anotia/Microtia
nr = not reported

China: CBDMN, Previous years rates 1996 - 2004

Birth prevalence rates: (LB+SB) * 10,000

	1974-79	1980-84	1985-89	1990-94	1995-99*	2000-04
Total births					**1,198,083**	**1,764,747**
Anencephaly					5.59	3.57
Spina bifida					7.58	6.78
Encephalocele					1.94	1.54
Microcephaly					0.22	0.23
Arhinencephaly / Holoprosencephaly					nr	nr
Hydrocephaly					6.63	7.07
Anophthalmos**					0.40	0.31
Microphthalmos					nr	nr
Unspecified Anophthalmos / Microphthalmos					---	---
Anotia***					2.98	3.01
Microtia					nr	nr
Unspecified Anotia / Microtia					---	---
Transposition of great vessels					nr	nr
Tetralogy of Fallot					nr	nr
Hypoplastic left heart syndrome					nr	nr
Coarctation of aorta					nr	nr
Choanal atresia, bilateral					nr	nr
Cleft palate without cleft lip					2.31	2.71
Cleft lip with or without cleft palate					13.89	14.26
Oesophageal atresia / stenosis with or without fistula					0.68	0.93
Small intestine atresia / stenosis					nr	nr
Anorectal atresia / stenosis					2.59	3.21
Undescended testis (36 weeks of gestation or later)					0.61	0.95
Hypospadias					3.30	4.75
Epispadias					nr	nr
Indeterminate sex					1.11	1.25*
Renal agenesis					0.24	0.39*
Cystic kidney					0.74	1.24
Bladder exstrophy					0.08	0.09
Polydactyly, preaxial					nr	13.43*
Total Limb reduction defects (include unspecified)					5.23	5.41
Transverse					nr	nr
Preaxial					nr	nr
Postaxial					nr	nr
Intercalary					nr	nr
Mixed					nr	nr
Unspecified					---	---
Diaphragmatic hernia					0.53	0.58
Omphalocele					1.38	1.53
Gastroschisis					2.75	2.69
Unspecified Omphalocele / Gastroschisis					---	---
Prune belly sequence					nr	nr
Trisomy 13					nr	0.05*
Trisomy 18					nr	0.08*
Down syndrome, all ages (include age unknown)					1.79	2.41
<20					0.00	6.66
20-24					1.09	1.25
25-29					1.40	2.04
30-34					2.47	2.82
35+					10.17	9.64
unknown					---	---

* data include less than 5 years

**= include Microphthalmos and Unspecified Anophthalmos/ Microphthalmos

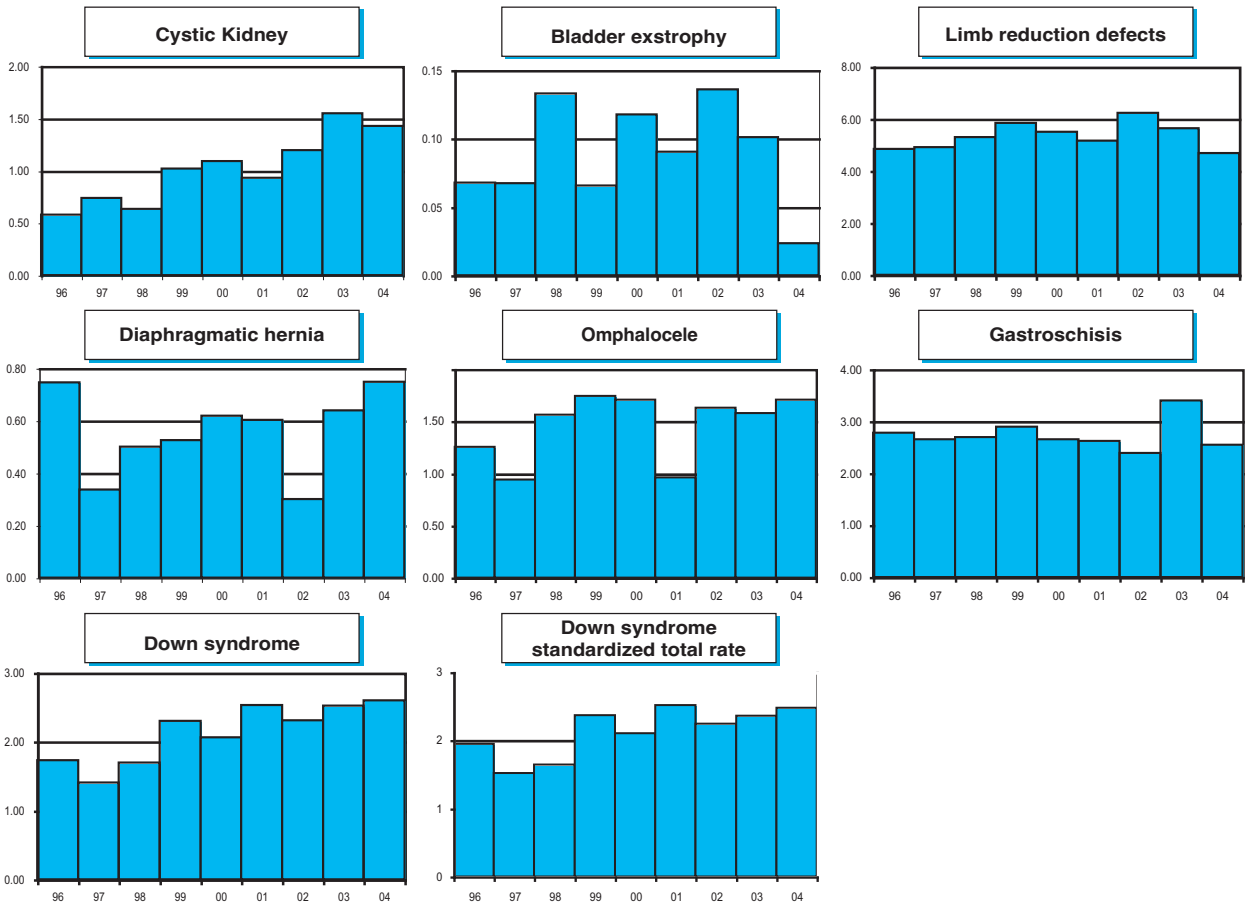
*** = include Microtia and Unspecified Anotia/Microtia

nr = not reported

China: CBDMN

Time trends 1996-2004 (Birth prevalence rates per 10,000)

Note: L+S rates



Note: L+S rates

Costa Rica: CREC
Costa Rican Birth Defects Register Center

History:
The registry was created in 1986, based in a government decret by which birth defects became subject of obligatory notification.
The program became an ICBDSR member in September 2003.

Size and coverage
The program is population based. Includes all births from the National Security System (CCSS) which covers about 98% of all births occurred in the country, and births of private hospitals.
There are approximately 75000 annual births in Costa Rica.

Legislation and funding
The Registry is financed by the government as a program of the Costa Rican Institute of Research and Training in Nutrition and Health (INCIENSA), Institute that depends from the Ministry of Health.

Sources and ascertainment
Reporting is made by neonatologists, pediatricians and physicians before newborns discharge from maternity services, with biostatistics personal collaboration.

Exposure information
None is routinely collected at present.

Background information
Linkage studies are possible with other statistical data from the National Statistics Center and the National Security System Statistical Center

Addresses and Staff:
Lila Umaña,
Costa Rican Birth Defects Register Centre (CREC)
Department of Genetics, Costa Rican Institute of Research and training in Nutrition and Health.
PO Box 4-2250 Tres Ríos, Cartago.
Costa Rica, Central America.

Phone: (506) 2799911

Fax: (506) 2795546

E-mail: lumana@inciensa.sa.cr

Costa Rica: CREC, 2004

Live births (LB)	nr
Stillbirths (SB)	nr
Total births	71,125
Number of terminations of pregnancy (ToP) for birth defects	not permitted

Birth Defects	Number of cases			Rates*10,000
	LB	SB	ToP	Total rate
Anencephaly	11	0		1.55
Spina bifida	24	0		3.37
Encephalocele	6	0		0.84
Microcephaly	8	0		1.12
Arhinencephaly / Holoprosencephaly	4	0		0.56
Hydrocephaly	20	0		2.81
Anophthalmos	2	0		0.28
Microphthalmos	5	0		0.70
Unspecified Anophthalmos/ Microphthalmos	0	0		---
Anotia	4	0		0.56
Microtia	12	0		1.69
Unspecified Anotia/Microtia	0	0		---
Transposition of great vessels	6	0		0.84
Tetralogy of Fallot	17	0		2.39
Hypoplastic left heart syndrome	2	0		0.28
Coarctation of aorta	3	0		0.42
Choanal atresia, bilateral	2	0		0.28
Cleft palate without cleft lip	17	0		2.39
Cleft lip with or without cleft palate	53	0		7.45
Oesophageal atresia / stenosis with or without fistula	8	0		1.12
Small intestine atresia / stenosis	3	0		0.42
Anorectal atresia / stenosis	20	0		2.81
Undescended testis (36 weeks of gestation or later)	76	0		10.69
Hypospadias	46	0		6.47
Epispadias	0	0		0.00
Indeterminate sex	5	0		0.70
Renal agenesis	1	0		0.14
Cystic kidney	3	0		0.42
Bladder exstrophy	1	0		0.14
Polydactyly, preaxial	6	0		0.84
Total Limb reduction defects (include unspecified)	36	0		5.06
Transverse	nr	nr		nr
Preaxial	nr	nr		nr
Postaxial	nr	nr		nr
Intercalary	nr	nr		nr
Mixed	nr	nr		nr
Unspecified	nr	nr		---
Diaphragmatic hernia	6	0		0.84
Omphalocele	3	0		0.42
Gastroschisis	11	0		1.55
Unspecified Omphalocele/Gastroschisis	0	0		---
Prune belly sequence	4	0		0.56
Trisomy 13	5	0		0.70
Trisomy 18	8	0		1.12
Down syndrome, all ages (include age unknown)	63	0		8.86
<20	10	0		6.90
20-24	10	0		4.66
25-29	5	0		3.00
30-34	4	0		3.65
35-39	19	0		33.46
40-44	10	0		68.97
45+	0	0		0.00
unknown	0	0		---

nr = not reported

Cuba: RECUMAC
Cuban Register of Congenital Malformation

History:
The program started in 1985 and has grown in size and coverage. The registry became a member of ICBDSR in 2003.

Size and coverage
Reports are obtained from hospitals distributed all over Cuba. The number of partecipating hospitals has grown in 1986 to 60 at the present time. The annual number of birth is approximately 121 000 representing almost 96% of all births.

Legislation and funding
RIt is a research programme with voluntary participation of hospitals. The registry is associated with the National Centre of Medical Genetics, and is financed by Health Public Ministery of Cuba.

Sources and ascertainment
Reports are obtained from delivery units paediatric departments of the participating hospitals. Mothers are also interviewed directly to gather information and fill in the RECUMAC standard protocols.

Exposure information
The mother of each reported infant and the mother of a control infant, the next non malformed infant born at the hospital with the same sex as the proband are interviewed on various exposures, including drug usage and parental occupation.

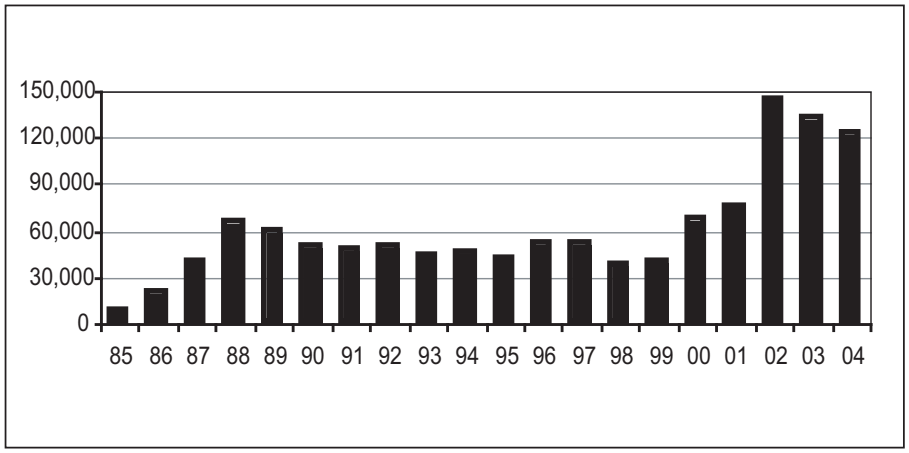
Background information
Total number of birth by sex and number of twin pairs in each participating hospital are known. Other background information is obtained partly from summarizing tables of births in each participating hospital, partly from the control material.

Address for further information
Maria Teresa Pérez Mateo, Recumac
Centro Nacional de Genética Medica ISCM-Habana Victoria de Giròn, C.P. 16000 Ciudad de la Habana. Cuba.

E-mail: mauro@infomed.sld.cu

Cuba: RECUMAC

Total births by year



Cuba: RECUMAC, 2004

Live births (LB)	119,841
Stillbirths (SB)	1,702
Total births	121,543
Number of terminations of pregnancy (ToP) for birth defects	772

Birth Defects	Number of cases			Rates*10,000
	LB	SB	ToP	Total rate
Anencephaly	0	1	76	6.30
Spina bifida	15	2	57	6.05
Encephalocele	1	2	15	1.47
Microcephaly	2	0	6	0.65
Arhinencephaly / Holoprosencephaly	0	0	3	0.25
Hydrocephaly	26	0	77	8.42
Anophthalmos	2	1	1	0.33
Microphthalmos	1	0	0	0.08
Unspecified Anophthalmos/ Microphthalmos	1	0	0	0.08
Anotia 2	0	0	0.16	
Microtia	3	0	0	0.25
Unspecified Anotia/Microtia	0	0	0	0.00
Transposition of great vessels	5	3	9	1.39
Tetralogy of Fallot	7	2	9	1.47
Hypoplastic left heart syndrome	6	0	10	1.31
Coarctation of aorta	2	0	2	0.33
Choanal atresia, bilateral	2	0	1	0.25
Cleft palate without cleft lip	16	0	1	1.39
Cleft lip with or without cleft palate	36	0	5	3.35
Oesophageal atresia / stenosis with or without fistula	18	2	5	2.04
Small intestine atresia / stenosis	7	2	10	1.55
Anorectal atresia / stenosis	10	2	1	1.06
Undescended testis (36 weeks of gestation or later)	15	0	0	1.23
Hypospadias	100	0	0	8.18
Epispadias	1	0	0	0.08
Indeterminate sex	3	0	2	0.41
Renal agenesis	3	2	4	0.74
Cystic kidney	8	0	22	2.45
Bladder exstrophy	0	0	1	0.08
Polydactyly, preaxial	10	0	0	0.82
Total Limb reduction defects (include unspecified)	15	0	9	1.96
Transverse	4	0	0	0.33
Preaxial	2	0	0	0.16
Postaxial	0	0	0	0.00
Intercalary	8	0	0	0.65
Mixed	0	0	0	0.00
Unspecified	1	0	9	0.82
Diaphragmatic hernia	6	1	13	1.64
Omphalocele	2	1	23	2.13
Gastroschisis	7	1	35	3.52
Unspecified Omphalocele/Gastroschisis	2	0	1	0.25
Prune belly sequence	0	0	1	0.08
Trisomy 13	5	2	7	1.14
Trisomy 18	4	1	8	1.06
Down syndrome, all ages (include age unknown)	117	2	14	10.87
<20	11	1	2	nr
20-24	18	1	1	nr
25-29	25	0	0	nr
30-34	32	0	0	nr
35-39	20	0	7	nr
40-44	9	0	3	nr
45+	0	0	1	nr
unknown	2	0	0	nr

nr = not reported

Cuba: RECUMAC, Previous years rates 1985 - 2004

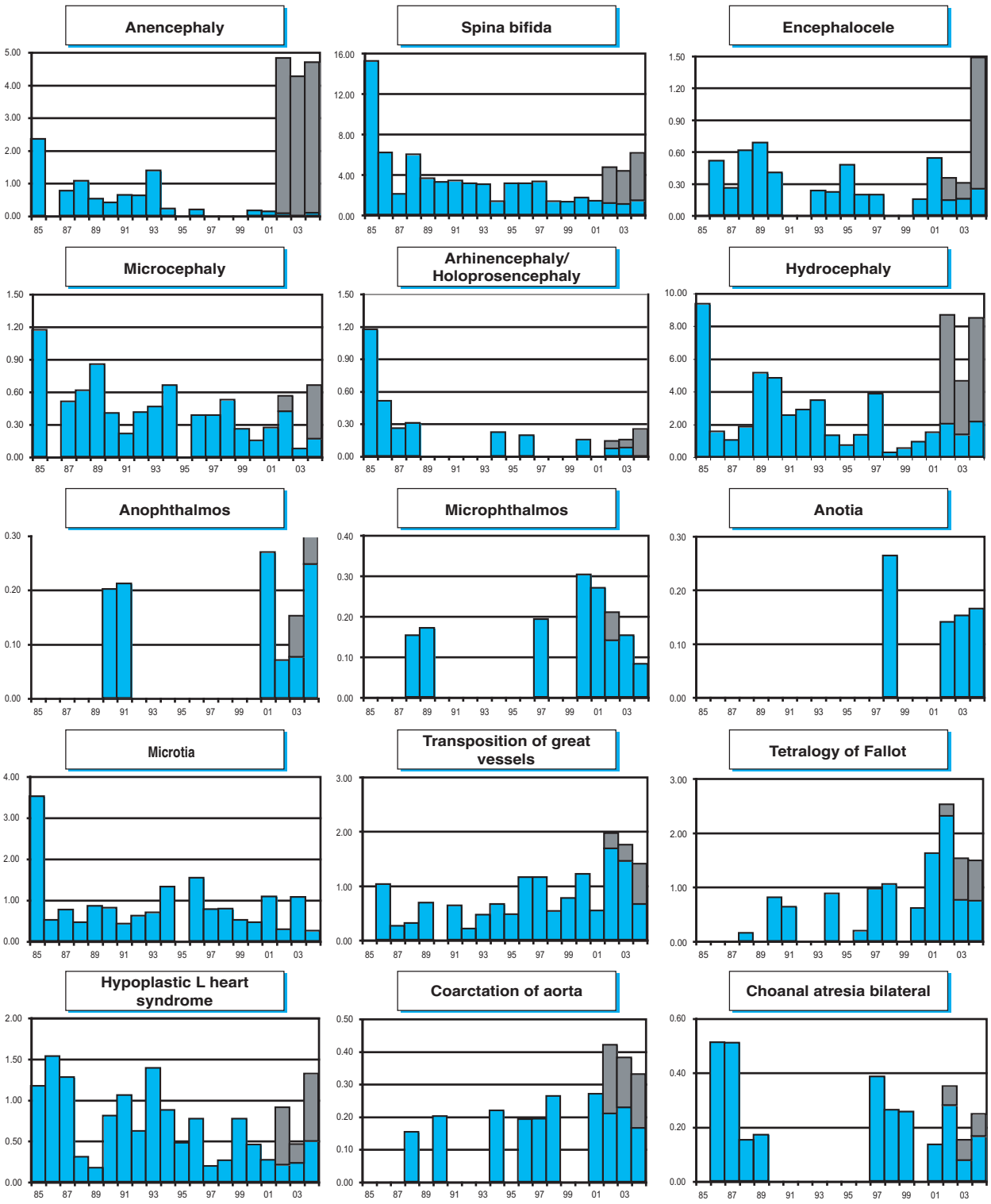
Birth prevalence rates: (LB+SB) * 10,000 until 2001
Birth prevalence rates: (LB+SB+TOP) * 10,000 from 2002

	1974-79	1980-84	1985-89	1990-94	1995-99	2000-04
Total births			**191,491**	**234,691**	**223,546**	**536,617**
Anencephaly			0.78	0.64	0.04	3.43
Spina bifida			4.86	2.81	2.51	3.76
Encephalocele			0.52	0.17	0.18	0.54
Microcephaly			0.63	0.43	0.31	0.30
Arhinencephaly / Holoprosencephaly			0.26	0.04	0.04	0.15
Hydrocephaly			2.98	3.03	1.48	5.63
Anophthalmos			0.00	0.09	0.00	0.13
Microphthalmos			0.10	0.00	0.04	0.19
Unspecified Anophthalmos / Microphthalmos			---	---	---	---
Anotia			0.00	0.00	0.04	0.11
Microtia			0.78	0.77	0.76	0.60
Unspecified Anotia / Microtia			---	---	---	---
Transposition of great vessels			0.47	0.38	0.85	1.34
Tetralogy of Fallot			0.05	0.47	0.45	1.55
Hypoplastic left heart syndrome			0.63	0.94	0.49	0.76
Coarctation of aorta			0.05	0.09	0.13	0.32
Choanal atresia, bilateral			0.26	0.00	0.18	0.20
Cleft palate without cleft lip			1.46	1.41	1.48	1.53
Cleft lip with or without cleft palate			4.80	5.37	6.26	4.88
Oesophageal atresia / stenosis with or without fistula			1.31	1.53	1.92	2.37
Small intestine atresia / stenosis			0.63	0.77	0.63	1.19
Anorectal atresia / stenosis			1.78	1.53	1.07	1.32
Undescended testis (36 weeks of gestation or later)			4.86	3.71	2.95	2.31
Hypospadias			12.79	15.13	10.33	7.72
Epispadias			0.26	0.30	0.13	0.09
Indeterminate sex			0.37	0.17	0.18	0.34
Renal agenesis			0.68	0.21	0.36	0.69
Cystic kidney			1.15	1.11	0.40	1.62
Bladder exstrophy			0.42	0.13	0.18	0.11
Polydactyly, preaxial			0.10	0.21	0.40	0.86
Total Limb reduction defects (include unspecified)			2.77	2.77	2.24	2.33
Transverse			1.15	0.89	0.67	0.60
Preaxial			0.00	0.00	0.00	0.06
Postaxial			0.00	0.00	0.00	0.04
Intercalary			0.00	0.00	0.00	0.09
Mixed			0.00	0.00	0.00	0.19
Unspecified			---	---	---	---
Diaphragmatic hernia			1.57	1.41	1.30	1.66
Omphalocele			0.68	0.81	0.31	1.34
Gastroschisis			0.31	0.43	0.40	2.22
Unspecified Omphalocele / Gastroschisis			---	---	---	---
Prune belly sequence			0.10	0.13	0.00	0.02
Trisomy 13			0.37	0.60	0.31	0.82
Trisomy 18			0.10	0.26	0.45	0.45
Down syndrome, all ages (include age unknown)			8.15	7.97	7.38	9.32
<20			nr	nr	nr	nr
20-24			nr	nr	nr	nr
25-29			nr	nr	nr	nr
30-34			nr	nr	nr	nr
35-39			nr	nr	nr	nr
40-44			nr	nr	nr	nr
45+			nr	nr	nr	nr
unknown			---	---	---	---

nr = not reported

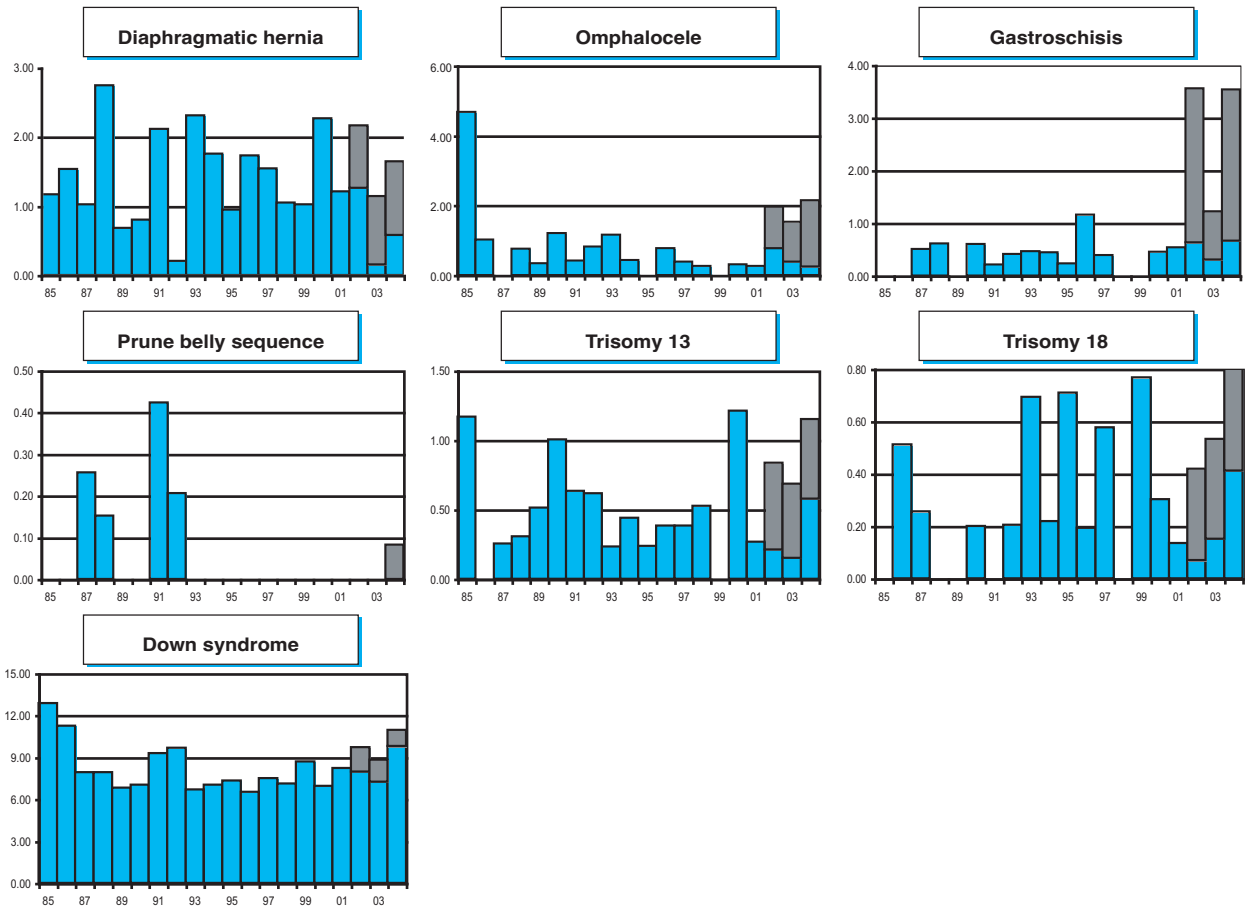
Cuba: RECUMAC

'Time trends 1985-2004 (Birth prevalence rates per 10,000)



Note: L+S rates, ToP rates

Note: L+S rates, ToP rates



Note: L+S rates, ToP rates

Czech Republic

Congenital Malformations Monitoring Program of the Czech Republic

History:
A registration of congenital malformation began in 1961 and regular monitoring started in 1975. The programme was a founding member of the Clearinghouse and is a full member.

Size and coverage:
All births in the Czech Republic (Bohemia, Moravia and Silesia regions) are covered, at present comprising approximately 90,000 annual births. Stillbirths weighting at least 1,000g are included.

Legislation and funding:
Reporting is compulsory. The registration is financed and run by the government in the Institute of Health Information and Statistics of the Czech Republic. Analysis of data is supported by Grant project NJ 7516-3 of Grant Agency Ministry of Health of the Czech Republic in the Institute for Care of Mother and Child.

Sources of ascertainment:
Reports are obtained from delivery units, neonatal, pediatric, child surgery, pathology departments and cytogenetic laboratories. Reporting to the central registry occurs via Regional Department of Institute of Health Information and Statistics.

Exposure information:
Some exposure information is available on malformed infants, at present none on controls.

Background information:
Information's on all births are available in the Institute of Health Information and Statistics of the Czech Republic.

Addresses and Staff:
Antonin Sipek, MD, Programme Director Institute for Care of Mother and Child Dept. of Population Teratology & Epidemiology Podolske nabrezi 157 147 10 Prague 4, Czech Republic

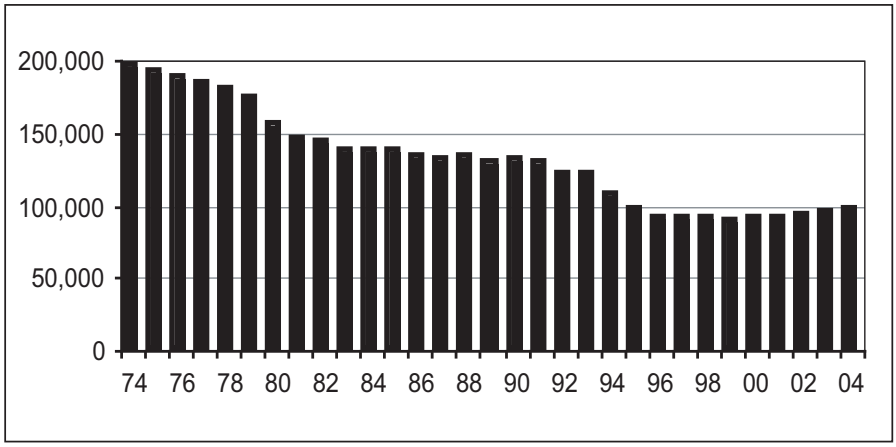
Phone: 420-2-61214341 ext 467

Fax: 420-2-61213851

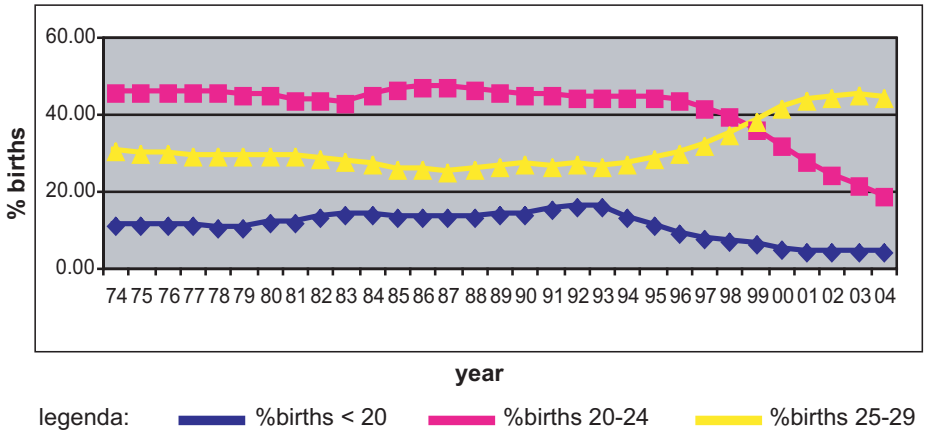
Email: AntoninSipek@seznam.cz

Czech Republic

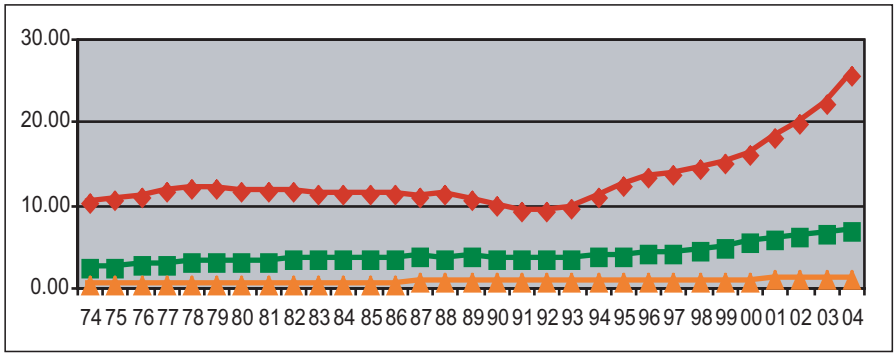
Total births by year



Percentage of births by maternal age



legenda: %births < 20 %births 20-24 %births 25-29



legenda: %births 30-34 %births 35-39 %births 40+

Czech Republic: 2004

Live births (LB)	97,664
Stillbirths (SB)	265
Total births	97,929
Number of terminations of pregnancy (ToP) for birth defects	598

Birth Defects	Number of cases			Rates*10,000
	LB	SB	ToP	Total rate
Anencephaly	2	0	23	2.54
Spina bifida	10	2	27	3.96
Encephalocele	6	0	12	1.83
Microcephaly	10	2	0	1.22
Arhinencephaly / Holoprosencephaly	2	3	6	1.12
Hydrocephaly	20	2	27	4.97
Anophthalmos	0	0	0	0.00
Microphthalmos	0	0	0	0.00
Unspecified Anophthalmos/ Microphthalmos	0	0	0	---
Anotia	4	0	0	0.41
Microtia	8	0	0	0.81
Unspecified Anotia/Microtia	0	0	0	---
Transposition of great vessels	39	0	3	4.26
Tetralogy of Fallot	27	0	5	3.25
Hypoplastic left heart syndrome	13	0	18	3.15
Coarctation of aorta	36	0	1	3.76
Choanal atresia, bilateral	4	1	0	0.51
Cleft palate without cleft lip	64	0	0	6.50
Cleft lip with or without cleft palate	91	0	20	11.27
Oesophageal atresia / stenosis with or without fistula	22	0	0	2.23
Small intestine atresia / stenosis	39	1	0	4.06
Anorectal atresia / stenosis	53	0	0	5.38
Undescended testis (36 weeks of gestation or later)	250	14	0	26.79
Hypospadias	338	0	0	34.31
Epispadias	4	0	0	0.41
Indeterminate sex	4	0	0	0.41
Renal agenesis	69	1	16	8.73
Cystic kidney	57	0	12	7.00
Bladder exstrophy	4	1	0	0.51
Polydactyly, preaxial	153	0	0	15.53
Total Limb reduction defects (include unspecified)	49	0	16	6.60
Transverse	nr	nr	nr	nr
Preaxial	nr	nr	nr	nr
Postaxial	nr	nr	nr	nr
Intercalary	nr	nr	nr	nr
Mixed	nr	nr	nr	nr
Unspecified	nr	nr	nr	---
Diaphragmatic hernia	22	0	9	3.15
Omphalocele	5	1	18	2.44
Gastroschisis	5	1	23	2.94
Unspecified Omphalocele/Gastroschisis	0	0	0	---
Prune belly sequence	2	1	0	0.30
Trisomy 13	7	0	19	2.64
Trisomy 18	6	0	37	4.36
Down syndrome, all ages (include age unknown)	53	1	122	17.86
<20	1	0	1	5.36
20-24	7	0	4	6.08
25-29	20	0	29	11.35
30-34	12	1	32	17.97
35-39	12	0	37	71.45
40-44	1	0	18	168.89
45+	0	0	1	243.90
unknown	0	0	0	---

nr = not reported

Czech Republic: Previous years rates 1974 - 2004

Birth prevalence rates: (LB+SB) * 10,000
Birth prevalence rates: (LB+SB+TOP) * 10,000 in different periods according to the malformation

	1974-79	1980-84	1985-89	1990-94	1995-99	2000-04
Total births	**1,112,785**	**718,080**	**664,018**	**610,039**	**458,693**	**467,116**
Anencephaly	2.99	3.61	2.73	3.66	2.92	2.65
Spina bifida	3.91	4.02	3.73	3.79	4.25	3.96
Encephalocele	0.45	0.82	0.62	1.10	0.87	1.07
Microcephaly	1.04	1.02	1.02	0.75	0.81	1.39
Arhinencephaly / Holoprosencephaly	nr	nr	nr	nr	0.11	0.43
Hydrocephaly	2.31	2.73	3.55	5.25	4.21	5.12
Anophthalmos	nr	nr	nr	nr	0.06*	0.04
Microphthalmos	nr	nr	nr	nr	0.17*	0.30
Unspecified Anophthalmos / Microphthalmos	---	---	---	---	---	---
Anotia	nr	nr	nr	nr	0.89*	0.73
Microtia	nr	nr	nr	nr	0.22*	0.51
Unspecified Anotia / Microtia	---	---	---	---	---	---
Transposition of great vessels	2.75	2.10	1.63	1.60*	2.83	4.32
Tetralogy of Fallot	nr	nr	nr	1.40*	2.96	3.45
Hypoplastic left heart syndrome	0.56	0.70	0.71	1.26*	2.18	2.95
Coarctation of aorta	nr	nr	nr	3.84*	3.62	4.65
Choanal atresia, bilateral	nr	nr	nr	0.19*	0.35	0.24
Cleft palate without cleft lip	5.70	6.71	5.81	5.48	6.47	7.39
Cleft lip with or without cleft palate	9.66	10.29	11.05	10.06	9.61	11.65
Oesophageal atresia / stenosis with or without fistula	1.15	1.24	1.22	1.31	2.31	2.83
Small intestine atresia / stenosis	nr	nr	nr	1.78*	2.14	3.06
Anorectal atresia / stenosis	1.35	1.31	0.63	1.69	2.77	3.72
Undescended testis (36 weeks of gestation or later)	nr	nr	nr	2.81*	10.03	23.55
Hypospadias	18.30	19.89	22.73	23.44	25.94	31.28
Epispadias	nr	nr	nr	0.28*	0.50	0.39
Indeterminate sex	nr	nr	nr	0.37*	0.44	0.49
Renal agenesis	1.62	1.50	1.25	1.84	2.75	6.08
Cystic kidney	2.57	2.41	2.67	2.52	3.64	6.12
Bladder exstrophy	0.16	0.11	0.03	0.00*	0.17	0.21
Polydactyly, preaxial	nr	nr	13.09*	12.21	13.06	14.19
Total Limb reduction defects (include unspecified)	4.34	5.07	4.61	5.66	4.82	6.14
Transverse	nr	nr	nr	nr	nr	nr
Preaxial	nr	nr	nr	nr	nr	nr
Postaxial	nr	nr	nr	nr	nr	nr
Intercalary	nr	nr	nr	nr	nr	nr
Mixed	nr	nr	nr	nr	nr	nr
Unspecified	nr	nr	nr	nr	nr	nr
Diaphragmatic hernia	2.62	2.52	2.27	1.57	2.53	2.83
Omphalocele	2.32	2.14	2.56	2.03	2.55	2.74
Gastroschisis	1.02	1.38	1.17	0.62	3.14	2.85
Unspecified Omphalocele / Gastroschisis	---	---	---	---	---	---
Prune belly sequence	nr	nr	nr	nr	nr	0.16*
Trisomy 13	nr	nr	nr	0.84*	0.92	1.80
Trisomy 18	nr	nr	nr	1.59*	3.03	4.13
Down syndrome, all ages (include age unknown)	8.35	8.15	8.25	10.74	14.32	16.78
<20	4.84	4.49	4.68	4.69	7.79	6.11
20-24	5.46	4.83	3.85	4.21	8.44	7.67
25-29	8.38	7.50	6.95	7.06	10.16	10.65
30-34	11.81	9.67	7.91	12.08	17.76	19.93
35-39	32.61	31.38	27.37	40.58	58.39	61.66
40-44	123.51	99.30	68.99	195.32	204.38	185.31
45+	207.47	360.36	404.04	608.70	454.55	635.84
unspecified	---	---	---	---	---	---

* data include less than 5 years

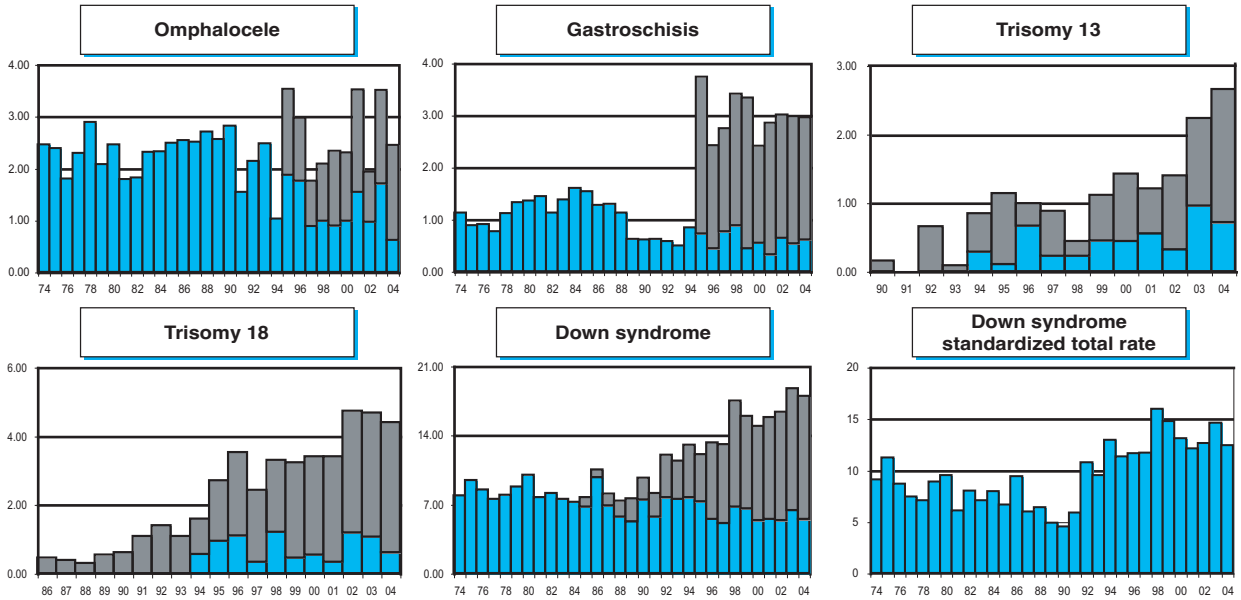
nr = not reported

Czech Republic

'Time trends 1974-2004 (Birth prevalence rates per 10,000)

Note: L+S rates, ToP rates

Note: L+S rates, ToP rates



Note: L+S rates, ToP rates

England and Wales
National Congenital Anomaly System (NCAS)

History:
The monitoring Programme was started in 1964. It was a founding member of the Clearinghouse and is a full member.

Size and coverage:
All births in England and Wales are covered, at present approximately 625,000 annually. Stillbirths of 24 weeks or more gestation are registered.

Legislation and funding:
Reporting is voluntary. The governmental Office for National Statistics finances the National Congenital Anomaly System.

Sources of ascertainment:
Reports are mainly based on notifications of births prepared by attendants at birth, either physicians or midwives by means of a paper form (the SD56 form). This form contains a written description of the anomaly and details of the birth, along with some demographic information about the parents. In areas covered by local congenital anomaly registers this information is supplemented by other reports from neonatal intensive care units, special care baby units etc.

It has long been recognised, however, that there is under reporting in NCAS. Therefore NCAS has embarked on an on-going programme of improving the level of reporting to the system. Since 1998, local congenital anomaly registers have begun to provide data to NCAS in each of the years detailed below:

1998 CARIS (Wales)
1999 East Midlands & South Yorkshire Congenital Anomaly Register
2000 North Thames (West) Congenital Malformation Register
2000 Merseyside and Cheshire Congenital Anomaly Survey
2002 Wessex Antenatally Detected Anomalies Register (WANDA)
2002 Congenital Anomaly Register for Oxfordshire, Berkshire & Buckinghamshire (Oxfordshire only prior to 2004)
2003 Northern Congenital Abnormality Survey
2003 South West Congenital Anomaly Register

In 2004, congenital anomaly notifications are now received for all births in Wales and 45 per cent of births in England. For areas for which NCAS relies solely on SD56 notification forms recording is likely to be less complete.

Reports of terminations of pregnancy have been compiled from notifications of abortions that are completed by the operating practitioners under the 1967 Abortion Act and are sent to the Chief Medical Officers of England and Wales. The tables sent to the International Clearinghouse only include notifications of abortions performed under Grounds E of the Act. An abortion may be performed under Grounds E if 'there is substantial risk that if the child were born it would suffer from such physical or mental abnormalities as to be seriously handicapped'. Since April 2002, the Department of Health has been responsible for the processing of the abortions notification forms and information has been made accessible to the Office for National Statistics (ONS) for statistical purposes.

Exposure information:
Parents' occupation is known.

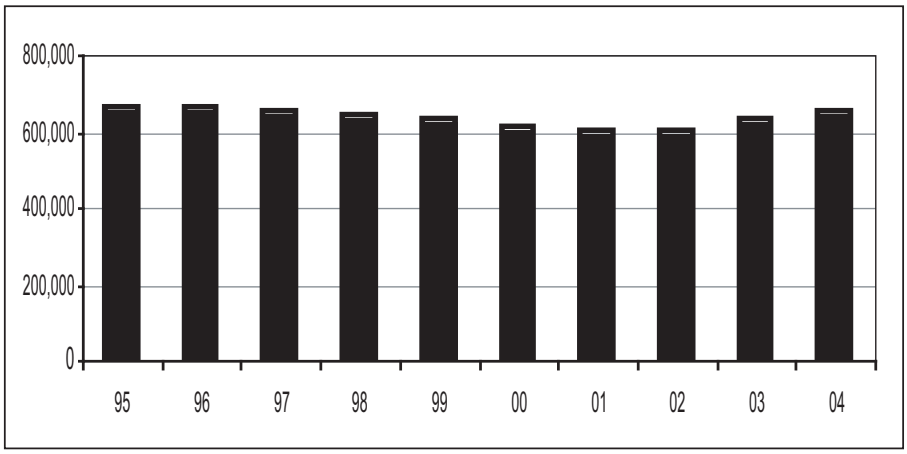
Addresses and Staff:
Vera Ruddock, Office for National Statistics, Room 2.164, Government Buildings, Cardiff Road, Newport, Gwent, NP10 8XG

Phone: 01633 812918

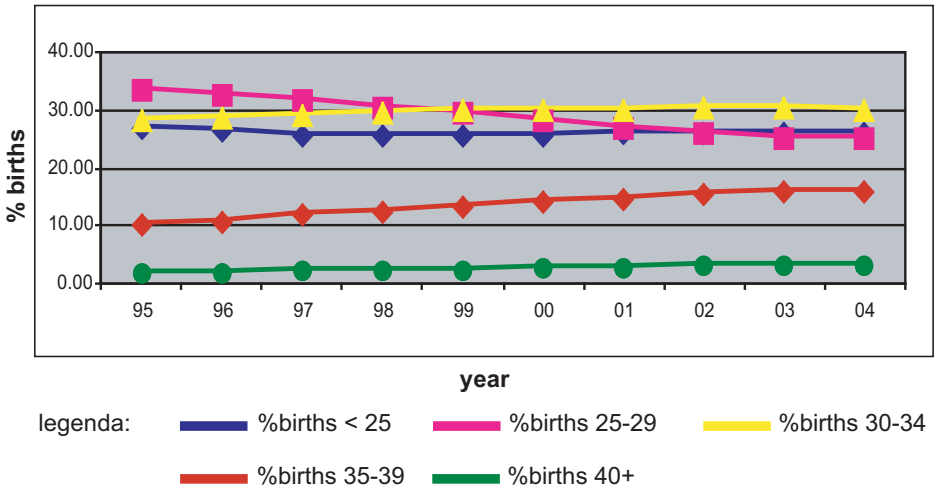
E-mail: ncas@ons.gsi.gov.uk

England and Wales

Total births by year



Percentage of births by maternal age



England and Wales: 2004

Live births (LB)	639,509
Stillbirths (SB)	3670
Total births	643,179
Number of terminations of pregnancy (ToP) for birth defects	1,894

Birth Defects	Number of cases			Rates*10,000
	LB	SB	ToP	Total rate
Anencephaly	12	10	145	2.60
Spina bifida	58	17	90	2.57
Encephalocele	12	2	19	0.51
Microcephaly	37	4	nr	0.64
Arhinencephaly / Holoprosencephaly	8	3	28	0.61
Hydrocephaly	52	20	52	1.93
Anophthalmos	3	1	nr	0.06
Microphthalmos	4	1	nr	0.08
Unspecified Anophthalmos/ Microphthalmos	0	0	nr	---
Anotia	7	0	nr	0.11
Microtia	3	0	nr	0.05
Unspecified Anotia/Microtia	0	0	nr	---
Transposition of great vessels	77	4	nr	1.26
Tetralogy of Fallot	50	5	nr	0.86
Hypoplastic left heart syndrome	32	8	38	1.21
Coarctation of aorta	75	3	nr	1.21
Choanal atresia, bilateral	12	1	nr	0.20
Cleft palate without cleft lip	157	7	nr	2.55
Cleft lip with or without cleft palate	326	9	nr	5.21
Oesophageal atresia / stenosis with or without fistula	61	4	nr	1.01
Small intestine atresia / stenosis	56	8	nr	1.00
Anorectal atresia / stenosis	78	3	nr	1.26
Undescended testis (36 weeks of gestation or later)	10	1	nr	0.17
Hypospadias	410	1	nr	6.39
Epispadias	12	0	nr	0.19
Indeterminate sex	22	4	nr	0.40
Renal agenesis	61	8	24	1.45
Cystic kidney	130	11	19	2.49
Bladder exstrophy	4	0	nr	0.06
Polydactyly, preaxial	32	0	nr	0.50
Total Limb reduction defects (include unspecified)	187	8	nr	3.03
Transverse	70	2	nr	1.12
Preaxial	15	1	nr	0.25
Postaxial	1	0	nr	0.02
Intercalary	54	3	nr	0.89
Mixed	16	2	nr	0.28
Unspecified	31	0	nr	0.48
Diaphragmatic hernia	74	9	18	1.57
Omphalocele	50	7	11	1.06
Gastroschisis	186	5	nr	2.97
Unspecified Omphalocele/Gastroschisis	29	1	nr	0.47
Prune belly sequence	3	0	nr	0.05
Trisomy 13	5	10	54	1.07
Trisomy 18	18	20	135	2.69
Down syndrome, all ages (include age unknown)	420	34	419	13.57
<25	45	5	18	2.99
25-29	51	3	29	3.36
30-34	94	7	94	5.28
35-39	134	11	175	14.11
40+	64	8	103	34.32
unknown	32	0	0	---

nr = not reported
* = Abortion data for 2003 and 2004 are provided on a much reduced scale to conform with the disclosure rules now in place (all conditions with less than 10 cases are suppressed).

England and Wales: Previous years rates 1995 - 2004

Birth prevalence rates: (LB+SB+TOP) * 10,000

	1974-79	1980-84	1985-89	1990-94	1995-99	2000-04
Births					**3,213,870**	**3,071,931**
Anencephaly					2.84	2.89
Spina bifida					2.89	3.03
Encephalocele					0.51	0.53
Microcephaly					0.58	0.78*
Arhinencephaly / Holoprosencephaly					0.48	0.60
Hydrocephaly					2.09	2.11
Anophthalmos					0.10	0.06*
Microphthalmos					0.21	0.27*
Unspecified Anophthalmos / Microphthalmos					---	---
Anotia					0.22	0.16*
Microtia					0.04	0.08*
Unspecified Anotia / Microtia					---	---
Transposition of great vessels					0.61	1.12*
Tetralogy of Fallot					0.71*	1.21*
Hypoplastic left heart syndrome					0.79	1.18
Coarctation of aorta					0.78*	1.26*
Choanal atresia, bilateral					0.11	0.21*
Cleft palate without cleft lip					3.03	3.44*
Cleft lip with or without cleft palate					6.23	6.32*
Oesophageal atresia / stenosis with or without fistula					0.87	1.20*
Small intestine atresia / stenosis					0.70	1.06*
Anorectal atresia / stenosis					1.35	1.57*
Undescended testis (36 weeks of gestation or later)					0.33	0.62*
Hypospadias					8.29*	9.86*
Epispadias					0.30	0.24*
Indeterminate sex					0.72	0.75*
Renal agenesis					1.16	1.77
Cystic kidney					1.96	2.85
Bladder exstrophy					0.18	0.16*
Polydactyly, preaxial					nr	nr
Total Limb reduction defects (include unspecified)					3.09	3.30*
Transverse					nr	nr
Preaxial					nr	nr
Postaxial					nr	nr
Intercalary					nr	nr
Mixed					nr	nr
Unspecified					---	---
Diaphragmatic hernia					1.07	1.59
Omphalocele					0.95	1.23*
Gastroschisis					1.66	2.02*
Unspecified Omphalocele / Gastroschisis					---	---
Prune belly sequence					0.06	0.07*
Trisomy 13					0.73	1.03
Trisomy 18					1.80	2.77
Down syndrome, all ages (include age unknown)					11.23	12.62
<25					4.66	4.24
25-29					5.20	5.17
30-34					9.48	9.95
35-39					29.35	28.99
40+					103.71	88.68
unspecified					---	---

* data include less than 5 years

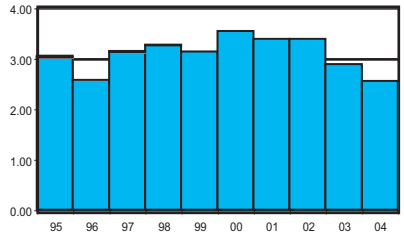
nr = not reported

England and Wales

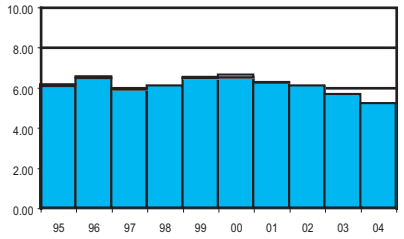
Time trends 1974-2004 (Birth prevalence rates per 10,000)

Note: L+S rates, ToP rates

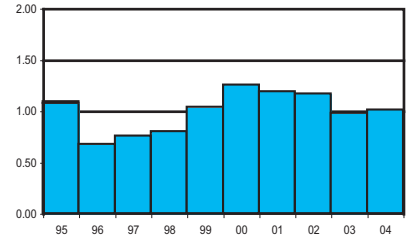
Cleft palate without cleft lip



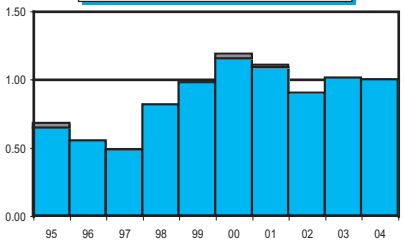
Cleft lip with or without cleft palate



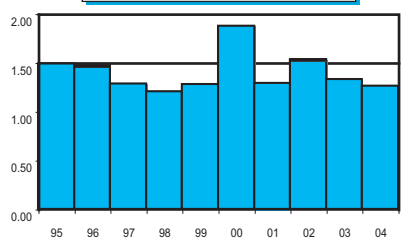
Oesophageal atresia/stenosis with or without fistula



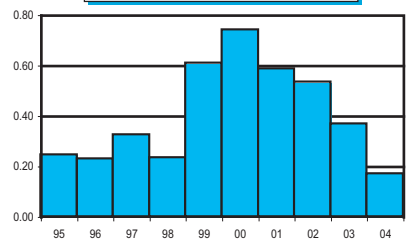
Small intestine atresia/stenosis



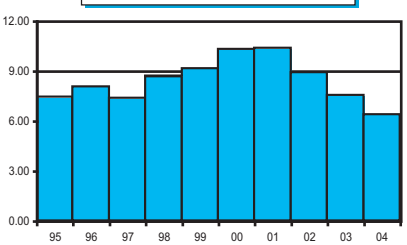
Anorectal atresia/stenosis



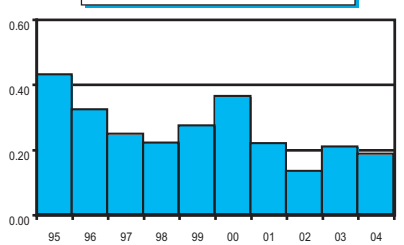
Undescended testis



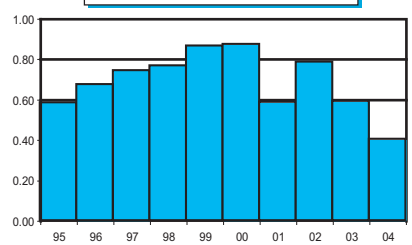
Hypospadias



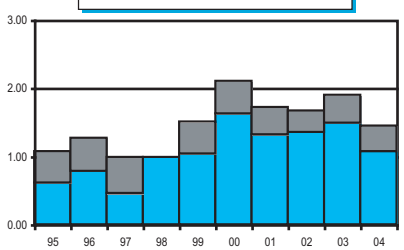
Epispadias



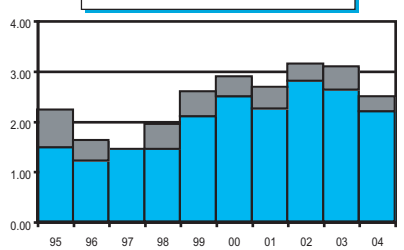
Indeterminate sex



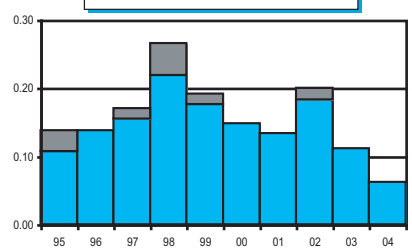
Renal agenesis



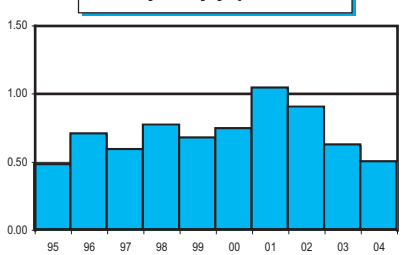
Cystic Kidney



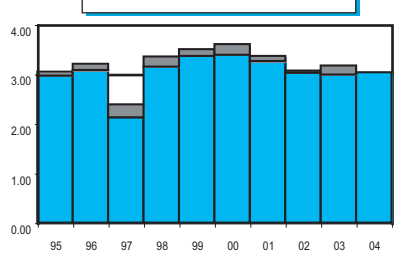
Bladder exstrophy



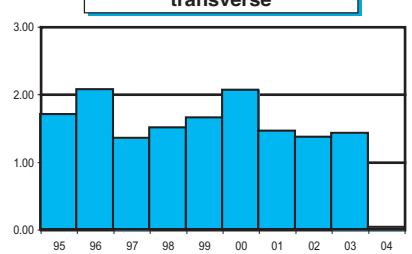
Polydactyly, preaxial



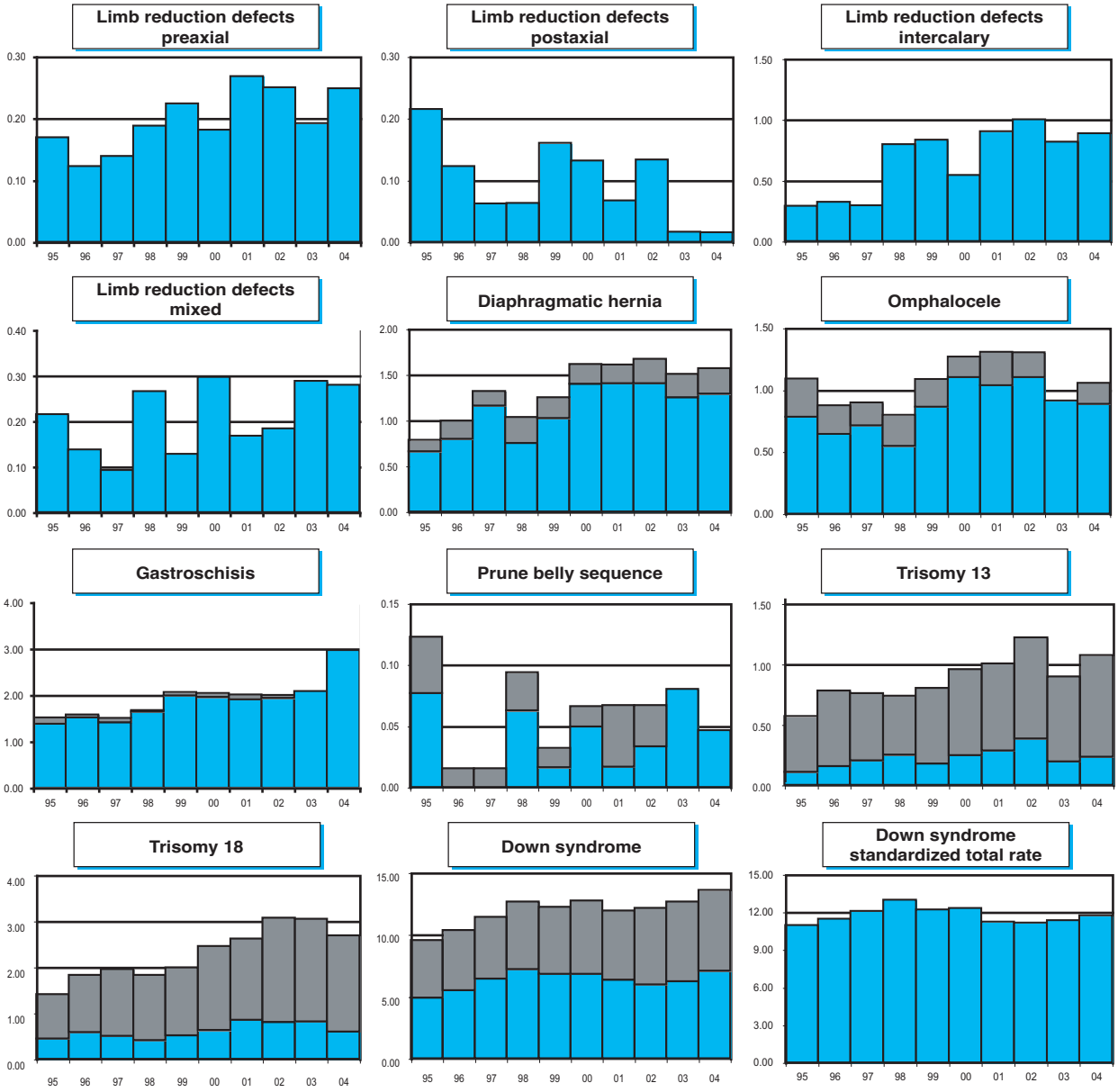
Limb reduction defects



Limb reduction defects transverse



Note: L+S rates, ToP rates



Note: L+S rates, ToP rates

Finland
The Finnish Register of Congenital Malformations

History:
The registry was established in 1963 and regular monitoring started in 1977. It was a founding member of the ICBDSR and is a full member. In 1998 the registry became an associate member of EUROCAT. The registry system (data collection etc.) has been changed twice, in 1985 and in 1993.

Size and coverage
The registry is national and population based. All births in Finland are covered, at present approximately 57,000 annually. Stillbirths of 22 weeks / 500 g or more are registered. Information on malformations is principally collected up to 1 year of age, but later information is also included. Selective terminations of pregnancy and spontaneous abortions with malformations have been included since 1993.

Legislation and funding
Reporting is compulsory. The registry is regulated by the act and statute on the national health care registers with personal data. The registry is run and financed by STAKES, the governmental National Research and Development Centre for Welfare and Health (under the Ministry of Social Affairs and Health).

Sources and ascertainment
Reports are obtained from delivery units, neonatal, paediatric and pathology departments, death certificates and cytogenetic laboratories. Case information is also received from the national Medical Birth Register, Abortion Register and Hospital Discharge Register. The diagnoses of the malformation cases received from other sources are confirmed from the hospitals.

Exposure information
Until 1986, extensive exposure information was obtained from maternity health centres and by personal interviews for cases with selected malformations and their controls. In 1987-1992 only parental occupation was reported. Exposure information, like maternal occupation, medication, X-rays and diseases, etc., has been obtained since 1993. Some exposure information on all births is also available in the Medical Birth Register since 1987.

Background information
Epidemiological background data are available on all births in the Medical Birth Register and in the Statistics Finland.

Address and Staff:
Annukka Ritvanen
The Finnish Register of Congenital Malformations
The National Research and Development Centre for Welfare and Health, STAKES Lintulahdenkuja 4, P.O. Box 220 SF 00531-Helsinki - Finland

Phone: +358-9-39672376

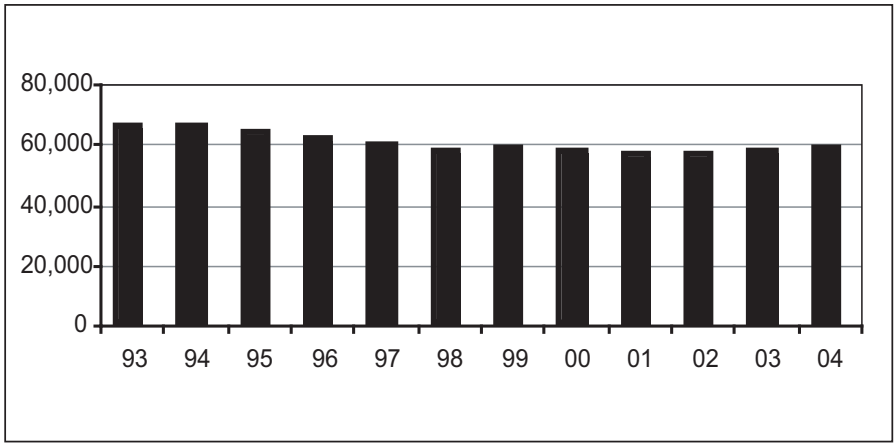
Fax: +358-9-39672459

E-mail: annukka.ritvanen@stakes.fi

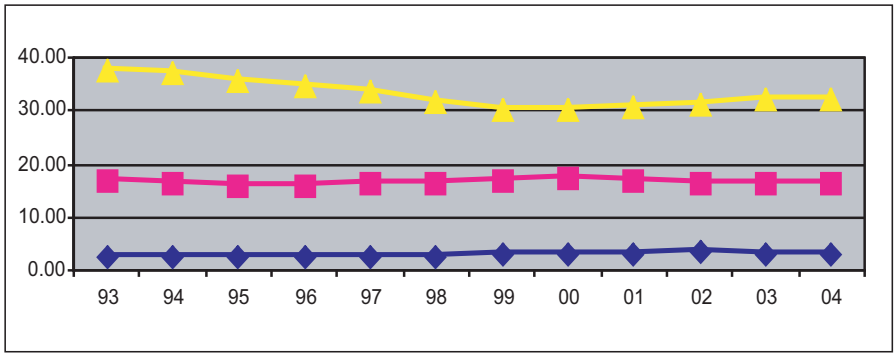
Website: http://www.stakes.fi/

Finland

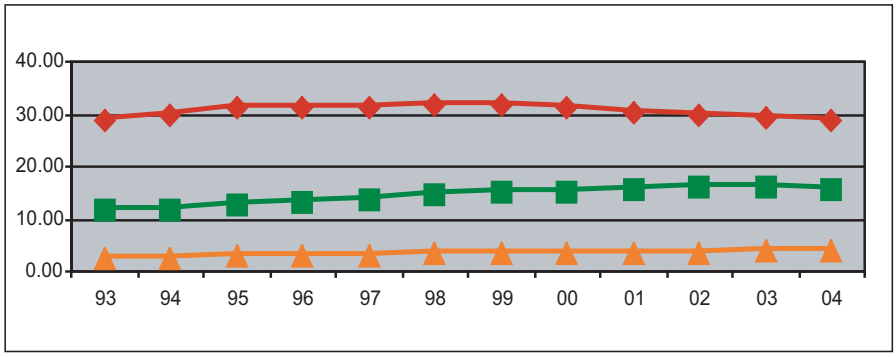
Total births by year



Percentage of births by maternal age



legenda: %births < 20 %births 20-24 %births 25-29



legenda: %births 30-34 %births 35-39 %births 40+

Finland: 2004

Live births (LB)	57,758
Stillbirths (SB)	187
Total births	57,945
Number of terminations of pregnancy (ToP) for birth defects	286

Birth Defects	Number of cases			Rates*10,000
	LB	SB	ToP	Total rate
Anencephaly	1	0	9	1.72
Spina bifida	12	1	8	3.61
Encephalocele	3	0	5	1.37
Microcephaly	5	1	0	1.04
Arhinencephaly / Holoprosencephaly	3	1	9	2.23
Hydrocephaly	9	1	9	3.28
Anophthalmos	4	0	1	0.86
Microphthalmos	3	0	1	0.69
Unspecified Anophthalmos / Microphthalmos	0	0	0	---
Anotia nr	nr	nr	nr	
Microtia	nr	nr	nr	nr
Unspecified Anotia / Microtia	nr	nr	nr	---
Transposition of great vessels	22	1	0	3.95
Tetralogy of Fallot	22	0	1	3.95
Hypoplastic left heart syndrome	22	0	2	4.12
Coarctation of aorta	53	2	3	10.01
Choanal atresia, bilateral	8	0	1	1.55
Cleft palate without cleft lip	80	0	5	14.67
Cleft lip with or without cleft palate	45	2	9	9.62
Oesophageal atresia / stenosis with or without fistula	23	0	0	3.95
Small intestine atresia / stenosis	6	3	0	1.55
Anorectal atresia / stenosis	28	1	1	5.18
Undescended testis (36 weeks of gestation or later)	nr	nr	nr	nr
Hypospadias	73	0	0	12.54
Hypospadias severe	15	0	0	2.58
Epispadias	2	0	0	0.34
Indeterminate sex	8	0	0	1.37
Renal agenesis	2	1	6	1.55
Cystic kidney	29	1	12	7.21
Bladder exstrophy	2	0	0	0.35
Polydactyly, preaxial	19	1	1	3.61
Total Limb reduction defects (include unspecified)	38	3	6	8.07
Transverse	23	1	2	4.46
Preaxial	8	1	0	1.55
Postaxial	1	0	0	0.17
Intercalary	1	1	1	0.52
Mixed	1	0	1	0.34
Unspecified	4	0	2	---
Diaphragmatic hernia	11	1	4	2.75
Omphalocele	10	3	18	5.32
Gastroschisis	9	0	6	2.58
Unspecified Omphalocele/Gastroschisis	0	1	1	---
Prune belly sequence	1	0	0	0.17
Trisomy 13	3	0	11	2.40
Trisomy 18	5	4	27	6.18
Down syndrome, all ages (include age unknown)	67	4	92	27.99
<20	1	0	0	6.01
20-24	1	0	7	8.46
25-29	9	1	13	12.28
30-34	12	1	12	14.87
35-39	24	1	34	64.36
40-44	18	1	25	209.32
45+	2	0	1	275.23
unknown	0	0	0	---

nr = not reported

Finland: Previous years rates 1993 - 2004

Prevalence rates: (LB+SB+TOP) * 10,000

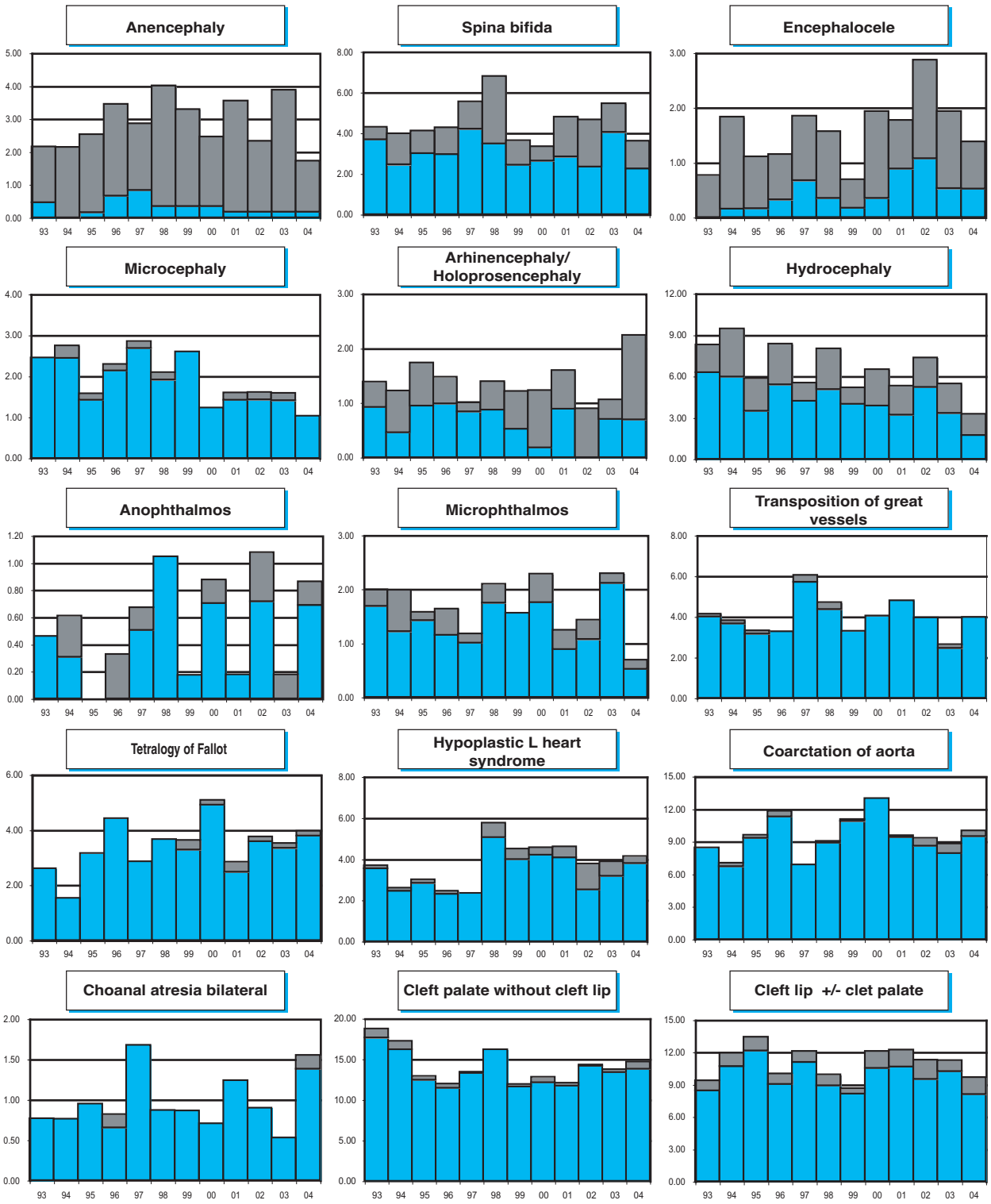
	1974-79	1980-84	1985-89	1990-94*	1995-99	2000-04
Births				**130,578**	**299,029**	**283,887**
Anencephaly				2.14	3.21	2.78
Spina bifida				4.14	4.85	4.37
Encephalocele				1.30	1.27	1.97
Microcephaly				2.60	2.27	1.41
Arhinencephaly / Holoprosencephaly				1.30	1.37	1.41
Hydrocephaly				8.88	6.59	5.57
Anophthalmos				0.54	0.43	0.63
Microphthalmos				1.99	1.61	1.59
Unspecified Anophthalmos / Microphthalmos				---	---	---
Anotia				nr	nr	nr
Microtia				nr	nr	nr
Unspecified Anotia / Microtia				---	---	---
Transposition of great vessels				3.98	4.11	3.87
Tetralogy of Fallot				2.07	3.54	3.84
Hypoplastic left heart syndrome				3.14	3.58	4.19
Coarctation of aorta				7.73	9.70	10.14
Choanal atresia, bilateral				0.77	1.04	0.99
Cleft palate without cleft lip				18.00	13.28	13.53
Cleft lip with or without cleft palate				10.64	10.87	11.31
Oesophageal atresia / stenosis with or without fistula				3.14	3.71	3.91
Small intestine atresia / stenosis				1.30	1.07	1.20
Anorectal atresia / stenosis				6.13	5.05	4.97
Undescended testis (36 weeks of gestation or later)				nr	nr	nr
Hypospadias				3.37	3.18	3.31
Epispadias				0.23	0.27	0.42
Indeterminate sex				0.84	1.04	1.90
Renal agenesis				1.99	1.74	1.44
Cystic kidney				6.51	5.95	7.68
Bladder exstrophy				0.54	0.47	0.70
Polydactyly, preaxial				4.90	4.18	4.05
Total Limb reduction defects (include unspecified)				7.73	6.76	7.64
Transverse				4.82	3.85	3.77
Preaxial				1.76	1.74	2.54
Postaxial				0.38	0.30	0.39
Intercalary				0.23	0.43	0.32
Mixed				0.31	0.37	0.18
Unspecified				---	---	---
Diaphragmatic hernia				2.53	2.54	2.71
Omphalocele				4.06	3.98	5.07
Gastroschisis				1.76	2.34	3.24
Unspecified Omphalocele / Gastroschisis				---	---	---
Prune belly sequence				0.31	0.27	0.25
Trisomy 13				2.14	2.14	2.11
Trisomy 18				4.98	5.72	6.69
Down syndrome, all ages (include age unknown)				23.28	22.57	25.50
<20				21.35	9.14	7.10
20-24				6.46	7.04	7.43
25-29				11.71	10.09	10.80
30-34				19.03	17.48	15.79
35-39				61.46	52.19	57.27
40-44				163.30	170.83	186.10
45+				465.12	210.53	401.53
unknown				---	---	---

* data include less than 5 years

nr = not reported

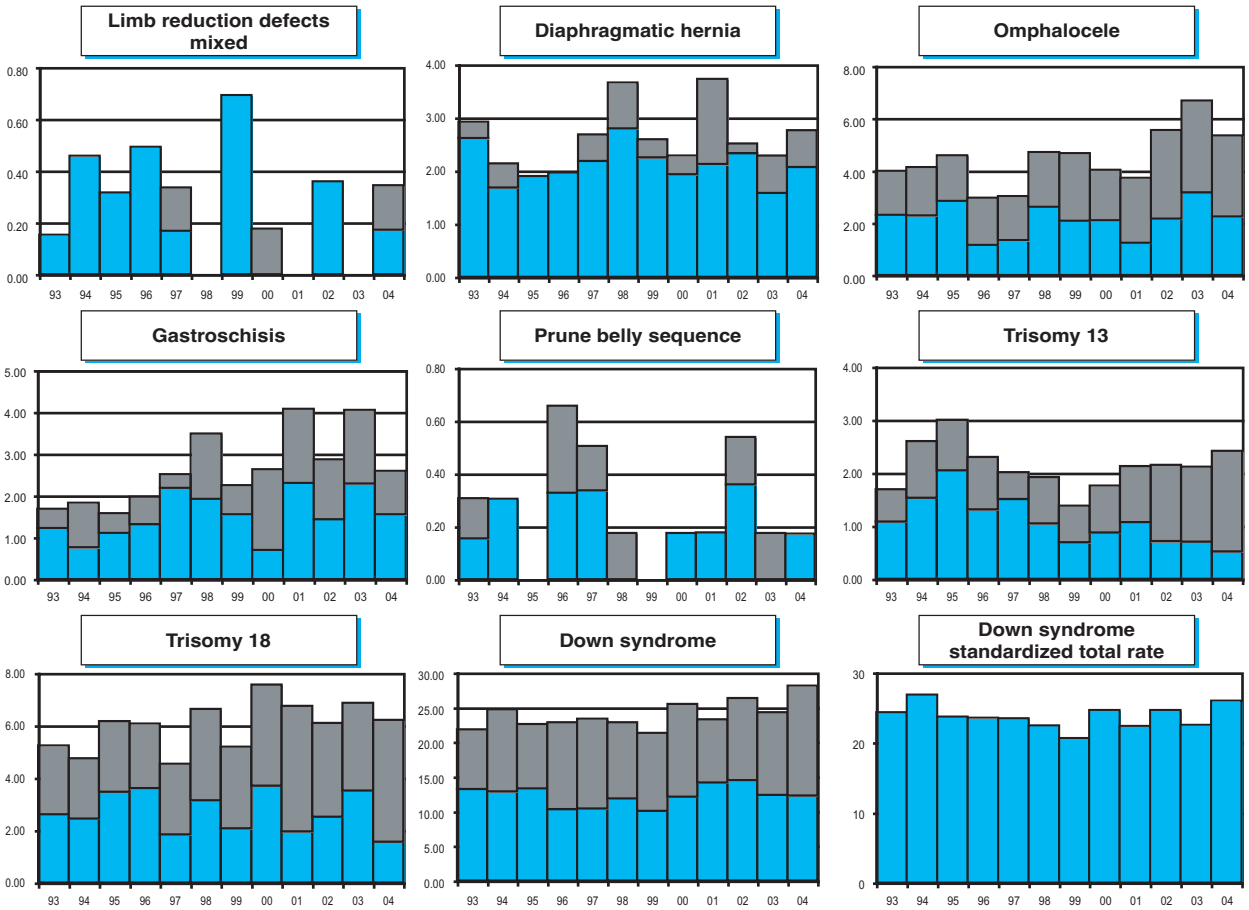
Finland

'Time trends 1974-2004 (Birth prevalence rates per 10,000)



Note: L+S rates, ToP rates

Note: L+S rates, ToP rates



Note: L+S rates, ToP rates

France: Central-East

Central-East France Register of Congenital Malformations.

History:
The registry began in 1973 within the Rhone-Alps area -the Auvergne region was added in 1983, the Jura area in 1985, the Côte d'Or & Nièvre in 1989 and Saône-et-Loire in 1990. The Programme was a founding member of the ICBDSR and is a full member. In 1998 the registry was split up and the Auvergne region, became financially independent, under the responsibility of Christine Francannet. The collaboration between Auvergne and the rest of the FCE-registry is maintained and common results are published.

Size and coverage:
The registry covers all births in the area approximately 100,000 births annually, which represents about 13% of all births in France. Stillbirths of 22 weeks or more gestation are included.

Legislation and funding:
It has multiple sources of funding: 33 % comes from an insurance company (GROUPAMA), 13% from the Ministry of Health, 5% from local health authorities, 10% from specific epi studies, 13 % from the French National Electricity Agency, and 26 % from contracts with drug companies for the surveillance of their products with respect to potential teratogenic effects.

Sources of ascertainment:
The registry is population based and covers Rhône-Alpes region, plus 4 French departments: Côte d'Or, Jura, Nièvre, Saône-et-Loire. Since 2000, Auvergne still uses the same database, but the registry is separated from an administrative point of view, and sends its own data to Eurocat separately. The approximate annual number of births covered is 90,000. Data collection is actively performed in large maternity wards and pediatric units but passive, on a voluntary basis, in small maternity and pediatric units. Other sources of information include cytogenetic laboratories, pathology laboratories, departments of medical genetics, birth certificates.
The maximum age at postnatal diagnosis is 1 year. For children born in year x, notifications are taken into account until March x+2. We have no follow-up procedure. Are excluded from registration: balanced chromosomal anomalies, pyloric stenosis, metabolic disorders, minor malformations (small angiomas or naevi, hip subdislocation, small foot deformities, ill-defined facial anomalies, inguinal and umbilical hernias).
We register terminations since year 1985. Our official stillbirth definition is 22 w (28 w before 1997), which is our lower gestational age limit to include early fetal deaths/spontaneous abortions.

Exposure information:
Our exposure data include drug intake in 1 st trimester of pregnancy, medically assisted procreation, occupation. Denominators information is obtained from National institute of Statistics. We collect no controls.

Background information:
Some background information is available from the general population statistics.

Addresses and Staff:
Emmanuelle Amar, Programme Director Registre Des Malformations en Rhone Alpes Faculté Laennec 7-9 rue Guillaume Paradin 69372 LYON - FRANCE

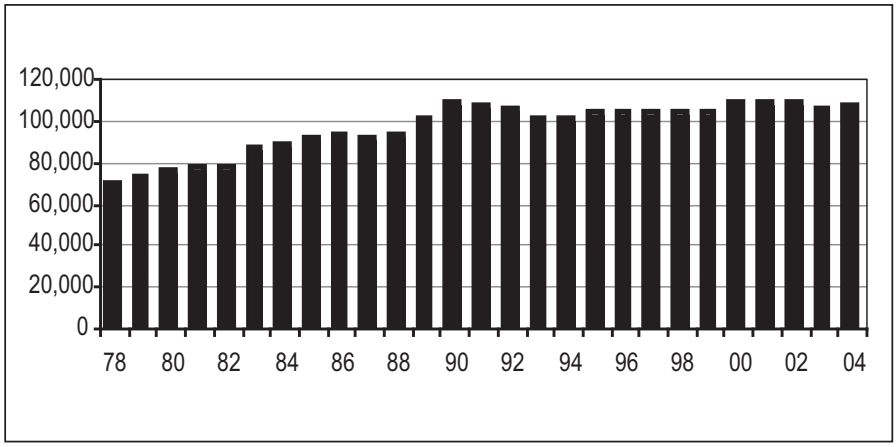
E-mail: emmanuelle.amar@orange.fr

Elisabeth Robert

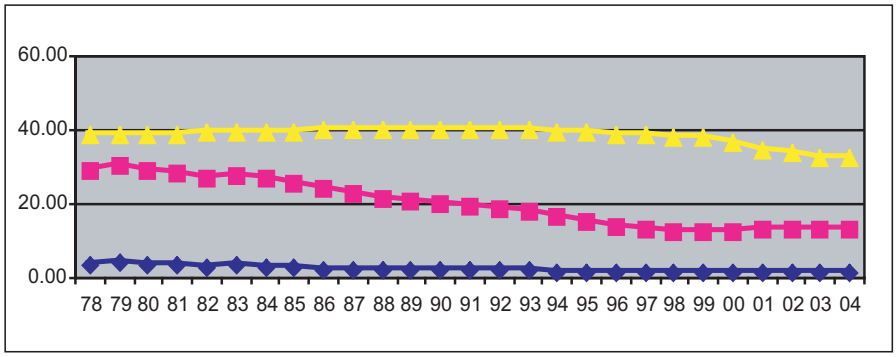
E-mail: elisabeth.robert-gnansia@afsset.fr

France: Central-East

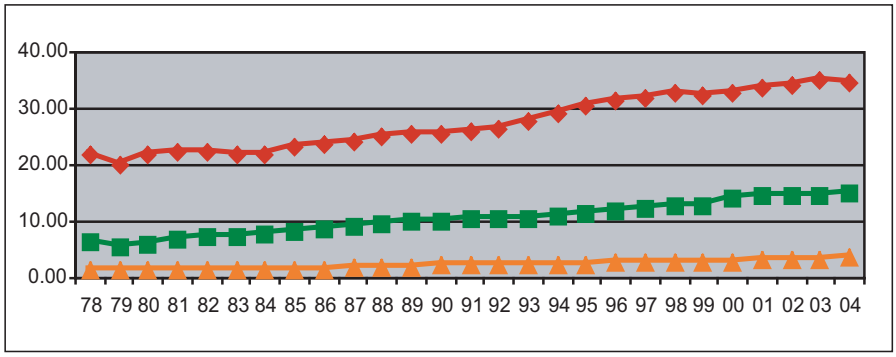
Total births by year



Percentage of births by maternal age



legenda: %births < 20 %births 20-24 %births 25-29



legenda: %births 30-34 %births 35-39 %births 40+

France: Central East, 2004

Live births (LB)	104,853
Stillbirths (SB)	878
Total births	105,731
Number of terminations of pregnancy (ToP) for birth defects	569

Birth Defects	Number of cases			Rates*10,000
	LB	SB	ToP	Total rate
Anencephaly	0	0	16	1.51
Spina bifida	11	1	29	3.86
Encephalocele	1	0	13	1.32
Microcephaly	13	0	15	2.63
Arhinencephaly / Holoprosencephaly	1	0	19	1.88
Hydrocephaly	25	2	38	6.11
Anophthalmos	1	1	1	0.28
Microphthalmos	2	0	4	0.56
Unspecified Anophthalmos/ Microphthalmos	0	0	0	---
Anotia	1	0	2	0.28
Microtia	5	0	3	0.75
Unspecified Anotia/Microtia	0	0	0	---
Transposition of great vessels	37	1	6	4.14
Tetralogy of Fallot	36	0	4	3.76
Hypoplastic left heart syndrome	6	2	15	2.16
Coarctation of aorta	41	0	2	4.05
Choanal atresia, bilateral	10	1	2	1.22
Cleft palate without cleft lip	42	1	3	4.33
Cleft lip with or without cleft palate	66	0	13	7.43
Oesophageal atresia / stenosis with or without fistula	19	0	1	1.88
Small intestine atresia / stenosis	28	1	5	3.20
Anorectal atresia / stenosis	33	3	4	3.76
Undescended testis (36 weeks of gestation or later)	nr	nr	nr	nr
Hypospadias	125	2	3	12.23
Epispadias	3	0	0	0.28
Indeterminate sex	5	1	3	0.85
Renal agenesis	2	1	11	1.32
Cystic kidney	34	0	11	4.23
Bladder exstrophy	2	1	2	0.47
Polydactyly, preaxial	14	0	1	1.41
Total Limb reduction defects (include unspecified)	36	2	18	5.27
Transverse	25	1	5	2.92
Preaxial	5	1	6	1.13
Postaxial	3	0	2	0.47
Intercalary	1	0	2	0.28
Mixed	2	0	3	0.47
Unspecified	0	0	0	---
Diaphragmatic hernia	30	1	11	3.95
Omphalocele	12	1	18	2.92
Gastroschisis	10	0	3	1.22
Unspecified Omphalocele/Gastroschisis	0	0	0	---
Prune belly sequence	0	0	2	0.19
Trisomy 13	1	0	19	1.88
Trisomy 18	5	1	50	5.27
Down syndrome, all ages (include age unknown)	60	2	180	22.77
<20	0	0	0	0.00
20-24	3	0	5	5.78
25-29	7	1	15	6.78
30-34	12	1	33	12.53
35-39	13	0	77	55.77
40-44	7	0	44	145.38
45+	1	0	1	122.70
unknown	17	0	5	---

nr = not reported

France: Central East, Previous years rates 1978 - 2004

Birth prevalence rates: (LB+SB) * 10,000 until 1993
Birth prevalence rates: (LB+SB+TOP) * 10,000 from 1994

	1974-79*	1980-84	1985-89	1990-94	1995-99	2000-04
Total births	**140,274**	**400,702**	**464,127**	**517,380**	**513,148**	**533,361**
Anencephaly	0.78	1.00	0.50	0.66	1.54	1.69
Spina bifida	4.56	3.54	2.48	2.84	3.66	3.75
Encephalocele	0.57	0.72	0.80	1.41	1.56	1.72
Microcephaly	1.35	2.00	2.48	1.84	1.79	2.19
Arhinencephaly / Holoprosencephaly	0.50	0.30	0.71	1.39	1.46	1.22
Hydrocephaly	1.64	2.45	3.34	2.90	5.01	6.26
Anophthalmos	0.21	0.17	0.17	0.21	0.18	0.17
Microphthalmos	1.14	0.80	1.31	1.04	1.33	0.86
Unspecified Anophthalmos / Microphthalmos	---	---	---	---	---	---
Anotia	0.14	0.32	0.45	0.31	0.45	0.41
Microtia	0.21	0.20	0.30	0.29	0.58	0.43
Unspecified Anotia / Microtia	---	---	---	---	---	---
Transposition of great vessels	3.14	3.09	3.47	3.25	3.29	3.22
Tetralogy of Fallot	1.57	2.35	2.39	2.20	2.30	2.57
Hypoplastic left heart syndrome	0.93	2.05	2.35	1.86	2.92	2.68
Coarctation of aorta	1.92	2.55	2.87	2.59	2.65	2.14
Choanal atresia, bilateral	0.71	0.65	0.86	0.62	0.80	1.09
Cleft palate without cleft lip	4.35	4.77	4.80	5.76	6.90	5.06
Cleft lip with or without cleft palate	7.06	6.86	5.69	7.29	8.54	6.71
Oesophageal atresia / stenosis with or without fistula	2.00	2.47	2.59	2.96	3.33	2.55
Small intestine atresia / stenosis	1.78	1.17	2.09	1.89	2.47	2.85
Anorectal atresia / stenosis	2.42	2.57	3.21	3.23	4.11	3.56
Undescended testis (36 weeks of gestation or later)	nr	nr	nr	nr	nr	nr
Hypospadias	5.87	6.11	9.22	9.55	12.02	12.04
Epispadias	0.29	0.12	0.26	0.21	0.23	0.19
Indeterminate sex	0.71	0.67	0.80	0.75	0.60	0.56
Renal agenesis	0.43	0.70	0.67	0.64	1.48	1.41
Cystic kidney	0.29	1.22	2.28	3.40	4.54	4.52
Bladder exstrophy	0.29	0.07	0.45	0.35	0.35	0.28
Polydactyly, preaxial	0.78	0.85	1.29	1.74	2.40	1.52
Total Limb reduction defects (include unspecified)	3.78	4.64	4.27	4.29	5.34	4.61
Transverse	2.21	2.15	2.48	2.30	2.63	2.31
Preaxial	0.43	0.77	0.71	0.52	0.78	1.09
Postaxial	0.36	0.30	0.37	0.39	0.33	0.49
Intercalary	0.29	0.65	0.32	0.52	0.43	0.34
Mixed	0.36	0.65	0.37	0.25	0.35	0.32
Unspecified	---	---	---	---	---	---
Diaphragmatic hernia	1.92	2.75	2.54	2.55	3.10	2.70
Omphalocele	0.93	1.20	1.16	1.24	2.34	2.51
Gastroschisis	0.36	0.77	0.88	1.18	1.29	1.39
Unspecified Omphalocele / Gastroschisis	---	---	---	---	---	---
Prune belly sequence	0.29	0.15	0.28	0.48	0.57	0.15
Trisomy 13	0.29	0.57	0.88	1.18	1.58	1.97
Trisomy 18	0.86	0.97	1.96	2.47	4.11	4.37
Down syndrome, all ages (include age unknown)	11.26	11.40	11.16	12.93	19.62	21.94
<20	7.76	4.72	5.68	6.10	10.98	4.78
20-24	6.52	7.48	4.93	6.49	7.54	7.56
25-29	4.78	5.27	7.47	5.79	7.83	8.43
30-34	11.60568	11.08497	9.493221	8.91	14.12	13.10
35-39	23.13969	31.34683	24.18351	27.94	45.30	46.50
40-44	131.1085	67.63898	55.19655	76.75	151.21	143.79
45+	119.0476	94.33962	158.371	243.90	248.06	229.11
unknown	---	---	---	---	---	---

* data include less than 5 years

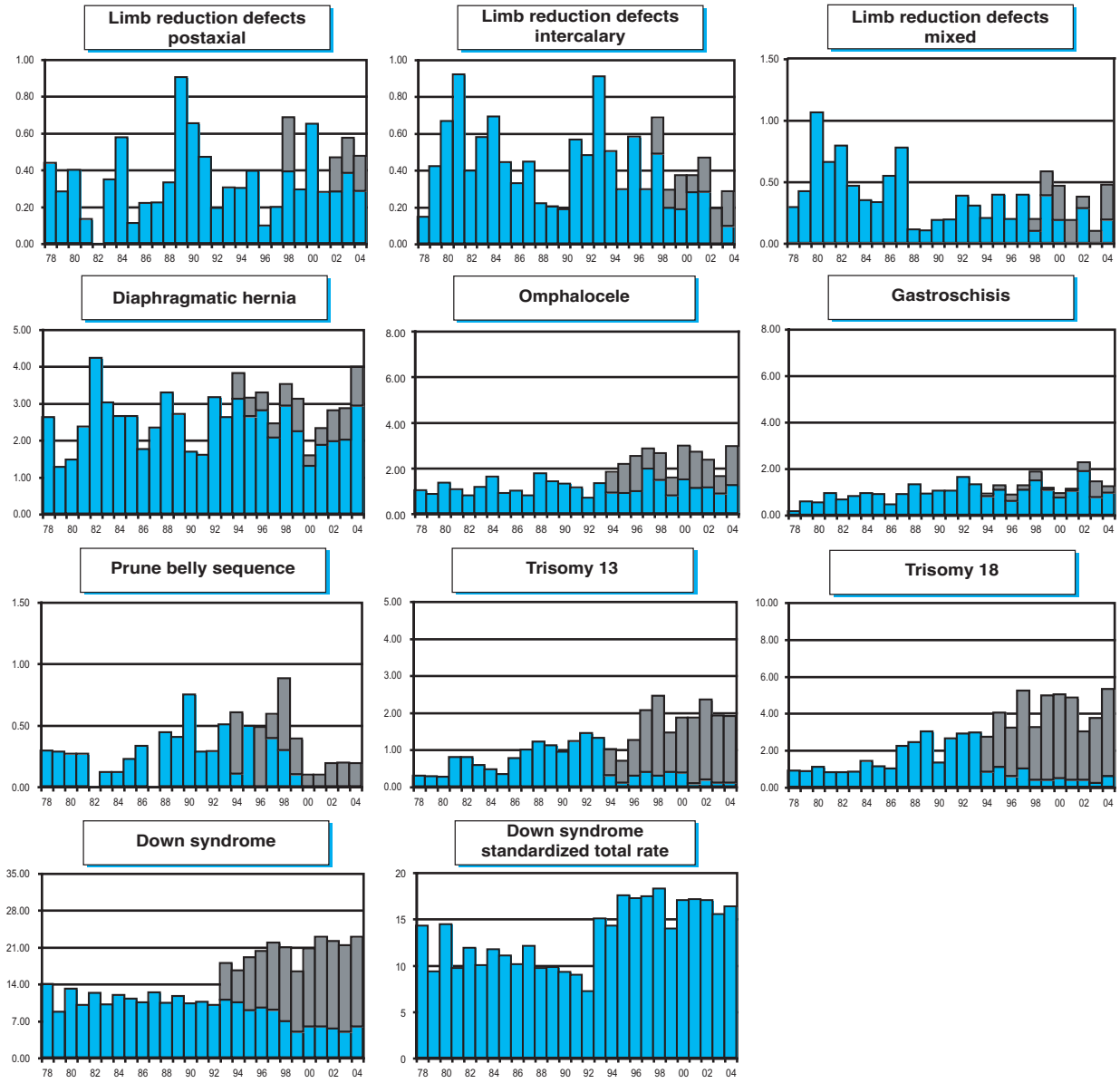
nr = not reported

France: Central-East

'Time trends 1978-2004 (Birth prevalence rates per 10,000)

Note: L+S rates, ToP rates

Note: L+S rates, ToP rates



Note: L+S rates, ToP rates

France: Paris

History:
The Programme was initiated in 1975, but the registry really started in 1981. It became an associate member of the Clearinghouse in 1982. It is also a member of EUROCAT.

Size and coverage:
The registry covers 38.000 annual births (about 5% of all births in France), that is all births (live and still births of 22 weeks or more) and terminations of pregnancy in the population of Greater Paris delivering in Paris maternity units. The estimation of the coverage of the registry is around 95%.

Legislation and funding:
Reporting is volontary. The registry is part of a research unit of INSERM (National Institute of Health and Medical Research). The registry has been officially recognized by the French National Comity of Registries, and is renewed for four years (2001-2004) and supported by an annual grant from INSERM and Institut de la Veille Sanitaire (Institute for Health Surveillance).

Sources of ascertainment:
Reports are actively collected from delivery units, pediatric departments, cytogenetic laboratories, pathology departments. Terminations of pregnancy are included. Case information is also received from the health certificates of the first week.

Exposure information:
Information on maternal drug use, maternal and paternal diseases and occupations, outcome of previous pregnancies, is available for the malformed cases. Data about techniques of prenatal screening (ultrasound, serum markers) and prenatal diagnosis are systematically collected.

Background information:
Background data on births are available from the National Institute of Statistics (INSEE).

Addresses and Staff:
Catherine De Vigan, MD, Programme Director
INSERM U149 16 av P Vaillant-Couturier 94807 Villejuif Cedex, France.

Phone: 33-1-45 59 50 09

Fax: 33-1-45 59 50 89

E-mail: devigan@vjf.inserm.fr

Ingrid Lemaire, statistician

Phone: 33-01- 45 59 50 09

Fax: 33-01-43268979

E-mail: regmalf@vjf.inserm.fr

Vèronique Vodovar, pediatric nurse, technical assistance

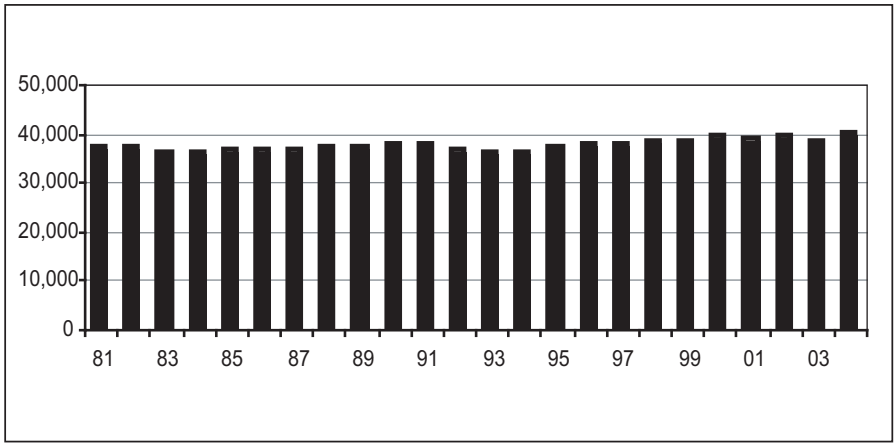
Phone: 33-01-45 59 50 09

Fax: 33-01-43268979

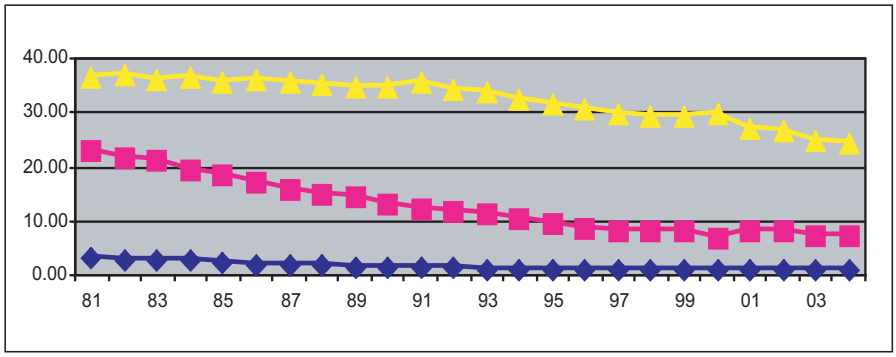
E-mail: regmalf@vjf.inserm.fr

France: Paris

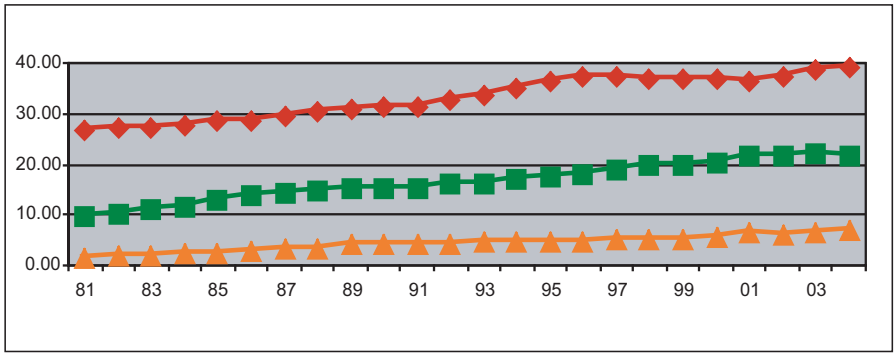
Total births by year



Percentage of births by maternal age



legenda: %births < 20 %births 20-24 %births 25-29



legenda: %births 30-34 %births 35-39 %births 40+

France: Paris, 2004

Live births (LB)	39,029
Stillbirths (SB)	503
Total births	39,532
Number of terminations of pregnancy (ToP) for birth defects	518

Birth Defects	Number of cases			Rates*10,000
	LB	SB	ToP	Total rate
Anencephaly	0	0	22	5.49
Spina bifida	6	0	10	4.00
Encephalocele	1	0	7	2.00
Microcephaly	5	0	11	4.00
Arhinencephaly / Holoprosencephaly	2	0	17	4.74
Hydrocephaly	22	0	26	11.99
Anophthalmos	1	0	1	0.50
Microphthalmos	2	0	3	1.25
Unspecified Anophthalmos/ Microphthalmos	0	0	0	---
Anotia	4	0	1	1.25
Microtia	0	0	2	0.50
Unspecified Anotia/Microtia	0	0	0	---
Transposition of great vessels	25	0	1	6.49
Tetralogy of Fallot	11	0	11	5.49
Hypoplastic left heart syndrome	3	1	15	4.74
Coarctation of aorta	10	0	2	3.00
Choanal atresia, bilateral	1	0	1	0.50
Cleft palate without cleft lip	12	1	6	4.74
Cleft lip with or without cleft palate	20	0	11	7.74
Oesophageal atresia / stenosis with or without fistula	12	0	5	4.24
Small intestine atresia / stenosis	13	0	3	4.00
Anorectal atresia / stenosis	7	0	10	4.24
Undescended testis (36 weeks of gestation or later)	nr	nr	nr	nr
Hypospadias	68	0	3	17.73
Epispadias	2	0	0	0.50
Indeterminate sex	2	0	2	1.00
Renal agenesis	1	0	10	2.75
Cystic kidney	23	0	17	9.99
Bladder exstrophy	1	0	1	0.50
Polydactyly, preaxial	6	0	2	2.00
Total Limb reduction defects (include unspecified)	10	0	22	7.99
Transverse	7	0	7	3.50
Preaxial	0	0	5	1.25
Postaxial	1	0	3	1.00
Intercalary	0	0	1	0.25
Mixed	2	0	3	1.25
Unspecified	0	0	3	---
Diaphragmatic hernia	15	0	4	4.74
Omphalocele	6	3	15	5.99
Gastroschisis	10	0	3	3.25
Unspecified Omphalocele/Gastroschisis	1	0	3	---
Prune belly sequence	1	0	0	0.25
Trisomy 13	2	0	16	4.49
Trisomy 18	6	4	50	14.98
Down syndrome, all ages (include age unknown)	20	1	141	40.45
<20	0	0	1	32.79
20-24	3	0	6	32.27
25-29	2	0	14	16.52
30-34	7	1	28	23.32
35-39	4	0	40	50.76
40-44	4	0	43	180.98
45+	0	0	9	473.68
uknown	0	0	0	---

nr = not reported

France: Paris, Previous years rates 1981 - 2004

Prevalence rates: (LB+SB) * 10,000 until 1993
Prevalence rates: (LB+SB+TOP) * 10,000 since 1994

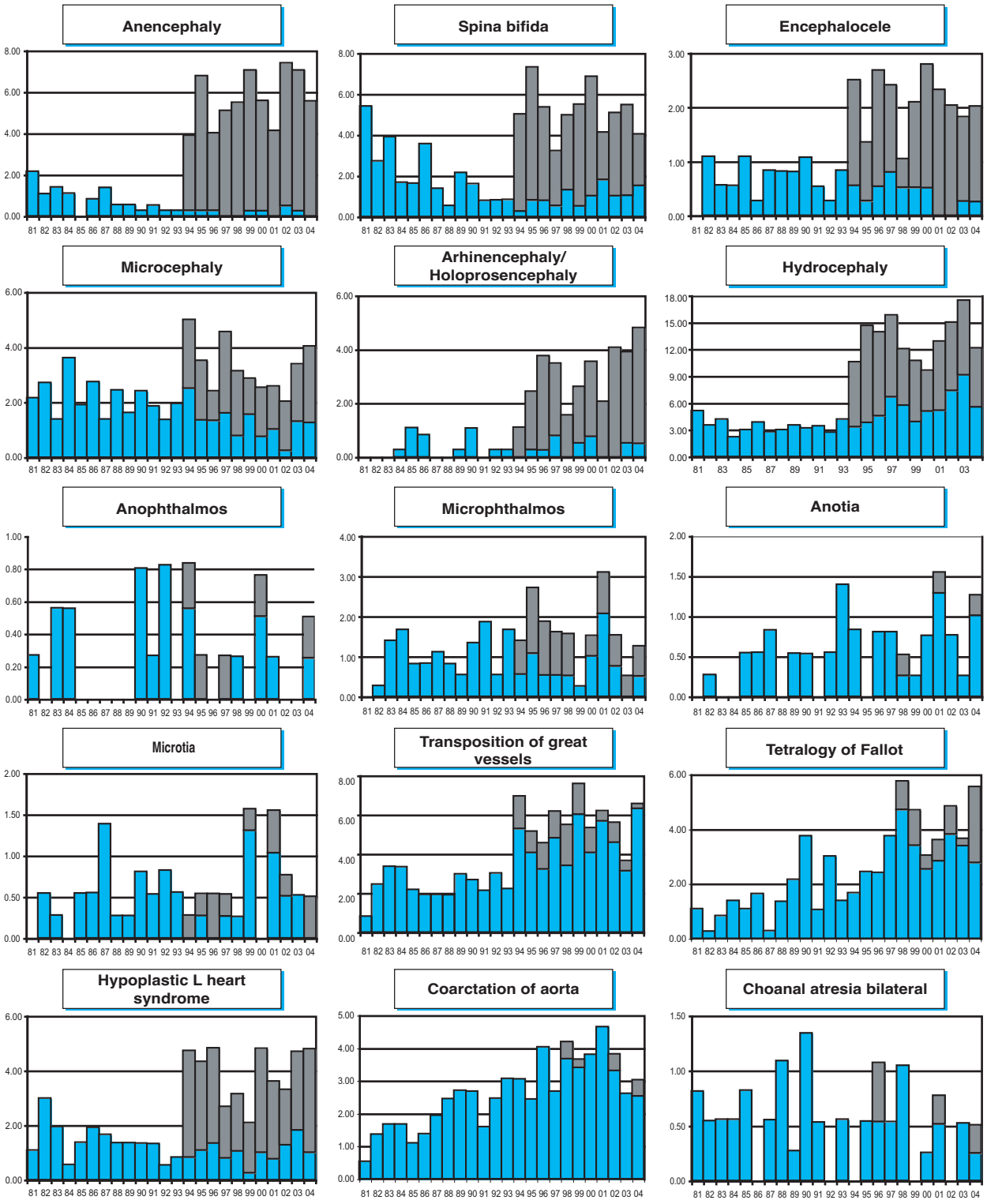
	1974-79	1980-84*	1985-89	1990-94	1995-99	2000-04
Births		**145,343**	**182,538**	**183,049**	**187,753**	**195,150**
Anencephaly		1.44	0.66	1.04	5.70	5.94
Spina bifida		3.44	1.86	1.80	5.27	5.12
Encephalocele		0.55	0.77	1.04	1.92	2.20
Microcephaly		2.48	2.03	2.51	3.30	2.92
Arhinencephaly / Holoprosencephaly		0.07	0.44	0.55	2.77	3.69
Hydrocephaly		3.78	3.23	4.81	13.42	13.43
Anophthalmos		0.34	0.00	0.55	0.16	0.31
Microphthalmos		0.83	0.82	1.37	1.60	1.59
Unspecified Anophthalmos / Microphthalmos		---	---	---	---	---
Anotia		0.07	0.49	0.66	0.48	0.92
Microtia		0.21	0.60	0.60	0.69	0.67
Unspecified Anotia / Microtia		---	---	---	---	---
Transposition of great vessels		2.48	2.19	3.39	5.81	5.48
Tetralogy of Fallot		0.89	1.31	2.19	3.83	4.15
Hypoplastic left heart syndrome		1.65	1.53	1.75	3.41	4.25
Coarctation of aorta		1.31	1.92	2.57	3.41	3.59
Choanal atresia, bilateral		0.62	0.55	0.49	0.64	0.41
Cleft palate without cleft lip		4.06	4.16	6.17	6.07	6.56
Cleft lip with or without cleft palate		6.67	7.29	9.67	9.11	7.89
Oesophageal atresia / stenosis with or without fistula		2.27	2.79	3.33	3.94	4.36
Small intestine atresia / stenosis		0.41	0.99	1.69	2.29	3.48
Anorectal atresia / stenosis		3.51	1.75	3.39	3.25	4.05
Undescended testis (36 weeks of gestation or later)		8.53	13.37	11.09	6.50	6.39*
Hypospadias		10.46	10.52	15.46	10.28	14.86
Epispadias		0.07	0.60	0.55	0.37	0.31
Indeterminate sex		1.58	1.15	1.42	1.23	1.23
Renal agenesis		1.03	1.04	1.20	3.41	2.36
Cystic kidney		1.58	3.07	5.30	9.05	10.86
Bladder exstrophy		0.21	0.33	0.60	0.69	0.36
Polydactyly, preaxial		0.62	0.82	1.69	2.45	1.43
Total Limb reduction defects (include unspecified)		nr	nr	nr	6.60	8.51
Transverse		nr	nr	nr	2.93	4.56
Preaxial		nr	nr	nr	0.85	1.43
Postaxial		nr	nr	nr	0.27	0.97
ntercalary		nr	nr	nr	0.48	0.46
Mixed		nr	nr	nr	0.27	0.77
Unspecified		---	---	---	---	---
Diaphragmatic hernia		2.00	3.18	3.11	5.59	5.38
Omphalocele		1.58	1.81	2.08	5.01	6.35
Gastroschisis		0.48	0.82	1.97	2.56	3.69
Unspecified Omphalocele / Gastroschisis		---	---	---	---	---
Prune belly sequence		0.07	0.16	0.00	0.16	0.15
Trisomy 13		0.41	0.55	0.87	3.52	4.56
Trisomy 18		1.65	0.93	2.51	8.10	12.55
Down syndrome, all ages (include age unknown)		11.49	12.65	17.48	34.46	38.79
<20		10.09	15.83	4.51	5.95	24.05
20-24		6.80	5.79	9.76	14.61	11.10
25-29		6.79	6.36	8.68	14.20	13.55
30-34		11.19308	12.99135	13.52	21.40	21.55
35-39		24.03846	30.1263	31.42	56.98	60.55
40-44		57.40528	25.53812	83.26	199.31	188.13
45+		220.9945	158.1028	161.73	334.93	400.57
unspecified		---	---	---	---	---

* data include less than 5 years

nr = not reported

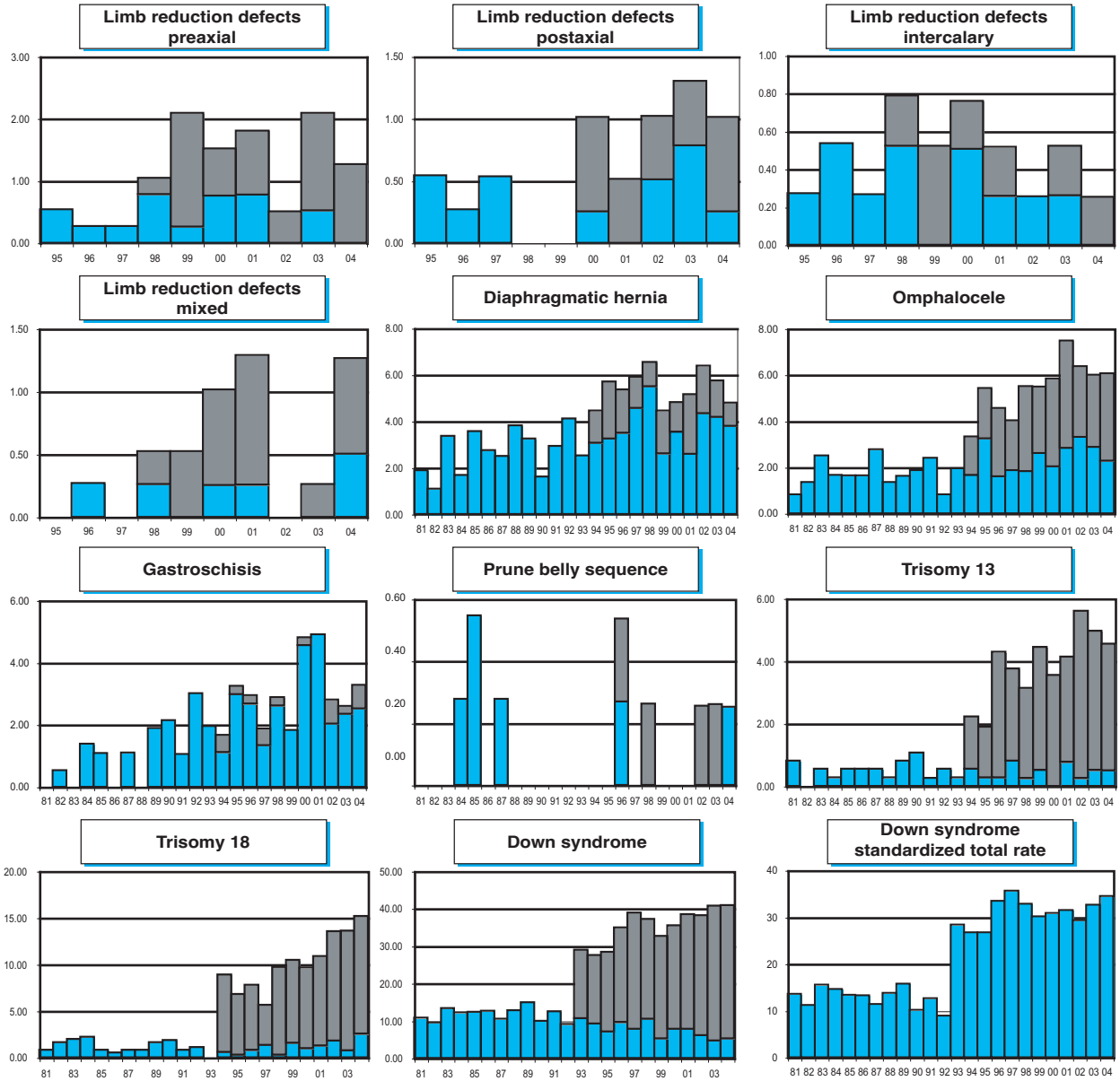
France: Paris

'Time trends 1981-2004 (Birth prevalence rates per 10,000)



Note: L+S rates, ToP rates

Note: L+S rates, ToP rates



Note: L+S rates, ToP rates

France: Strasbourg

Strasbourg Prospective Study of Congenital Malformations.

History:
The registry was started in 1979. The Programme became an associate member of the Clearinghouse in 1982.

Size and coverage:
All births in an area including and around Strasbourg and the Bas-Rhin are covered -13,000 to 13,500 annually, or 1,8% of all births in France.

Legislation and funding:
The Programme is a research Programme, recognised by the local health authorities and funded by Social Security, Ministry of Health, and INSERM.

Sources of ascertainment:
Reports are obtained from pediatricians examining the newborn infants. A control infant is selected for each malformed one: the next infant of the same sex as the proband born at that hospital.

Exposure information:
Detailed information on various exposures is obtained by interview of the mothers of the malformed infants and their controls. The children are followed to the age of one year.

Background information:
General demographic information is obtained from the National Institute of Statistics. Further information is obtained from Social Security Records and Health Sheets.

Addresses and Staff:
Claude Stoll, Programme Director Laboratoire de Génétique Médicale Faculté de Médecine 11 rue Humann 67085 Strasbourg Cedex, France

Phone: 33 (0) 3.90.24.32.07

E-mail: Claude.Stoll@medecine.u-strasbg.fr

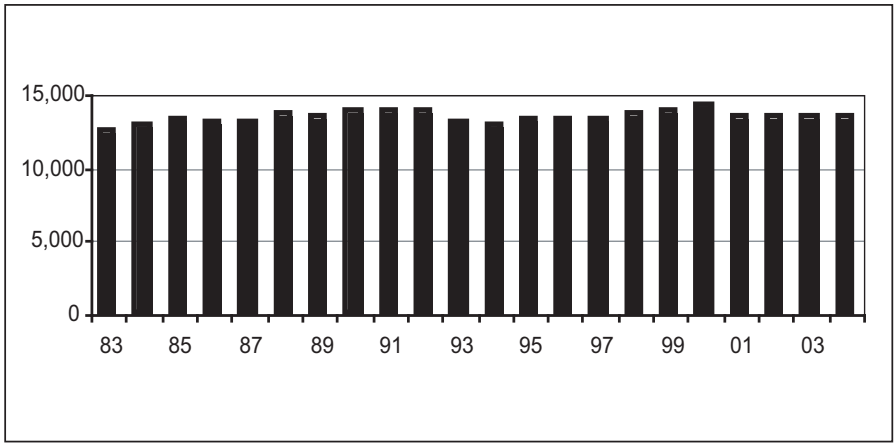
Dott Beatrice, MD Centre Hospitalo-Universitaire

E-mail: Beatrice.Dott@chru-strasbourg.fr

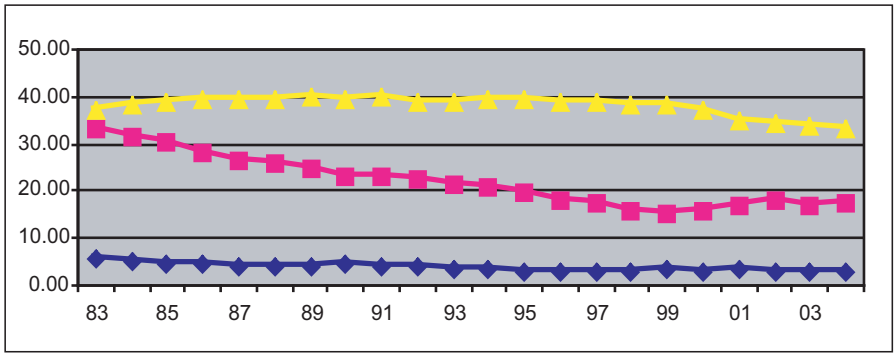
Alembik Yves, MD Centre Hospitalo-Universitaire

France: Strasbourg

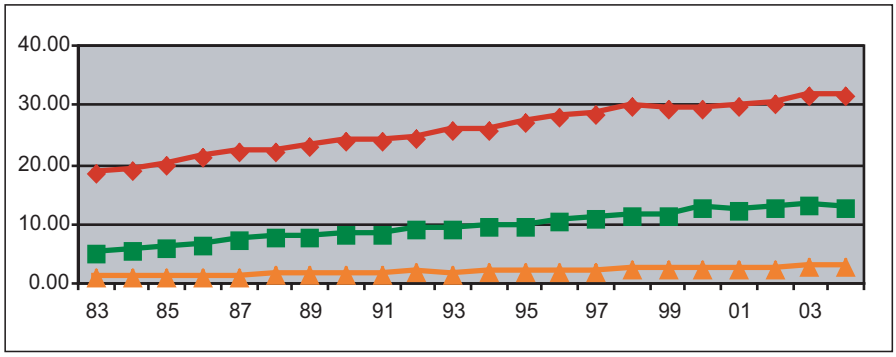
Total births by year



Percentage of births by maternal age



legenda: %births < 20 %births 20-24 %births 25-29



legenda: %births 30-34 %births 35-39 %births 40+

France: Strasbourg, 2004

Live births (LB)	13,291
Stillbirths (SB)	117
Total births	13,408
Number of terminations of pregnancy (ToP) for birth defects	91

Birth Defects	Number of cases			Rates*10,000
	LB	SB	ToP	Total rate
Anencephaly	0	0	10	7.41
Spina bifida	0	0	7	5.19
Encephalocele	0	0	2	1.48
Microcephaly	2	0	0	1.48
Arhinencephaly / Holoprosencephaly	0	0	3	2.22
Hydrocephaly	2	0	9	8.15
Anophthalmos	0	0	0	0.00
Microphthalmos	2	0	0	1.48
Unspecified Anophthalmos/ Microphthalmos	0	0	0	---
Anotia	2	0	0	1.48
Microtia	0	0	0	0.00
Unspecified Anotia/Microtia	0	0	0	---
Transposition of great vessels	6	0	4	7.41
Tetralogy of Fallot	6	0	0	4.44
Hypoplastic left heart syndrome	0	0	3	2.22
Coarctation of aorta	6	0	0	4.44
Choanal atresia, bilateral	1	0	0	0.74
Cleft palate without cleft lip	11	0	0	8.15
Cleft lip with or without cleft palate	13	0	2	11.11
Oesophageal atresia / stenosis with or without fistula	1	1	3	3.70
Small intestine atresia / stenosis	2	0	1	2.22
Anorectal atresia / stenosis	1	0	2	2.22
Undescended testis (36 weeks of gestation or later)	nr	nr	nr	nr
Hypospadias	20	0	0	14.82
Epispadias	1	0	0	0.74
Indeterminate sex	0	0	1	0.74
Renal agenesis	4	0	6	7.41
Cystic kidney	6	0	2	5.93
Bladder exstrophy	0	0	0	0.00
Polydactyly, preaxial	1	0	0	0.74
Total Limb reduction defects (include unspecified)	7	0	1	5.93
Transverse	3	0	0	2.22
Preaxial	0	0	1	0.74
Postaxial	0	0	0	0.00
Intercalary	0	0	0	0.00
Mixed	4	0	0	2.96
Unspecified	0	0	0	---
Diaphragmatic hernia	7	0	0	5.19
Omphalocele	1	0	1	1.48
Gastroschisis	1	0	0	0.74
Unspecified Omphalocele/Gastroschisis	0	0	0	---
Prune belly sequence	0	0	1	0.74
Trisomy 13	1	0	4	3.70
Trisomy 18	2	0	7	6.67
Down syndrome, all ages (include age unknown)	11	0	26	27.41
<20	0	0	0	0.00
20-24	2	0	1	13.02
25-29	0	0	1	2.25
30-34	5	0	7	28.40
35-39	0	0	13	76.97
40-44	4	0	4	232.56
45+	0	0	0	0.00
unknown	0	0	0	---

nr = not reported

France: Strasbourg, Previous years rates 1983 - 2004

Birth prevalence rates: (LB+SB) * 10,000 until 1993
Birth prevalence rates: (LB+SB+TOP) * 10,000 from 1994

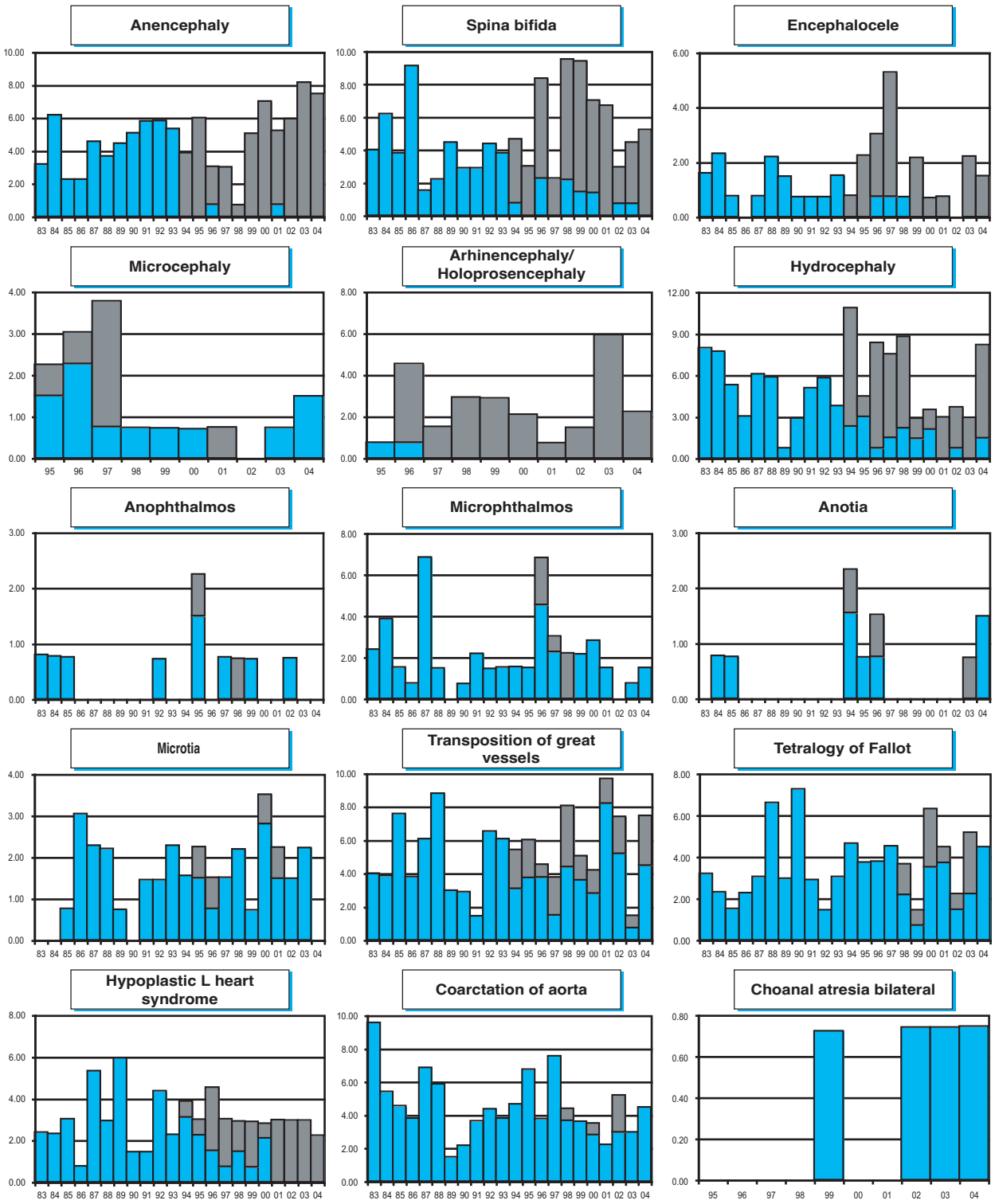
	1974-79	1980-84*	1985-89	1990-94	1995-99	2000-04
Births		**25,449**	**66,504**	**67,271**	**67,218**	**68,028**
Anencephaly		4.72	3.46	5.20	3.57	6.76
Spina bifida		5.11	4.21	3.72	6.55	5.29
Encephalocele		1.96	1.05	0.89	2.68	1.03
Microcephaly		nr	nr	nr	2.08	0.73
Arhinencephaly / Holoprosencephaly		nr	nr	nr	2.53	2.50
Hydrocephaly		7.86	4.21	5.65	6.40	4.26
Anophthalmos		0.79	0.15	0.15	0.89	0.15
Microphthalmos		3.14	2.11	1.49	3.12	1.32
Unspecified Anophthalmos / Microphthalmos		---	---	---	---	---
Anotia		0.39	0.15	0.45	0.45	0.44
Microtia		0.00	1.80	1.34	1.64	1.91
Unspecified Anotia / Microtia		---	---	---	---	---
Transposition of great vessels		3.93	5.86	4.46	5.50	6.03
Tetralogy of Fallot		2.75	3.31	3.86	3.42	4.56
Hypoplastic left heart syndrome		2.36	3.61	2.68	3.27	2.79
Coarctation of aorta		7.47	4.51	3.72	5.21	3.67
Choanal atresia, bilateral		nr	nr	nr	0.15	0.44
Cleft palate without cleft lip		10.22	8.87	9.07	7.14	7.06
Cleft lip with or without cleft palate		9.43	8.72	13.23	12.65	13.38
Oesophageal atresia / stenosis with or without fistula		1.96	2.56	2.97	2.68	4.12
Small intestine atresia / stenosis		nr	nr	nr	2.23	2.06
Anorectal atresia / stenosis		4.72	4.81	5.20	6.55	5.00
Undescended testis (36 weeks of gestation or later)		nr	nr	nr	1.52*	nr
Hypospadias		13.36	24.06	26.46	22.02	22.34
Epispadias		nr	nr	nr	0.37*	0.15
Indeterminate sex		nr	nr	nr	0.15	1.03
Renal agenesis		nr	nr	2.33	4.31	8.82
Cystic kidney		nr	nr	nr	8.93	7.20
Bladder exstrophy		nr	nr	nr	0.60	0.29
Polydactyly, preaxial		nr	nr	nr	3.72	4.26
Total Limb reduction defects (include unspecified)		6.29	6.77	6.99	12.05	8.08
Transverse		4.32	4.36	3.86	5.80	3.38
Preaxial		1.96	1.50	1.49	1.64	0.73
Postaxial		0.00	0.45	0.45	0.45	0.44
Intercalary		0.00	0.00	0.45	0.89	0.29
Mixed		0.00	0.45	0.30	0.74	1.76
Unspecified		---	---	---	---	---
Diaphragmatic hernia		3.14	4.06	5.35	4.46	4.56
Omphalocele		2.75	3.16	4.01	4.61	2.79
Gastroschisis		0.79	2.41	2.08	2.83	1.47
Unspecified Omphalocele / Gastroschisis		---	---	---	---	---
Prune belly sequence		nr	nr	nr	0.45	0.88
Trisomy 13		nr	nr	nr	1.79	2.65
Trisomy 18		nr	nr	nr	3.57	6.91
Down syndrome, all ages (include age unknown)		9.43	15.79	22.15	30.05	24.55
<20		0.00	18.21	15.49	15.58	4.76
20-24		7.31	8.37	10.79	11.22	11.33
25-29		4.16	7.64	14.02	10.39	5.51
30-34		10.57	16.78	13.90	26.11	15.45
35-39		53.07	47.42	77.08	101.90	77.49
40-44		109.89	269.46	221.37	282.80	150.94
45+		0.00	232.56	243.90	0.00	0.00
unspecified		---	---	---	---	---

* data include less than 5 years

nr = not reported

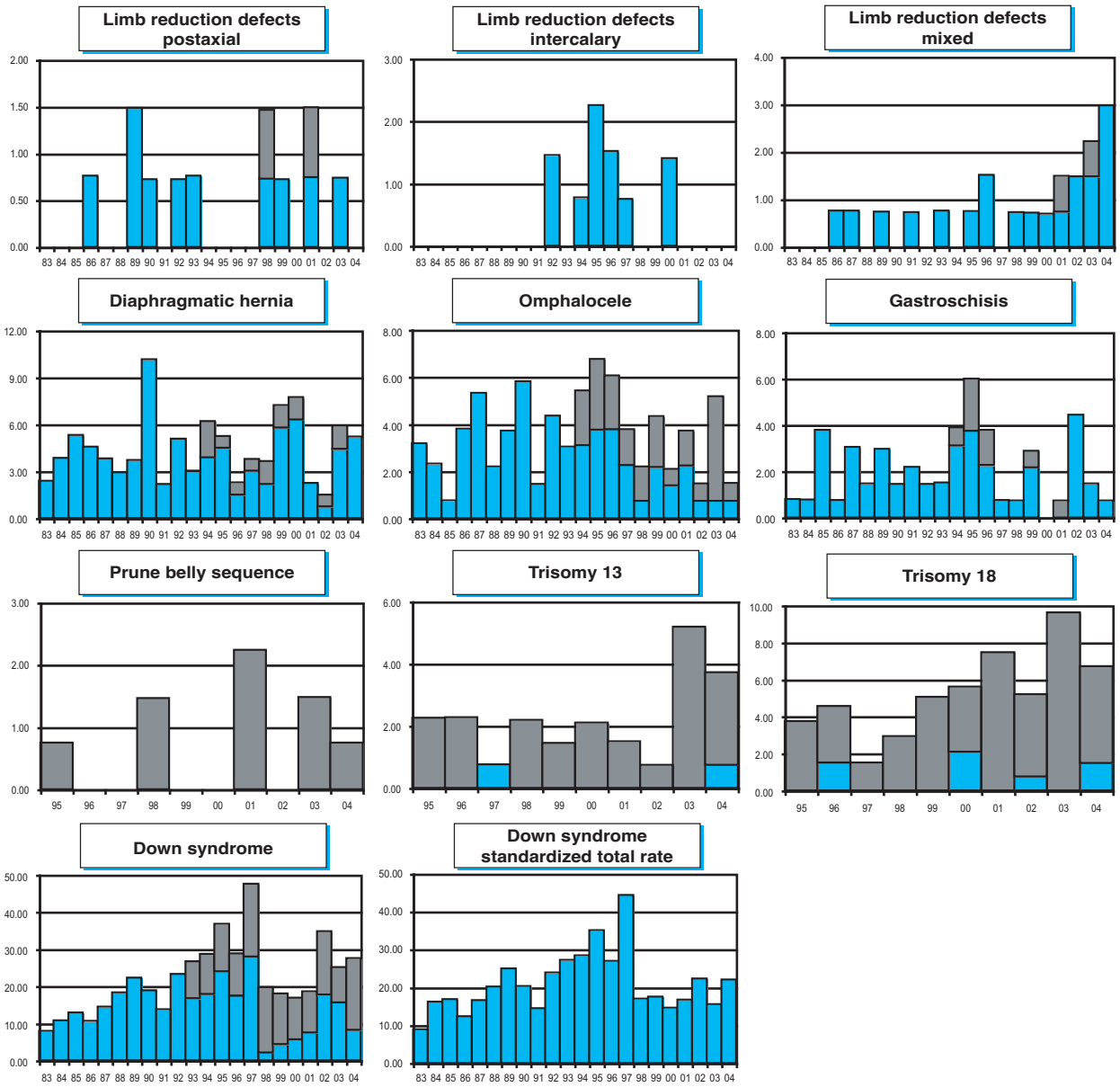
France: Strasbourg

'Time trends 1983-2004 (Birth prevalence rates per 10,000)



Note: L+S rates, ToP rates

Note: L+S rates, ToP rates



Note: L+S rates, ToP rates

Germany: Saxony-Anhalt
Malformation Monitoring Saxony-Anhalt

History:
Since 1980 in the city of Magdeburg all live- and stillbirths, abortions after the 16th week of gestation (spontaneous and induced abortions according to medical evidence based on prenatal diagnoses of congenital defects), and postnatal anomalies or congenital defects have been recorded up to the first week of life. After the reunification of Germany and the creation of the Federal state of Saxony-Anhalt, the survey of congenital defects included approximately two-thirds of all births with postnatal anomalies and congenital defects in the same federal state. Since 1 January 2000 the survey region includes the entire state of Saxony-Anhalt. Saxony-Anhalt has 2.49 million inhabitants (31.12.2004) and annual births at a rate of about 17 500 children (2004). The survey system is multi-centric and based on population.

Legislation and funding
1980 to 1989: Ministry of Health of the former German Democratic Republic
1990 to 1992: Medical Faculty, Magdeburg
1993 to 1995: Ministry of Health, Federal Republic of Germany
since 1995: Ministry of Labour, Women, Health and Social Security of the Federal State of Saxony-Anhalt. The Malformation Monitoring is working in order of Ministry of Labour, Women, Health and Social Security of the Federal State of Saxony-Anhalt

Sources of ascertainment
The co-operation partner are:
- 32 obstetrics departments
- 29 children hospitals
- 10 institutions of prenatal diagnostic
- 6 departments of pathology

Exposure information
Maternal and paternal occupation (in groups); occupation risk; drugs in pregnancy (ATC-code); alcohol, nicotine, drug abuse.

Background information
Population based registry (Federal State Saxony-Anhalt); written informed consent of the mother (parents); name and address don't registered; two healthy "controls" per one malformed child; inclusion of terminations of pregnancy, spontaneous abortions after 16th week of gestation, live and stillborn babies; definition of stillbirth: >/= 500 grams; maximum age to include diagnoses: 1 year (almost 1th week of life); annual report (in German).

Addresses and staff
Dr. Simone Pötzsch, Program Director (Paediatrician) Malformation Monitoring Saxony-Anhalt, Faculty of Medicine, Otto-von-Guericke University, Leipziger Straße 44, Haus 39, D-39120 Magdeburg, Germany

Phone: 49-(0)391- 6714174

Fax: 49-(0)391-6714176

E-mail:
simone.poetzsch@medizin.uni-magdeburg.de

Dr. Jana Hoyer-Schuschke – Paediatrician, Assistent physician

E-mail:
jana.hoyer-schuschke@medizin.uni-magdeburg.de

Dipl.-Ges.-Wirt Andrea Köhn – Scientific staff

E-mail:
andrea.koehn@medizin.uni-magdeburg.de

Dipl. Ing. Dorit Götz - Computer expert

E-mail:
dorit.goetz@medizin.uni-magdeburg.de

Cornelia Vogt - Documentation assistant

E-mail:
cornelia.vogt@medizin.uni-magdeburg.de

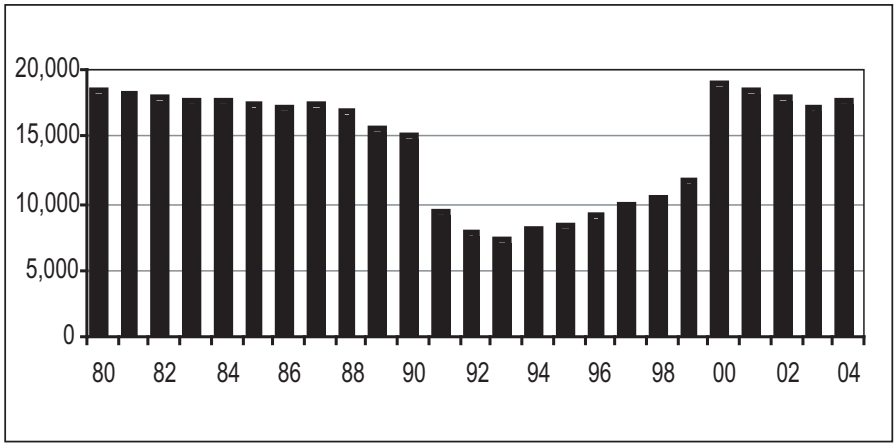
Marion Haase – Secretary

E-mail:
marion.haase@medizin.uni-magdeburg.de

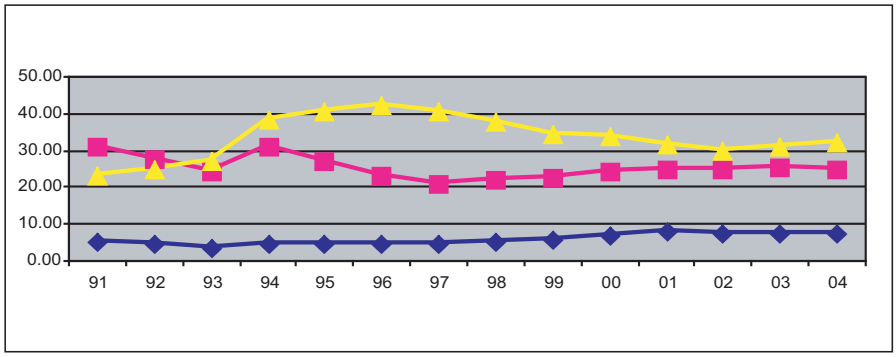
Web site:
http://www.med.uni-magdeburg.de/fme/zkh/mz/

Germany: Saxony-Anhalt

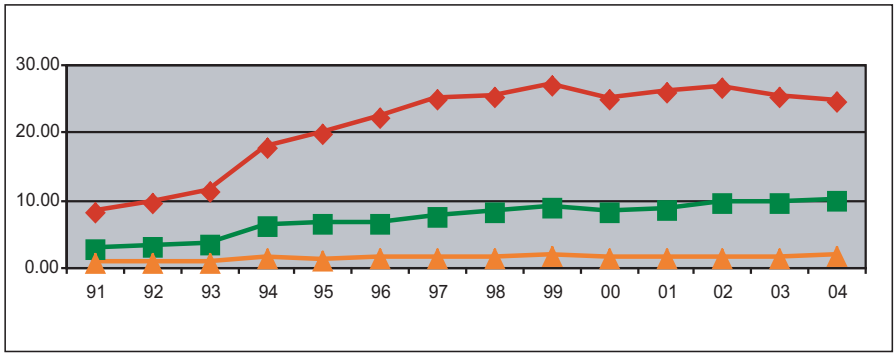
Total births by year



Percentage of births by maternal age



legenda: %births < 20 %births 20-24 %births 25-29



legenda: %births 30-34 %births 35-39 %births 40+

Germany: Saxony Anhalt, 2004

Live births (LB)	17,337
Stillbirths (SB)	77
Total births	17,414
Number of terminations of pregnancy (ToP) for birth defects	73

Birth Defects	Number of cases			Rates*10,000
	LB	SB	ToP	Total rate
Anencephaly	0	0	5	2.86
Spina bifida	8	0	10	10.29
Encephalocele	2	0	2	2.29
Microcephaly	27	0	0	15.44
Arhinencephaly / Holoprosencephaly	0	0	5	2.86
Hydrocephaly	4	0	6	5.72
Anophthalmos	0	0	0	0.00
Microphthalmos	0	0	0	0.00
Unspecified Anophthalmos/ Microphthalmos	0	0	0	---
Anotia	0	0	1	0.57
Microtia	0	0	1	0.57
Unspecified Anotia/Microtia	0	0	0	---
Transposition of great vessels	7	0	0	4.00
Tetralogy of Fallot	4	0	0	2.29
Hypoplastic left heart syndrome	3	1	0	2.29
Coarctation of aorta	7	0	1	4.57
Choanal atresia, bilateral	1	0	0	0.57
Cleft palate without cleft lip	12	0	3	8.58
Cleft lip with or without cleft palate	25	1	5	17.73
Oesophageal atresia / stenosis with or without fistula	4	0	2	3.43
Small intestine atresia / stenosis	5	0	1	3.43
Anorectal atresia / stenosis	8	0	0	4.57
Undescended testis (36 weeks of gestation or later)	13	0	0	7.43
Hypospadias	18	0	0	10.29
Epispadias	1	0	0	0.57
Indeterminate sex	1	0	0	0.57
Renal agenesis	0	0	6	3.43
Cystic kidney	13	0	3	9.15
Bladder exstrophy	0	0	0	0.00
Polydactyly, preaxial	3	1	0	2.29
Total Limb reduction defects (include unspecified)	11	0	5	9.15
Transverse	3	0	3	3.43
Preaxial	1	0	1	1.14
Postaxial	0	0	0	0.00
Intercalary	3	0	1	2.29
Mixed	4	0	0	2.29
Unspecified	0	0	0	---
Diaphragmatic hernia	4	0	2	3.43
Omphalocele	2	0	6	4.57
Gastroschisis	14	0	1	8.58
Unspecified Omphalocele/Gastroschisis	0	0	0	---
Prune belly sequence	0	0	2	1.14
Trisomy 13	0	0	0	0.00
Trisomy 18	1	1	8	5.72
Down syndrome, all ages (include age unknown)	16	0	14	17.16
<20	1	0	0	7.94
20-24	3	0	0	7.04
25-29	3	0	2	9.02
30-34	2	0	3	11.77
35-39	4	0	4	45.69
40-44	2	0	5	243.06
45+	0	0	0	0.00
unknown	1	0	0	---

Germany: Saxony Anhalt, Previous years rates 1980 - 2004

Prevalence rates: (LB+SB+TOP) * 10,000

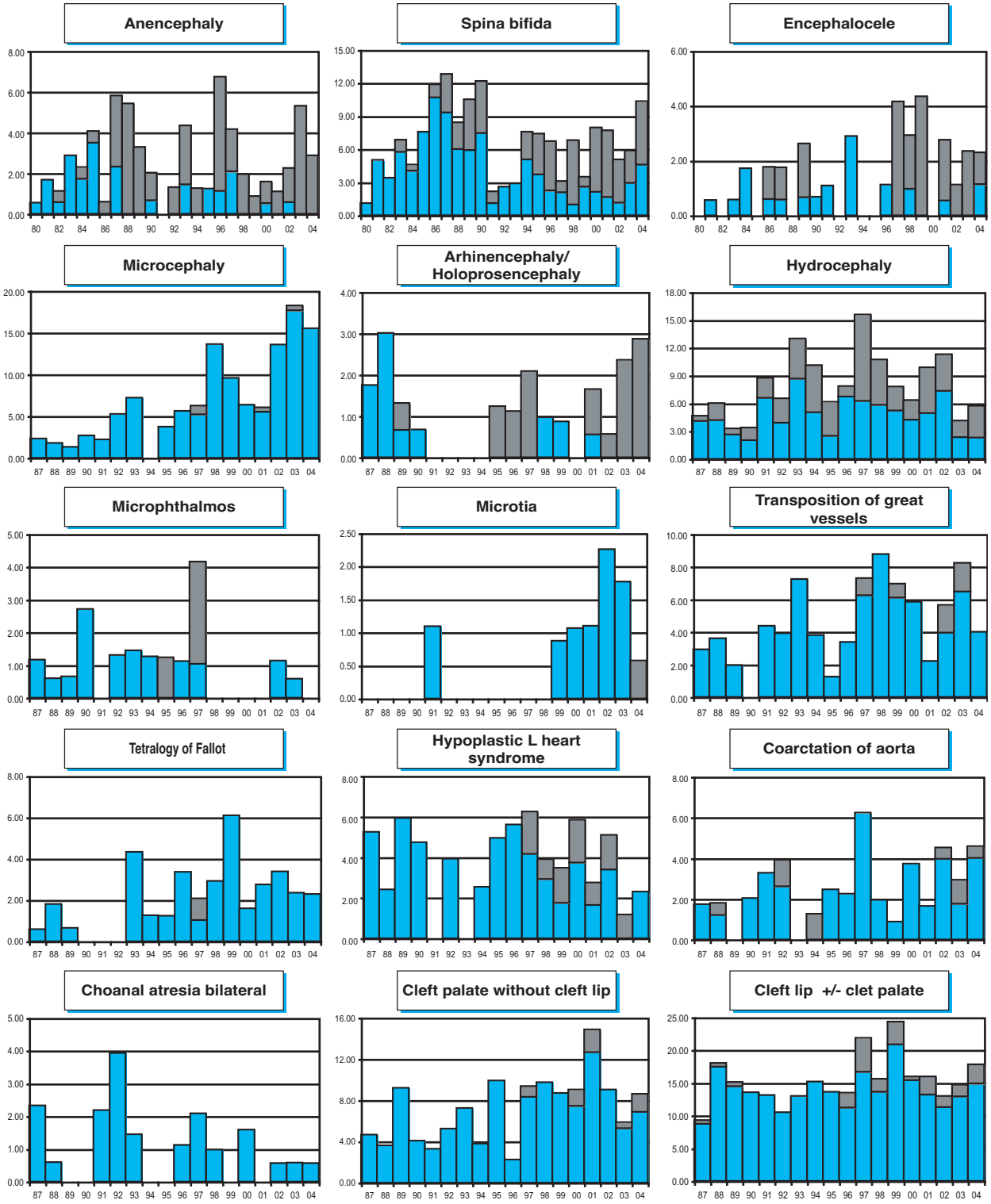
	1974-79	1980-84*	1985-89	1990-94	1995-99	2000-04
Births		**88,477**	**83,005**	**46,342**	**48,335**	**89,025**
Anencephaly		1.70	3.86	1.73	2.90	2.58
Spina bifida		4.18	10.24	6.47	5.38	7.41
Encephalocele		0.57	1.20	0.86	2.69	1.68
Microcephaly		nr	1.84*	3.24	8.07	11.79
Arhinencephaly / Holoprosencephaly		nr	2.04*	0.22	1.24	1.46
Hydrocephaly		nr	4.69*	7.55	9.72	7.53
Anophthalmos		nr	0.00*	0.86	0.00	0.11
Microphthalmos		nr	0.82*	1.51	1.24	0.34
Unspecified Anophthalmos / Microphthalmos		----	----	----	----	----
Anotia		nr	0.00*	0.22	0.00	0.22
Microtia		nr	0.00*	0.22	0.21	1.35
Unspecified Anotia / Microtia		----	----	----	----	----
Transposition of great vessels		nr	2.86*	3.24	5.79	5.17
Tetralogy of Fallot		nr	1.02*	0.86	3.31	2.47
Hypoplastic left heart syndrome		nr	4.49*	2.59	4.76	3.48
Coarctation of aorta		nr	1.22*	2.16	2.69	3.48
Choanal atresia, bilateral		nr	1.02*	1.29	0.83	0.67
Cleft palate without cleft lip		nr	5.72*	4.53	8.07	9.55
Cleft lip with or without cleft palate		nr	14.08*	13.16	18.21	15.50
Oesophageal atresia / stenosis with or without fistula		nr	2.45*	2.37	2.90	2.92
Small intestine atresia / stenosis		nr	0.82*	3.24	1.24	2.58
Anorectal atresia / stenosis		nr	3.47*	3.45	2.69	2.92
Undescended testis (36 weeks of gestation or later)		nr	11.84*	18.77	12.21	9.44
Hypospadias		nr	14.49*	17.05	17.79	8.87
Epispadias		nr	0.20*	0.43	0.62	0.34
Indeterminate sex		nr	0.61*	0.00	0.83	0.79
Renal agenesis		nr	1.84*	1.08	3.10	2.36
Cystic kidney		nr	2.04*	3.67	4.14	5.28
Bladder exstrophy		nr	0.82*	0.22	0.62	0.00
Polydactyly, preaxial		nr	0.00*	1.94	4.34	3.48
Total Limb reduction defects (include unspecified)		nr	4.29*	7.34	8.28	7.86
Transverse		nr	nr	nr	nr	3.37
Preaxial		nr	nr	nr	nr	0.56
Postaxial		nr	nr	nr	nr	0.00
Intercalary		nr	nr	nr	nr	1.68
Mixed		nr	nr	nr	nr	1.80
Unspecified		----	----	----	----	----
Diaphragmatic hernia		nr	1.43*	1.51	1.24	2.81
Omphalocele		nr	5.72*	2.37	2.90	3.15
Gastroschisis		nr	1.02*	2.37	3.72	3.71
Unspecified Omphalocele / Gastroschisis		----	----	----	----	----
Prune belly sequence		nr	0.00*	0.86	1.24	0.79
Trisomy 13		0.23	0.36	0.65	2.48	0.67
Trisomy 18		0.90	0.84	0.86	1.66	3.37
Down syndrome, all ages (include age unknown)		8.59	8.19	10.36	16.96	16.40
<20		nr	nr	nr	nr	6.15
20-24		nr	nr	nr	nr	6.36
25-29		nr	nr	nr	nr	9.60
30-34		nr	nr	nr	nr	12.83
35-39		nr	nr	nr	nr	53.03
40-44		nr	nr	nr	nr	174.19
45+		nr	nr	nr	nr	392.16
unspecified		----	----	----	----	----

* data include less than 6 and 5 years

nr = not reported

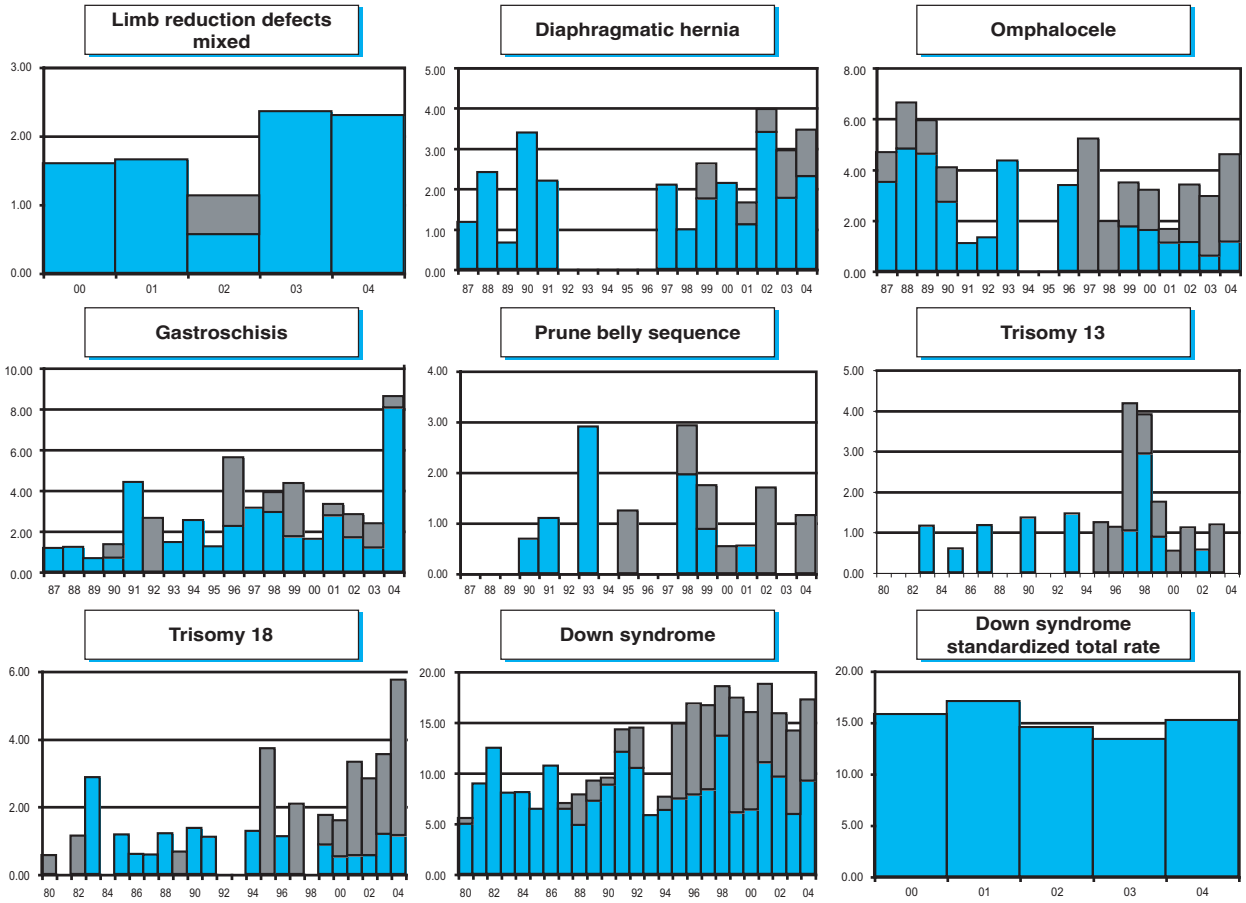
Germany: Saxony Anhalt

'Time trends 1980-2004 (Birth prevalence rates per 10,000)



Note: L+S rates, ToP rates

Note: L+S rates, ToP rates



Note: L+S rates, ToP rates

Hungary
Hungarian Congenital Abnormality Registry

History:
Centralized registration of congenital abnormalities began in Hungary in 1962, and came under our coordination in 1970. Monitoring began in 1973. The Programme was a founding member of the International Clearinghouse and is a full member.

Size and coverage
The registry covers all births in Hungary, approximately 100,000 annually. Criteria to define stillbirth was changed in 1998. At present, stillbirths of at least 24 weeks gestation or 500 grams are registered. Prenatally diagnosed and terminated fetuses are also registered.

Legislation and funding
Reporting is compulsory. The registry is run and financed by the governmental National Center for Epidemiology (formerly the National Institute of Public Health).

Sources of ascertainment
Reports are obtained from multiple sources, such as delivery units, neonatal and pediatric surgery, pathology, and prenatal diagnostic centers. Abnormalities detected before the age of one are reported. Variations in figures (especially in the1990s) may reflect incomplete notification.

Exposure information
Exposure information has been available since 1980, when a case-control system was initiated. Mothers of selected malformed infants and controls are interviewed by community nurses to collect information.

Background information
General background information on all births is available from central statistics.

Addresses and staff
Hungarian Congenital Abnormality Registry
National Center for Healthcare Audit and Improvement Gyali ut 2-6, H-1966 Budapest, Pf. 64., Hungary.

Phone: +36-1-476 1129

Fax: +36-1-476 1389

Melinda Csaky-Szunyogh, MSc

E-mail:
szunyoghm@oek.antsz.hu;
csaky-szunyogh@oszmk.hu

Erzsebet Horvath-Puho, MSc

E-mail: puhoe@oek.antsz.hu

Julia Metneki, PhD

E-mail: metnekij@oek.antsz.hu

Janos Sandor, MD, PhD

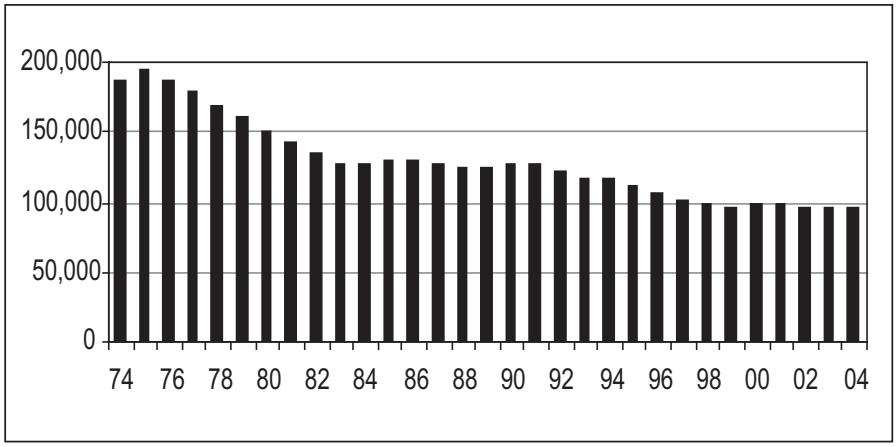
E-mail: janos.sandor@etk.pte.hu

Csaba Siffel, MD, PhD

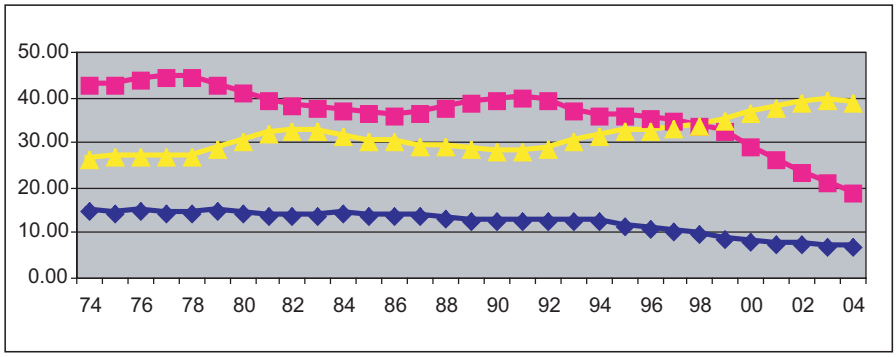
E-mail: csiffel@cdc.gov

Hungary

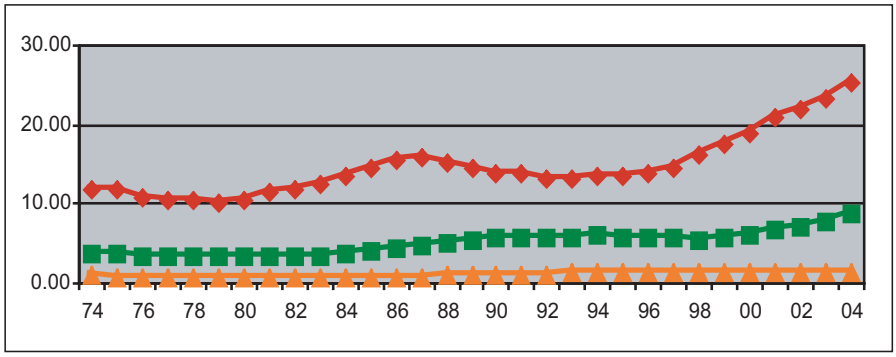
Total births by year



Percentage of births by maternal age



legenda: %births < 20 %births 20-24 %births 25-29



legenda: %births 30-34 %births 35-39 %births 40+

Hungary: 2004

Live births (LB)	95,137
Stillbirths (SB)	476
Total births	95,613
Number of terminations of pregnancy (ToP) for birth defects	274

Birth Defects	Number of cases			Rates*10,000
	LB	SB	ToP	Total rate
Anencephaly	1	0	15	1.67
Spina bifida	12	0	14	2.71
Encephalocele	3	0	4	0.73
Microcephaly	8	0	0	0.83
Arhinencephaly / Holoprosencephaly	7	0	2	0.94
Hydrocephaly	19	0	20	4.07
Anophthalmos	0	0	0	0.00
Microphthalmos	2	0	0	0.21
Unspecified Anophthalmos / Microphthalmos	0	0	0	---
Anotia	4	0	0	0.42
Microtia	0	0	0	0.00
Unspecified Anotia / Microtia	0	0	0	---
Transposition of great vessels	8	0	2	1.04
Tetralogy of Fallot	14	0	0	1.46
Hypoplastic left heart syndrome	14	0	2	1.67
Coarctation of aorta	21	0	1	2.29
Choanal atresia, bilateral	0	0	0	0.00
Cleft palate without cleft lip	38	0	0	3.96
Cleft lip with or without cleft palate	77	0	3	8.34
Oesophageal atresia / stenosis with or without fistula	6	0	0	0.63
Small intestine atresia / stenosis	13	0	0	1.36
Anorectal atresia / stenosis	10	0	0	1.04
Undescended testis (36 weeks of gestation or later)	172	0	0	17.94
Hypospadias	233	0	0	24.30
Epispadias	nr	nr	nr	nr
Indeterminate sex	7	0	0	0.73
Renal agenesis	6	0	5	1.15
Cystic kidney	36	0	3	4.07
Bladder exstrophy	0	0	1	0.10
Polydactyly, preaxial	83	0	0	8.66
Total Limb reduction defects (include unspecified)	35	0	5	4.17
Transverse	nr	nr	nr	nr
Preaxial	nr	nr	nr	nr
Postaxial	nr	nr	nr	nr
Intercalary	nr	nr	nr	nr
Mixed	nr	nr	nr	nr
Unspecified	nr	nr	nr	---
Diaphragmatic hernia	5	0	3	0.83
Omphalocele	4	0	8	1.25
Gastroschisis	2	0	2	0.42
Unspecified Omphalocele / Gastroschisis	0	0	0	---
Prune belly sequence	0	0	0	0.00
Trisomy 13	5	0	9	1.46
Trisomy 18	8	0	14	2.29
Down syndrome, all ages (include age unknown)	74	0	58	13.77
<20	5	0	0	7.58
20-24	4	0	3	3.92
25-29	21	0	7	7.54
30-34	26	0	15	16.96
35-39	12	0	18	35.53
40+	5	0	15	137.08
unknown	1	0	0	---

NOTE1: Epispadias included in Hypospadias
NOTE2: Only isolated birth defects are reported
nr = not reported

Hungary: Previous years rates 1974 - 2004

Birth prevalence rates: (LB+SB) * 10,000 until 1993
Birth prevalence rates: (LB+SB+TOP) * 10,000 from 1994

	1974-79	1980-84	1985-89	1990-94	1995-99	2000-04
Births	**1,081,073**	**682,788**	**635,779**	**609,857**	**511,919**	**483,849**
Anencephaly	7.08	6.12	2.08	0.69	1.37	1.65
Spina bifida	10.63	8.38	6.23	2.44	2.60	2.93
Encephalocele	1.15*	1.80	1.37	0.61	0.39	0.70
Microcephaly	nr	1.39	0.91	0.72	0.55	0.62
Arhinencephaly / Holoprosencephaly	nr	0.21	0.27	0.10	0.41	0.95
Hydrocephaly	6.86	4.66	3.21	1.74	2.21	3.06
Anophthalmos	0.13	0.06	0.06	0.05	0.06	0.04
Microphthalmos	0.21	0.21	0.02	0.11	0.08	0.08
Unspecified Anophthalmos / Microphthalmos	---	---	---	---	---	---
Anotia	0.09	0.09	0.24	0.21	0.33	0.62
Microtia	0.05	0.03	0.02	0.02	0.02	0.10
Unspecified Anotia / Microtia	---	---	---	---	---	---
Transposition of great vessels	0.54*	1.57	1.71	1.39	1.05	1.45
Tetralogy of Fallot	1.22	1.42	1.04	0.82	1.50	1.98
Hypoplastic left heart syndrome	0.33*	0.47	0.74	0.62	0.70	1.01
Coarctation of aorta	1.23	1.89	2.71	1.59	1.66	1.69
Choanal atresia, bilateral	nr	0.15	0.14	0.16	0.02	0.06
Cleft palate without cleft lip	3.83	4.54	3.92	3.26	2.87	3.35
Cleft lip with or without cleft palate	10.84	11.50	9.39	9.02	6.27	7.27
Oesophageal atresia / stenosis with or without fistula	2.07*	1.74	1.71	1.41	0.92	0.93
Small intestine atresia / stenosis	nr	1.51	1.18	1.07	0.51	0.95
Anorectal atresia / stenosis	2.20*	2.28	1.84	1.57	0.90	0.95
Undescended testis (36 weeks of gestation or later)	nr	17.81	16.28	15.05	10.67	12.83
Hypospadias	15.60	20.93	21.17	21.37	19.42	22.07
Epispadias	nr	nr	nr	nr	nr	nr
Indeterminate sex		0.29	0.36	0.18	0.16	0.41
Renal agenesis	1.27*	1.04	1.04	0.89	0.20	0.35
Cystic kidney	nr	0.00	0.19	0.49	1.35	2.46
Bladder exstrophy	nr	0.34	0.44	0.07	0.06	0.10
Polydactyly, preaxial	nr	0.94	1.90	1.31	4.98	8.18
Total Limb reduction defects (include unspecified)	nr	4.01*	4.29	2.77	3.05	3.27
Transverse	nr	nr	nr	nr	nr	nr
Preaxial	nr	nr	nr	nr	nr	nr
Postaxial	nr	nr	nr	nr	nr	nr
Intercalary	nr	nr	nr	nr	nr	nr
Mixed	nr	nr	nr	nr	nr	nr
Unspecified	---	---	---	---	---	---
Diaphragmatic hernia	1.81	2.62	2.00	1.75	0.92	0.43
Omphalocele	nr	2.13*	1.27	0.85	0.74	1.12
Gastroschisis	nr	0.49*	0.50	0.61	0.61	0.70
Unspecified Omphalocele / Gastroschisis	---	---	---	---	---	---
Prune belly sequence	nr	nr	nr	0.00*	0.10	0.00
Trisomy 13	nr	0.15*	0.25	0.20	0.20	0.72
Trisomy 18	nr	0.23*	0.30	0.25	0.47	1.65
Down syndrome, all ages (include age unknown)	8.93	8.17	8.26	8.53	6.82	14.14
<20	nr	1.85*	1.91	1.45	1.92	6.24
20-24	nr	1.50*	3.03	2.31	2.89	6.71
25-29	nr	3.15*	4.50	3.06	2.76	8.82
30-34	nr	4.53*	5.41	4.50	5.33	13.87
35-39	nr	9.71*	14.43	22.94	13.58	39.32
40-44	nr	74.36*	48.29	86.88	79.82	167.65
45+	nr	nr	nr	nr	nr	nr
unspecified	---	---	---	---	---	---

* data include less than 5 and 6 years

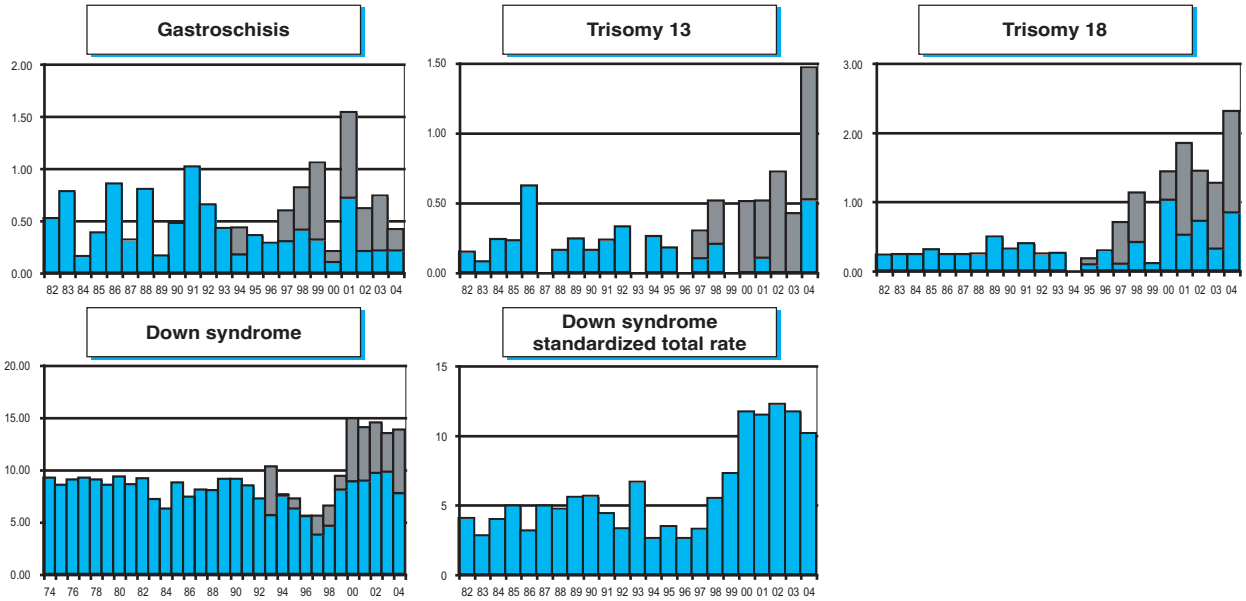
nr = not reported

Hungary

'Time trends 1974-2004 (Birth prevalence rates per 10,000)

Note: L+S rates, ToP rates

Note: L+S rates, ToP rates



Note: L+S rates, ToP rates

Iran: TROCA

Tabriz Registry of Congenital Anomalies (TROCA)

History:
The programme was initiated in 2000, but the registry started in 2003. It was then accepted as a member of the ICBDSR in the 2006 annual meeting in Uppsala, Sweden.

Size and coverage
TROCA is a hospital-based registry and situated in the North-West of Iran covering all births and children in three university hospitals in the city of Tabriz. This city is one of the three major cities in the country. The programme is based on approximately 60-70% of all births (15000 births per year) in the area.

Legislation and funding
The programme has been financially supported by the National Public Health Management Centre (NPMC) as a research grant. TROCA is located in the Alzahra University hospital of Tabriz University of Medical Sciences.

Exposure information
Some exposure information are currently available of mothers of all malformed infants. Other women giving births in all university hospitals with normal newborns routinely complete a similar form. They might be considered as matched control group.

Background information
General epidemiological data and basic characteristic information are available for all births.

Addresses and staff

Saeed Dastgiri, PhD, Programme Director
Department of Community and Family Medicine
School of Medicine Tabriz University of Medical Sciences Tabriz, Iran

Phone: 98 914 415 7039

Fax: 98 411 336 4668

E-mail: saeed.dastgiri@gmail.com

Web: http://www.tbzmed.ac.ir/troca

Iran: TROCA, 2004

Live births (LB)	18,785
Stillbirths (SB)	207
Total births	18,992
Number of terminations of pregnancy (ToP) for birth defects	46

Birth Defects	Number of cases			Rates*10,000
	LB	SB	ToP	Total rate
Anencephaly	4	6	35	23.64
Spina bifida	6	nr	nr	3.19
Encephalocele	1	nr	nr	0.53
Microcephaly	nr	nr	nr	nr
Arhinencephaly / Holoprosencephaly	nr	nr	nr	nr
Hydrocephaly	8	nr	4	6.37
Anophthalmos	2	nr	nr	1.06
Microphthalmos	nr	nr	nr	nr
Unspecified Anophthalmos/ Microphthalmos	2	nr	nr	1.06
Anotia	nr	nr	nr	nr
Microtia	nr	nr	nr	nr
Unspecified Anotia/Microtia	nr	nr	nr	nr
Transposition of great vessels	nr	nr	nr	nr
Tetralogy of Fallot	nr	nr	nr	nr
Hypoplastic left heart syndrome	nr	nr	nr	nr
Coarctation of aorta	nr	nr	nr	nr
Choanal atresia, bilateral	nr	nr	nr	nr
Cleft palate without cleft lip	13	nr	1	7.43
Cleft lip with or without cleft palate	22	nr	nr	11.71
Oesophageal atresia / stenosis with or without fistula	nr	nr	nr	nr
Small intestine atresia / stenosis	nr	nr	nr	nr
Anorectal atresia / stenosis	nr	nr	nr	nr
Undescended testis (36 weeks of gestation or later)	24	nr	nr	12.78
Hypospadias	11	nr	nr	5.86
Epispadias	1	nr	nr	0.53
Indeterminate sex	nr	nr	nr	nr
Renal agenesis	3	nr	1	2.12
Cystic kidney	1	nr	1	1.06
Bladder exstrophy	nr	nr	nr	nr
Polydactyly, preaxial	6	nr	nr	3.19
Total Limb reduction defects (include unspecified)	48	3	nr	49.58
Transverse	nr	nr	nr	nr
Preaxial	nr	nr	nr	nr
Postaxial	nr	nr	nr	nr
Intercalary	nr	nr	nr	nr
Mixed	nr	nr	nr	nr
Unspecified	nr	nr	nr	nr
Diaphragmatic hernia	2	1	1	2.10
Omphalocele	5	nr	2	3.72
Gastroschisis	5	nr	1	3.19
Unspecified Omphalocele/Gastroschisis	nr	nr	nr	nr
Prune belly sequence	nr	nr	nr	nr
Trisomy 13	nr	nr	nr	nr
Trisomy 18	nr	nr	nr	nr
Down syndrome, all ages (include age unknown)	nr	nr	nr	nr
<20	4	nr	nr	2.13
20-24	nr	nr	nr	nr
25-29	1	nr	nr	0.53
30-34	1	nr	nr	0.53
35-39	1	nr	nr	0.53
40-44	nr	nr	nr	nr
45+	1	nr	nr	0.53
unknown	nr	nr	nr	nr

nr= not reported
§ i tassi sono calcolati con il corrispondente denominatore

Ireland: Dublin
Dublin EUROCAT Registry

History:
Register began in September 1979 and joined EUROCAT at the same time. Joined International Clearinghouse in 1997.

Size and coverage
The Registry is population-based and situated in the East of Ireland covering the counties of Dublin, Wicklow and Kildare. About one third (22,000 births) of all births in Ireland occur in this region.

Legislation and funding
The Registry is located within the Population Health Directorate of the Health Service Executive. Staffing includes a full time Research Nurse and a part time secretary and is led by a Specialist in Public Health Medicine. Funding is provided by the Department of Health through the Health Service Executive. There is a Steering Committee comprised of specialists from each of Maternity and Paediatric Hospitals in the catchment plus a representative from the Department of Health.

Exposure information
For each malformed infant reported, very limited information is given on certain exposures. No information is available on controls.

Sources of ascertainment
All live and still births included. Termination of pregnancy is not legal in Ireland.

Addresses and staff

Bob McDonnell, MD, Programme Director Population Health Directorate Health Service Executive Dr. Steeven's Hospital Dublin 8 - Ireland

Phone: 353-1-6352752

Fax: 353-1-6353745

E-mail: bob.mcdonnell@mailf.hse.ie

Virginia Delany, Registry Co-ordinator/Research nurse Population Health Directorate Health Service Executive Dr. Steeven's Hospital Dublin 8 - Ireland

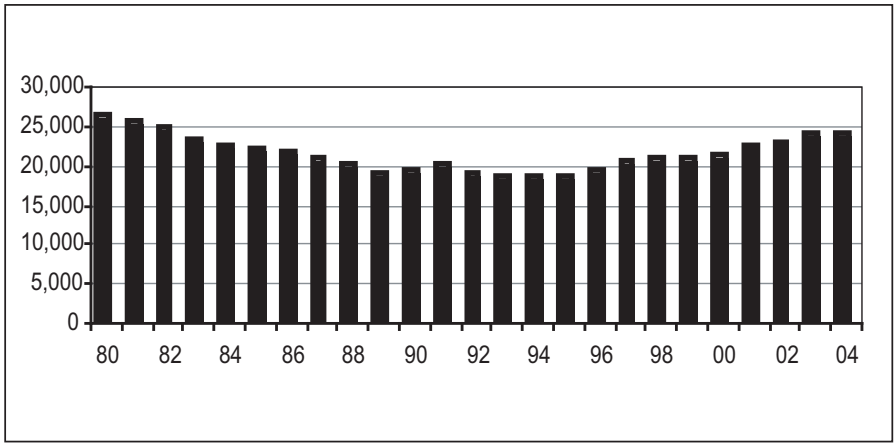
Phone: 353-1-6352751

Fax: 353-1-6353745

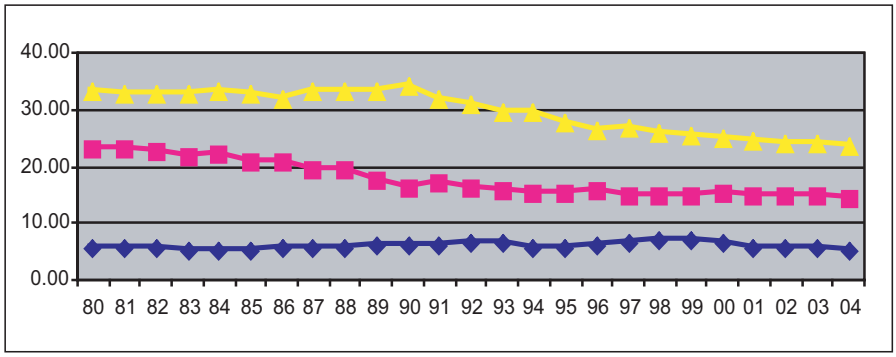
E-mail: virginia.delaney@mailf.hse.ie

Ireland: Dublin

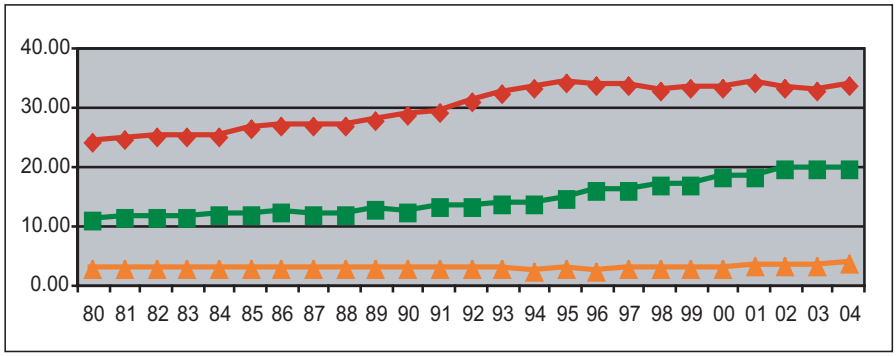
Total births by year



Percentage of births by maternal age



legenda: %births < 20 %births 20-24 %births 25-29



legenda: %births 30-34 %births 35-39 %births 40+

Ireland: Dublin, 2004

Live births (LB)*	23,786
Stillbirths (SB)*	130
Total births*	23,916
Number of terminations of pregnancy (ToP) for birth defects	not permitted

Birth Defects	Number of cases			Rates*10,000
	LB	SB	ToP	Total rate
Anencephaly	3	2		2.09
Spina bifida	9	1		4.18
Encephalocele	4	2		2.51
Microcephaly	5	1		2.51
Arhinencephaly / Holoprosencephaly	1	0		0.42
Hydrocephaly	4	2		2.51
Anophthalmos	0	0		0.00
Microphthalmos	3	0		1.25
Unspecified Anophthalmos/ Microphthalmos	0	0		---
Anotia	0	0		0.00
Microtia	0	0		0.00
Unspecified Anotia/Microtia	0	0		---
Transposition of great vessels	11	0		4.60
Tetralogy of Fallot	4	1		2.09
Hypoplastic left heart syndrome	3	0		1.25
Coarctation of aorta	18	0		7.53
Choanal atresia, bilateral	1	0		0.42
Cleft palate without cleft lip	17	0		7.11
Cleft lip with or without cleft palate	12	1		5.44
Oesophageal atresia / stenosis with or without fistula	6	0		2.51
Small intestine atresia / stenosis	4	1		2.09
Anorectal atresia / stenosis	7	0		2.93
Undescended testis (36 weeks of gestation or later)	nr	nr		nr
Hypospadias	24	0		10.04
Epispadias	nr	nr		nr
Indeterminate sex	1	0		0.42
Renal agenesis	2	2		1.67
Cystic kidney	8	0		3.35
Bladder exstrophy	1	0		0.42
Polydactyly, preaxial	11	0		4.60
Total Limb reduction defects (include unspecified)	7	0		2.93
Transverse	nr	nr		nr
Preaxial	nr	nr		nr
Postaxial	nr	nr		nr
Intercalary	nr	nr		nr
Mixed	nr	nr		nr
Unspecified	nr	nr		---
Diaphragmatic hernia	7	1		3.35
Omphalocele	5	2		2.93
Gastroschisis	10	0		4.18
Unspecified Omphalocele/Gastroschisis	0	0		---
Prune belly sequence	2	0		0.84
Trisomy 13	1	0		0.42
Trisomy 18	5	2		2.93
Down syndrome, all ages (include age unknown)	40	7		19.65
<20	2	0		16.53
20-24	3	0		8.83
25-29	6	0		10.76
30-34	4	1		6.21
35-39	21	3		51.31
40-44	4	3		87.36
45+	0	0		0.00
unknown	0	0		---

Note: stillbirths by maternal age not available
* estimated

Ireland: Dublin, Previous years rates 1980 - 2004

Birth prevalence rates: (LB+SB) * 10,000

	1974-79	1980-84	1985-89	1990-94	1995-99	2000-04
Total births		**121,428**	**102,584**	**94,474**	**99,207**	**113,437**
Anencephaly		14.99	8.48	5.61	3.12	3.00
Spina bifida		14.08	11.41	6.35	5.44	4.14
Encephalocele		2.72	1.46	2.43	1.71	1.15
Microcephaly		3.79	3.51	3.49	4.54	3.26
Arhinencephaly / Holoprosencephaly		0.33	0.39	0.42	0.91	1.23
Hydrocephaly		nr	nr	nr	2.10*	2.20
Anophthalmos		0.25	0.10	0.32	0.71	0.26
Microphthalmos		0.58	1.46	1.06	2.72	0.88
Unspecified Anophthalmos / Microphthalmos		---	---	---	---	---
Anotia		nr	nr	nr	nr	0.00*
Microtia		nr	nr	nr	nr	0.00*
Unspecified Anotia / Microtia		---	---	---	---	---
Transposition of great vessels		nr	nr	nr	5.32*	4.50
Tetralogy of Fallot		2.72	2.73	3.07	3.93	2.12
Hypoplastic left heart syndrome		2.31	1.66	2.65	1.92	2.12
Coarctation of aorta		4.45	6.63	5.29	7.16	6.35
Choanal atresia, bilateral		0.41	0.58	0.74	1.92	1.15
Cleft palate without cleft lip		7.16	6.92	7.94	7.96	8.29
Cleft lip with or without cleft palate		10.29	7.31	8.68	9.17	7.14
Oesophageal atresia / stenosis with or without fistula		3.71	4.00	2.96	3.53	2.29
Small intestine atresia / stenosis		2.55	3.02	2.12	2.12	2.38
Anorectal atresia / stenosis		3.46	3.80	3.07	2.42	2.29
Undescended testis (36 weeks of gestation or later)		nr	nr	nr	nr	nr
Hypospadias		15.24	10.04	12.70	18.35	12.96
Epispadias		nr	nr	nr	nr	0.00*
Indeterminate sex		0.16	0.19	0.21	0.40	0.18
Renal agenesis		4.86	4.58	4.34	4.23	2.64
Cystic kidney		3.71	1.85	4.23	3.63	2.73
Bladder exstrophy		nr	nr	nr	0.62*	0.44
Polydactyly, preaxial		6.75	5.26	6.03	6.45	8.37
Total Limb reduction defects (include unspecified)		4.28	3.41	4.45	4.64	3.44
Transverse		nr	nr	nr	nr	nr
Preaxial		nr	nr	nr	nr	nr
Postaxial		nr	nr	nr	nr	nr
Intercalary		nr	nr	nr	nr	nr
Mixed		nr	nr	nr	nr	nr
Unspecified		---	---	---	---	---
Diaphragmatic hernia		2.96	4.09	5.08	3.83	4.32
Omphalocele		2.72	2.63	1.69	2.52	3.70
Gastroschisis		0.16	0.58	0.85	2.02	3.00
Unspecified Omphalocele / Gastroschisis		---	---	---	---	---
Prune belly sequence		0.08	0.39	0.32	0.60	0.44
Trisomy 13		1.07	1.27	0.42	2.52	2.64
Trisomy 18		2.39	1.56	3.39	3.83	3.88
Down syndrome, all ages (include age unknown)		18.20	19.11	20.75	21.67	20.80
<20		nr	nr	17.58*	11.07	4.77
20-24		nr	nr	10.39*	7.41	6.59
25-29		nr	nr	10.23*	8.45	8.43
30-34		nr	nr	15.75*	19.50	12.73
35-39		nr	nr	44.06*	46.40	47.47
40-44		nr	nr	192.74*	128.88	133.02
45+		nr	nr	1153.85*	400.00	285.71
unknown		---	---	---	---	---

* data include less than 5 years

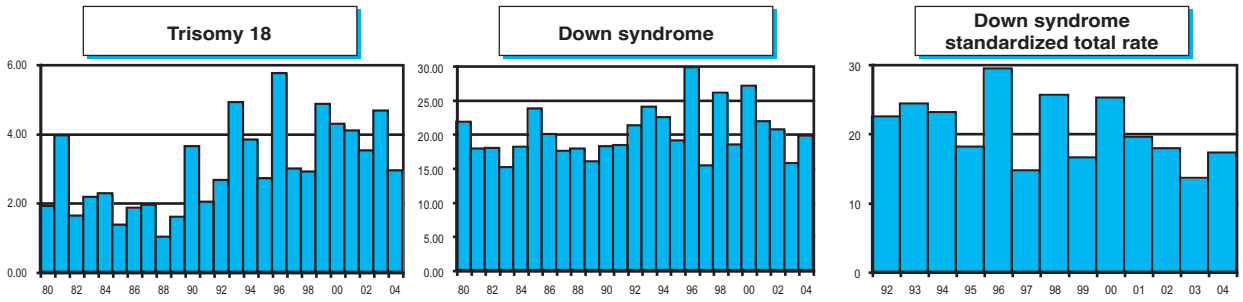
nr = not reported

Ireland: Dublin

Time trends 1980-2004 (Birth prevalence rates per 10,000)

Note: L+S rates

Note: L+S rates



Note: L+S rates

Israel: IBDSP

Israel Birth Defects Surveillance Program

History:
The program started in one hospital in 1966 and was a founder member of the Clearinghouse. It was a full member until 1986, when it becamean associate member.

Size and coverage
Reports are now obtained from five hospitals: three are located in the central region of the country, one in the south (with the largest number of births in the country) and one in the north. The total number of births is now around 40,000 per year (27% of all births in Israel).
Stillbirths of 20 weeks gestation or more and 500 grams or more are included.
The registry of termination of pregnancy began in 1995.

Legislation and funding
The Registry is a research program supported by research grants without any governmental support.

Sources of ascertainment
Reporting is voluntary. Reports are obtained from the Departments of Neonatology in the participating hospitals. The included hospitals are:
Rabin Medical Center , Beilinson Hospital , Petah Tikva (Prof Sirota, Prof Linder)
Lis Medical Center, Tel-Aviv (Prof Dolberg);
these two hospitals are affiliated to Sackler School of Medicine, Tel-Aviv University.
Kaplan Hospital, Rehovot (Prof Shinwell) affiliated to Hebrew University,Jerusalem.
The hospital in the south is Soroka Medical Center, Beer Sheva (Dr Landau, Prof Zmora) affiliated to Ben-Gurion University of the Negev, Beer-Sheva.
The hospital in the north is Bnai Zion Medical Center, Haifa (Dr Bader), affiliated to Faculty of Medicine, Technion, Haifa.

Exposure information
Complete anamneses are obtained by interviews of mothers of all malformed infants. The socio-demographic data of all the other women with normal newborns are also obtained.

Background information
Epidemiological information on all births occurring in the participating hospitals is available.

Addresses and Staff:
Prof Paul Merlob Rabin Medical Center and Schneider Children Hospital, Petah-Tikva, 49100

Phone: 972- 052 – 567 22 32

Fax: 972 – 3- 922 00 68

E-mail: merlobp@post.tau.ac.il

Dr Daniela Landau Department of Neonatology Soroka Medical Center Beer-Sheva

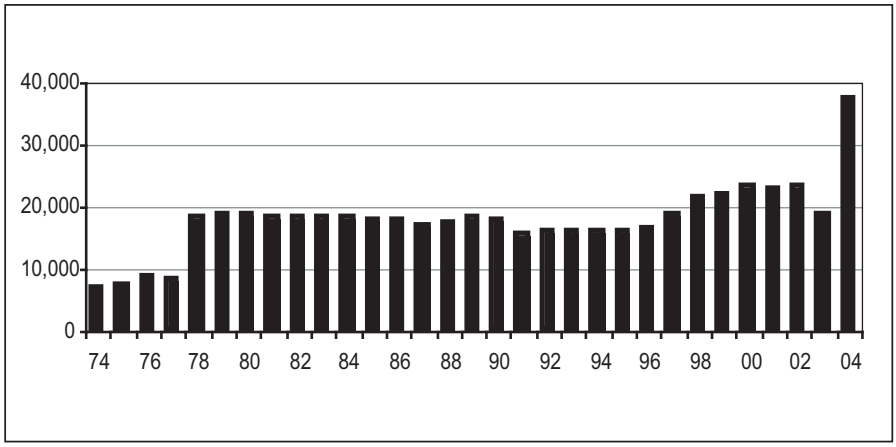
Phone: 972 – 8- 6400272

Fax: 972 – 8 – 6400545

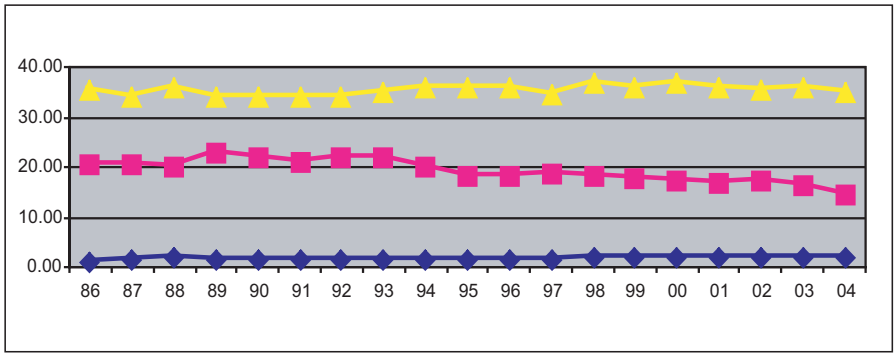
E-mail: landaud@bgumail.bgu.ac.il

Israel: IBDSP

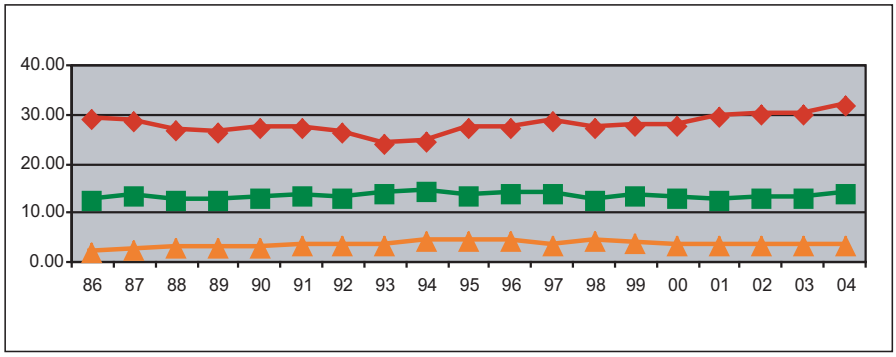
Total births by year



Percentage of births by maternal age



legenda: %births < 20 %births 20-24 %births 25-29



legenda: %births 30-34 %births 35-39 %births 40+

Israel: IBDSP, 2004

Live births (LB)	37,205
Stillbirths (SB)	284
Total births	37,489
Number of terminations of pregnancy (ToP) for birth defects	47

Birth Defects	Number of cases			Rates*10,000
	LB	SB	ToP	Total rate
Anencephaly	8	2	6	4.26
Spina bifida	13	1	5	5.06
Encephalocele	1	0	2	0.80
Microcephaly	6	0	0	1.60
Arhinencephaly / Holoprosencephaly	0	0	1	0.27
Hydrocephaly	17	0	6	6.13
Anophthalmos	0	0	0	0.00
Microphthalmos	2	0	0	0.53
Unspecified Anophthalmos/ Microphthalmos	0	0	0	---
Anotia	0	0	0	0.00
Microtia	5	0	0	1.33
Unspecified Anotia/Microtia	0	0	0	---
Transposition of great vessels	7	0	2	2.40
Tetralogy of Fallot	18	2	0	5.33
Hypoplastic left heart syndrome	8	0	0	2.13
Coarctation of aorta	6	0	0	1.60
Choanal atresia, bilateral	0	0	0	0.00
Cleft palate without cleft lip	7	0	0	1.86
Cleft lip with or without cleft palate	13	1	2	4.26
Oesophageal atresia / stenosis with or without fistula	5	0	0	1.33
Small intestine atresia / stenosis	8	0	0	2.13
Anorectal atresia / stenosis	7	0	0	1.86
Undescended testis (36 weeks of gestation or later)	nr	nr	nr	nr
Hypospadias	113	0	0	30.10
Epispadias	0	0	0	0.00
Indeterminate sex	1	0	0	0.27
Renal agenesis	5	0	1	1.60
Cystic kidney	3	0	1	1.07
Bladder exstrophy	0	0	1	0.27
Polydactyly, preaxial	3	0	0	0.80
Total Limb reduction defects (include unspecified)	12	1	1	3.73
Transverse	5	1	0	1.60
Preaxial	6	0	1	1.86
Postaxial	0	0	0	0.00
Intercalary	1	0	0	0.27
Mixed	0	0	0	0.00
Unspecified	0	0	0	---
Diaphragmatic hernia	7	0	1	2.13
Omphalocele	1	1	0	0.53
Gastroschisis	0	0	0	0.00
Unspecified Omphalocele/Gastroschisis	0	0	0	---
Prune belly sequence	0	0	0	0.00
Trisomy 13	2	0	1	0.80
Trisomy 18	0	0	2	0.53
Down syndrome, all ages (include age unknown)	25	0	14	10.39
<20	1	0	1	32.21
20-24	3	0	1	7.32
25-29	2	0	3	3.81
30-34	6	0	3	7.54
35-39	4	0	5	17.64
40-44	8	0	1	79.93
45+	1	0	0	76.34
unknown	0	0	0	---

nr = not reported

Israel: IBDSP, Previous years rates 1974 - 2004

Birth prevalence rates: (LB+SB) * 10,000 until 1985
Birth prevalence rates: (LB+SB+TOP) * 10,000 since 1986

	1974-79	1980-84	1985-89	1990-94	1995-99	2000-04
Births	**68,298**	**91,287**	**87,639**	**80,496**	**93,848**	**125,179**
Anencephaly	5.27	4.71	2.62	0.75	0.85	2.24
Spina bifida	2.78	5.59	4.91	2.61	2.02	3.99
Encephalocele	0.44	0.33	0.57	0.87	0.32	0.48
Microcephaly	nr	nr	nr	nr	0.92*	2.80
Arhinencephaly / Holoprosencephaly	nr	0.00*	0.23	0.50	0.00	0.08
Hydrocephaly	3.66	3.61	2.74	3.85	5.86	6.07
Anophthalmos	0.00	0.00	0.00	0.00	0.00	0.16
Microphthalmos	0.88	0.11	0.57	0.50	0.96	0.56
Unspecified Anophthalmos / Microphthalmos	---	---	---	---	---	---
Anotia	0.00	0.00	0.00	0.12	0.00	0.00
Microtia	0.59	1.10	0.91	2.36	1.81	1.04
Unspecified Anotia / Microtia	---	---	---	---	---	---
Transposition of great vessels	nr	nr	3.01*	3.60	3.84	3.91
Tetralogy of Fallot	nr	0.77	2.40	3.35	3.20	4.39
Hypoplastic left heart syndrome	nr		2.01*	2.36	2.45	2.08
Coarctation of aorta	nr	0.33	1.03	2.86	2.45	2.80
Choanal atresia, bilateral	nr	0.00*	0.34	0.25	0.11	0.16
Cleft palate without cleft lip	3.66	4.49	5.36	5.59	4.26	3.51
Cleft lip with or without cleft palate	5.12	4.38	6.05	4.97	4.58	5.51
Oesophageal atresia / stenosis with or without fistula	1.46	1.53	2.74	3.73	3.09	1.44
Small intestine atresia / stenosis	nr	0.55*	1.14	1.61	0.53	1.04
Anorectal atresia / stenosis	1.61	3.40	3.31	3.60	2.98	1.60
Undescended testis (36 weeks of gestation or later)	nr	nr	nr	nr	nr	nr
Hypospadias	29.14	26.73	30.24	43.85	34.52	37.63
Epispadias	0.15	0.00	0.11	0.25	0.11	0.32
Indeterminate sex	nr	nr	nr	nr	nr	0.16*
Renal agenesis	nr	nr	0.72*	0.99	0.53	0.64
Cystic kidney	0.59	0.55	1.48	0.99	1.49	2.08
Bladder exstrophy	0.15	0.22	0.68	0.37	0.32	0.24
Polydactyly, preaxial	0.29	0.66	0.46	0.37	1.07	0.96
Total Limb reduction defects (include unspecified)	3.51	2.85	2.51	3.60	1.60	2.16
Transverse	nr	0.55*	1.03	1.99	0.43	0.88
Preaxial	nr	0.55*	0.68	0.12	0.75	0.96
Postaxial	nr	0.55*	0.11	0.37	0.32	0.08
Intercalary	nr	0.37*	0.34	0.25	0.11	0.16
Mixed	nr	0.73*	0.34	0.87	0.00	0.08
Unspecified	---	---	---	---	---	---
Diaphragmatic hernia	2.15*	2.52	1.83	2.98	1.49	1.92
Omphalocele	1.90	2.52	0.80	1.12	0.43	0.88
Gastroschisis	0.00*	0.44	0.57	0.00	0.11	0.40
Unspecified Omphalocele / Gastroschisis	---	---	---	---	---	---
Prune belly sequence	0.44	0.22	0.00	0.12	0.11	0.08
Trisomy 13	nr	1.10*	0.34	0.50	0.32	0.56
Trisomy 18	nr	0.55*	0.80	0.50	0.85	1.52
Down syndrome, all ages (include age unknown)	11.27	9.64	12.32	7.58	8.31	10.39
<20	nr	nr	nr	0.00*	0.00	13.27
20-24	nr	nr	nr	0.00*	2.37	3.95
25-29	nr	nr	nr	2.74*	5.06	5.38
30-34	nr	nr	nr	7.50*	6.57	7.73
35-39	nr	nr	nr	15.27*	14.50	22.07
40-44	nr	nr	nr	39.76*	56.84	70.98
45+	nr	nr	nr	57.80*	99.26	121.95
unknown	---	---	---	---	---	---

* data include less than 5 years

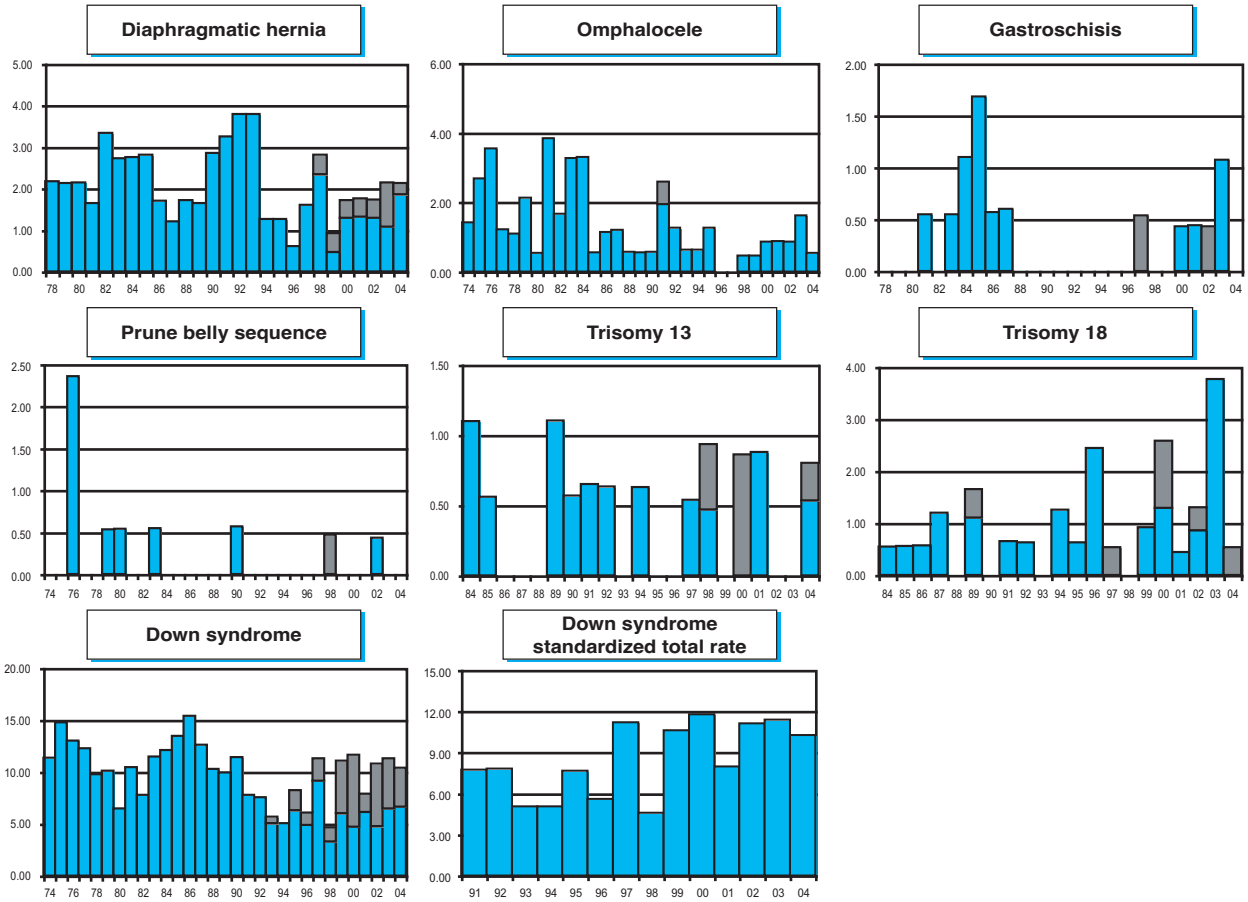
nr= not reported

Israel: IBDSP

'Time trends 1974-2004 (Birth prevalence rates per 10,000)

Note: L+S rates, ToP rates

Note: L+S rates, ToP rates



Note: L+S rates, ToP rates

Italy: BDRCam

Birth Defects Registry of Campania (BDRCam)

History:
The Registry started in 1991and became a full member of the ICBDSR in 1996.

Size and coverage
The Registry is based on reporting from hospitals distributed in Campania, a region in southern Italy. Naples is the main city. Initially 38 hospitals reported and the annual number of births was 38.000. Until 2001 the registry is hospital-based covering approximately 50.000 annual births. Actually beginning from 2002, the registry is population based covering approximately 100% of all births. Stillbirths and induced abortions are included. In 2002 is started officially a link with birth regional registry.

Legislation and funding
The Registry is a surveillance Programme supported by grants from Regional Health Authorities. Participation was voluntary up to 1995. From 1996 participation is mandatory.

Sources of ascertainment
Reports are obtained from delivery units and pediatric clinics at the participating hospitals. For selected malformations multiple sources are used with follow-up to one year using specific records from pediatric specialties departments dealing with malformed infants.

Exposure information
For each malformed infant reported, information is given on certain exposures, including maternal drug usage and parental occupation. Beginning from 2002 informations on controls are available but only partially on induced abortions.

Background information
Always from 2002 background information is given on certain exposures, including maternal drug usage and parental occupation. Informations on controls are available.

Addresses and staff

Gioacchino Scarano, Registro Campano Difetti Congeniti (BDRCam), Medical Genetics Division, Azienda Ospedaliera "G. Rummo", Via dell'Angelo 1, 82100 Benevento, Italy

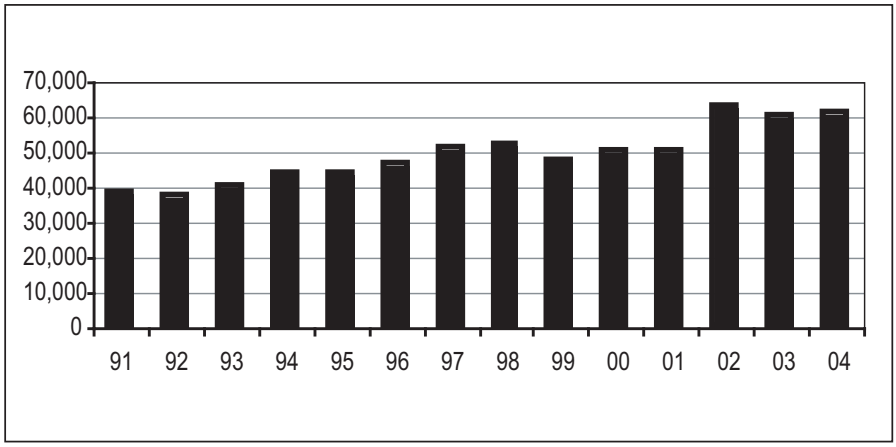
Phone: +39- 0824-57374

Fax: + 39-0824-57495

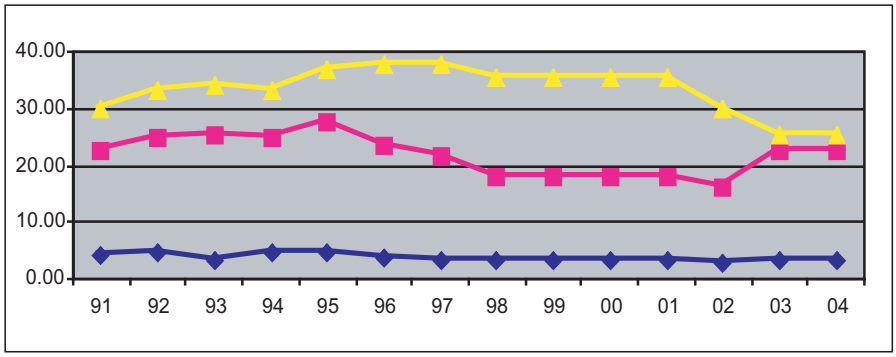
E-mail: giorecam@tin.it

Italy: BDRCam

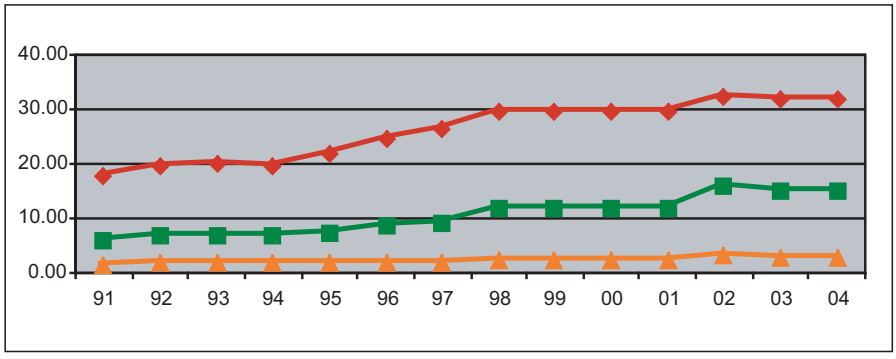
Total births by year



Percentage of births by maternal age



legenda: %births < 20 %births 20-24 %births 25-29



legenda: %births 30-34 %births 35-39 %births 40+

Italy: BDRCAM, 2004

Live births (LB)	60,781
Stillbirths (SB)	125
Total births	60,906
Number of terminations of pregnancy (ToP) for birth defects	288

Birth Defects	Number of cases			Rates*10,000
	LB	SB	ToP	Total rate
Anencephaly	1	1	18	3.27
Spina bifida	4	0	19	3.76
Encephalocele	0	0	2	0.33
Microcephaly	1	0	2	0.49
Arhinencephaly / Holoprosencephaly	1	0	7	1.31
Hydrocephaly	2	0	51	8.66
Anophthalmos	0	0	0	0.00
Microphthalmos	2	0	0	0.33
Unspecified Anophthalmos/ Microphthalmos	0	0	0	---
Anotia	2	0	0	0.33
Microtia	5	0	0	0.82
Unspecified Anotia/Microtia	0	0	0	---
Transposition of great vessels	2	0	3	0.82
Tetralogy of Fallot	8	0	7	2.45
Hypoplastic left heart syndrome	1	0	13	2.29
Coarctation of aorta	4	1	0	0.82
Choanal atresia, bilateral	1	0	0	0.16
Cleft palate without cleft lip	17	1	2	3.27
Cleft lip with or without cleft palate	29	1	10	6.54
Oesophageal atresia / stenosis with or without fistula	7	0	2	1.47
Small intestine atresia / stenosis	4	1	2	1.14
Anorectal atresia / stenosis	8	0	6	2.29
Undescended testis (36 weeks of gestation or later)	nr	nr	nr	nr
Hypospadias*	29	0	1	4.90
Epispadias	0	0	0	0.00
Indeterminate sex	4	0	0	0.65
Renal agenesis	5	0	9	2.29
Cystic kidney	3	0	10	2.12
Bladder exstrophy	0	0	0	0.00
Polydactyly, preaxial	6	0	0	0.98
Total Limb reduction defects (include unspecified)	19	0	6	4.09
Transverse	8	0	4	1.96
Preaxial	5	0	1	0.98
Postaxial	6	0	0	0.98
Intercalary	0	0	1	0.16
Mixed	0	0	0	0.00
Unspecified	0	0	0	---
Diaphragmatic hernia	5	0	2	1.14
Omphalocele	4	0	9	2.12
Gastroschisis	0	0	1	0.16
Unspecified Omphalocele/Gastroschisis	0	0	0	---
Prune belly sequence	0	0	1	0.16
Trisomy 13	1	0	5	0.98
Trisomy 18	1	1	11	2.12
Down syndrome, all ages (include age unknown)	29	0	64	15.20
<20	0	0	1	5.13
20-24	0	0	2	1.47
25-29	4	0	1	3.27
30-34	11	0	11	11.39
35-39	7	0	26	36.50
40-44	3	0	17	154.32
45+	1	0	1	46.84
unknown	3	0	5	---

* = excluded glandular hypospadia
nr = not reported

Italy: BDRCAM, Previous years rates 1991 - 2004

Birth prevalence rates: (LB+SB) * 10,000 until 1993
Birth prevalence rates: (LB+SB+TOP) * 10,000 from 1994

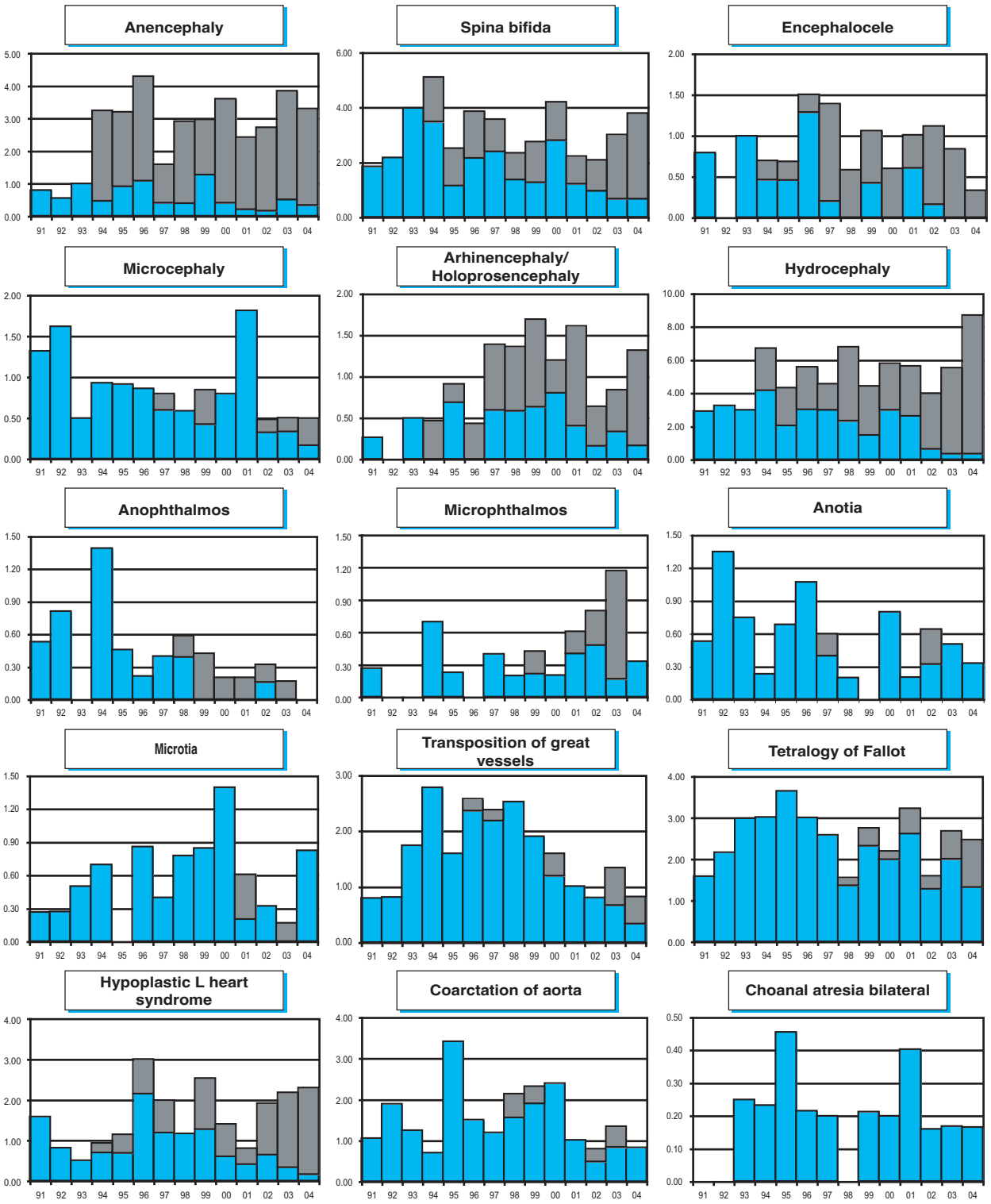
	1974-79	1980-84	1985-89	1990-94*	1995-99	2000-04
Total births				**158,531**	**240,032**	**283,377**
Anencephaly				1.45	2.96	3.18
Spina bifida				3.34	3.00	3.03
Encephalocele				0.63	1.04	0.78
Microcephaly				1.07	0.79	0.78
Arhinencephaly / Holoprosencephaly				0.32	1.17	1.09
Hydrocephaly				4.04	5.17	5.93
Anophthalmos				0.69	0.42	0.18
Microphthalmos				0.25	0.25	0.64
Unspecified Anophthalmos / Microphthalmos				---	---	---
Anotia				0.69	0.50	0.49
Microtia				0.44	0.58	0.64
Unspecified Anotia / Microtia				---	---	---
Transposition of great vessels				1.58	2.21	1.09
Tetralogy of Fallot				2.46	2.67	2.40
Hypoplastic left heart syndrome				0.95	1.96	1.76
Coarctation of aorta				1.20	2.08	1.24
Choanal atresia, bilateral				0.13	0.21	0.21
Cleft palate without cleft lip				4.73	4.29	4.13
Cleft lip with or without cleft palate				6.81	7.17	5.75
Oesophageal atresia / stenosis with or without fistula				2.33	2.25	1.62
Small intestine atresia / stenosis				1.83	2.17	1.48
Anorectal atresia / stenosis				2.84	3.04	2.65
Undescended testis (36 weeks of gestation or later)				nr	nr	nr
Hypospadias				3.66	2.79	5.75
Epispadias				0.25	0.21	0.11
Indeterminate sex				0.25	0.67	0.85
Renal agenesis				1.32	3.25	3.67
Cystic kidney				1.64	2.71	2.01
Bladder exstrophy				0.19	0.33	0.00
Polydactyly, preaxial				1.77	1.75	1.45
Total Limb reduction defects (include unspecified)				5.30	4.79	4.52
Transverse				3.78	2.42	2.72
Preaxial				0.63	0.92	0.81
Postaxial				0.25	0.54	0.53
Intercalary				0.32	0.46	0.35
Mixed				0.19	0.12	0.04
Unspecified				---	---	---
Diaphragmatic hernia				1.77	2.29	2.33
Omphalocele				1.32	2.00	2.08
Gastroschisis				0.32	0.62	0.60
Unspecified Omphalocele / Gastroschisis				---	---	---
Prune belly sequence				0.00	0.13*	0.09*
Trisomy 13				0.82	0.71	0.67
Trisomy 18				0.76	1.79	1.80
Down syndrome, all ages (include age unknown)				12.05	13.71	12.67
<20				4.80	3.52	4.60
20-24				6.48	4.46	2.00
25-29				7.35	6.02	4.96
30-34				13.47	11.00	7.38
35-39				38.39	31.86	29.46
40-44				72.76	140.29	101.80
45+				185.19	232.56	116.73
unknown				---	---	---

* data include less than 5 years

nr = not reported

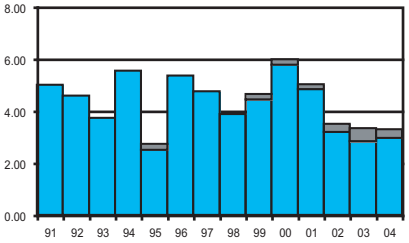
Italy: BDRCam

'Time trends 1991-2004 (Birth prevalence rates per 10,000)

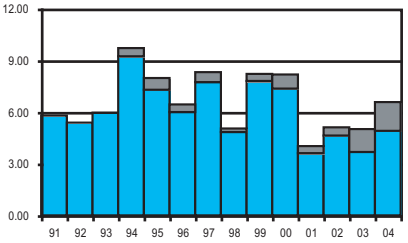


Note: L+S rates, ToP rates

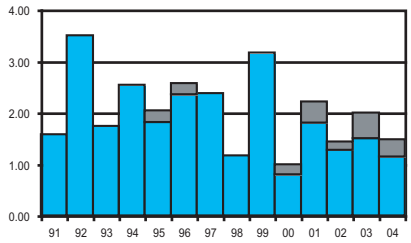
Cleft palate without cleft lip



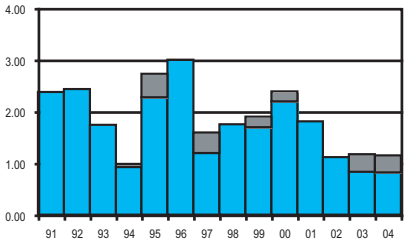
Cleft lip with or without cleft palate



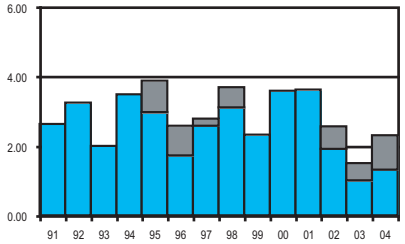
Oesophageal atresia/stenosis with or without fistula



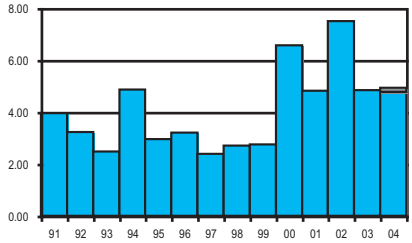
Small intestine atresia/stenosis



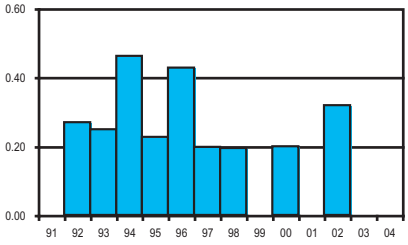
Anorectal atresia/stenosis



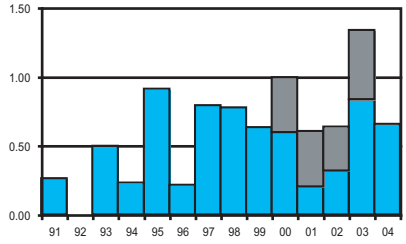
Hypospadias



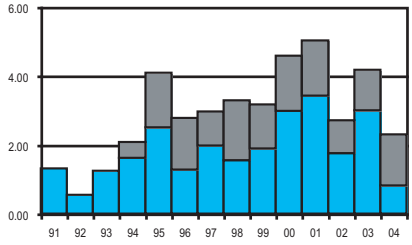
Epispadias



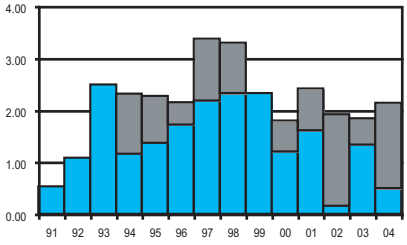
Indeterminate sex



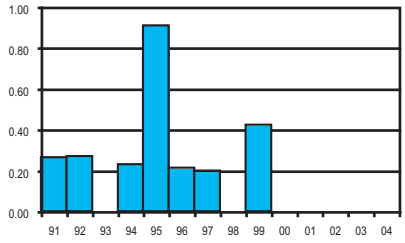
Renal agenesis



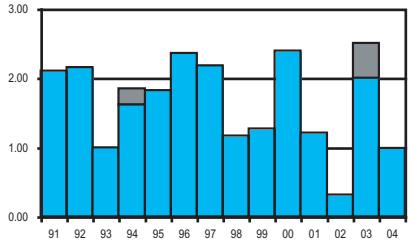
Cystic Kidney



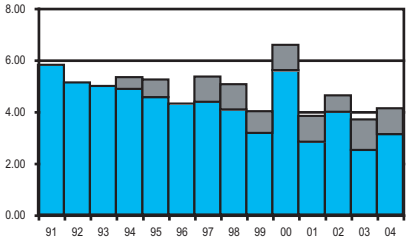
Bladder exstrophy



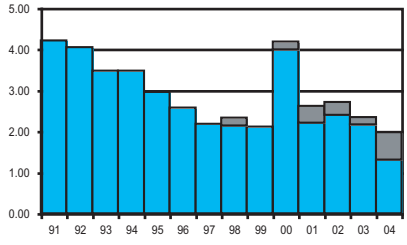
Polydactyly, preaxial



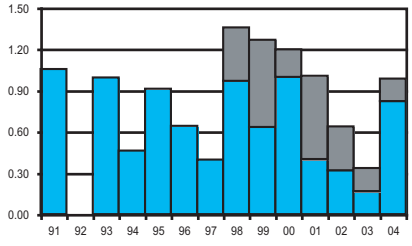
Limb reduction defects



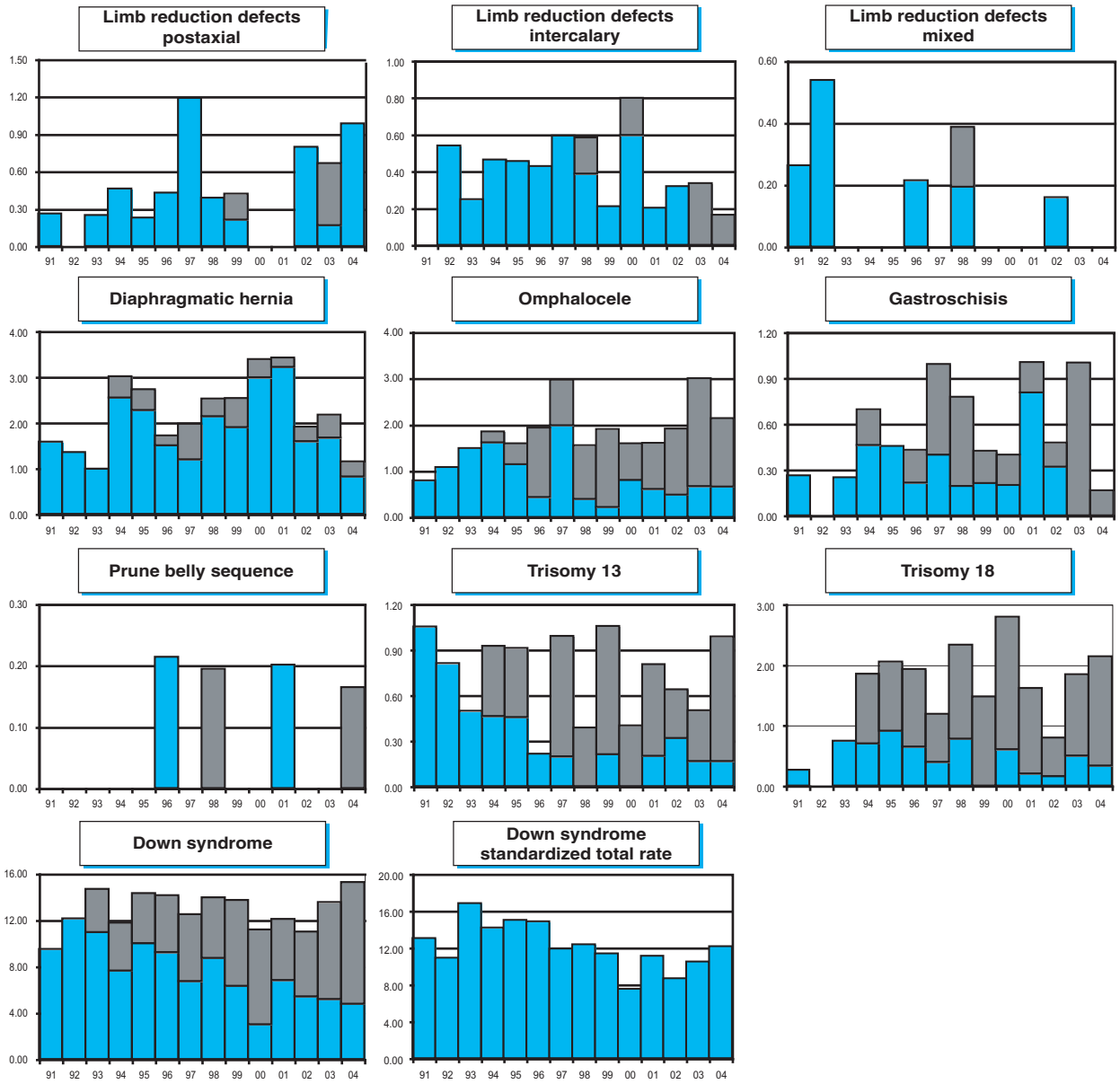
Limb reduction defects transverse



Limb reduction defects preaxial



Note: L+S rates, ToP rates



Note: L+S rates, ToP rates

Italy: IMER

Emilia-Romagna Registry of Congenital Malformations (IMER)

History:
The registry was started in 1978 in a few hospitals and has increased in size to now include 45 delivery units. The Programme became an associate member of the Clearinghouse in 1985.

Size and coverage:
The Programme is based on approximately 90% of all births in the Emilia-Romagna region, or approximately 25,000 annual births (4% of all births in Italy). Stillbirths of 28 weeks or more gestation are included.

Legislation and funding:
The Programme is recognised and financed by the health authorities, the National Research Council, and the Regional Health Council. Hospital participation is voluntary.

Sources of ascertainment:
Reporting is made by neonatologists and pediatricians during the first week of the infant's life. Selected malformations are followed up.

Exposure information:
Detailed exposure information is obtained by interviews of the mothers of malformed infants. For each malformed infant, a control is chosen (the baby born before or after the malformed case in the same hospital) and its mother is interviewed in a similar way.

Background information:
Some general demographic information is known for all births in the area. For each participating hospital, the number of livebirths and stillbirths are known.

Addresses and Staff:
Guido Cocchi, MD, Programme Director Istituto Pediatria Preventiva/Neonatologia Universita' di Bologna Via Massarenti 11, 40138 Bologna, Italy

Phone: 39-051-342754 /6364654

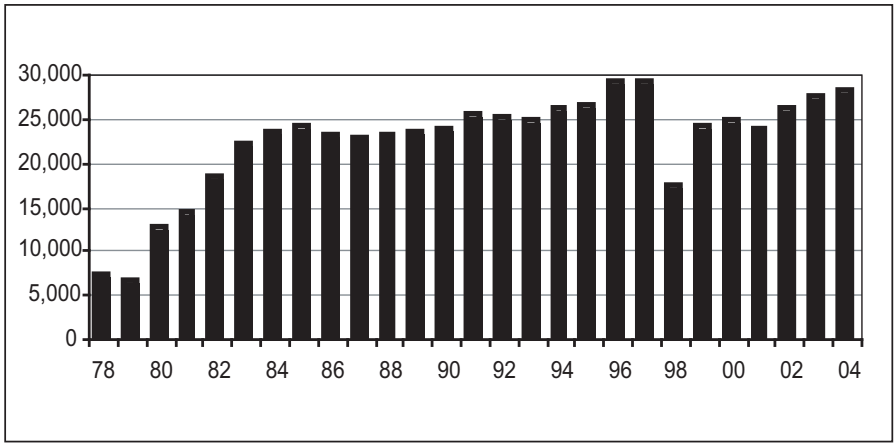
Fax: 39-051-342754

E-mail: cocchi@med.unibo.it

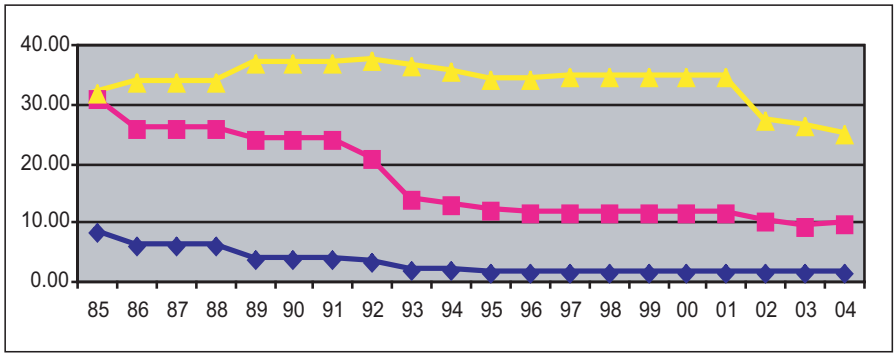
Website: www.unife.it/imer

Italy: IMER

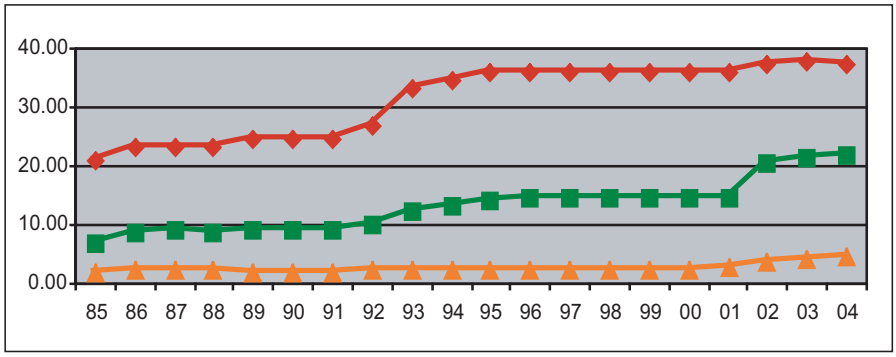
Total births by year



Percentage of births by maternal age



legenda: %births < 20 %births 20-24 %births 25-29



legenda: %births 30-34 %births 35-39 %births 40+

Italy: IMER, 2004

Live births (LB)	27,885
Stillbirths (SB)	77
Total births	27,962
Number of terminations of pregnancy (ToP) for birth defects	213

Birth Defects	Number of cases			Rates*10,000
	LB	SB	ToP	Total rate
Anencephaly	0	0	3	1.06
Spina bifida	4	0	14	6.39
Encephalocele	1	0	3	1.42
Microcephaly	3	0	1	1.42
Arhinencephaly / Holoprosencephaly	1	0	10	3.90
Hydrocephaly	3	0	22	8.87
Anophthalmos	2	0	0	0.71
Microphthalmos	5	0	2	2.48
Unspecified Anophthalmos/ Microphthalmos	0	0	0	0.00
Anotia	4	0	0	1.42
Microtia	0	0	0	0.00
Unspecified Anotia/Microtia	0	0	0	0.00
Transposition of great vessels	16	0	4	7.10
Tetralogy of Fallot	6	0	1	2.48
Hypoplastic left heart syndrome	4	0	4	2.84
Coarctation of aorta	9	0	1	3.55
Choanal atresia, bilateral	0	0	0	0.00
Cleft palate without cleft lip	11	0	0	3.90
Cleft lip with or without cleft palate	13	0	6	6.74
Oesophageal atresia / stenosis with or without fistula	8	0	1	3.19
Small intestine atresia / stenosis	13	0	0	4.61
Anorectal atresia / stenosis	8	0	10	6.39
Undescended testis (36 weeks of gestation or later)	nr	nr	nr	nr
Hypospadias	38	0	2	14.20
Epispadias	0	0	0	0.00
Indeterminate sex	1	0	0	0.35
Renal agenesis	9	0	3	4.26
Cystic kidney	6	0	8	4.97
Bladder exstrophy	0	0	0	0.00
Polydactyly, preaxial	6	0	0	2.13
Total Limb reduction defects (include unspecified)	9	0	11	7.10
Transverse	3	0	2	1.77
Preaxial	4	0	2	2.13
Postaxial	1	0	1	0.71
Intercalary	0	0	2	0.71
Mixed	0	0	1	0.35
Unspecified	1	0	3	1.42
Diaphragmatic hernia	10	0	12	7.81
Omphalocele	4	0	7	3.90
Gastroschisis	7	0	1	2.84
Unspecified Omphalocele/Gastroschisis	0	0	0	0.00
Prune belly sequence	0	0	0	0.00
Trisomy 13	1	0	7	2.84
Trisomy 18	3	0	14	6.03
Down syndrome, all ages (include age unknown)	16	0	43	20.94
<20	0	0	0	0.00
20-24	1	0	0	3.65
25-29	3	0	2	7.18
30-34	1	0	8	8.64
35-39	9	0	21	48.54
40-44	1	0	10	92.83
45+	1	0	0	188.68
unknown	0	0	2	952.38

nr = not reported

Italy: IMER, Previous years rates 1978 - 2004

Birth prevalence rates: (LB+SB) * 10,000 until 1993
Birth prevalence rates: (LB+SB+TOP) * 10,000 from 1994

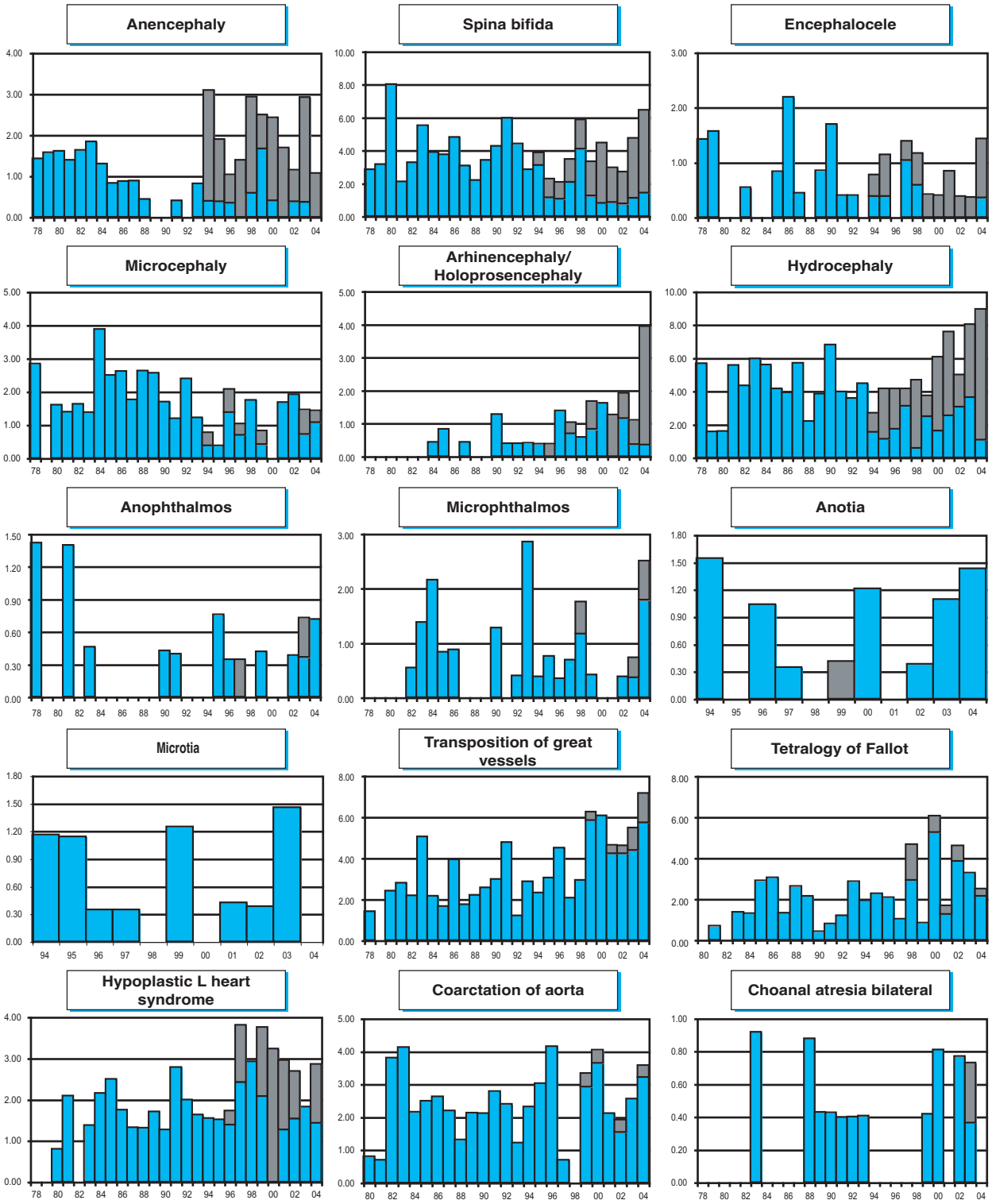
	1974-79*	1980-84	1985-89	1990-94	1995-99	2000-04
Total births	**13,407**	**90,077**	**115,805**	**124,129**	**125,188**	**129,766**
Anencephaly	1.49	1.55	0.60	0.89	1.84	1.85
Spina bifida	2.98	4.44	3.45	4.27	3.20	4.32
Encephalocele	1.49	0.11	0.86	0.64	0.80	0.69
Microcephaly	1.49	2.11	2.42	1.45	1.20	1.31
Arhinencephaly / Holoprosencephaly	0.00	0.11	0.26	0.56	1.04	2.00
Hydrocephaly	3.73	4.88	3.97	4.27	4.15	7.17
Anophthalmos	0.75	0.33	0.00	0.16	0.40	0.39
Microphthalmos	0.00	1.00	0.35	0.97	0.72	0.77
Unspecified Anophthalmos / Microphthalmos	---	---	---	---	---	---
Anotia	nr	nr	nr	1.55*	0.40	0.85
Microtia	nr	nr	nr	1.16*	0.64	0.46
Unspecified Anotia / Microtia	---	---	---	---	---	---
Transposition of great vessels	0.75	3.00	2.42	2.82	3.75	5.63
Tetralogy of Fallot	nr	0.78	2.42	1.45	2.00	3.62
Hypoplastic left heart syndrome	0.00	1.33	1.73	1.85	2.72	2.70
Coarctation of aorta	nr	2.55	2.16	2.18	2.40	2.85
Choanal atresia, bilateral	0.00	0.22	0.26	0.32	0.08	0.46
Cleft palate without cleft lip	3.73	4.66	6.56	5.40	4.47	3.93
Cleft lip with or without cleft palate	5.97	7.66	6.82	7.01	5.75	5.70
Oesophageal atresia / stenosis with or without fistula	3.73	3.77	3.97	3.63	3.20	3.47
Small intestine atresia / stenosis	1.49	2.22	3.28	3.79	3.04	2.85
Anorectal atresia / stenosis	0.75	3.55	2.68	3.14	2.24	4.16
Undescended testis (36 weeks of gestation or later)	nr	nr	nr	nr	nr	nr
Hypospadias	12.68	20.20	20.38	18.12*	16.09*	16.95
Epispadias	nr	nr	nr	nr	0.00	0.00
Indeterminate sex	nr	nr	nr	nr	0.16	0.39
Renal agenesis	4.48	1.00	1.64	1.45	2.48	4.39
Cystic kidney	0.75	0.33	1.04	0.24	3.91	3.62
Bladder exstrophy	0.75	0.33	0.78	0.08	0.16	0.23
Polydactyly, preaxial	8.95	8.66	8.29	7.41	3.59	2.31
Total Limb reduction defects (include unspecified)	nr	nr	5.53	5.64	3.99	4.70
Transverse	nr	nr	3.28	2.82	1.92	1.62
Preaxial	nr	nr	0.52	0.97	0.88	0.92
Postaxial	nr	nr	0.69	0.48	0.40	0.69
Intercalary	nr	nr	0.52	0.73	0.48	0.62
Mixed	nr	nr	0.26	0.48	0.08	0.15
Unspecified	---	---	---	---	---	---
Diaphragmatic hernia	0.00	1.67	1.81	3.38	2.80	4.08
Omphalocele	2.24	1.33	2.25	2.18	1.28	3.08
Gastroschisis	0.00	1.00	0.69	1.05	0.48	1.39
Unspecified Omphalocele / Gastroschisis	---	---	---	---	---	---
Prune belly sequence	0.00	0.33	0.43	0.24	0.40	0.23
Trisomy 13	0.75	1.55	0.78	0.73	0.96	1.93
Trisomy 18	0.75	1.55	0.78	0.89	2.16	4.93
Down syndrome, all ages (include age unknown)	21.63	13.32	13.30	13.45	19.73	18.73
<20	nr	nr	1.40	8.59	12.63	5.57
20-24	nr	nr	4.89	5.97	8.30	7.49
25-29	nr	nr	10.97	7.71	7.70	5.30
30-34	nr	nr	15.77	13.98	13.74	11.48
35-39	nr	nr	35.62	31.95	43.16	39.00
40-44	nr	nr	71.70	79.17	177.78	112.31
45+	nr	nr	52.36	158.73	135.14	181.82
unknown	---	---	---	---	---	---

* data include less than 6 or 5 years

nr = not reported

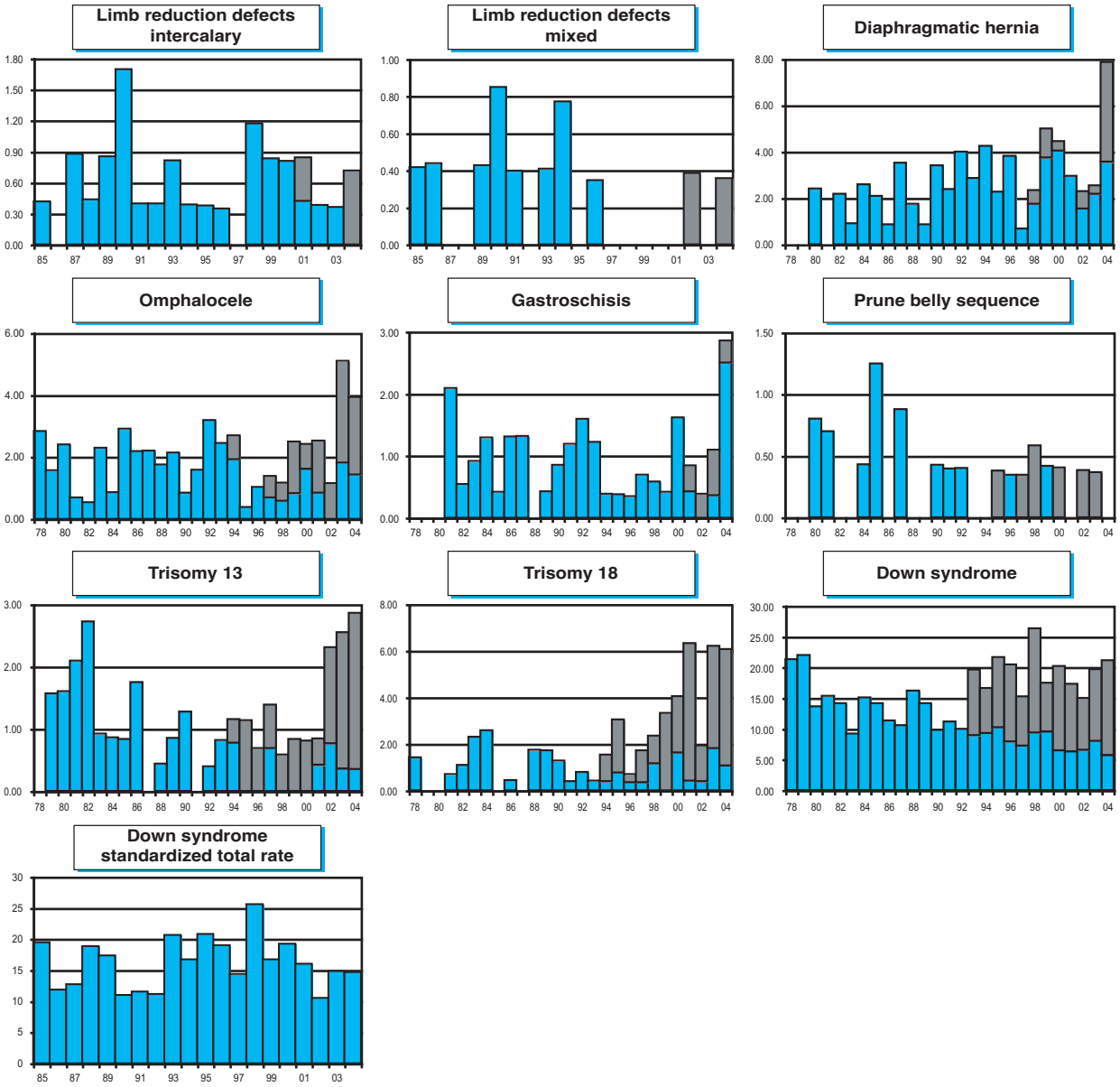
Italy: IMER

Time trends 1978-2003 (Birth prevalence rates per 10,000)



Note: L+S rates, ToP rates

Note: L+S rates, ToP rates



Note: L+S rates, ToP rates

Italy: ISMAC

Sicilian Registry of Congenital Malformations (ISMAC)

History:
The registry started in 1991 and became an associate member of the Clearinghouse in 1996.

Size and coverage
The Programme is hospital based and actually collaborates with four south-east provinces (Catania, Enna, Ragusa and Siracusa) of the nine Sicilian provinces, with a covering rate higher than 75% and with more than 19,000 controlled newborns for year. Stillbirths are included.

Legislation and funding
The Programme is a surveillance Programme with a voluntary participation, supported by ASMAC-Associazione per la Prevenzione Sociale e per il Trattamento delle Malformazioni Congenite.

Sources of ascertainment
Reports are obtained from delivery units, pediatric units and other specialistic departments.

Exposure information
for each malformed infant reported, information is given on certain exposures, including maternal drug usange and parental occupation. Up to now no information on controls is available.

Background information:
Up to now little background information is available and no information on controls is available.

Addresses and Staff:
Sebastiano Bianca, MD, Programme Director Sicilian Registry of Congenital Malformations (ISMAC) Via XXXI Maggio 51, 95123 Catania, Italy

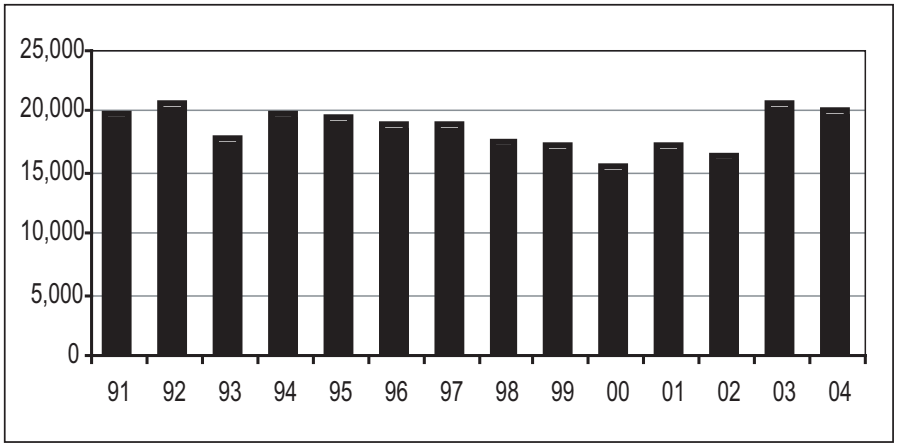
Phone: +39-095-3781111

Fax: +39-095-222532

E-mail: sebastiano.bianca@tiscali.it

Italy: ISMAC

Total births by year



Italy: ISMAC, 2004

Live births (LB)	19,880
Stillbirths (SB)	0
Total births	19,880
Number of terminations of pregnancy (ToP) for birth defects	nr

Birth Defects	Number of cases			Rates*10,000
	LB	SB	ToP	Total rate
Anencephaly	2	0	4	3.02
Spina bifida	2	0	3	2.52
Encephalocele	0	0	0	0.00
Microcephaly	3	0	0	1.51
Arhinencephaly / Holoprosencephaly	0	0	0	0.00
Hydrocephaly	3	0	8	5.53
Anophthalmos	0	0	0	0.00
Microphthalmos	2	0	0	1.01
Unspecified Anophthalmos/ Microphthalmos	0	0	0	0.00
Anotia	1	0	0	0.50
Microtia	0	0	0	0.00
Unspecified Anotia/Microtia	0	0	0	0.00
Transposition of great vessels	5	0	3	4.02
Tetralogy of Fallot	3	0	5	4.02
Hypoplastic left heart syndrome	1	0	4	2.52
Coarctation of aorta	4	0	0	2.01
Choanal atresia, bilateral	2	0	0	1.01
Cleft palate without cleft lip	6	0	0	3.02
Cleft lip with or without cleft palate	7	0	2	4.53
Oesophageal atresia / stenosis with or without fistula	7	0	0	3.52
Small intestine atresia / stenosis	3	0	0	1.51
Anorectal atresia / stenosis	3	0	0	1.51
Undescended testis (36 weeks of gestation or later)	12	0	0	6.04
Hypospadias	34	0	0	17.10
Epispadias	0	0	0	0.00
Indeterminate sex	0	0	0	0.00
Renal agenesis	2	0	3	2.52
Cystic kidney	1	0	2	1.51
Bladder exstrophy	0	0	0	0.00
Polydactyly, preaxial	3	0	0	1.51
Total Limb reduction defects (include unspecified)	3	0	3	3.02
Transverse	0	0	1	0.50
Preaxial	0	0	0	0.00
Postaxial	0	0	0	0.00
Intercalary	0	0	0	0.00
Mixed	0	0	0	0.00
Unspecified	3	0	2	2.52
Diaphragmatic hernia	4	0	3	3.52
Omphalocele	0	0	0	0.00
Gastroschisis	0	0	2	1.01
Unspecified Omphalocele/Gastroschisis	0	0	0	0.00
Prune belly sequence	0	0	0	0.00
Trisomy 13	2	0	0	1.01
Trisomy 18	1	0	3	2.01
Down syndrome, all ages (include age unknown)	11	0	8	9.56
<20	0	0	0	nr
20-24	0	0	0	nr
25-29	0	0	2	nr
30-34	0	0	2	nr
35-39	10	0	3	nr
40-44	1	0	1	nr
45+	0	0	0	nr
unknown	0	0	0	nr

nr = not reported

Italy: ISMAC, Previous years rates 1991 - 2004

Birth prevalence rates: (LB+SB) * 10,000
Birth prevalence rates: (LB+SB+TOP) * 10,000 only for 2000-2001-2004

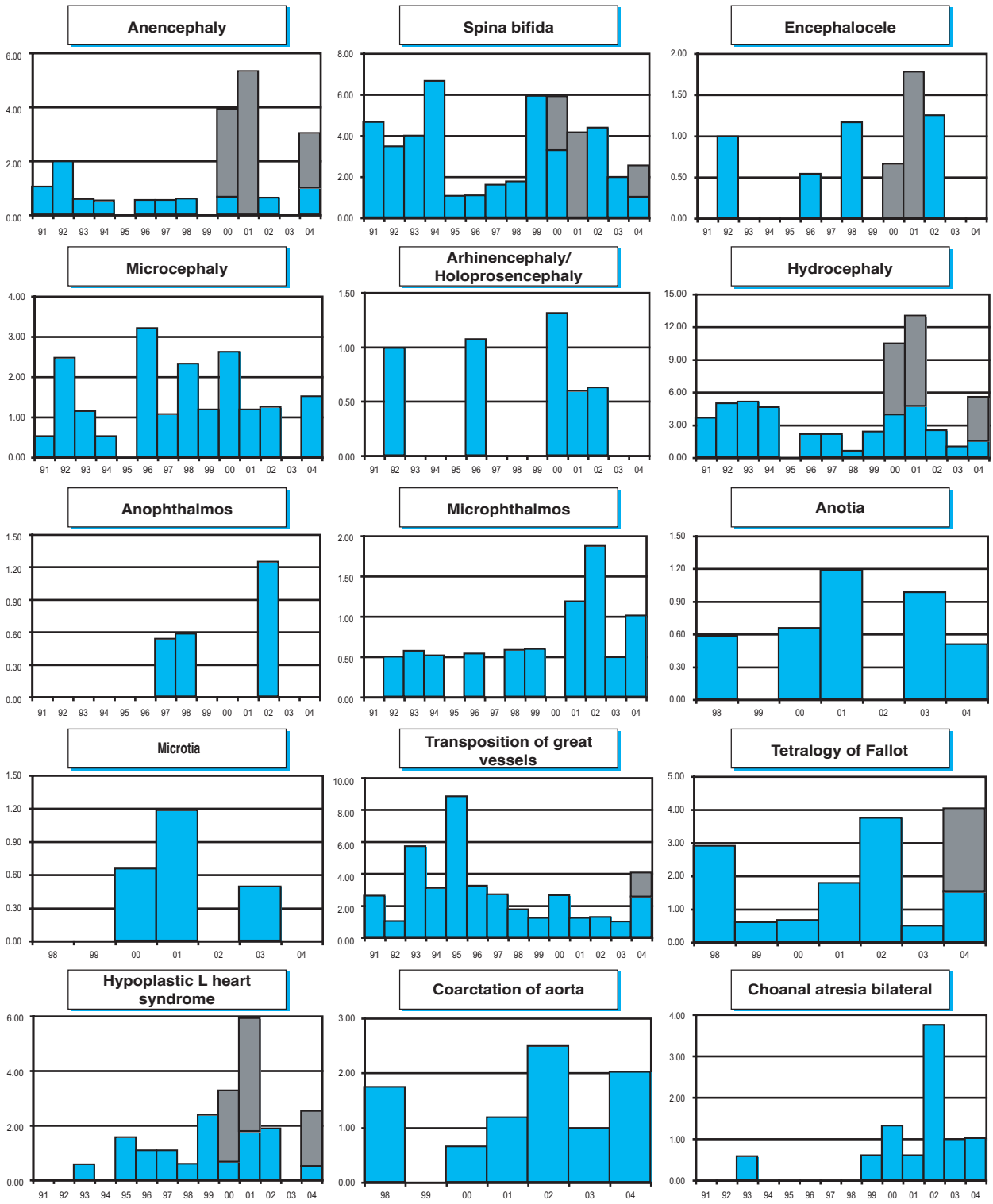
	1974-79	1980-84	1985-89	1990-94*	1995-99	2000-04
Total births				**76,898**	**91,033**	**88,656**
Anencephaly				1.04	0.33	2.48
Spina bifida				4.68	2.20	3.61
Encephalocele				0.26	0.33	0.68
Microcephaly				1.17	1.54	1.24
Arhinencephaly / Holoprosencephaly				0.26	0.22	0.45
Hydrocephaly				4.55	1.43	6.20
Anophthalmos				0.00	0.22	0.23
Microphthalmos				0.39	0.33	0.90
Unspecified Anophthalmos / Microphthalmos				---	---	---
Anotia				nr	0.29*	0.68
Microtia				nr	0.00*	0.45
Unspecified Anotia / Microtia				---	---	---
Transposition of great vessels				2.99	3.63	2.03
Tetralogy of Fallot				nr	1.75*	2.14
Hypoplastic left heart syndrome				0.13	1.32	2.59
Coarctation of aorta				nr	0.88*	1.47
Choanal atresia, bilateral				0.13	0.14*	1.47
Cleft palate without cleft lip				5.46	4.17	4.40
Cleft lip with or without cleft palate				6.50	7.14	4.40
Oesophageal atresia / stenosis with or without fistula				3.12	2.97	2.26
Small intestine atresia / stenosis				8.71	1.54	2.14
Anorectal atresia / stenosis				3.77	1.87	1.92
Undescended testis (36 weeks of gestation or later)				6.24	7.58	17.14
Hypospadias				nr	16.04*	22.00
Epispadias				0.00	0.28*	0.34
Indeterminate sex				0.52	0.11	0.68
Renal agenesis				0.39	2.20	1.24
Cystic kidney				0.78	1.32	2.71
Bladder exstrophy				0.13	0.41*	0.34
Polydactyly, preaxial				0.26	1.52*	2.59
Total Limb reduction defects (include unspecified)				3.25	2.75	3.61
Transverse				nr	1.46*	2.93
Preaxial				nr	0.00*	0.00
Postaxial				nr	0.58*	0.00
Intercalary				nr	0.00*	0.00
Mixed				nr	0.00*	0.00
Unspecified				---	---	---
Diaphragmatic hernia				1.69	1.65	2.71
Omphalocele				2.08	0.88	1.35
Gastroschisis				0.91	0.77	2.14
Unspecified Omphalocele / Gastroschisis				---	---	---
Prune belly sequence				0.00	0.14*	0.00
Trisomy 13				0.13	0.41*	1.92
Trisomy 18				0.52	0.55	1.69
Down syndrome, all ages (include age unknown)				13.78	11.64	10.83
<20				nr	nr	nr
20-24				nr	nr	nr
25-29				nr	nr	nr
30-34				nr	nr	nr
35-39				nr	nr	nr
40-44				nr	nr	nr
45+				nr	nr	nr
unknown				---	---	---

* data include less than 5 years

nr = not reported

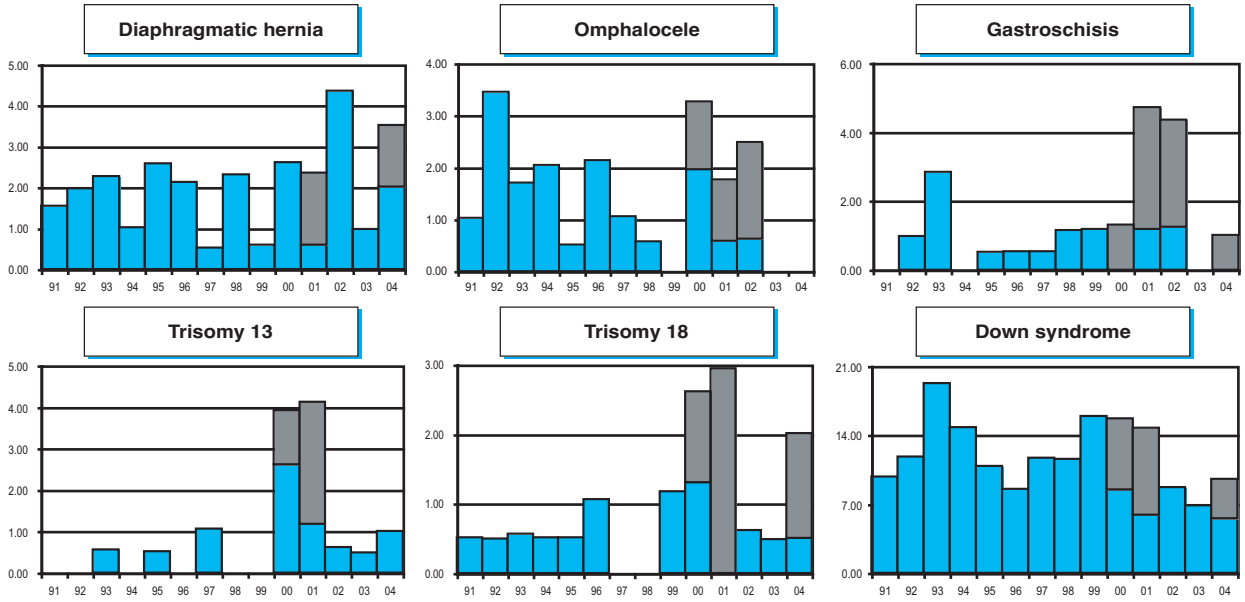
Italy: ISMAC

'Time trends 1991-2004 (Birth prevalence rates per 10,000)



Note: L+S rates, ToP rates

Note: L+S rates, ToP rates



Note: L+S rates, ToP rates

Italy: North East

North East Italy Registry of Congenital Malformations

History
The Registry was established in 1981 to include the Veneto, Friuli Venezia Giulia and Trentino Alto Adige regions. The Registry became a member of Eurocat in 1985, and an associate member of Clearinghouse in 1997.

Size and coverage
Reports are obtained from 78 participating hospitals, with a total of approximately 57,000 annual births; the actual coverage is estimated at 99%.

Legislation and funding
Reporting is voluntary. The Programme is partly run by privately funded research organisations and partly by Regional Health Authorities.

Sources of ascertainment
Reports are obtained on specific forms from delivery units, induced abortion units, pediatric, cardiology, ophthalmology and pathology departments, regional induced abortion database and cytogenetic laboratories. 32 selected malformations are recorded within 7 days from birth (within 3 years of age for cardiovascular and ophthalmological anomalies only). In induced abortions all fetal anomalies are recorded. Two control infants are selected for each malformed one.

Exposure information
Detailed information on various exposures, including maternal or paternal occupation, diseases and drug use is obtained by interview of the mothers at the birth of the malformed nfants and controls. Only selected malformations are colected.

Background information
Some epidemiological background data of all births are available. For each participating hospital the number of livebirths and stillbirths by sex and number of twin pairs are known.

Addresses and Staff:
Romano Tenconi, MD, Programme Director Genetica Medica Dipartimento Pediatria Via Giustiniani, 3 35128 Padova, Italy

Phone: +39-049-8213513

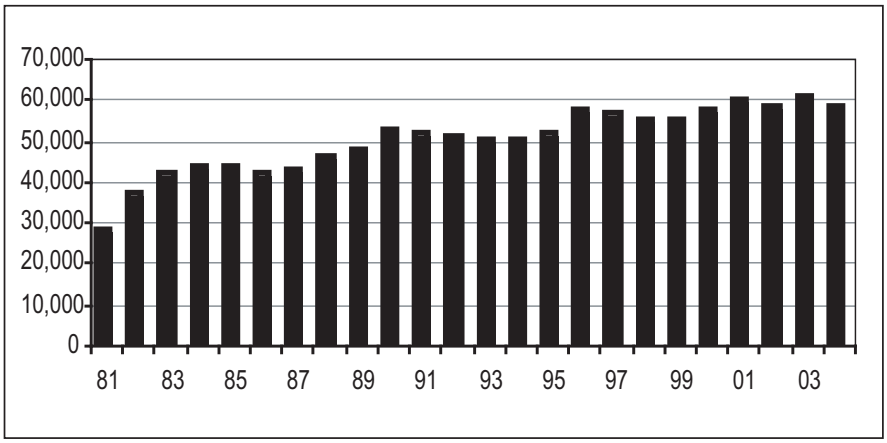
Fax: +39-049-8211425

E-mail: tenconi@child.pedi.unipd.it

Website: www.genetica.pediatria.unipd.it

Italy: North East

Total births by year



Italy: North East, 2004

Live births (LB)	51,806
Stillbirths (SB)	116
Total births	51,922
Number of terminations of pregnancy (ToP) for birth defects	130

Birth Defects	Number of cases			Rates*10,000
	LB	SB	ToP	Total rate
Anencephaly	2	0	5	1.34
Spina bifida	2	0	8	1.92
Encephalocele	0	0	2	0.38
Microcephaly	4	0	0	0.77
Arhinencephaly / Holoprosencephaly	0	0	1	0.19
Hydrocephaly	0	0	4	0.77
Anophthalmos	1	0	0	0.19
Microphthalmos	3	0	2	0.96
Unspecified Anophthalmos/ Microphthalmos	0	0	0	---
Anotia	3	0	0	0.58
Microtia	7	0	0	1.34
Unspecified Anotia/Microtia	1	0	0	---
Transposition of great vessels	5	0	0	0.96
Tetralogy of Fallot	13	0	1	2.69
Hypoplastic left heart syndrome	1	0	3	0.77
Coarctation of aorta	5	0	0	0.96
Choanal atresia, bilateral	2	0	1	0.58
Cleft palate without cleft lip	29	0	2	5.96
Cleft lip with or without cleft palate	29	0	4	6.34
Oesophageal atresia / stenosis with or without fistula	9	0	1	1.92
Small intestine atresia / stenosis	11	0	0	2.11
Anorectal atresia / stenosis	7	0	1	1.54
Undescended testis (36 weeks of gestation or later)	nr	nr	nr	nr
Hypospadias	23	0	0	4.42
Epispadias	0	0	0	0.00
Indeterminate sex	nr	nr	nr	nr
Renal agenesis	0	0	0	0.00
Cystic kidney	1	0	2	0.58
Bladder exstrophy	1	0	0	0.19
Polydactyly, preaxial	9	0	0	1.73
Total Limb reduction defects (include unspecified)	11	0	2	2.50
Transverse	4	0	0	0.77
Preaxial	0	0	1	0.19
Postaxial	2	0	0	0.38
Intercalary	0	0	0	0.00
Mixed	2	0	0	0.38
Unspecified	3	0	1	---
Diaphragmatic hernia	9	0	2	2.11
Omphalocele	2	0	4	1.15
Gastroschisis	6	0	0	1.15
Unspecified Omphalocele/Gastroschisis	0	0	0	---
Prune belly sequence	0	0	0	0.00
Trisomy 13	1	0	4	0.96
Trisomy 18	1	0	11	2.31
Down syndrome, all ages (include age unknown)	36	0	36	13.83
<20	0	0	0	nr
20-24	2	0	1	nr
25-29	2	0	0	nr
30-34	7	0	5	nr
35-39	14	0	16	nr
40-44	3	0	13	nr
45+	0	0	1	nr
unknown	8	0	0	---

nr = not reported

Italy: North East, Previous years rates 1981 - 2004

Birth prevalence rates: (LB+SB) * 10,000 until 1995
Birth prevalence rates: (LB+SB+TOP) * 10,000 from 1996

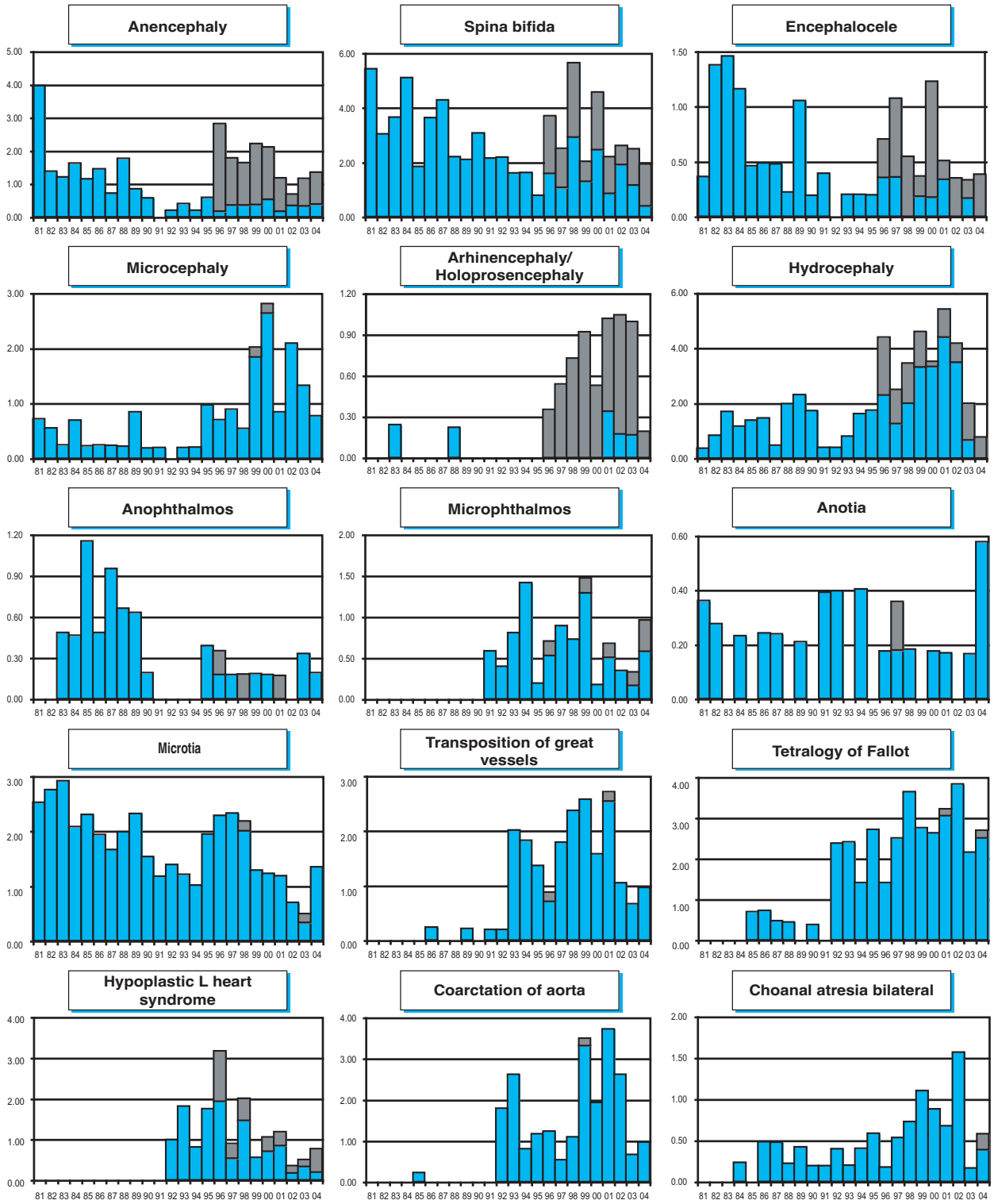
	1974-79	1980-84*	1985-89	1990-94	1995-99	2000-04
Total births		**148,426**	**219,536**	**252,816**	**273,566**	**285,598**
Anencephaly		1.89	1.18	0.28	1.83	1.30
Spina bifida		4.24	2.78	2.14	2.96	2.77
Encephalocele		1.15	0.55	0.20	0.58	0.56
Microcephaly		0.54	0.36	0.16	1.02	1.58
Arhinencephaly / Holoprosencephaly		0.07	0.05	0.00	0.51	0.77
Hydrocephaly		1.08	1.55	0.99	3.36	3.22
Anophthalmos		0.27	0.77	0.04	0.26	0.18
Microphthalmos		0.00	0.00	0.63	0.80	0.49
Unspecified Anophthalmos / Microphthalmos		---	---	---	---	---
Anotia		0.20	0.14	0.24	0.15	0.21
Microtia		2.56	2.05	1.27	2.01	0.98
Unspecified Anotia / Microtia		---	---	---	---	---
Transposition of great vessels		0.00	0.09	0.83	1.79	1.40
Tetralogy of Fallot		0.00	0.46	1.31	2.60	2.91
Hypoplastic left heart syndrome		0.00	0.00	0.71	1.68	0.77
Coarctation of aorta		0.00	0.05	1.03	1.50	2.00
Choanal atresia, bilateral		0.07	0.32	0.28	0.62	0.77
Cleft palate without cleft lip		3.50	6.33	4.47	4.39	4.94
Cleft lip with or without cleft palate		8.83	8.70	6.45	7.75	6.34
Oesophageal atresia / stenosis with or without fistula		2.29	2.23	2.73	2.41	2.77
Small intestine atresia / stenosis		0.40	0.59	1.23	0.73	1.47
Anorectal atresia / stenosis		2.49	3.19	2.10	2.60	2.38
Undescended testis (36 weeks of gestation or later)		nr	nr	nr	nr	nr
Hypospadias		6.87	6.65	5.85	6.84	7.39
Epispadias		0.07	0.14	0.12	0.22	0.04
Indeterminate sex		nr	nr	nr	nr	nr
Renal agenesis		0.74	0.64	0.32	0.69	0.28
Cystic kidney		0.00	0.00	0.08	0.22	1.09
Bladder exstrophy		0.34	0.14	0.44	0.26	0.18
Polydactyly, preaxial		1.62	2.32	2.45	2.01	1.68
Total Limb reduction defects (include unspecified)		5.79	5.78	4.90	4.86	3.78
Transverse		3.44	3.10	2.65	2.85	1.72
Preaxial		0.00	0.05	0.28	0.40	0.35
Postaxial		0.00	0.14	0.08	0.18	0.28
Intercalary		0.54	0.73	0.63	0.33	0.35
Mixed		1.82	1.78	1.27	0.48	0.39
Unspecified		---	---	---	---	---
Diaphragmatic hernia		0.34	0.64	0.16	0.51	1.58
Omphalocele		1.21	1.41	0.63	1.06	1.05
Gastroschisis		0.67	0.73	0.28	0.33	0.77
Unspecified Omphalocele / Gastroschisis		---	---	---	---	---
Prune belly sequence		0.07	0.00	0.04	0.00	0.07
Trisomy 13		0.81	0.68	0.51	0.62	1.23
Trisomy 18		0.94	1.32	0.75	1.43	2.28
Down syndrome, all ages (include age unknown)		13.88	15.12	14.20	16.92	16.46
<20		nr	nr	nr	nr	nr
20-24		nr	nr	nr	nr	nr
25-29		nr	nr	nr	nr	nr
30-34		nr	nr	nr	nr	nr
35-39		nr	nr	nr	nr	nr
40-44		nr	nr	nr	nr	nr
45+		nr	nr	nr	nr	nr
unknown		---	---	---	---	---

* data include less than 5 years

nr = not reported

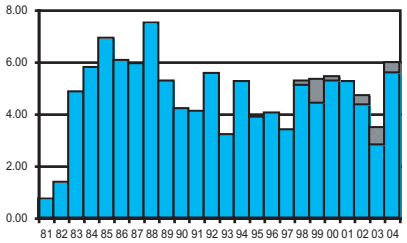
Italy: North East

'Time trends 1981-2004 (Birth prevalence rates per 10,000)

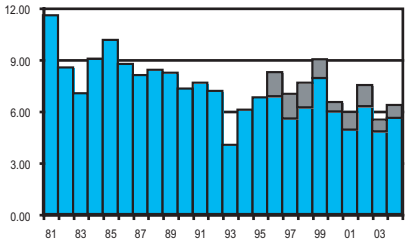


Note: L+S rates, ToP rates

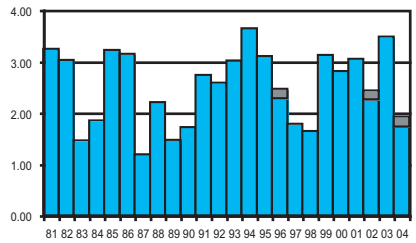
Cleft palate without cleft lip



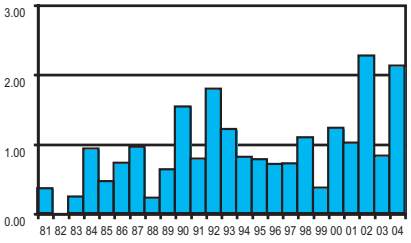
Cleft lip with or without cleft palate



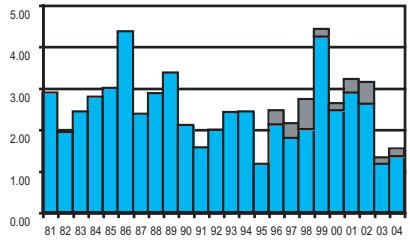
Oesophageal atresia/stenosis with or without fistula



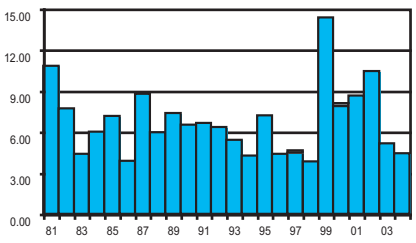
Small intestine atresia/stenosis



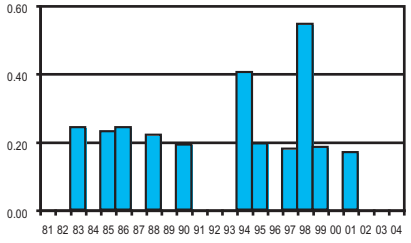
Anorectal atresia/stenosis



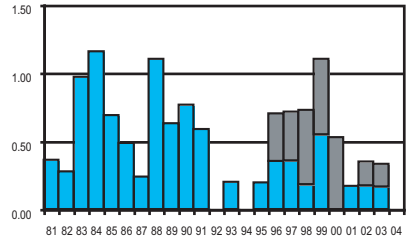
Hypospadias



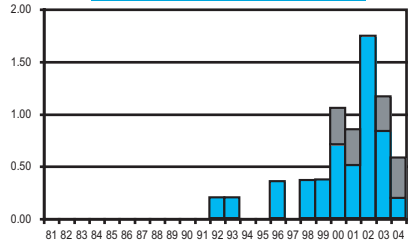
Epispadias



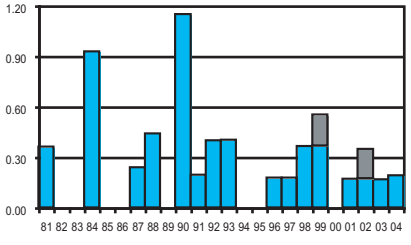
Renal agenesis



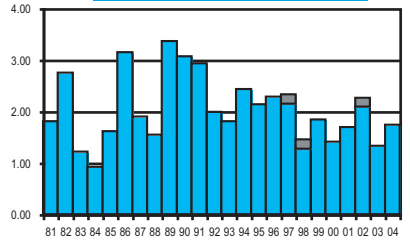
Cystic Kidney



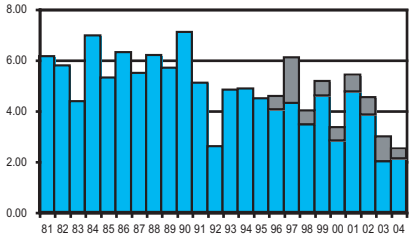
Bladder exstrophy



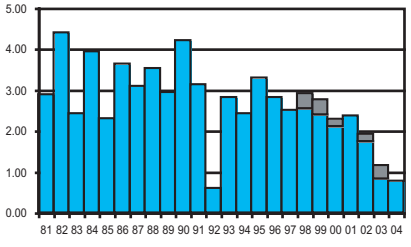
Polydactyly, preaxial



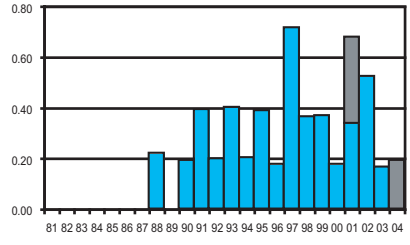
Limb reduction defects



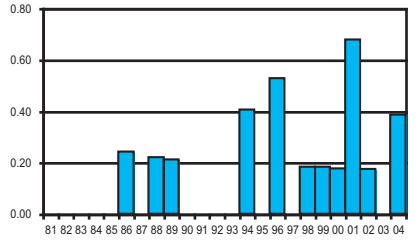
Limb reduction defects transverse



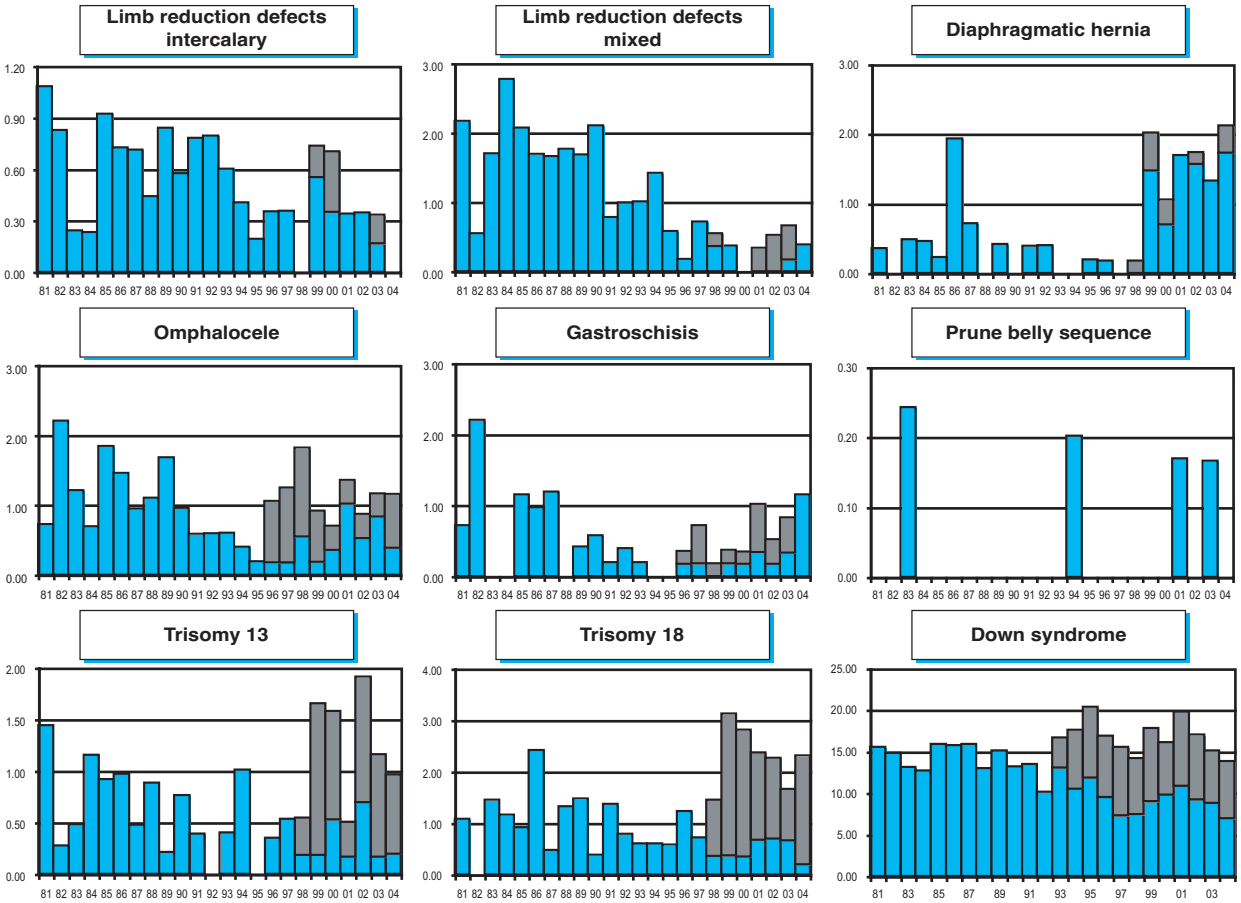
Limb reduction defects preaxial



Limb reduction defects postaxial



Note: L+S rates, ToP rates



Note: L+S rates, ToP rates

Italy: Tuscany

Tuscany Registry of Congenital Defects (RTDC)

History:
The registry started in 1979 in the province of Florence and from 1992 in the whole Tuscany region. The Programme became a full member of the Clearinghouse in 1998.

Size and coverage:
The Programme is population based, involves all the regional hospitals and the coverage is around 95% of all births in the Tuscany region (approximately 3.5 millions inhabitants and 25,000 births/year). Stillbirths of 20 weeks or more gestation and induced abortions after prenatal diagnosis of birth defects are systematically included. Malformed babies diagnosed within the first year of life are also registered.

Legislation and funding:
The Registry is a surveillance Programme included in the Regional Statistics System; it is formally recognised and supported by the Tuscany Region Health Authority.

Sources and ascertainment:
Multiple sources are used to ascertain malformed infants; records are obtained from all obstetrical and maternity units, pediatric departments, neonatal and pediatric surgery units, prenatal diagnostic centers and pathology services. Mothers are interviewed by using a standardized questionnaire.

Exposure information:
Exposure information on maternal and paternal occupation, life-style, and socio-economical characteristics are obtained by interviews of mothers of malformed infants.

Background information:
Vital statistics and other epidemiological information are obtained by the birth medical records collected by the Regional Bureau of Statistics. Selected information is obtained from the control material collected.

Addresses and Staff:
Fabrizio Bianchi, MD, Programme Director IFC-Unit of Epidemiology CNR Area di Ricerca di San Cataldo Via Alfieri 1 - 56010 Ghezzano-Pisa

Phone: 39-050-3152100

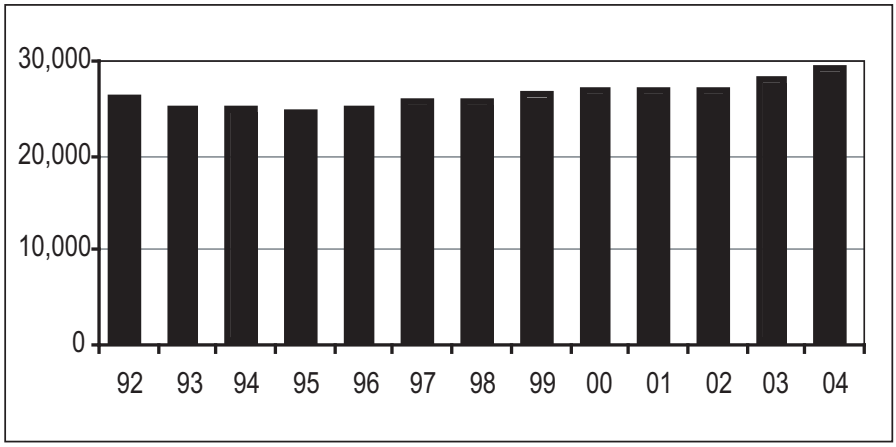
Fax: 39-050 3152095

E-mail: fabriepi@ifc.cnr.it

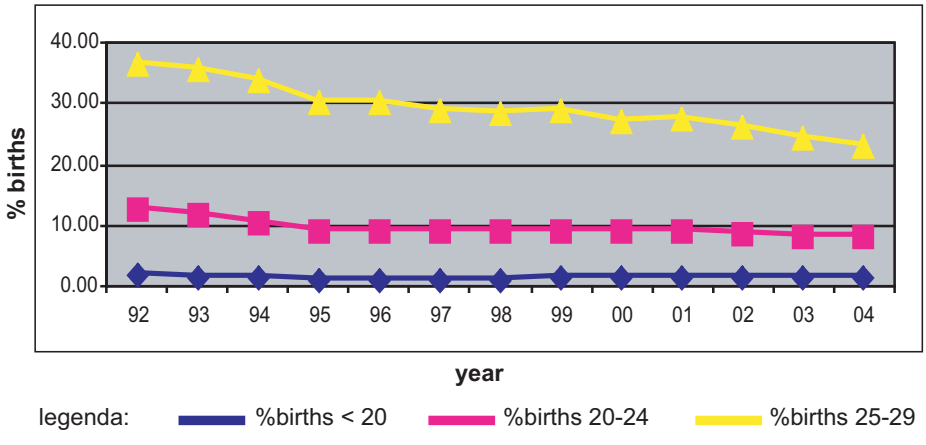
Website: www.rtdc.it

Italy: Tuscany

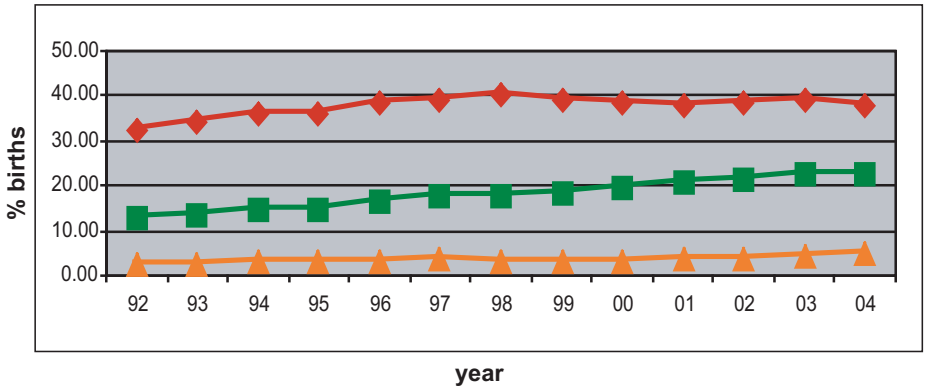
Total births by year



Percentage of births by maternal age



legenda: %births < 20 %births 20-24 %births 25-29



legenda: %births 30-34 %births 35-39 %births 40+

Italy: Tuscany, 2004

Live births (LB)	28,874
Stillbirths (SB)	105
Total births	28,979
Number of terminations of pregnancy (ToP) for birth defects	124

Birth Defects	Number of cases			Rates*10,000
	LB	SB	ToP	Total rate
Anencephaly	0	0	1	0.34
Spina bifida	5	1	8	4.81
Encephalocele	0	0	1	0.34
Microcephaly	2	0	0	0.69
Arhinencephaly / Holoprosencephaly	2	0	4	2.06
Hydrocephaly	2	1	6	3.09
Anophthalmos	0	0	0	0.00
Microphthalmos	1	0	1	0.69
Unspecified Anophthalmos/ Microphthalmos	0	0	0	---
Anotia	1	0	0	0.34
Microtia	1	0	0	0.34
Unspecified Anotia/Microtia	0	0	0	---
Transposition of great vessels	10	0	1	3.78
Tetralogy of Fallot	6	0	0	2.06
Hypoplastic left heart syndrome	4	0	3	2.41
Coarctation of aorta	6	0	0	2.06
Choanal atresia, bilateral	1	0	0	0.34
Cleft palate without cleft lip	14	0	2	5.50
Cleft lip with or without cleft palate	12	0	1	4.47
Oesophageal atresia / stenosis with or without fistula	6	1	0	2.41
Small intestine atresia / stenosis	4	0	0	1.37
Anorectal atresia / stenosis	10	1	2	4.47
Undescended testis (36 weeks of gestation or later)	15	0	0	5.15
Hypospadias	18	0	0	6.18
Epispadias	1	0	0	0.34
Indeterminate sex	0	0	0	0.00
Renal agenesis	0	0	2	0.69
Cystic kidney	7	0	8	5.15
Bladder exstrophy	0	0	0	0.00
Polydactyly, preaxial	4	0	0	1.37
Total Limb reduction defects (include unspecified)	19	1	3	7.90
Transverse	15	1	1	5.84
Preaxial	0	0	1	0.34
Postaxial	0	0	0	0.00
Intercalary	1	0	1	0.69
Mixed	0	0	0	0.00
Unspecified	3	0	0	---
Diaphragmatic hernia	6	1	2	3.09
Omphalocele	2	0	2	1.37
Gastroschisis	1	0	3	1.37
Unspecified Omphalocele/Gastroschisis	0	0	0	---
Prune belly sequence	0	0	0	0.00
Trisomy 13	3	1	3	2.41
Trisomy 18	1	1	10	4.12
Down syndrome, all ages (include age unknown)	12	0	45	19.59
<20	0	0	0	0.00
20-24	0	0	0	0.00
25-29	1	0	0	1.50
30-34	2	0	6	7.33
35-39	4	0	23	40.90
40-44	5	0	15	153.26
45+	0	0	1	270.27
unknown	0	0	0	---

Italy: Tuscany, Previous years rates 1992 - 2004

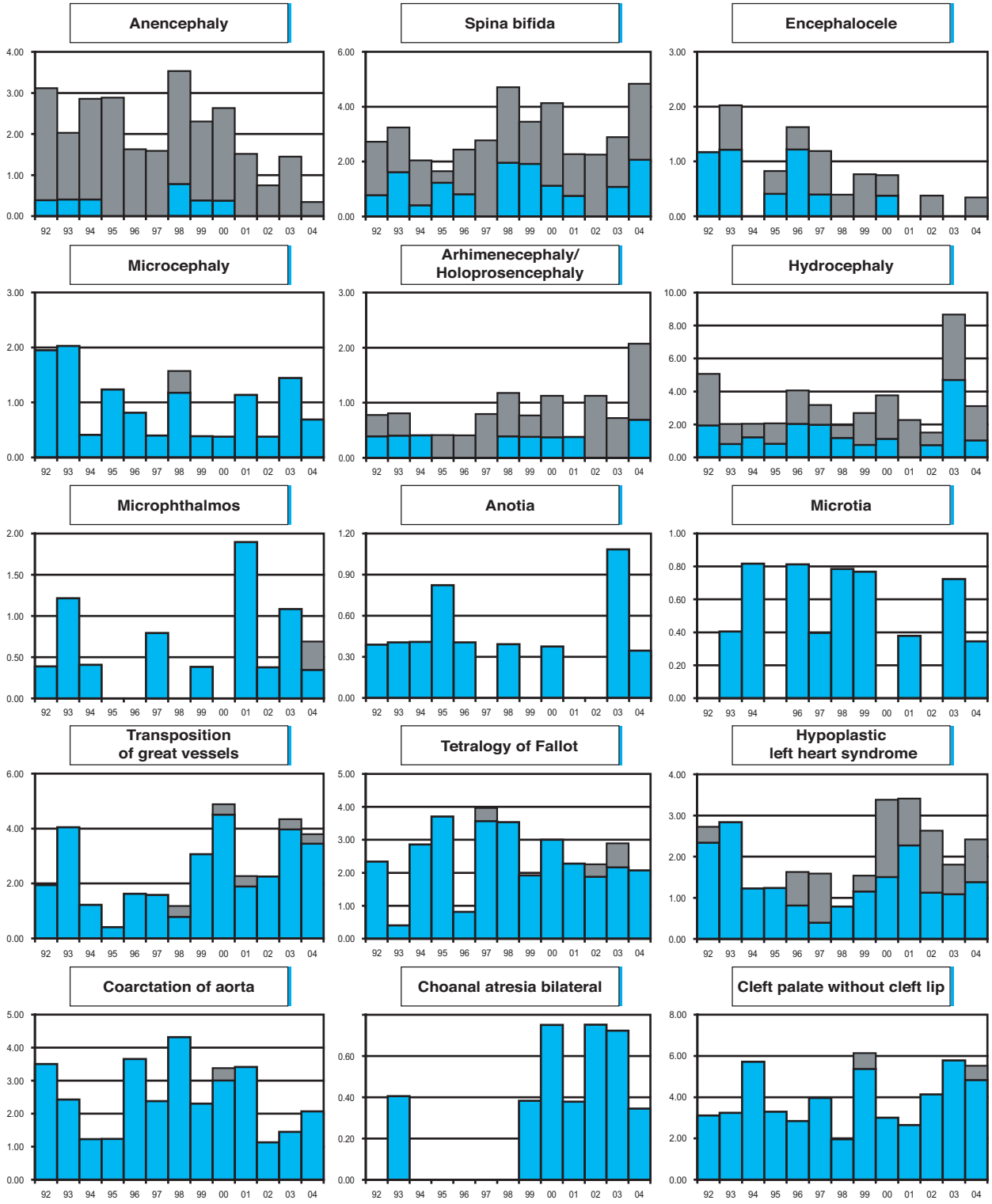
Birth prevalence rates: (LB+SB+TOP) * 10,000
since 1992 except for LRD subcategories (ToPs reported since 1998)

	1974-79	1980-84	1985-89	1990-94*	1995-99	2000-04
Total births				**74,844**	**125,638**	**136,241**
Anencephaly				2.67	2.39	1.32
Spina bifida				2.67	3.02	3.30
Encephalocele				1.07	0.96	0.29
Microcephaly				1.47	0.88	0.81
Arhinencephaly / Holoprosencephaly				0.67	0.72	1.10
Hydrocephaly				3.07	2.79	3.89
Anophthalmos				0.00	0.08	0.22
Microphthalmos				0.67	0.24	0.81
Unspecified Anophthalmos / Microphthalmos				---	---	---
Anotia				0.40	0.32	0.37
Microtia				0.40	0.72	0.29
Unspecified Anotia / Microtia				---	---	---
Transposition of great vessels				2.41	1.59	3.52
Tetralogy of Fallot				1.87	2.79	2.50
Hypoplastic left heart syndrome				2.27	1.35	2.72
Coarctation of aorta				2.41	2.79	2.28
Choanal atresia, bilateral				0.13	0.08	0.59
Cleft palate without cleft lip				4.01	3.66	4.26
Cleft lip with or without cleft palate				8.42	5.49	6.61
Oesophageal atresia / stenosis with or without fistula				2.14	2.55	2.35
Small intestine atresia / stenosis				1.07	0.64	1.17
Anorectal atresia / stenosis				1.60	1.99	3.16
Undescended testis (36 weeks of gestation or later)				3.47	6.53	10.20
Hypospadias				5.75	3.18	6.75
Epispadias				0.27	0.24	0.22
Indeterminate sex				0.94	0.64	0.51
Renal agenesis				1.87	1.43	0.88
Cystic kidney				3.34	3.26	4.40
Bladder exstrophy				0.40	0.16	0.22
Polydactyly, preaxial				0.80	0.96	1.39
Total Limb reduction defects (include unspecified)				5.08	4.78	6.53
Transverse				4.01	2.79	4.40
Preaxial				0.13	0.32	0.66
Postaxial				0.13	0.32	0.15
Intercalary				0.27	0.56	0.59
Mixed				0.27	0.48	0.29
Unspecified				---	---	---
Diaphragmatic hernia				1.20	1.43	2.57
Omphalocele				2.27	1.27	1.83
Gastroschisis				0.53	0.40	0.59
Unspecified Omphalocele / Gastroschisis				---	---	---
Prune belly sequence				0.13	0.16	0.00
Trisomy 13				0.40	0.80	1.32
Trisomy 18				2.81	3.34	2.79
Down syndrome, all ages (include age unknown)				13.49	16.79	16.59
<20				0.00	0.00	0.00
20-24				3.49	8.04	4.35
25-29				8.35	8.71	3.17
30-34				10.11	13.65	9.24
35-39				31.57	28.31	26.99
40-44				66.19	104.63	156.86
45+				236.22	45.45	154.64
uknown				---	---	---

* data include less than 5 years

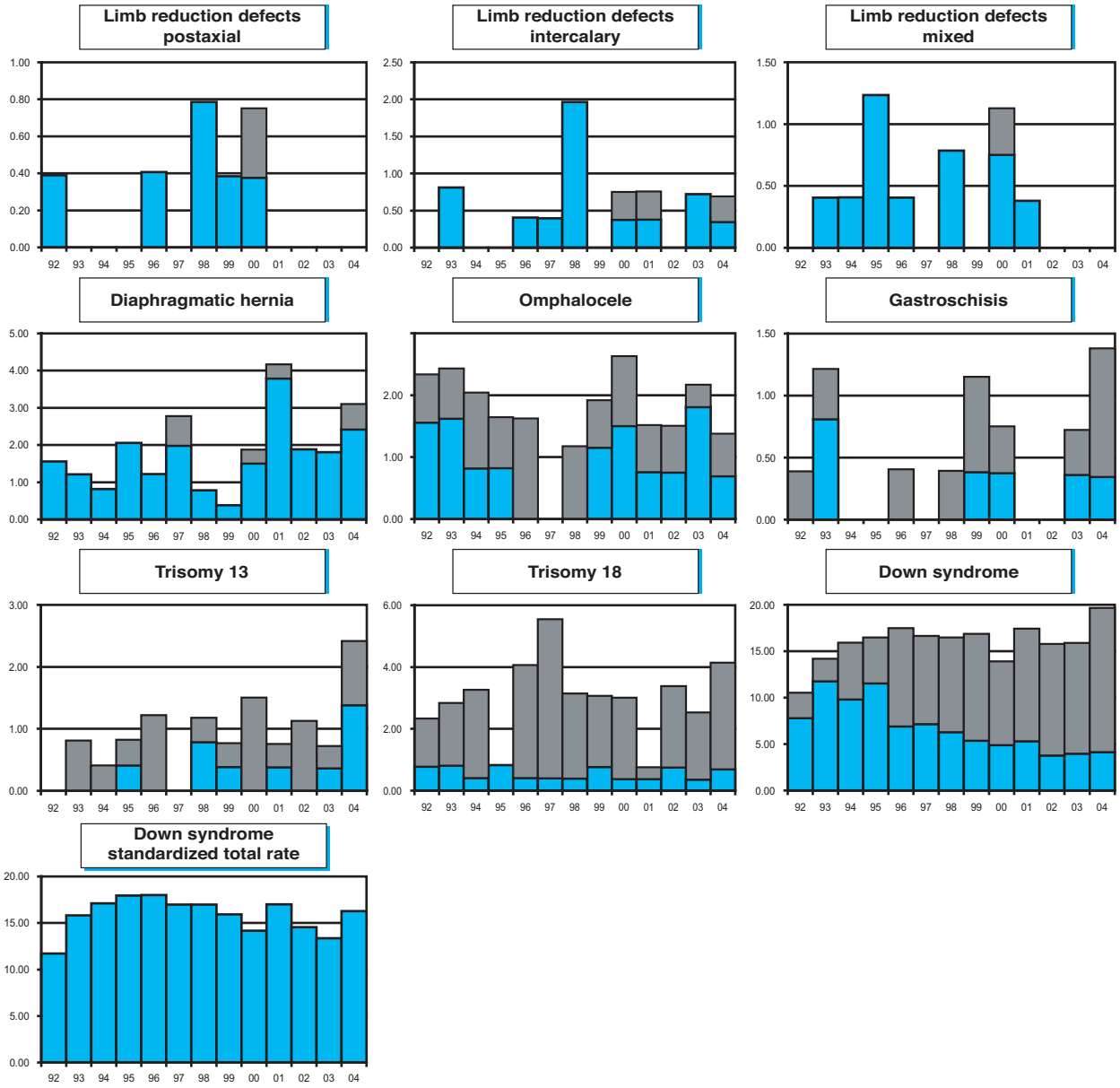
Italy: Tuscany

Time trends 1992-2004 (Birth prevalence rates per 10,000)



Note: L+S rates, ToP rates

Note: L+S rates, ToP rates



Note: L+S rates, ToP rates

Japan: JAOG

Japan Association of Obstetricians and Gynaecologists (JAOG)

History:
The Programme started in 1972 and became a full member of the Clearinghouse in 1988.

Size and coverage:
The Programme is based on reports from 270 hospitals throughout Japan. At present approximately 100,000 births are covered, representing about 9% of all Japanese births. Stillbirths of 22 weeks or more gestation are included.

Legislation and funding:
The Programme is a research Programme acknowledged by the Ministry of Welfare and supported by the Japanese Association of Obstetricians and Gynecologists.

Sources of ascertainment:
Reports are obtained from delivery units and pediatric clinics of the participating hospitals.

Exposure information:
Exposure to drugs, X-ray and viral infections are available.

Background informations:
Basic epidemiological information on all births is available from each participating hospital.

Addresses and Staff:
Fumiki Hirahara, MD Yokohama City University Hospital Dept. OB V GYN 3-9 Fukuura, Kanazawaku, Yokohama, 236-0004, Japan

Phone: 81-45-787-2689

Fax: 81-45-787-2689

E-mail: hirafu@med.yokohama-cu.ac.jp

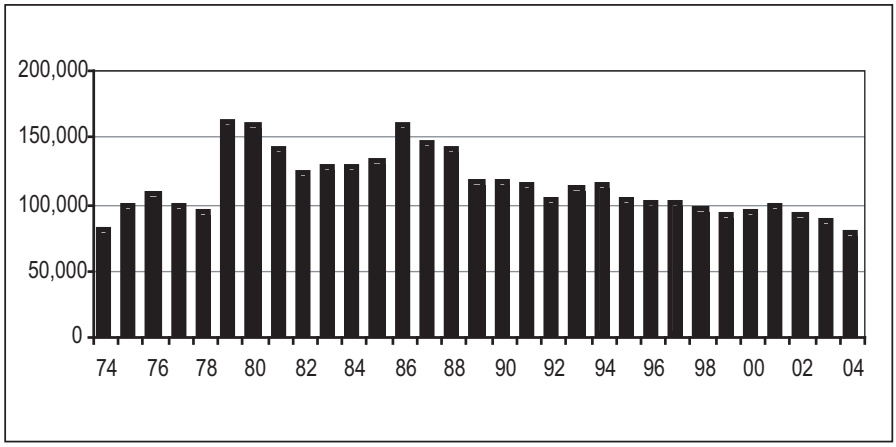
Michiko Yamanaka, MD Kanawaga Children's Medical Center Division of Obstetrics and Gynecology 2-138-4 Mutsukawa, Minami-ku Yokohama City, 232-8555 , Japan

Phone: 81-45-716-5366

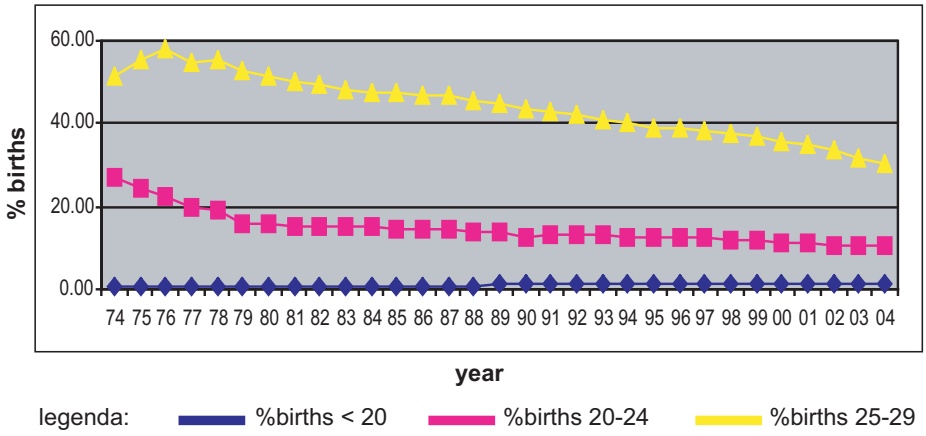
E-mail: michikoy@yha.att.ne.jp

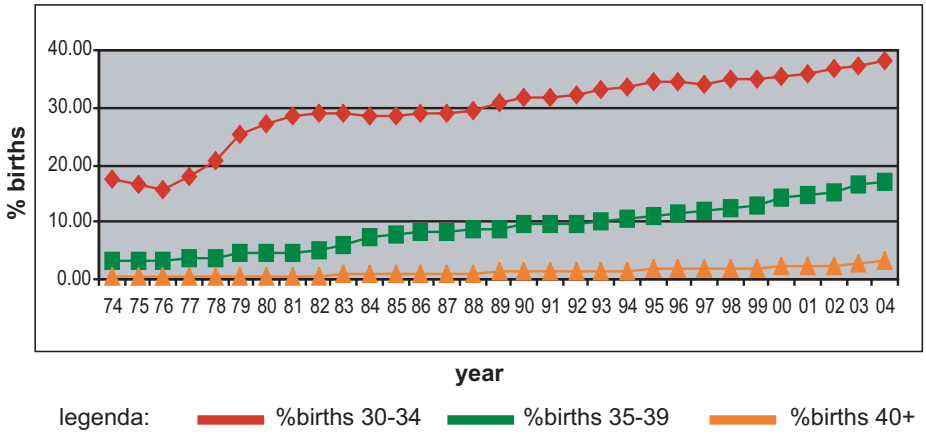
Japan: JAOG

Total births by year



Percentage of births by maternal age





Japan: JAOG, 2004

Live births (LB)	76,705
Stillbirths (SB)	528
Total births	77,233
Number of terminations of pregnancy (ToP) for birth defects	nr

Birth Defects	Number of cases			Rates*10,000
	LB	SB	ToP	Total rate
Anencephaly	5	7	nr	1.55
Spina bifida	37	2	nr	5.05
Encephalocele	1	2	nr	0.39
Microcephaly	9	2	nr	1.42
Arhinencephaly / Holoprosencephaly	9	1	nr	1.29
Hydrocephaly	45	5	nr	6.47
Anophthalmos	5	1	nr	0.78
Microphthalmos	6	2	nr	1.04
Unspecified Anophthalmos / Microphthalmos	0	0	nr	---
Anotia	nr	nr	nr	nr
Microtia	7	1	nr	1.04
Unspecified Anotia / Microtia	nr	nr	nr	---
Transposition of great vessels	33	1	nr	4.40
Tetralogy of Fallot	39	1	nr	5.18
Hypoplastic left heart syndrome	18	6	nr	3.11
Coarctation of aorta	36	0	nr	4.66
Choanal atresia, bilateral	nr	nr	nr	nr
Cleft palate without cleft lip	25	0	nr	3.24
Cleft lip with or without cleft palate	149	7	nr	20.20
Oesophageal atresia / stenosis with or without fistula	40	5	nr	5.83
Small intestine atresia / stenosis	36	2	nr	4.92
Anorectal atresia / stenosis	44	3	nr	6.09
Undescended testis (36 weeks of gestation or later)	nr	nr	nr	nr
Hypospadias	32	0	nr	4.14
Epispadias	nr	nr	nr	nr
Indeterminate sex	nr	nr	nr	nr
Renal agenesis	19	3	nr	2.85
Cystic kidney	22	3	nr	3.24
Bladder exstrophy	2	0	nr	0.26
Polydactyly, preaxial	53	4	nr	7.38
Total Limb reduction defects (include unspecified)	18	6	nr	3.11
Transverse	3	1	nr	0.52
Preaxial	2	1	nr	0.39
Postaxial	2	0	nr	0.26
Intercalary	3	0	nr	0.39
Mixed	6	2	nr	1.04
Unspecified	2	2	nr	---
Diaphragmatic hernia	34	6	nr	5.18
Omphalocele	22	2	nr	3.11
Gastroschisis	19	2	nr	2.72
Unspecified Omphalocele / Gastroschisis	0	0	nr	---
Prune belly sequence	0	0	nr	0.00
Trisomy 13	14	4	nr	2.33
Trisomy 18	45	20	nr	8.42
Down syndrome, all ages (include age unknown)	88	2	nr	11.65
<20	0	0	nr	0.00
20-24	4	0	nr	5.03
25-29	14	0	nr	6.02
30-34	24	1	nr	8.52
35-39	28	0	nr	21.24
40+	18	1	nr	82.11
unknown	0	0	nr	---

nr = not reported

Japan: JAOG, Previous years rates 1974 - 2004

Prevalence rates: (LB+SB) * 10,000

	1974-79	1980-84	1985-89	1990-94	1995-99	2000-04
Births	**630,508**	**668,362**	**685,695**	**551,995**	**484,401**	**439,875**
Anencephaly	9.01	9.32	7.03	4.06	2.09	1.27
Spina bifida	1.90	2.35	3.19	3.55	3.74	5.32
Encephalocele	1.03	1.03	1.34	0.94	1.05	0.70
Microcephaly	0.84	1.24	1.15	1.63	1.36	1.34
Arhinencephaly / Holoprosencephaly	nr	nr	nr	nr	0.95	1.20
Hydrocephaly	2.78	3.65	5.44	7.25	6.92	7.91
Anophthalmos	0.76	0.84	0.61	0.54	0.14	0.32
Microphthalmos	0.57	0.64	0.53	0.53	0.52	0.52
Unspecified Anophthalmos / Microphthalmos	---	---	---	---	---	---
Anotia	nr	nr	nr	nr	nr	nr
Microtia	1.06	1.14	1.09	1.07	1.34	1.30
Unspecified Anotia / Microtia	---	---	---	---	---	---
Transposition of great vessels	nr	nr	nr	nr	1.94*	3.48
Tetralogy of Fallot	nr	nr	nr	nr	2.15*	4.21
Hypoplastic left heart syndrome	nr	nr	nr	nr	1.52*	2.43
Coarctation of aorta	nr	nr	nr	nr	1.48*	2.91
Choanal atresia, bilateral	nr	nr	nr	nr	nr	nr
Cleft palate without cleft lip	12.74	8.71	5.18	5.53	4.38	3.82
Cleft lip with or without cleft palate	14.62	12.79	14.25	15.47	16.78	18.89
Oesophageal atresia / stenosis with or without fistula	1.19*	1.03	1.60	2.25	2.81	4.39
Small intestine atresia / stenosis	nr	nr	nr	nr	4.44*	5.66
Anorectal atresia / stenosis	4.04	3.55	4.24	4.17	4.38	5.46
Undescended testis (36 weeks of gestation or later)	nr	nr	nr	nr	nr	nr
Hypospadias	1.74	2.21	2.36	3.10	2.93	4.18
Epispadias	nr	nr	nr	nr	nr	nr
Indeterminate sex	nr	nr	nr	nr	nr	nr
Renal agenesis	nr	nr	0.95	1.58	1.49	2.14
Cystic kidney	nr	nr	nr	nr	2.79*	4.30
Bladder exstrophy	0.17*	0.13	0.16	0.13	0.17	0.30
Polydactyly, preaxial	nr	nr	5.28	6.54	6.19	6.48
Total Limb reduction defects (include unspecified)	nr	nr	nr	3.23*	3.32	3.43
Transverse	nr	nr	nr	0.36*	0.37	0.32
Preaxial	nr	nr	nr	0.58*	0.50	0.66
Postaxial	nr	nr	nr	0.18*	0.33	0.27
Intercalary	nr	nr	nr	1.39*	1.09	0.86
Mixed	nr	nr	nr	0.40*	0.64	0.82
Unspecified	---	---	---	---	---	---
Diaphragmatic hernia	nr	nr	2.25	2.92	3.67	6.16
Omphalocele	0.97	1.38	2.63	3.32	3.24	3.52
Gastroschisis	1.08	0.84	1.18	1.59	1.88	2.55
Unspecified Omphalocele / Gastroschisis	---	---	---	---	---	---
Prune belly sequence	nr	nr	nr	nr	0.04	0.02
Trisomy 13	nr	nr	nr	0.36*	0.78	1.34
Trisomy 18	nr	nr	nr	2.33*	3.34	7.66
Down syndrome, all ages (include age unknown)	3.37*	4.94	5.94	6.05	8.26	9.64
<20	nr	nr	nr	8.33*	0.00	7.32
20-24	nr	nr	nr	2.07*	3.18	3.17
25-29	nr	nr	nr	4.08*	5.24	4.93
30-34	nr	nr	nr	4.96*	8.38	8.07
35-39	nr	nr	nr	16.65*	17.37	20.57
40+	nr	nr	nr	67.33*	50.31	57.46
unknown	---	---	---	---	---	---

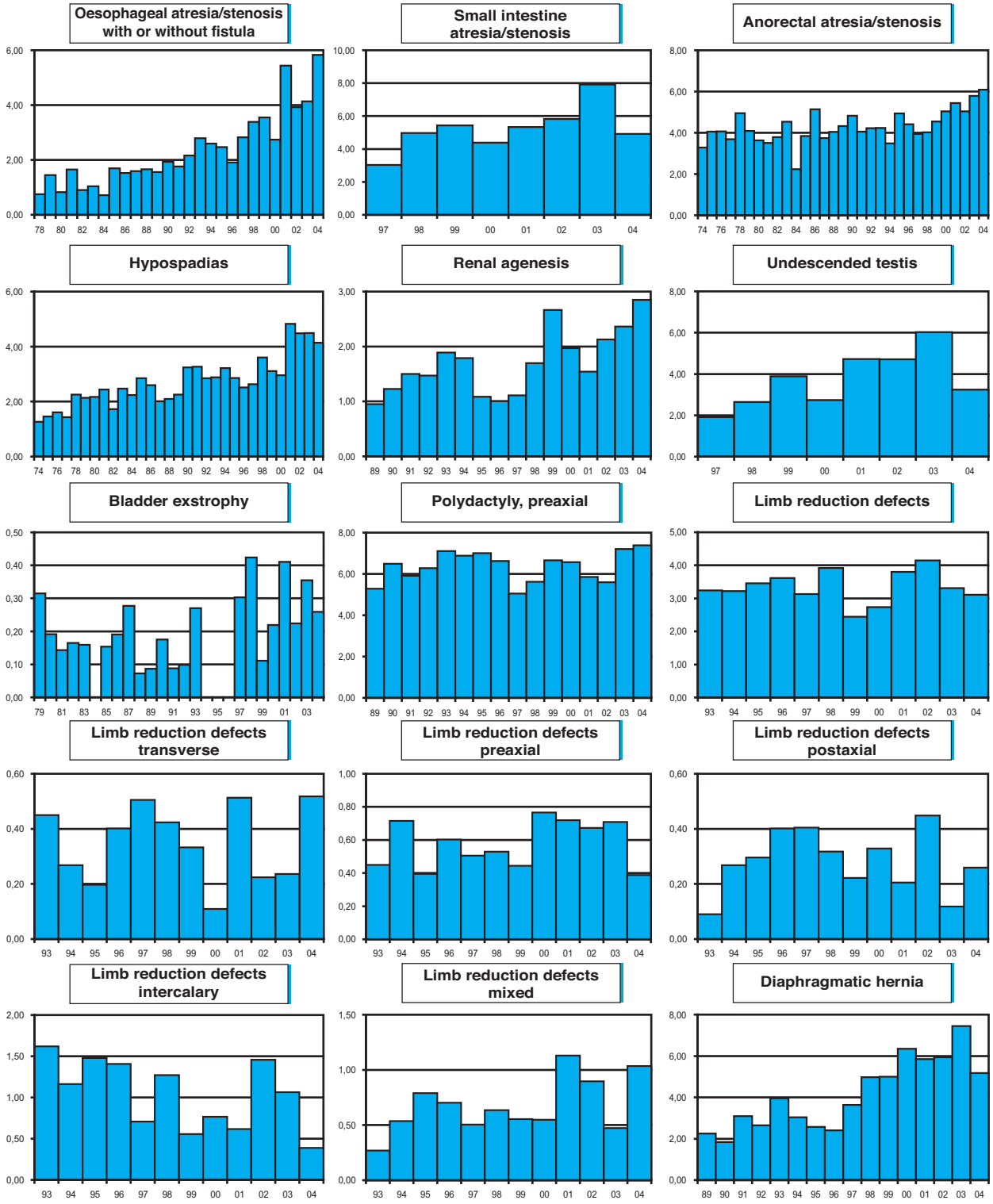
* data include less than 5 years

nr = not reported

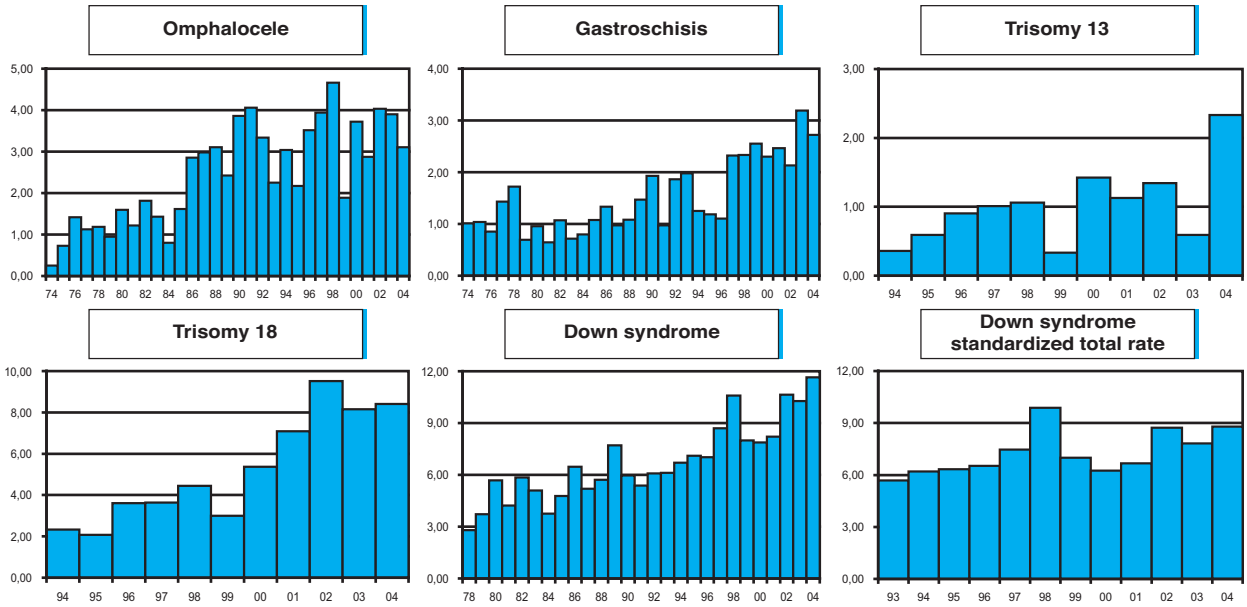
Japan: JAOG

Time trends 1974-2004 (Birth prevalence rates per 10,000)

Note: L+S rates



Note: L+S rates



Note: L+S rates

Malta

Malta Congenital Anomalies Register (MCAR)

History:
The register started in 1985 as a research project of the University of Malta. It started as a hospital based register collecting data regarding congenital anomalies diagnosed in babies born at the main general hospital. It became a member of EUROCAT in 1986. Funding for the research project was stopped in 1995 and in 1997 the Department of Health Information assumed the functions of data collection increasing coverage to all hospitals on the islands making it a population based register. The Register was accepted as an associate member of the International Clearinghouse in 2000.

Size and Coverage:
The registry is population based and covers just under 5000 births per year.

Legislation and Funding:
The registry is run and funded by the state Department of Health Information. Reporting is not statutory.

Sources of ascertainment:
The registry employs active data collection from multiple sources including labour and postnatal wards, doctors' reporting, cardiac lab records, genetics clinic records, National Mortality Register, National Obstetric Systems database, Hospital Activity Analysis database, National Cancer Register and the hypothyroid screening Programme.

Exposure information:
Information regarding maternal exposure to medicinal drugs, smoking, alcohol and drug abuse as well as parental occupation are collected for all malformed infants.

Background information:
Epidemiological background data on all births are available from the National Obstetric Information Systems database and vital statistics.

Addresses and Staff:
Miriam Gatt, MD, Programme Director Malta Congenital Anomalies Registry Department of Health Information 95, Guardamangia Hill Guardamangia PTA 1313 Malta

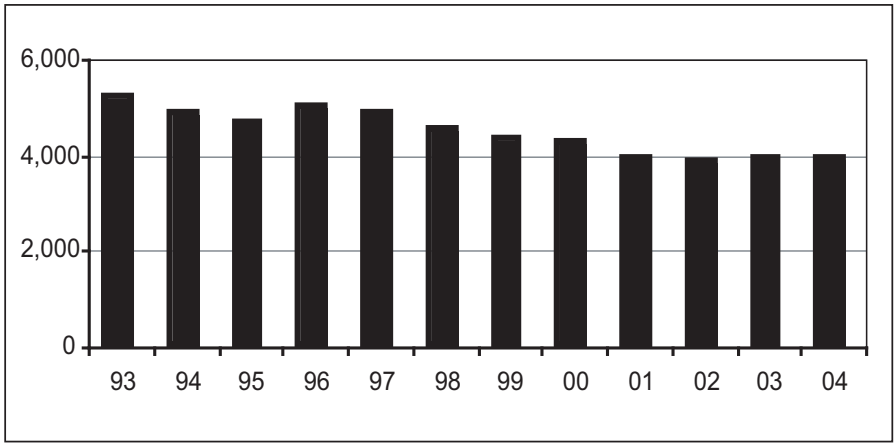
Phone: 356 25599000

Fax: 356 25599385

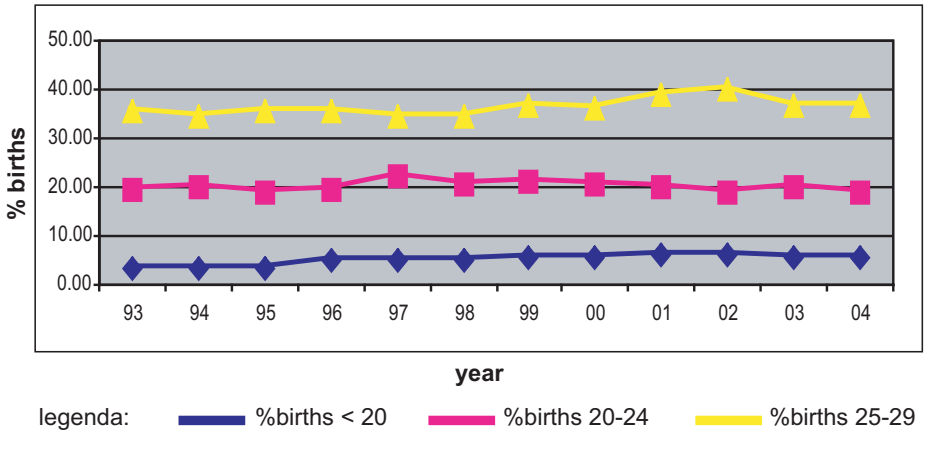
E-mail: miriam.gatt@gov.mt

Malta

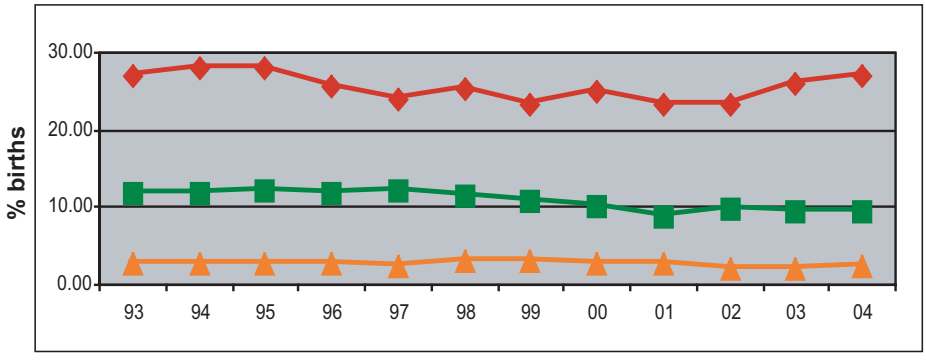
Total births by year



Percentage of births by maternal age



legenda: %births < 20 %births 20-24 %births 25-29



legenda: %births 30-34 %births 35-39 %births 40+

Malta: 2004

Live births (LB)	3,887
Stillbirths (SB)	15
Total births	3,902
Number of terminations of pregnancy (ToP) for birth defects	not permitted

Birth Defects	Number of cases			Rates*10,000
	LB	SB	ToP	Total rate
Anencephaly	0	1		2.56
Spina bifida	2	0		5.13
Encephalocele	0	0		0.00
Microcephaly	1	0		2.56
Arhinencephaly / Holoprosencephaly	0	0		0.00
Hydrocephaly	2	0		5.13
Anophthalmos	0	0		0.00
Microphthalmos	1	0		2.56
Unspecified Anophthalmos/ Microphthalmos	0	0		---
Anotia	0	0		0.00
Microtia	0	0		0.00
Unspecified Anotia/Microtia	0	0		---
Transposition of great vessels	2	0		5.13
Tetralogy of Fallot	1	0		2.56
Hypoplastic left heart syndrome	1	0		2.56
Coarctation of aorta	4	0		10.25
Choanal atresia, bilateral	0	0		0.00
Cleft palate without cleft lip	4	0		10.25
Cleft lip with or without cleft palate	2	0		5.13
Oesophageal atresia / stenosis with or without fistula	0	0		0.00
Small intestine atresia / stenosis	1	0		2.56
Anorectal atresia / stenosis	1	0		2.56
Undescended testis (36 weeks of gestation or later)	nr	nr		nr
Hypospadias	6	0		15.38
Epispadias	0	0		0.00
Indeterminate sex	1	0		2.56
Renal agenesis	1	0		2.56
Cystic kidney	0	1		2.56
Bladder exstrophy	0	0		0.00
Polydactyly, total	5	2		17.94
Total Limb reduction defects (include unspecified)	1	0		2.56
Transverse	nr	nr		nr
Preaxial	nr	nr		nr
Postaxial	nr	nr		nr
Intercalary	nr	nr		nr
Mixed	nr	nr		nr
Unspecified	nr	nr		---
Diaphragmatic hernia	0	0		0.00
Omphalocele	1	0		2.56
Gastroschisis	1	0		2.56
Unspecified Omphalocele/Gastroschisis	0	0		---
Prune belly sequence	0	0		0.00
Trisomy 13	1	0		2.56
Trisomy 18	3	0		7.69
Down syndrome, all ages (include age unknown)	4	0		10.25
<20	0	0		0.00
20-24	0	0		0.00
25-29	1	0		6.95
30-34	1	0		9.53
35-39	1	0		27.25
40-44	1	0		116.28
45+	0	0		0.00
unknown	0	0		---

nr = not reported

Malta: Previous years rates 1993 - 2004

Birth prevalence rates: (LB+SB) * 10,000

	1974-79	1980-84	1985-89	1990-94*	1995-99	2000-04
Births				**10,035**	**23,325**	**19,803**
Anencephaly				3.99	4.72	1.51
Spina bifida				6.98	7.29	5.05
Encephalocele				1.99	1.71	2.02
Microcephaly				4.98	3.00	3.53
Arhinencephaly / Holoprosencephaly				1.00	1.29	0.00
Hydrocephaly				6.98	5.57	3.03
Anophthalmos				1.00	0.00	0.00
Microphthalmos				0.00	1.71	1.01
Unspecified Anophthalmos / Microphthalmos				---	---	---
Anotia				0.00	0.00	0.00
Microtia				0.00	0.00	0.00
Unspecified Anotia / Microtia				---	---	---
Transposition of great vessels				2.99	4.72	4.54
Tetralogy of Fallot				1.99	4.72	3.03
Hypoplastic left heart syndrome				1.00	1.71	3.53
Coarctation of aorta				7.97	4.29	6.06
Choanal atresia, bilateral				1.99	1.29	1.01
Cleft palate without cleft lip				14.95	15.86	7.07
Cleft lip with or without cleft palate				9.97	8.57	8.58
Oesophageal atresia / stenosis with or without fistula				3.99	1.71	2.02
Small intestine atresia / stenosis				0.00	2.14	2.52
Anorectal atresia / stenosis				1.99	5.14	5.05
Undescended testis (36 weeks of gestation or later)				nr	nr	nr
Hypospadias				11.96	17.15	17.67
Epispadias				1.99	0.86	0.00
Indeterminate sex				1.99	0.86	1.01
Renal agenesis				1.00	1.29	2.02
Cystic kidney				3.99	4.29	2.02
Bladder exstrophy				0.00	0.00	0.00
Polydactyly, preaxial				13.95	16.72	17.17
Total Limb reduction defects (include unspecified)				6.98	4.72	7.07
Transverse				nr	nr	nr
Preaxial				nr	nr	nr
Postaxial				nr	nr	nr
Intercalary				nr	nr	nr
Mixed				nr	nr	nr
Unspecified				---	---	---
Diaphragmatic hernia				3.99	6.43	4.04
Omphalocele				2.99	2.14	1.51
Gastroschisis				1.99	0.43	1.51
Unspecified Omphalocele / Gastroschisis				---	---	---
Prune belly sequence				1.00	0.43	0.00
Trisomy 13				0.00	0.00	1.01
Trisomy 18				1.00	3.86	5.05
Down syndrome, all ages (include age unknown)				21.92	16.29	19.19
<20				0.00	9.12	8.64
20-24				0.00	0.00	0.00
25-29				8.55	3.65	9.35
30-34				14.54	22.12	22.32
35-39				84.32	44.26	64.10
40-44				204.92	130.72	155.21
45+				0.00	400.00	0.00
unknown				---	---	---

* data include less than 5 years

nr = not reported

Malta

'Time trends 1993-2004 (Birth prevalence rates per 10,000)

Note: L+S rates

Note: L+S rates

Mexico: RYVEMCE

Mexican Registry and Epidemiological Surveillance of External Congenital Malformations (RYVEMCE)

History:
The Programme was started in 1978. The Programme became a full member of the ICBD-SR in 1980.

Size and coverage:
Reports are obtained from 15 hospitals in 11 cities in Mexico. Participation is voluntary. The annual number of births is approximately 40.000, about 3.5% of all births in Mexico. Stillbirths of 20 weeks or more gestation and/or at least 500g birthweight are included.

Legislation and funding:
The Programme is a research Programmeand is funded by research grants.

Sources of ascertainment:
Reports are obtained from the delivery units and pediatric departments of the participating hospitals.

Exposure information:
The mother of each reported infant and the mother of a control infant-the next non-malformed infant born at that hospital with the same sex as the proband - are interviewed on various exposures, including drug usage and parental occupation

Background information:
The total number of births in the hospitals is known.

Addresses and Staff:
Osvaldo Mutchinick, MD, Programme Director RYVEMCE Departamento de Genetica Inst. Nacional de Nutricion Tlalpan 1400 Mexico DF, Mexico

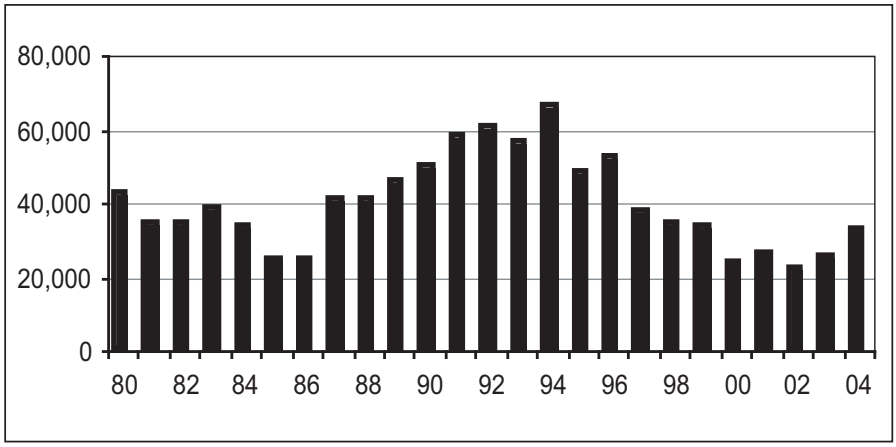
Phone: 52-5-5731200/ 52-5-5730611
52-5-5737333 (ext 2426, 2425)

Fax: 52-5-6556138

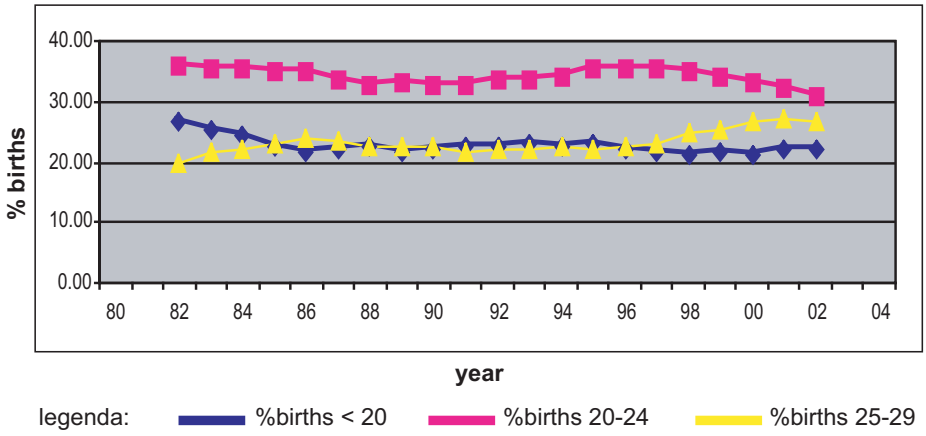
E-mail: osvaldo@servidor.unam.mx

Mexico: RYVEMCE

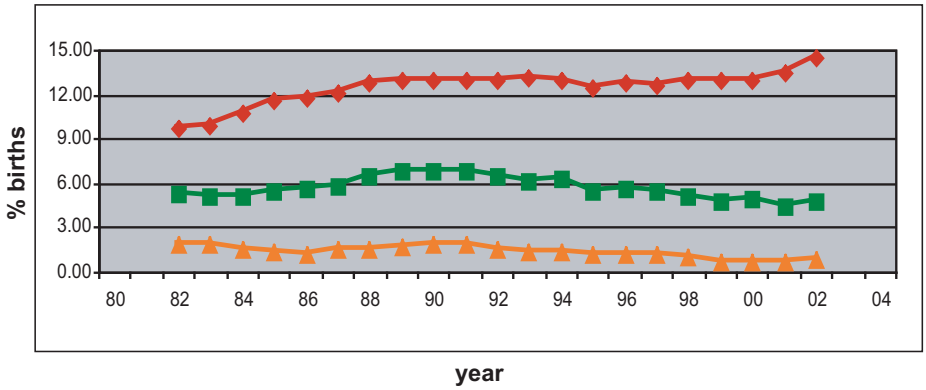
Total births by year



Percentage of births by maternal age



legenda: %births < 20 %births 20-24 %births 25-29



legenda: %births 30-34 %births 35-39 %births 40+

Mexico: RYVEMCE, 2004

Live births (LB)	32,319
Stillbirths (SB)	450
Total births	32,769
Number of terminations of pregnancy (ToP) for birth defects	not permitted

Birth Defects	Number of cases			Rates*10,000
	LB	SB	ToP	Total rate
Anencephaly	5	9		4.27
Spina bifida	23	1		7.32
Encephalocele	3	3		1.83
Microcephaly	7	2		2.75
Arhinencephaly / Holoprosencephaly	5	1		1.83
Hydrocephaly	22	4		7.93
Anophthalmos*	8	0		2.44
Microphthalmos	nr	nr		nr
Unspecified Anophthalmos/ Microphthalmos	nr	nr		---
Anotia**	29	3		9.77
Microtia	nr	nr		nr
Unspecified Anotia/Microtia	nr	nr		---
Transposition of great vessels	1	0		0.31
Tetralogy of Fallot	0	0		0.00
Hypoplastic left heart syndrome	0	1		0.31
Coarctation of aorta	0	0		0.00
Choanal atresia, bilateral	0	0		0.00
Cleft palate without cleft lip	9	0		2.75
Cleft lip with or without cleft palate	51	3		16.48
Oesophageal atresia / stenosis with or without fistula	10	0		3.05
Small intestine atresia / stenosis	6	0		1.83
Anorectal atresia / stenosis	11	1		3.66
Undescended testis (36 weeks of gestation or later)	nr	nr		nr
Hypospadias	9	0		2.75
Epispadias	0	0		0.00
Indeterminate sex	3	2		1.53
Renal agenesis	1	0		0.31
Cystic kidney	1	2		0.92
Bladder exstrophy	0	0		0.00
Polydactyly, preaxial	33	1		10.38
Total Limb reduction defects (include unspecified)	21	3		7.32
Transverse	12	0		3.66
Preaxial	4	1		1.53
Postaxial	1	0		0.31
Intercalary	1	2		0.92
Mixed	2	0		0.61
Unspecified	1	0		---
Diaphragmatic hernia	4	0		1.22
Omphalocele	6	0		1.83
Gastroschisis	14	1		4.58
Unspecified Omphalocele/Gastroschisis	0	0		---
Prune belly sequence	0	0		0.00
Trisomy 13	0	0		0.00
Trisomy 18	1	0		0.31
Down syndrome, all ages (include age unknown)	35	3		11.60
<20	6	1		9.67
20-24	7	0		6.93
25-29	7	1		9.12
30-34	3	0		6.30
35-39	9	0		58.49
40-44	2	1		99.34
45+	1	0		nc
unknown	0	0		---

* = include Microphthalmos and Unspecified Anophthalmos/ Microphthalmos
** = include Microtia and Unspecified Anotia/Microtia
nr = not reported
nc = not calculable

Mexico: RYVEMCE, Previous years rates 1980 - 2004

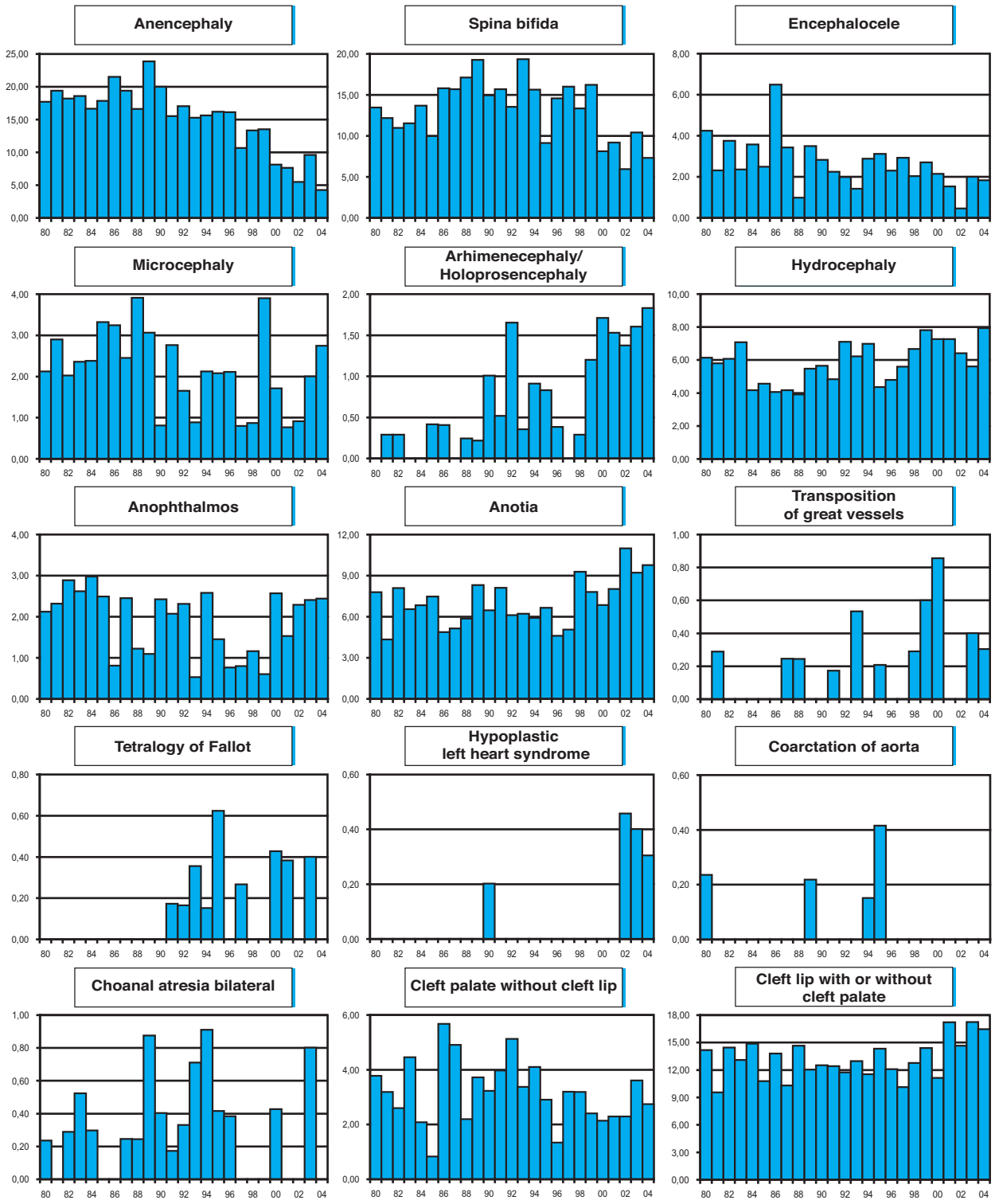
Birth prevalence rates: (LB+SB) * 10,000

	1974-79	1980-84	1985-89	1990-94	1995-99	2000-04
Total births		**183,228**	**176,079**	**290,075**	**205,529**	**129,044**
Anencephaly		18.12	19.99	16.58	14.26	6.90
Spina bifida		12.39	16.19	15.82	13.62	8.21
Encephalocele		3.27	3.18	2.28	2.63	1.63
Microcephaly		2.35	3.18	1.69	1.95	1.70
Arhinencephaly / Holoprosencephaly		0.11	0.23	0.90	0.54	1.63
Hydrocephaly		5.89	4.49	6.21	5.64	6.97
Anophthalmos**		2.57	1.59	2.00	0.97	2.25
Microphthalmos		nr	nr	nr	nr	nr
Unspecified Anophthalmos / Microphthalmos		---	---	---	---	---
Anotia***		6.77	6.42	6.55	6.47	8.99
Microtia		nr	nr	nr	nr	nr
Unspecified Anotia / Microtia		---	---	---	---	---
Transposition of great vessels		0.05	0.11	0.14	0.19	0.31
Tetralogy of Fallot		0.00	0.00	0.17	0.19	0.23
Hypoplastic left heart syndrome		0.00	0.00	0.03	0.00	0.23
Coarctation of aorta		0.05	0.06	0.03	0.10	0.00
Choanal atresia, bilateral		0.27	0.34	0.52	0.19	0.23
Cleft palate without cleft lip		3.27	3.52	4.00	2.53	2.63
Cleft lip with or without cleft palate		13.26	12.32	12.20	12.75	15.50
Oesophageal atresia / stenosis with or without fistula		1.26	1.76	2.41	1.99	2.79
Small intestine atresia / stenosis		0.65	0.85	1.34	1.22	2.17
Anorectal atresia / stenosis		3.93	4.43	5.27	4.28	5.11
Undescended testis (36 weeks of gestation or later)		nr	nr	nr	nr	nr
Hypospadias		4.20	3.75	5.17	3.89	3.64
Epispadias		nr	nr	nr	nr	0.00
Indeterminate sex		1.80	1.70	2.69	2.04	2.79
Renal agenesis		0.38	0.40	0.66	0.39	0.54
Cystic kidney		0.33	0.57	0.90	0.78	1.78
Bladder exstrophy		0.49	0.40	0.45	0.44	0.23
Polydactyly, preaxial		11.84	13.80	12.20	12.65	13.41
Total Limb reduction defects (include unspecified)		6.11	6.87	6.07	5.16	6.66
Transverse		nr	nr	nr	nr	3.56
Preaxial		nr	nr	nr	nr	1.32
Postaxial		nr	nr	nr	nr	0.31
Intercalary		nr	nr	nr	nr	0.54
Mixed		nr	nr	nr	nr	0.85
Unspecified		---	---	---	---	---
Diaphragmatic hernia		0.55	0.80	1.10	0.92	1.32
Omphalocele		1.58	1.65	1.69	1.46	2.40
Gastroschisis		0.71	1.19	2.07	2.63	5.11
Unspecified Omphalocele / Gastroschisis		---	---	---	---	---
Prune belly sequence		1.20	1.08	1.07	0.73	0.70
Trisomy 13		0.33	0.34	0.14	0.15	0.31
Trisomy 18		0.65	0.57	0.38	0.15	0.31
Down syndrome, all ages (include age unknown)		12.88	13.01	14.44	12.21	10.85
<20		6.63*	8.45	9.63	6.86	5.30
20-24		5.83*	6.23	8.08	5.27	6.39
25-29		4.00*	8.41	12.17	8.93	5.49
30-34		16.65*	16.56	14.60	11.85	12.70
35-39		44.09*	45.37	40.24	48.93	56.33
40-44		171.72*	134.23	130.13	231.87	174.15
45+		530.30*	185.19	223.55	346.53	0.00*
unknown		---	---	---	---	---

* data include less than 5 years
**= include Microphthalmos and Unspecified Anophthalmos/ Microphthalmos
*** = include Microtia and Unspecified Anotia/Microtia
nr = not reported
nc= not calculable

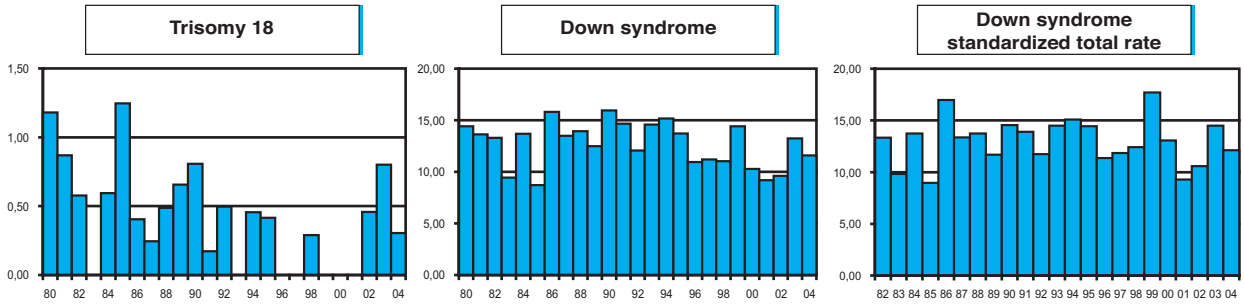
Mexico: RYVEMCE

'Time trends 1980-2004 (Birth prevalence rates per 10,000)



Note: L+S rates

Note: L+S rates



Note: L+S rates

New Zealand

New Zealand Congenital Anomalies Monitoring Programme

History:
The Programme began in 1975 and became a full member of the ICBDSR in 1979

Size and coverage:
The Programme covers all livebirths (approximately 56,000 per year) delivered or treated in a New Zealand publicly funded hospital. Only these data are included in the quarterly and annual reports to the ICBDSR. Data on stillbirths are retrospectively added to the database together with additional cases derived from the national perinatal and mortality databases. In late 1995 the definition of stillbirth was changed from 28 weeks completed gestation to 20 weeks or more gestation and/or 400g birthweight.

Legislation and funding:
The Programme is run and funded by the Public Health Directorate, Ministry of Health.

Sources of ascertainment:
Ascertainment is from discharge records of publicly funded hospitals and stillbirth notification forms. Data on voluntary terminations of pregnancy are being added to the database.

Exposure information:
No exposure information are currently available, but attempts are being made to obtain such data as well as increase the level of ascertainment.

Background information:
General epidemiological characteristics for all births are available.

Addresses and Staff:
Barry Borman, PhD, Programme Director Public Health Intelligence Public Health Directorate Ministry of Health, PO Box 5013, Wellington, New Zealand

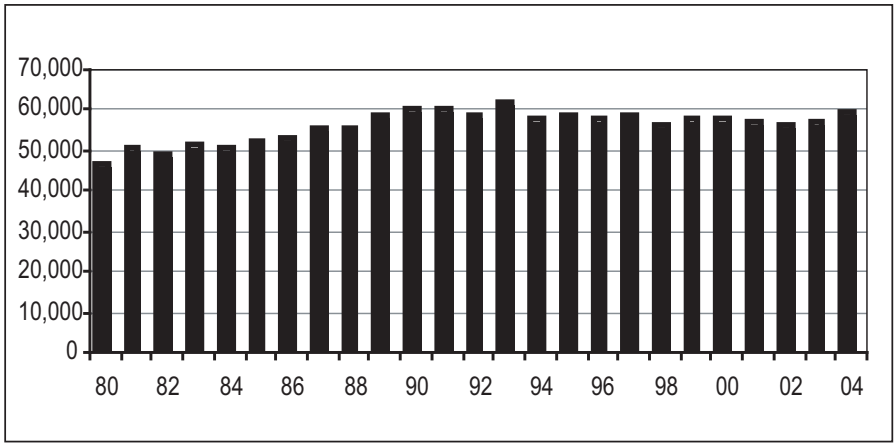
Phone: 64-4-496-2445

Fax: 64-4-496-2340

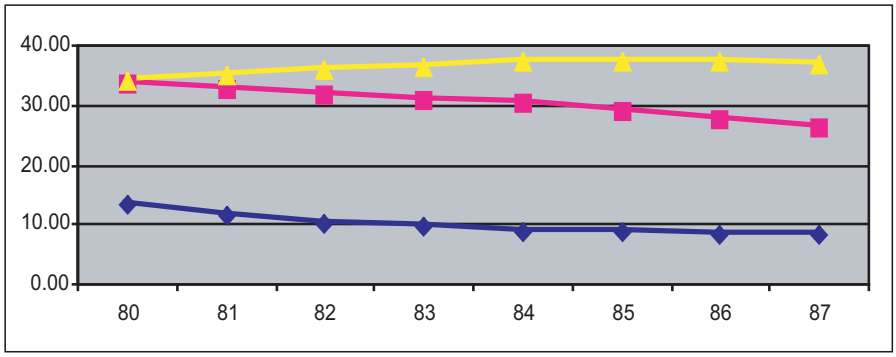
E-mail: barry_borman@moh.govt.nz

New Zealand

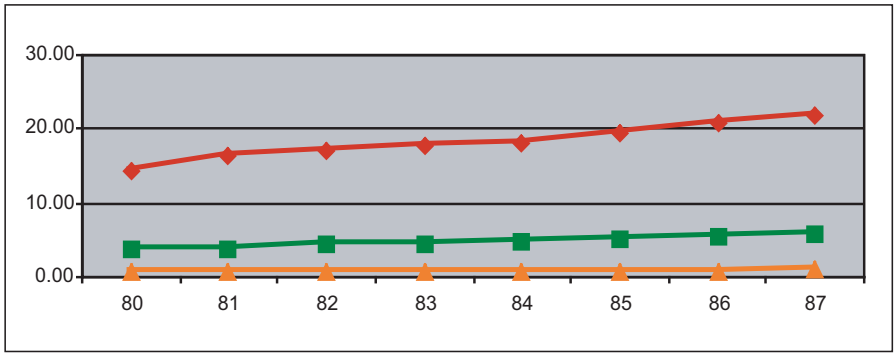
Total births by year



Percentage of births by maternal age



legenda: %births < 20 %births 20-24 %births 25-29



legenda: %births 30-34 %births 35-39 %births 40+

New Zealand: 2004

Live births (LB)	58,073
Stillbirths (SB)	483
Total births	58,556
Number of terminations of pregnancy (ToP) for birth defects	nr

Birth Defects	Number of cases			Rates*10,000
	LB	SB	ToP	Total rate
Anencephaly	3	nr	nr	0.52
Spina bifida	13	nr	nr	2.24
Encephalocele	3	nr	nr	0.52
Microcephaly	17	nr	nr	2.93
Arhinencephaly / Holoprosencephaly	nr	nr	nr	nr
Hydrocephaly	19	nr	nr	3.27
Anophthalmos	0	nr	nr	0.00
Microphthalmos	1	nr	nr	0.17
Unspecified Anophthalmos/ Microphthalmos	0	nr	nr	0.00
Anotia	nr	nr	nr	nr
Microtia	nr	nr	nr	nr
Unspecified Anotia/Microtia	nr	nr	nr	nr
Transposition of great vessels	30	nr	nr	5.17
Tetralogy of Fallot	24	nr	nr	4.13
Hypoplastic left heart syndrome	7	nr	nr	1.21
Coarctation of aorta	23	nr	nr	3.96
Choanal atresia, bilateral	2	nr	nr	0.34
Cleft palate without cleft lip	50	nr	nr	8.61
Cleft lip with or without cleft palate	48	nr	nr	8.27
Oesophageal atresia / stenosis with or without fistula	9	nr	nr	1.55
Small intestine atresia / stenosis	14	nr	nr	2.41
Anorectal atresia / stenosis	7	nr	nr	1.21
Undescended testis (36 weeks of gestation or later)	430	nr	nr	74.04
Hypospadias + epispadias	166	nr	nr	28.58
Epispadias	nr	nr	nr	nr
Indeterminate sex	4	nr	nr	0.69
Renal agenesis	20	nr	nr	3.44
Cystic kidney	30	nr	nr	5.17
Bladder exstrophy	0	nr	nr	0.00
Polydactyly, preaxial	52	nr	nr	8.95
Total Limb reduction defects (include unspecified)	17	nr	nr	2.93
Transverse	nr	nr	nr	nr
Preaxial	nr	nr	nr	nr
Postaxial	nr	nr	nr	nr
Intercalary	nr	nr	nr	nr
Mixed	nr	nr	nr	nr
Unspecified	nr	nr	nr	nr
Diaphragmatic hernia	10	nr	nr	1.72
Omphalocele	nr	nr	nr	nr
Gastroschisis	nr	nr	nr	nr
Unspecified Omphalocele/Gastroschisis	nr	nr	nr	nr
Prune belly sequence	nr	nr	nr	nr
Trisomy 13	4	nr	nr	0.69
Trisomy 18	5	nr	nr	0.86
Down syndrome, all ages (include age unknown)	60	nr	nr	10.33
<20	nr	nr	nr	nr
20-24	nr	nr	nr	nr
25-29	nr	nr	nr	nr
30-34	nr	nr	nr	nr
35-39	nr	nr	nr	nr
40-44	nr	nr	nr	nr
45+	nr	nr	nr	nr
unknown	nr	nr	nr	nr

nr = not reported

New Zealand: Previous years rates 1980 - 2004

Birth prevalence rates: (LB) * 10,000

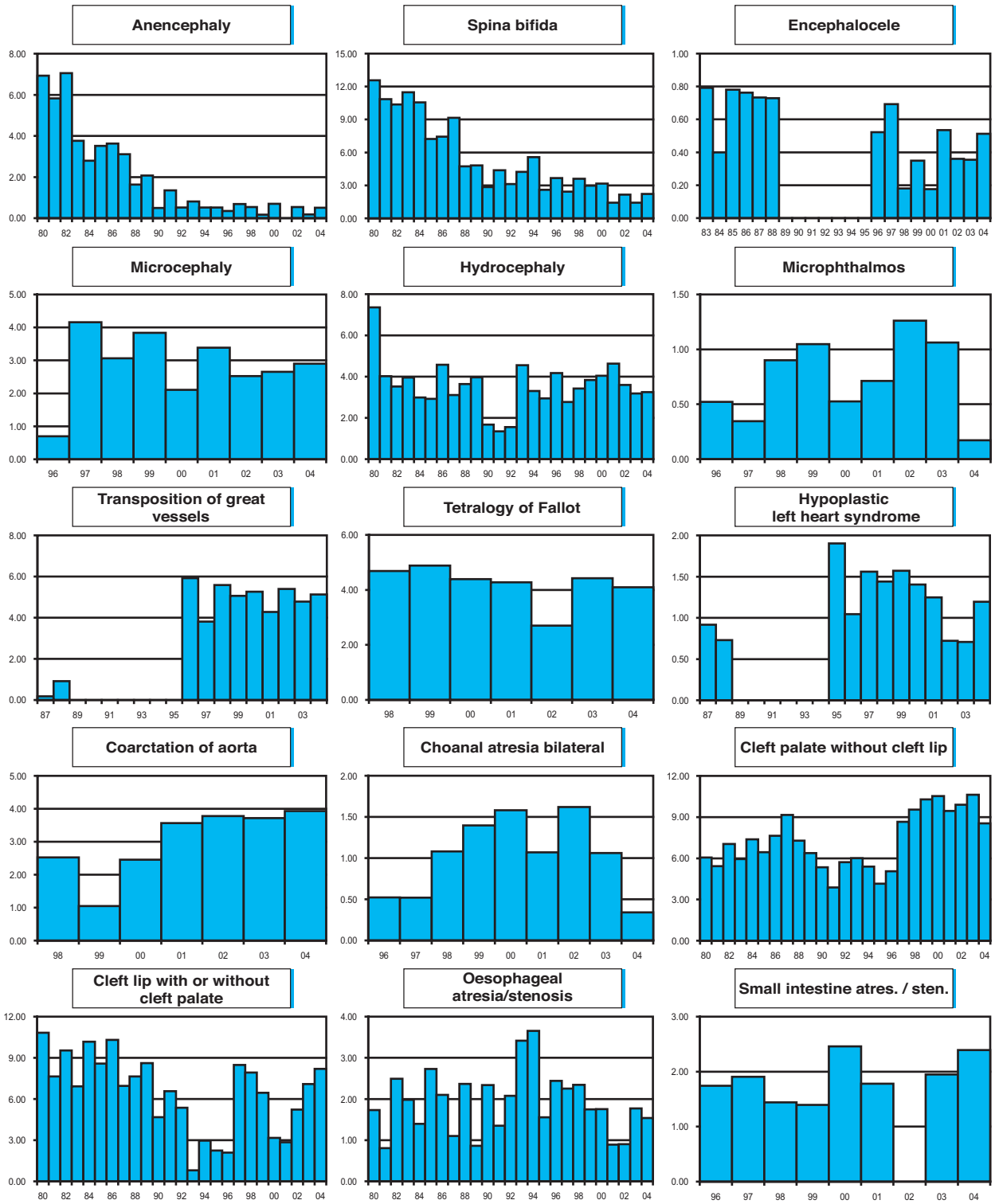
	1974-79	1980-84	1985-89	1990-94	1995-99	2000-04
Total births		**244,840**	**271,170**	**295,872**	**285,789**	**283,649**
Anencephaly		5.23	2.77	0.74	0.45	0.39
Spina bifida		11.15	6.64	4.02	3.04	2.08
Encephalocele		0.60*	0.75*	nr	0.35	0.39
Microcephaly		nr	nr	nr	2.94*	2.71
Arhinencephaly / Holoprosencephaly		nr	nr	nr	nr	nr
Hydrocephaly		4.33	3.65	2.50	3.43	3.74
Anophthalmos		nr	nr	nr	0.00*	0.04
Microphthalmos		nr	nr	nr	0.70*	0.74
Unspecified Anophthalmos / Microphthalmos		nr	nr	nr	0.00*	0.00
Anotia		nr	nr	nr	nr	nr
Microtia		nr	nr	nr	nr	nr
Unspecified Anotia / Microtia		nr	nr	nr	nr	nr
Transposition of great vessels		nr	0.55*	nr	5.09*	4.97
Tetralogy of Fallot		nr	nr	nr	4.79*	3.98
Hypoplastic left heart syndrome		nr	0.82*	nr	1.50	1.06
Coarctation of aorta		nr	nr	nr	1.77*	3.49
Choanal atresia, bilateral		nr	nr	nr	0.88*	1.13
Cleft palate without cleft lip		6.37	7.38	5.27	7.52	9.80
Cleft lip with or without cleft palate		8.99	8.41	4.06	5.42	5.32
Oesophageal atresia / stenosis with or without fistula		1.67	1.81	2.57	2.06	1.37
Small intestine atresia / stenosis		nr	nr	nr	1.62*	2.15*
Anorectal atresia / stenosis		2.33	2.47	2.87	2.34	2.22
Undescended testis (36 weeks of gestation or later)		nr	nr	nr	62.32*	77.84
Hypospadias		13.15	13.39	11.66*	20.97*	29.68
Epispadias		nr	nr	nr	nr	nr
Indeterminate sex		nr	nr	nr	0.47*	0.63
Renal agenesis		0.40*	0.33*	nr	3.52*	3.10
Cystic kidney		nr	nr	nr	5.61*	6.03
Bladder exstrophy		nr	nr	nr	0.35*	0.28
Polydactyly, preaxial		nr	nr	nr	5.99*	10.06*
Total Limb reduction defects (include unspecified)		3.72	3.10	2.43	2.41	2.93
Transverse		nr	nr	nr	nr	nr
Preaxial		nr	nr	nr	nr	nr
Postaxial		nr	nr	nr	nr	nr
Intercalary		nr	nr	nr	nr	nr
Mixed		nr	nr	nr	nr	nr
Unspecified		nr	nr	nr	nr	nr
Diaphragmatic hernia		1.39*	1.55*	nr	2.22*	2.54
Omphalocele		2.72*	1.59	2.20		
Gastroschisis		0.05*	0.75*	nr	nr	nr
Unspecified Omphalocele / Gastroschisis		0.00*	0.38*	nr	nr	nr
Prune belly sequence		nr	nr	nr	nr	nr
Trisomy 13		nr	nr	nr	0.44*	0.42
Trisomy 18		nr	nr	nr	0.88*	1.30
Down syndrome, all ages (include age unknown)		8.82	9.62	9.69*	10.22	11.49
<20		5.42	5.96*	nr	nr	nr
20-24		4.50	3.68*	nr	nr	nr
25-29		8.12	9.18*	nr	nr	nr
30-34		10.46	8.28*	nr	nr	nr
35-39		32.23	33.29*	nr	nr	nr
40-44		86.39	232.37*	nr	nr	nr
45+		138.89	147.06*	nr	nr	nr
unspecified		5.76	4.47*	nr	nr	nr

* data include less than 5 years

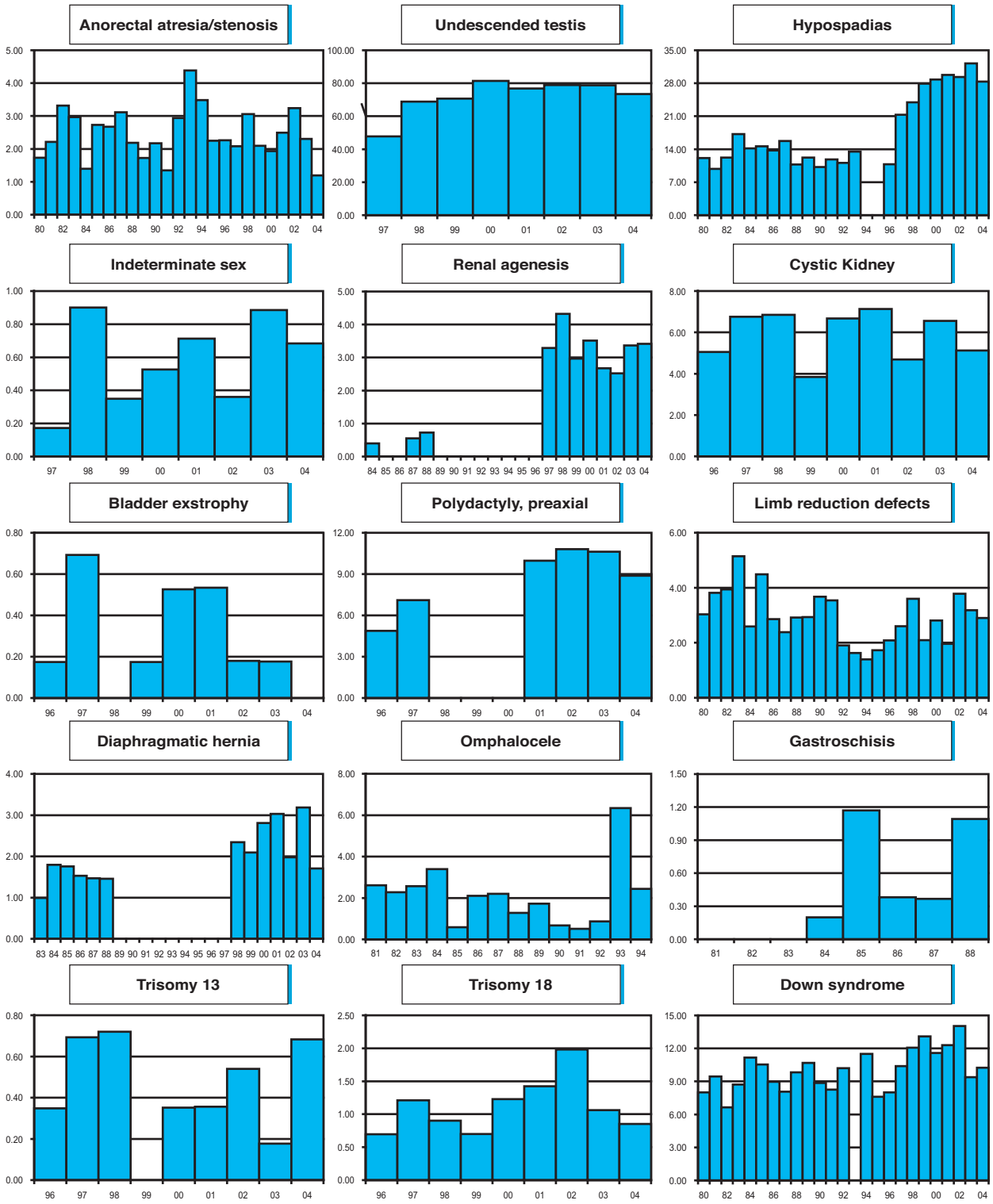
nr = not reported

New Zealand

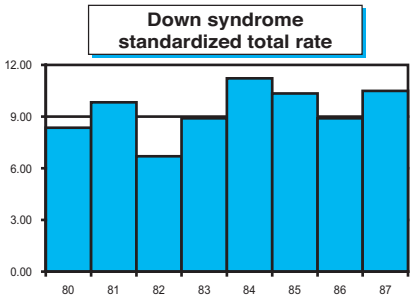
Time trends 1980-2004 (Birth prevalence rates per 10,000)



Note: L+S rates



Note: L+S rates



Note: L+S rates

Northern Netherlands

EUROCAT registration Northern Netherlands

History:
The Programme started in 1981, and became a Clearinghouse member in 1993.

Size and coverage
In the beginning the Programme covered 7,500 births annually in the province of Groningen and northern Drenthe. Coverage was gradually increased to 20,000 births annually in the provinces Groningen, Friesland and Drenthe from 1989 onwards. Home deliveries (35% of births) are included.

Legislation and funding
The Programme is funded by the Dutch Ministry of Public Health, Welfare and Sports. The registry is carried out in the Department of Genetics of the University Medical Center Groningen of the University of Groningen.

Sources of ascertainment
Children and foetuses with congenital anomalies are reported on a voluntary basis by various sources: obstetricians, pediatricians, clinical geneticists, surgeons, general practitioners, midwives, well-baby clinics, pathologists and the national obstetric registry. Registry personnel is also actively involved in data collection. Children and foetuses with congenital anomalies diagnosed before or after birth are eligible for registration at the EUROCAT registry, if the mother lived in the region at the time of birth and the child has not reached the age of 16 at notification. There is no lower limit for gestational age. Spontaneous and induced abortions are included. A number of frequently occurring mild anomalies is not registered, unless they occur in combination with other congenital anomalies. Informed consent of the parents is needed.

Exposure information
Since 1997 parents are asked to fill out a questionnaire including questions on occupational activities and medication use. Besides, pharmacy data are collected routinely and the actual use of the reported medications is verified with the mother.

Background information
General statistics are available from the Dutch Central Bureau of Statistics (CBS).

Addresses and Staff:

Marian Bakker, Programme Director Department of Genetics University Medical Centre Groningen University of Groningen, PO Box 30001 9700 RB Groningen, The Netherlands

Phone: +31-50-3617110 / 3617115

Fax: +31-50-3617232

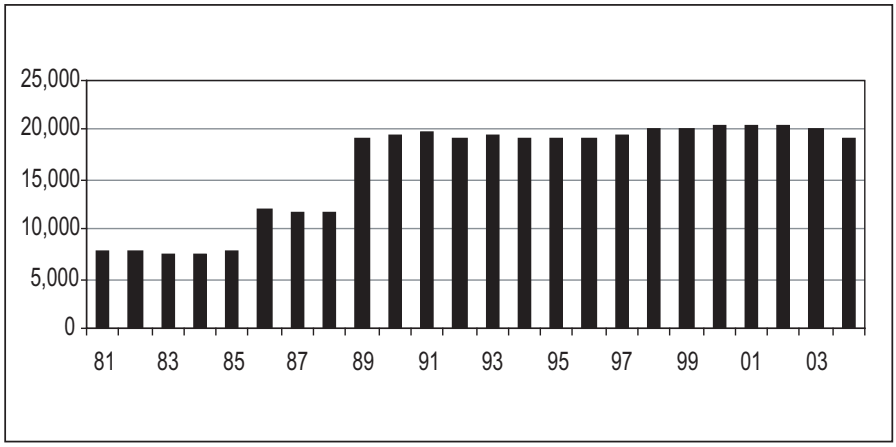
E-mail: m.k.bakker@medgen.umcg.nl

E-mail: h.e.k.de.Walle@medgen.umcg.nl

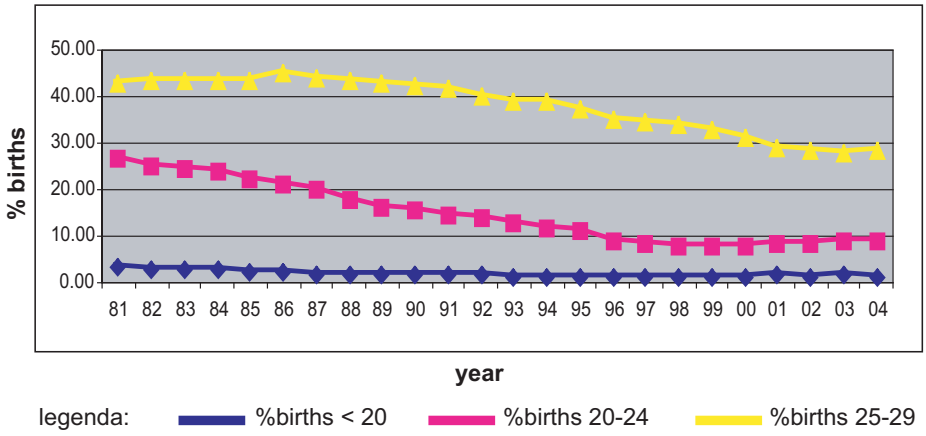
Website: www.eurocatnederland.nl

Northern Netherlands

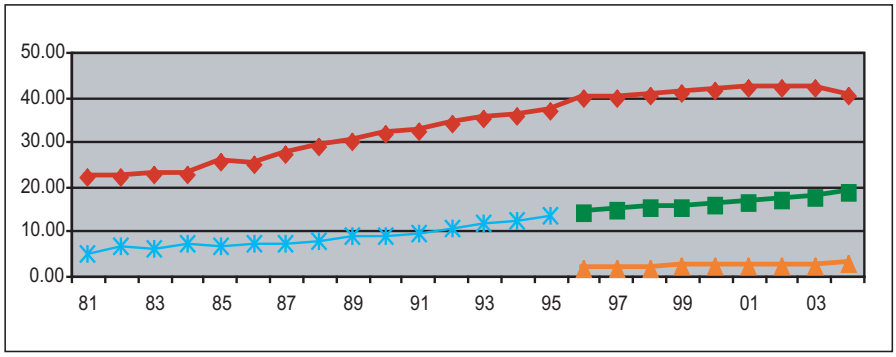
Total births by year



Percentage of births by maternal age



legenda: %births < 20 %births 20-24 %births 25-29



legenda: %births 30-34 %births 35-39 %births 40+

Northern Netherlands: 2004

Live births (LB)	19,012
Stillbirths (SB)	121
Total births	19,133
Number of terminations of pregnancy (ToP) for birth defects	23

Birth Defects	Number of cases			Rates*10,000
	LB	SB	ToP	Total rate
Anencephaly	0	0	1	0.52
Spina bifida	6	1	2	4.70
Encephalocele	0	0	1	0.52
Microcephaly	4	0	0	2.09
Arhinencephaly / Holoprosencephaly	1	0	0	0.52
Hydrocephaly	2	0	2	2.09
Anophthalmos	0	0	0	0.00
Microphthalmos	2	0	0	1.04
Unspecified Anophthalmos/ Microphthalmos	0	0	0	---
Anotia	1	0	0	0.52
Microtia	1	0	0	0.52
Unspecified Anotia/Microtia	0	0	0	---
Transposition of great vessels	8	0	0	4.18
Tetralogy of Fallot	6	0	0	3.13
Hypoplastic left heart syndrome	3	0	1	2.09
Coarctation of aorta	12	0	0	6.26
Choanal atresia, bilateral	0	0	0	0.00
Cleft palate without cleft lip	10	0	0	5.22
Cleft lip with or without cleft palate	19	0	1	10.44
Oesophageal atresia / stenosis with or without fistula	6	0	0	3.13
Small intestine atresia / stenosis	0	0	0	0.00
Anorectal atresia / stenosis	4	0	0	2.09
Undescended testis (36 weeks of gestation or later)	0	0	0	0.00
Hypospadias	22	0	0	11.48
Epispadias	0	0	0	0.00
Indeterminate sex	0	0	0	0.00
Renal agenesis	2	0	0	1.04
Cystic kidney	5	0	1	3.13
Bladder exstrophy	0	0	0	0.00
Polydactyly, preaxial	0	0	0	0.00
Total Limb reduction defects (include unspecified)	10	0	1	5.74
Transverse	7	0	0	3.65
Preaxial	1	0	0	0.52
Postaxial	4	0	0	2.09
Intercalary	0	0	0	0.00
Mixed	0	0	0	0.00
Unspecified	0	0	1	---
Diaphragmatic hernia	2	0	0	1.04
Omphalocele	3	0	0	1.57
Gastroschisis	1	0	1	1.04
Unspecified Omphalocele/Gastroschisis	0	0	0	---
Prune belly sequence	0	0	0	0.00
Trisomy 13	1	0	0	0.52
Trisomy 18	5	1	1	3.65
Down syndrome, all ages (include age unknown)	17	1	11	15.14
<20	0	0	0	0.00
20-24	1	0	0	5.93
25-29	6	0	1	12.92
30-34	5	0	2	9.09
35-39	3	0	4	19.71
40-44	2	1	4	125.22
45+	0	0	0	0.00
unknown	0	0	0	---

Northern Netherlands: Previous years rates 1981 - 2004

Prevalence rates: (LB+SB+TOP) * 10,000

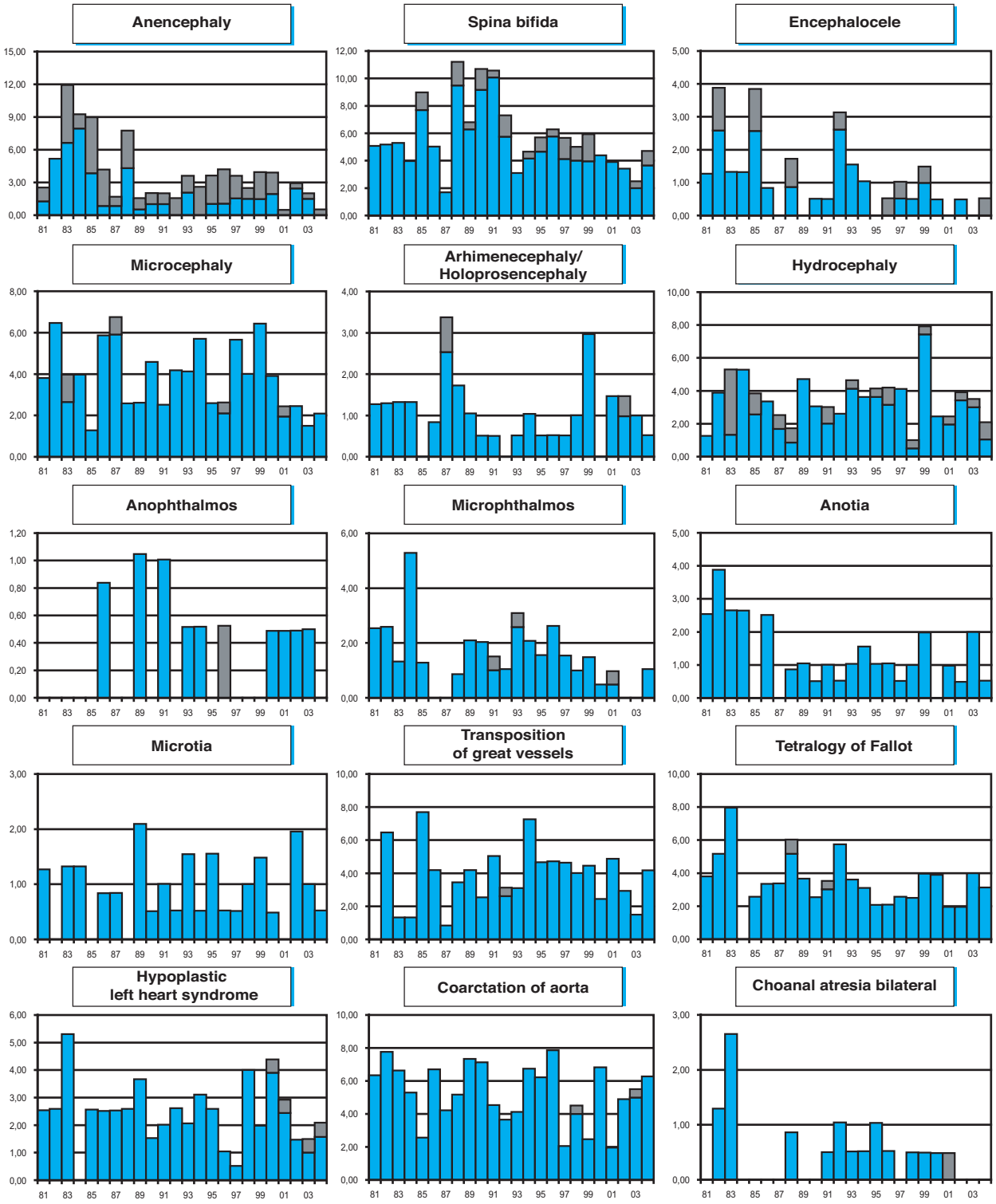
	1974-79	1980-84*	1985-89	1990-94	1995-99	2000-04
Births		**30,709**	**62,263**	**97,298**	**97,969**	**100,552**
Anencephaly		7.16	4.18	2.36	3.57	1.99
Spina bifida		4.88	6.58	7.30	5.72	3.78
Encephalocele		1.95	0.96	1.34	0.71	0.30
Microcephaly		4.56	3.85	4.21	4.29	2.49
Arhinencephaly / Holoprosencephaly		1.30	1.45	0.51	1.12	0.90
Hydrocephaly		3.91	3.37	3.39	4.29	2.88
Anophthalmos		0.00	0.48	0.41	0.10	0.40
Microphthalmos		2.93	0.96	1.95	1.63	0.50
Unspecified Anophthalmos / Microphthalmos		---	---	---	---	---
Anotia		2.93	0.96	0.92	1.12	0.80
Microtia		0.98	0.96	0.82	1.02	0.80
Unspecified Anotia / Microtia		---	---	---	---	---
Transposition of great vessels		2.28	3.85	4.21	4.49	3.18
Tetralogy of Fallot		4.23	3.85	3.70	2.65	2.98
Hypoplastic left heart syndrome		2.61	2.89	2.26	2.04	2.49
Coarctation of aorta		6.51	5.62	5.24	4.59	5.07
Choanal atresia, bilateral		0.98	0.16	0.51	0.51	0.20
Cleft palate without cleft lip		7.49	6.75	8.32	6.12	6.86
Cleft lip with or without cleft palate		16.28	15.10	15.21	14.80	13.33
Oesophageal atresia / stenosis with or without fistula		2.28	3.21	2.67	3.98	3.68
Small intestine atresia / stenosis		2.93	2.89	2.36	2.45	1.59
Anorectal atresia / stenosis		1.95	3.69	2.98	3.57	3.58
Undescended testis (36 weeks of gestation or later)		2.28	2.09	1.54	1.22	1.39
Hypospadias		17.58	9.64	10.07	13.07	13.82
Epispadias		0.33	0.64	0.41	0.51	0.40
Indeterminate sex		0.00	0.32	0.21	0.51	0.40
Renal agenesis		3.26	4.82	4.32	4.39	4.18
Cystic kidney		1.63	5.30	5.04	4.18	3.18
Bladder exstrophy		0.33	0.16	0.21	0.10	0.30
Polydactyly, preaxial		1.95	1.77	1.95	2.25	1.09
Total Limb reduction defects (include unspecified)		8.47	4.98	7.30	5.41	5.97
Transverse		5.54	2.57	4.21	3.37	3.58
Preaxial		1.30	0.64	0.92	0.61	0.90
Postaxial		0.65	0.80	1.75	1.12	1.49
Intercalary		0.00	0.00	0.31	0.10	0.10
Mixed		0.00	0.32	0.51	0.10	0.20
Unspecified		---	---	---	---	---
Diaphragmatic hernia		2.28	3.05	2.16	3.47	1.99
Omphalocele		1.95	1.77	3.49	2.76	1.49
Gastroschisis		0.98	0.96	0.31	1.12	0.90
Unspecified Omphalocele / Gastroschisis		---	---	---	---	---
Prune belly sequence		0.33	0.16	0.62	0.20	0.30
Trisomy 13		0.33	1.12	1.54	0.82	1.09
Trisomy 18		2.61	2.25	1.54	3.78	3.88
Down syndrome, all ages (include age unknown)		10.75	15.26	13.57	15.62	15.41
<20		0.00	0.00	0.00	0.00	0.00
20-24		13.01	4.25	9.04	5.89	4.68
25-29		3.76	14.78	5.86	7.38	9.65
30-34		13.05	8.67	14.00	12.67	9.54
35-39		30.85	52.40	36.07	38.33	29.67
40-44		nr	nr	nr	100.60*	118.46
45+		nr	nr	nr	217.39*	114.94
unknown		---	---	---	---	---

* data include less than 5 years

nr = not reported

Northern Netherlands

'Time trends 1981-2004 (Birth prevalence rates per 10,000)



Note: L+S rates, ToP rates

Note: L+S rates, ToP rates

Note: L+S rates, ToP rates

Norway
Medical Birth Registry of Norway

History:
The Programme was started in 1967. The Programme was a founding member of the ICBD-SR and is a full member.

Size and coverage
The Programme covers all births in Norway, approximately 60,000 annual births. Stillbirths of 16 weeks or more gestation are included.

Legislation and funding
The Programme is run and funded by the governmental National Institute of Public Health. Reporting is compulsory.

Sources of ascertainment
The registry is based on the notification of births from the delivery units and since 1999 also from the neonatal units.

Exposure information
Some basic information, such as maternal disease and since 1999:smoking and occupation, is collected on all infants, malformed or not.

Background information
All information available for the reported malformed infants is also available for the total population of births.

Addresses and Staff:

Lorentz M. Irgens, MD, Programme Director
Department of Medical Birth Registry of Norway
University of Bergen Kalfarveien, 31 NO-5018 Bergen - Norway

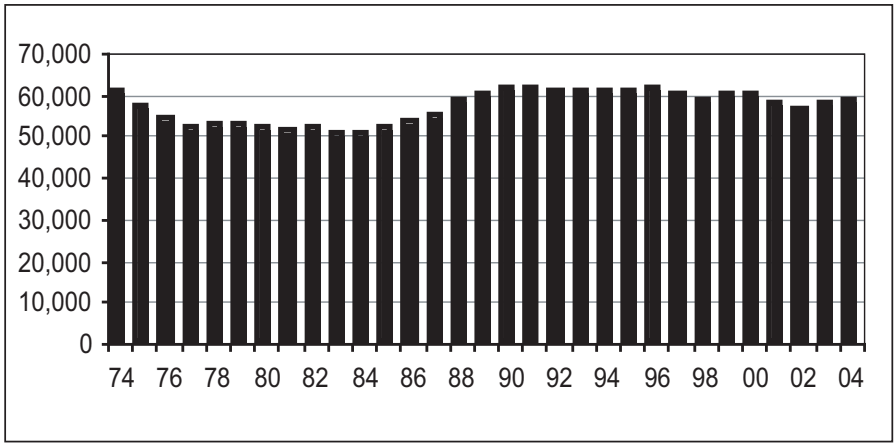
Phone: +47- 22 04 2702

Fax: +47 - 22 04 2701

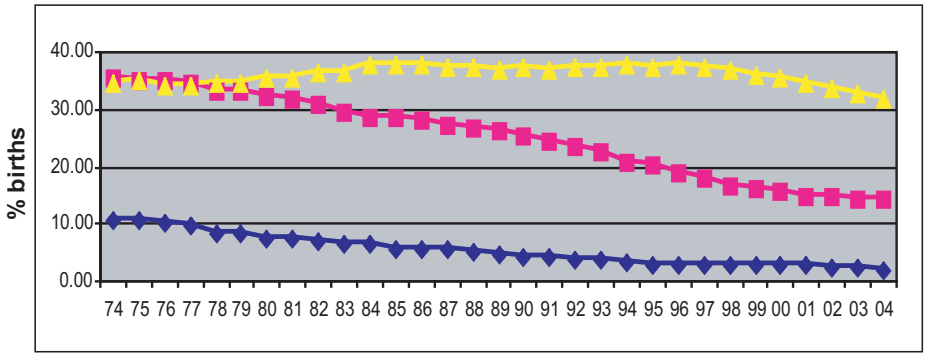
E-mail: lorentz.irgens@mfr.uib.no

Norway

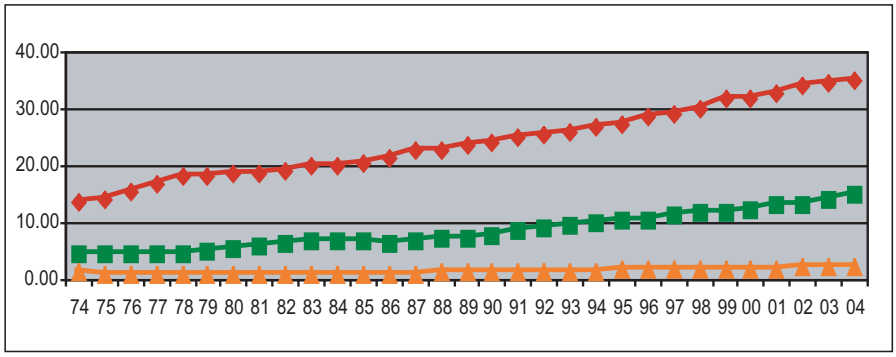
Total births by year



Percentage of births by maternal age



legenda: %births < 20 %births 20-24 %births 25-29



legenda: %births 30-34 %births 35-39 %births 40+

Norway: 2004

Live births (LB)	57,622
Stillbirths (SB)	469
Total births	58,091
Number of terminations of pregnancy (ToP) for birth defects	190

Birth Defects	Number of cases			Rates*10,000
	LB	SB	ToP	Total rate
Anencephaly	1	5	22	4.80
Spina bifida	20	3	9	5.49
Encephalocele	2	0	5	1.20
Microcephaly	0	0	0	0.00
Arhinencephaly / Holoprosencephaly	3	1	5	1.54
Hydrocephaly	14	5	11	5.15
Anophthalmos	0	0	0	0.00
Microphthalmos	4	0	1	0.86
Unspecified Anophthalmos/ Microphthalmos	nr	nr	nr	---
Anotia	0	0	0	0.00
Microtia	6	0	0	1.03
Unspecified Anotia/Microtia	nr	nr	nr	---
Transposition of great vessels	14	2	0	2.75
Tetralogy of Fallot	13	0	2	2.57
Hypoplastic left heart syndrome	13	0	6	3.26
Coarctation of aorta	11	1	1	2.23
Choanal atresia, bilateral	2	0	0	0.34
Cleft palate without cleft lip	45	1	1	8.06
Cleft lip with or without cleft palate	61	0	3	10.98
Oesophageal atresia / stenosis with or without fistula	12	0	0	2.06
Small intestine atresia / stenosis	7	0	0	1.20
Anorectal atresia / stenosis	18	0	1	3.26
Undescended testis (36 weeks of gestation or later)	174	0	0	29.86
Hypospadias	83	0	0	14.24
Epispadias	1	0	0	0.17
Indeterminate sex	2	0	0	0.34
Renal agenesis	2	0	3	0.86
Cystic kidney	15	1	7	3.95
Bladder exstrophy	1	0	0	0.17
Polydactyly, preaxial	48	0	2	8.58
Total Limb reduction defects (include unspecified)	21	2	2	4.29
Transverse	9	1	1	1.89
Preaxial	5	0	0	0.86
Postaxial	0	0	0	0.00
Intercalary	0	0	0	0.00
Mixed	9	1	1	1.89
Unspecified	nr	nr	nr	---
Diaphragmatic hernia	9	0	5	2.40
Omphalocele	9	0	1	1.72
Gastroschisis	8	0	1	1.54
Unspecified Omphalocele/Gastroschisis	nr	nr	nr	---
Prune belly sequence	6	0	2	1.37
Trisomy 13	2	0	1	0.51
Trisomy 18	3	0	15	3.09
Down syndrome, all ages (include age unknown)	90	6	28	21.28
<20	1	0	0	8.45
20-24	6	1	1	9.70
25-29	12	0	2	7.61
30-34	29	1	4	16.81
35-39	32	0	10	48.68
40-44	9	4	11	183.07
45+	1	0	0	200.00
unknown	0	0	0	---

nr = not reported

Norway: Previous years rates 1974 - 2004

Birth prevalence rates: (LB+SB) * 10,000 until 1993
Birth prevalence rates: (LB+SB+TOP) * 10,000 since 1994

	1974-79	1980-84	1985-89	1990-94	1995-99	2000-04
Total births	**327,455**	**255,156**	**276,487**	**304,426**	**300,214**	**289,409**
Anencephaly	4.12	3.29	2.21	1.94	3.10	4.56
Spina bifida	4.83	5.72	4.77	3.88	5.06	4.94
Encephalocele	0.40	0.82	0.54	0.56	0.57	1.21
Microcephaly	0.70	0.55	0.61	0.56	0.83	0.38
Arhinencephaly / Holoprosencephaly	0.03	0.27	0.43	0.59	1.03	0.79
Hydrocephaly	4.24	3.49	3.36	3.32	3.63	5.11
Anophthalmos	0.00	0.16	0.18	0.10	0.07	0.03
Microphthalmos	0.12	0.27	0.29	0.30	0.20	0.38
Unspecified Anophthalmos / Microphthalmos	---	---	---	---	---	---
Anotia	0.12	0.27	0.14	0.20	0.13	0.24
Microtia	nr	nr	1.47*	0.53	0.43	0.55
Unspecified Anotia / Microtia	---	---	---	---	---	---
Transposition of great vessels	0.40	0.74	1.56	1.84	2.40	2.70
Tetralogy of Fallot	0.12	0.27	0.51	1.08	1.53	2.83
Hypoplastic left heart syndrome	nr	nr	0.70*	1.64	2.13	2.76
Coarctation of aorta	0.38*	0.47	0.69	0.92	1.13	1.90
Choanal atresia, bilateral	0.21	0.43	0.65	0.43	0.40	0.66
Cleft palate without cleft lip	4.61	5.09	5.71	5.12	5.90	6.15
Cleft lip with or without cleft palate	14.08	14.03	14.76	13.21	12.96	12.61
Oesophageal atresia / stenosis with or without fistula	2.05	1.72	2.17	2.43	1.87	2.45
Small intestine atresia / stenosis	0.86	1.25	1.05	1.45	1.63	0.83
Anorectal atresia / stenosis	1.50	2.08	2.28	2.56	2.00	2.52
Undescended testis (36 weeks of gestation or later)	18.14	14.85	15.08	18.07	15.96	27.75
Hypospadias	12.73	13.76	16.17	15.96	14.46	16.24
Epispadias	0.27	0.35	0.47	0.16	0.33	0.24
Indeterminate sex	2.23	3.96	3.98	5.45	7.26	0.59
Renal agenesis	0.12	0.78	1.34	1.41	1.53	0.97
Cystic kidney	0.46	0.82	1.70	2.43	3.13	5.04
Bladder exstrophy	0.24	0.55	0.29	0.20	0.40	0.24
Polydactyly, preaxial	nr	nr	nr	nr	8.34*	8.22
Total Limb reduction defects (include unspecified)	8.83	6.47	7.34	6.50	6.66	4.01
Transverse	nr	nr	2.68*	3.61	3.03	2.04
Preaxial	nr	nr	0.84*	0.66	0.37	0.62
Postaxial	nr	nr	1.00*	0.56	0.40	0.03
Intercalary	nr	nr	0.17*	0.33	0.53	0.03
Mixed	nr	nr	0.00*	0.62	1.33	1.45
Unspecified	---	---	---	---	---	---
Diaphragmatic hernia	1.92	2.39	2.53	2.37	2.93	2.38
Omphalocele	2.38	2.04	2.03	2.00	2.10	2.28
Gastroschisis	1.25	1.69	1.74	2.14	2.96	2.42
Unspecified Omphalocele / Gastroschisis	---	---	---	---	---	---
Prune belly sequence	nr	nr	nr	nr	1.50*	1.24
Trisomy 13	nr	nr	nr	nr	0.33*	0.76
Trisomy 18	nr	nr	nr	nr	1.00*	1.52
Down syndrome, all ages (include age unknown)	9.80	9.76	11.32	10.12	12.92	16.69
<20	1.90	4.00	5.46	0.86	5.97	5.82
20-24	6.21	6.16	8.00	5.24	3.34	5.87
25-29	8.09	6.70	6.12	6.81	6.85	7.11
30-34	10.48	13.20	13.81	11.78	10.67	13.59
35-39	37.32	31.18	39.10	22.81	37.31	45.51
40-44	134.91	76.30	62.69	83.99	128.45	152.67
45+	161.29	185.19	315.79	322.58	196.08	363.64
unknown	---	---	---	---	---	---

* data include less than 5 years

nr = not reported

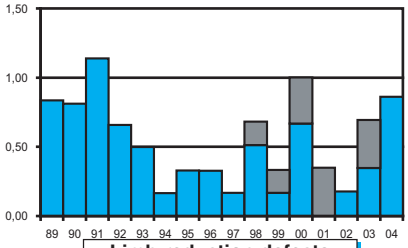
Norway

'Time trends 1974-2004 (Birth prevalence rates per 10,000)

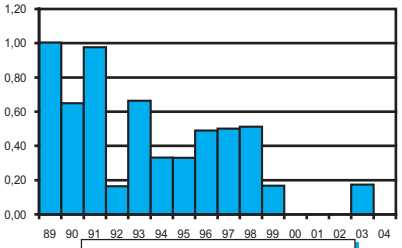
Note: L+S rates, ToP rates

Note: L+S rates, ToP rates

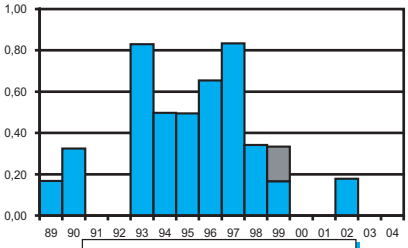
Limb reduction defects preaxial



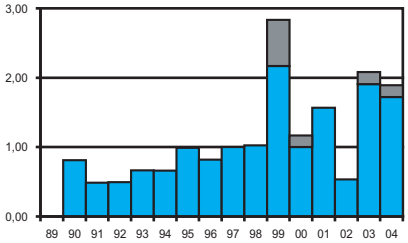
Limb reduction defects postaxial



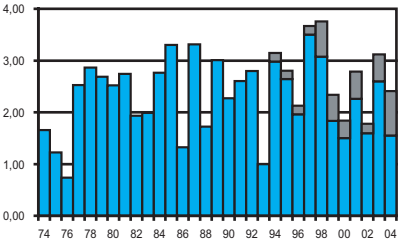
Limb reduction defects intercalary



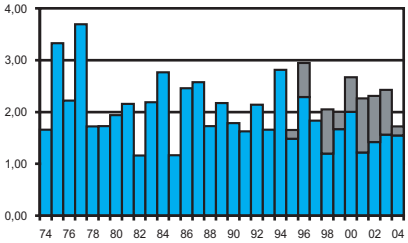
Limb reduction defects mixed



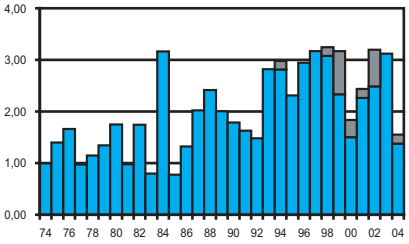
Diaphragmatic hernia



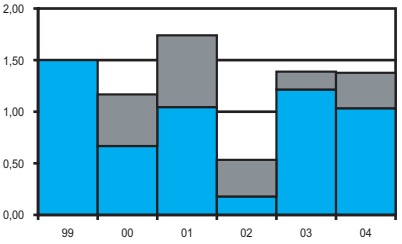
Omphalocele



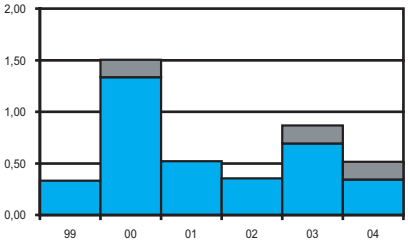
Gastroschisis



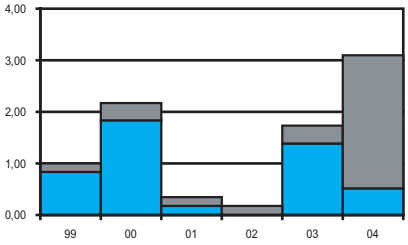
Prune belly sequence



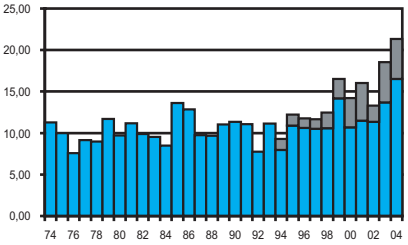
Trisomy 13



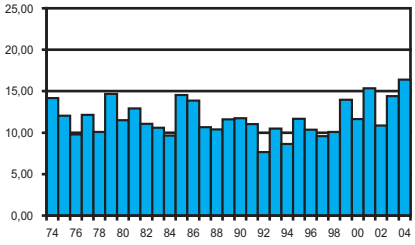
Trisomy 18



Down syndrome



Down syndrome standardized total rate



Note: L+S rates, ToP rates

Russia: MRRCM

Moscow Regional Registry of Congenital malformation (MRRCM)

History:
Moscow Regional Registry of Congenital malformation started the activity in 1999 and legally defined by the order of the Ministry of Health Care of Russian Federation. MRRCM became a Member of ICBDSR in 2001

Size and coverage:
MRRCM be located as a section of Moscow Regional Medical genetic consultation by The Moscow Regional Research institute of obstetrics and gynecology (MONIIAG). Director of the MONIIAG is Professor Vladislav Krasnopolsky. The Head of the Moscow Regional Medical genetic consultation and Director of the Programme of MRRCM is Ludmila Jouthenko. The Programme of Monitoring of Birth defects covers all births in Moscow Region. In 1999 MRRCM observed 45 000 birth. There are about 55 000 births today (2002). The information about babies and fetuses with Birth defects collect from 54 maternity hospitals also from all women consultations and clinics, children clinics. Prenatal diagnosed and terminated fetuses are register also.

Legislation and funding:
Monitoring of the birth of fetuses and babies with congenital malformations is legally defined by the Order of the Ministry of Health Care of Russian Federation in 1999.

Sources of ascertainment:
Reporting is made by neonatologist during the first week of the infant's life in maternity hospitals and by pediatricians during the first year – in pediatric clinics and departments. Reports are collected form cytogenetic laboratories, pathology departments.

Exposure information:
No exposure information is routinely collected in the registry.

Background information:
Background information on all births is available from statistics department.

Addresses and Staff:
Ludmila Joutchenko, MD, Programme Director
Moscow Regional Research Scientific Institute of Obstetric and Gynecology, 22a, Pokrovka St. 101000 Moscow Russia

Phone: 007-0959246228

Fax: 007-0959215398

E-mail: mrrcm@mail.ru

Russia: Moscow Region, 2004

Live births (LB)	56,828
Stillbirths (SB)	364
Total births	57,192
Number of terminations of pregnancy (ToP) for birth defects	159

Birth Defects	Number of cases			Rates*10,000
	LB	SB	ToP	Total rate
Anencephaly	0	1	16	2.96
Spina bifida	21	0	4	4.36
Encephalocele	1	0	1	0.35
Microcephaly	3	0	1	0.70
Arhinencephaly / Holoprosencephaly	0	0	1	0.17
Hydrocephaly	12	1	5	3.14
Anophthalmos	1	0	0	0.17
Microphthalmos	2	0	0	0.35
Unspecified Anophthalmos/ Microphthalmos	0	0	0	---
Anotia	1	0	0	0.17
Microtia	5	0	0	0.87
Unspecified Anotia/Microtia	0	0	0	---
Transposition of great vessels	11	1	1	2.27
Tetralogy of Fallot	6	0	0	1.05
Hypoplastic left heart syndrome	2	0	1	0.52
Coarctation of aorta	6	0	1	1.22
Choanal atresia, bilateral	2	0	0	0.35
Cleft palate without cleft lip	32	0	0	5.58
Cleft lip with or without cleft palate	36	0	2	6.63
Oesophageal atresia / stenosis with or without fistula	11	0	0	1.92
Small intestine atresia / stenosis	7	0	1	1.39
Anorectal atresia / stenosis	11	1	1	2.27
Undescended testis (36 weeks of gestation or later)	95	0	0	16.56
Hypospadias	65	0	0	11.33
Epispadias	0	0	0	0.00
Indeterminate sex	2	0	0	0.35
Renal agenesis	2	0	5	1.22
Cystic kidney	14	0	9	4.01
Bladder exstrophy	0	0	0	0.00
Polydactyly, preaxial	64	0	0	11.16
Total Limb reduction defects (include unspecified)	15	0	0	2.62
Transverse	9	0	3	2.09
Preaxial	0	0	0	0.00
Postaxial	0	0	0	0.00
Intercalary	1	0	0	0.17
Mixed	2	0	3	0.87
Unspecified	3	0	3	---
Diaphragmatic hernia	10	0	1	1.92
Omphalocele	3	1	5	1.57
Gastroschisis	15	1	2	3.14
Unspecified Omphalocele/Gastroschisis	0	0	0	---
Prune belly sequence	0	0	0	0.00
Trisomy 13	0	0	1	0.17
Trisomy 18	0	0	1	0.17
Down syndrome, all ages (include age unknown)	61	0	5	11.51
<20	4	0	0	5.40
20-24	9	0	1	4.78
25-29	14	0	0	8.59
30-34	19	0	1	23.31
35-39	8	0	0	24.10
40-44	7	0	3	148.59
45+	0	0	0	0.00
unknown	0	0	0	---

Slovak Republic

Congenital Malformations Monitoring Program of the Slovak Republic

History:
In Slovakia the collection of reports from delivery units and processing of data performs the National Health Information Center SR, former the Institution of Health Information and Statistics (IHIS). The obligation of reporting all groups of congenital malformations results from valid legislation norms. Reporting of congenital malformations began in 1964 year.
The programme of Slovak Teratological Information Center (STIC) was established in 2003 year and consists in cooperation of the Slovak Medical University, IHIS and the Center of Medical Genetics. Research collaboration began from 1995 year, under the responsibility of Elena Szabova.

Size and coverage
The registry covers all births in the area approximately 55.000 births annually according to the announcements of birth defects from delivery units. The detail informations about cases of CM are collected in the Center of Medical Genetics, Bratislava from western regions of Slovakia (cca 15.000 births) by Eva Veghova, or under the running research projects at the Slovak Medical University.

Legislation and funding
Reporting is compulsory. Analysis of data is supported by grant projects.

Sources of ascertainment
Reports are received from delivery units, neonatal, pediatric clinics, or departments of clinical genetics.

Exposure Information
Detail information on maternal and paternal occupation, drug use, etc. are collected by interviews of case´s and control´s mothers only according to research projects.

Backround information
Some background information is available from the general population statistics..

Addresses and Staff:

Elena Szabova Slovak Medical University Limbova 12, 833 03 Bratislava, Slovak Republic

Phone: 00421 2 59369241

E-mail: elena.szabova@szu.sk

Contact for the Slovak Teratologic Informaton Center (STIC):

Elena Szabova

E-mail: elena.szabova@szu.sk

Eva Veghova

Phone: 00421 2 52968855

E-mail: evaveghova@pobox.sk

Daniela Brasenova

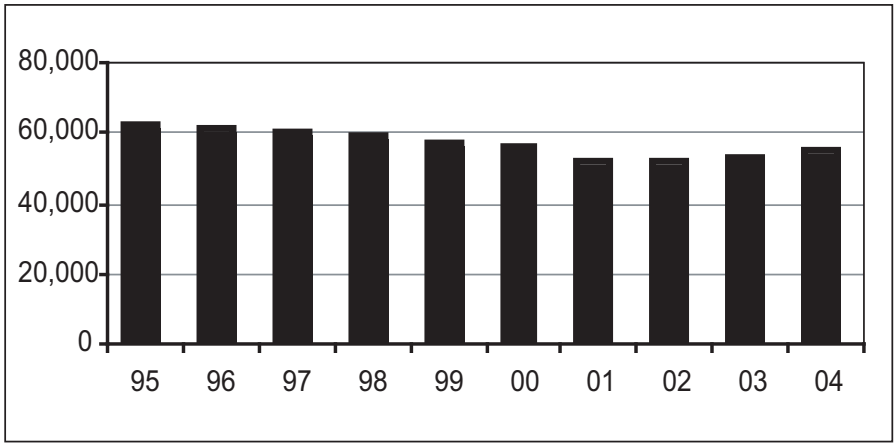
Phone: 00421 2 57269301

Dagmar Zeljenkova

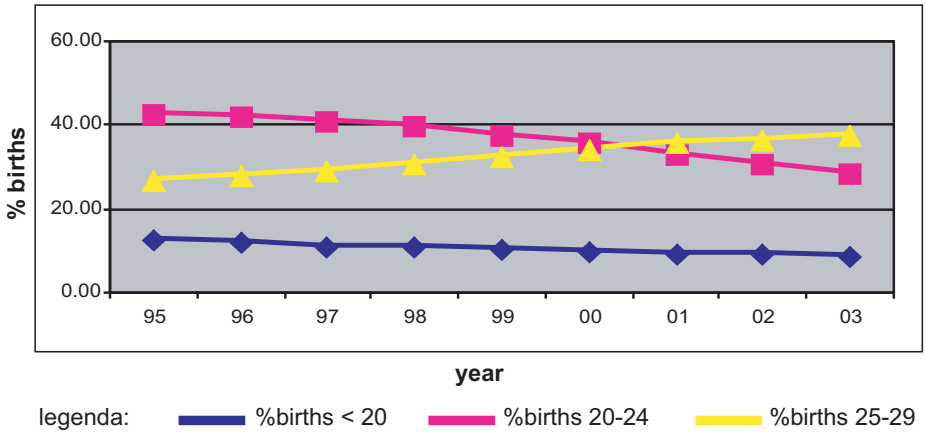
Phone: 00421 2 59369379

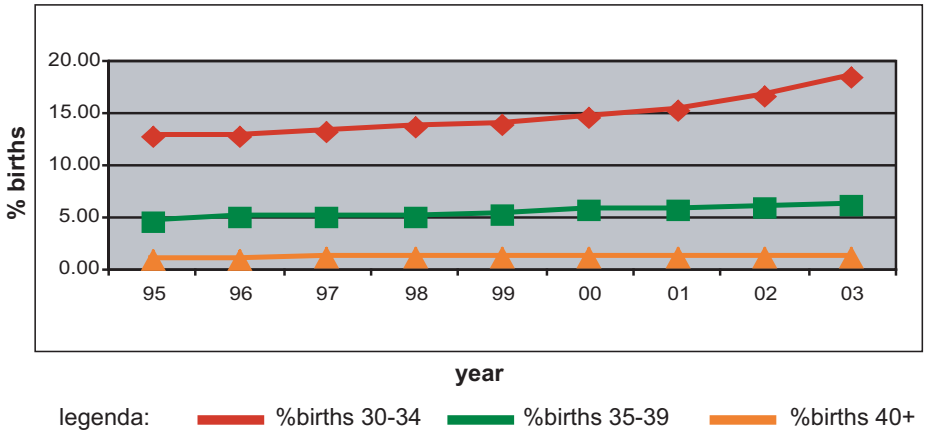
Slovak Republic

Total births by year



Percentage of births by maternal age





Slovak Republic: 2004

Live births (LB)	53,747
Stillbirths (SB)	211
Total births	53,958
Number of terminations of pregnancy (ToP) for birth defects	33

Birth Defects	Number of cases			Rates*10,000
	LB	SB	ToP	Total rate
Anencephaly	0	0	1	0.19
Spina bifida	17	0	1	3.33
Encephalocele	4	1	1	1.11
Microcephaly	3	1	1	0.93
Arhinencephaly / Holoprosencephaly	1	0	1	0.37
Hydrocephaly	15	1	9	4.63
Anophthalmos	0	0	0	0.00
Microphthalmos	2	0	0	0.37
Unspecified Anophthalmos/ Microphthalmos	nr	nr	nr	---
Anotia	0	0	0	0.00
Microtia	1	0	0	0.19
Unspecified Anotia/Microtia	2	0	0	---
Transposition of great vessels	7	0	0	1.30
Tetralogy of Fallot	9	0	0	1.67
Hypoplastic left heart syndrome	12	0	0	2.22
Coarctation of aorta	5	0	0	0.93
Choanal atresia, bilateral	1	0	0	0.19
Cleft palate without cleft lip	24	1	2	5.00
Cleft lip with or without cleft palate	42	1	0	7.96
Oesophageal atresia / stenosis with or without fistula	9	0	0	1.67
Small intestine atresia / stenosis	7	1	0	1.48
Anorectal atresia / stenosis	14	1	0	2.78
Undescended testis (36 weeks of gestation or later)	53	1	0	10.00
Hypospadias	145	4	0	27.60
Epispadias	0	0	0	0.00
Indeterminate sex	2	0	0	0.37
Renal agenesis	28	0	0	5.19
Cystic kidney	10	0	0	1.85
Bladder exstrophy	2	0	0	0.37
Polydactyly, preaxial	22	0	0	4.07
Total Limb reduction defects (include unspecified)	17	0	0	3.15
Transverse	nr	nr	nr	nr
Preaxial	nr	nr	nr	nr
Postaxial	nr	nr	nr	nr
Intercalary	nr	nr	nr	nr
Mixed	nr	nr	nr	nr
Unspecified	nr	nr	nr	---
Diaphragmatic hernia	5	0	0	0.93
Omphalocele	3	1	0	0.74
Gastroschisis	5	0	0	0.93
Unspecified Omphalocele/Gastroschisis	nr	nr	nr	---
Prune belly sequence	2	0	0	0.37
Trisomy 13	0	0	0	0.00
Trisomy 18	3	0	0	0.56
Down syndrome, all ages (include age unknown)	45	0	5	9.26
<20	1	0	0	nr
20-24	7	0	1	nr
25-29	11	0	0	nr
30-34	9	0	0	nr
35-39	10	0	3	nr
40-44	6	0	1	nr
45+	1	0	0	nr
unknown	0	0	0	---

nr = not reported

Slovak Republic: Previous years rates 1995 - 2004

Birth prevalence rates: (LB+SB+TOP) * 10,000

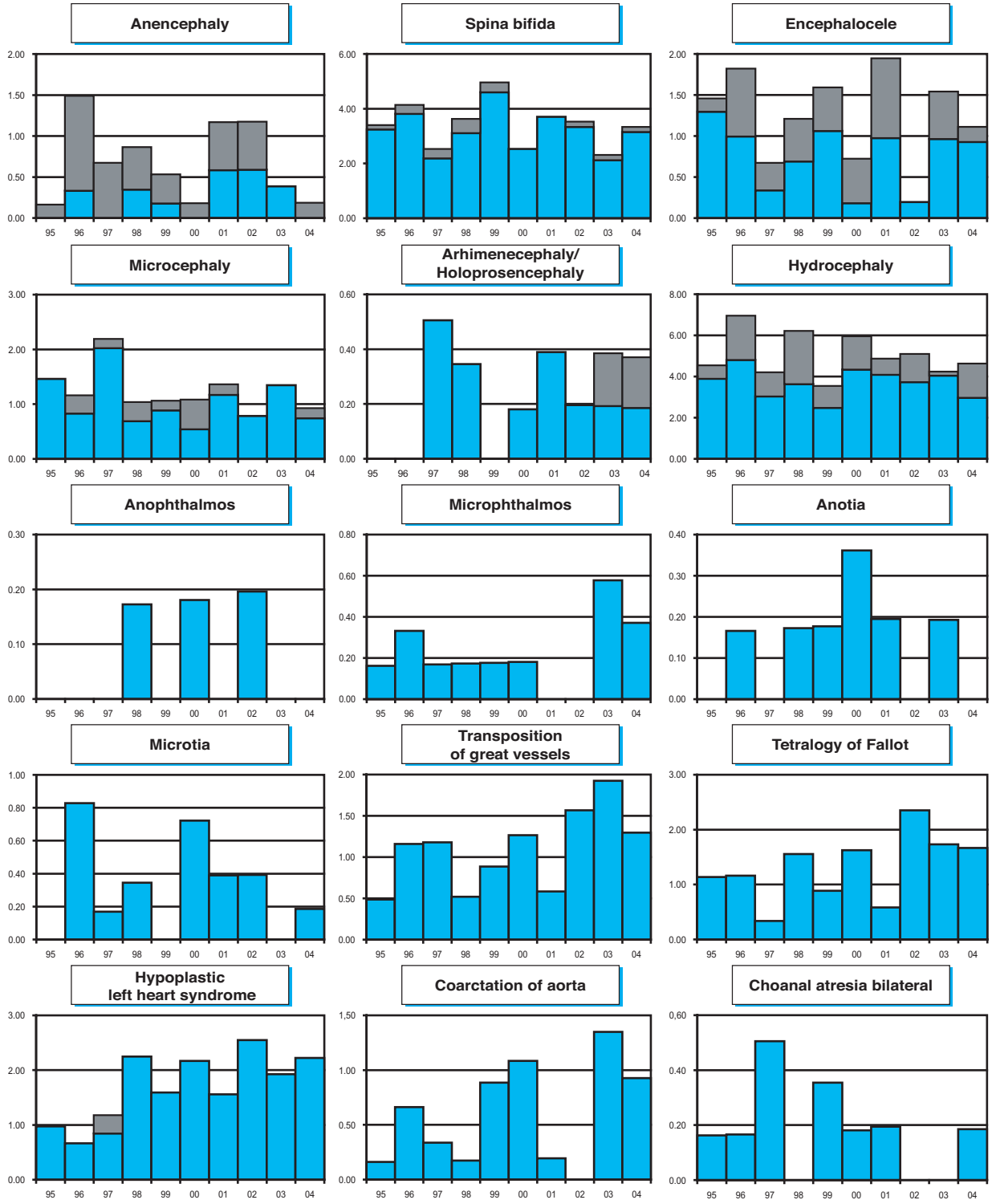
	1974-79	1980-84	1985-89	1990-94	1995-99	2000-04
Births					**295,732**	**263,632**
Anencephaly					0.74	0.61
Spina bifida					3.72	3.07
Encephalocele					1.35	1.10
Microcephaly					1.39	1.10
Arhinencephaly / Holoprosencephaly					0.17	0.30
Hydrocephaly					5.11	4.97
Anophthalmos					0.03	0.08
Microphthalmos					0.20	0.23
Unspecified Anophthalmos / Microphthalmos					---	---
Anotia					0.10	0.15
Microtia					0.27	0.34
Unspecified Anotia / Microtia					---	---
Transposition of great vessels					0.85	1.33
Tetralogy of Fallot					1.01	1.59
Hypoplastic left heart syndrome					1.32	2.09
Coarctation of aorta					0.44	0.72
Choanal atresia, bilateral					0.24	0.11
Cleft palate without cleft lip					5.28	5.73
Cleft lip with or without cleft palate					10.35	10.09
Oesophageal atresia / stenosis with or without fistula					1.01	1.52
Small intestine atresia / stenosis					1.59	1.67
Anorectal atresia / stenosis					1.56	2.96
Undescended testis (36 weeks of gestation or later)					6.12	8.34
Hypospadias					23.26	23.63
Epispadias					0.17	0.15
Indeterminate sex					0.51	0.34
Renal agenesis					2.50	5.46
Cystic kidney					0.85	1.63
Bladder exstrophy					0.17	0.19
Polydactyly, preaxial					1.59	3.57
Total Limb reduction defects (include unspecified)					3.52	3.64
Transverse					nr	nr
Preaxial					nr	nr
Postaxial					nr	nr
Intercalary					nr	nr
Mixed					nr	nr
Unspecified					---	---
Diaphragmatic hernia					1.28	1.48
Omphalocele					0.57	0.68
Gastroschisis					0.78	1.21
Unspecified Omphalocele / Gastroschisis					0.00	0.00
Prune belly sequence					0.00	0.23
Trisomy 13					0.27	0.34
Trisomy 18					0.27	0.46
Down syndrome, all ages (include age unknown)					9.37	10.28
<20					6.97	5.30*
20-24					6.50	3.88*
25-29					6.17	7.59*
30-34					12.54	12.95*
35-39					34.56	45.52*
40-44					71.60	104.12*
45+					173.91	300.00*
unspecified					---	---

* data include less than 5 years

nr = not reported

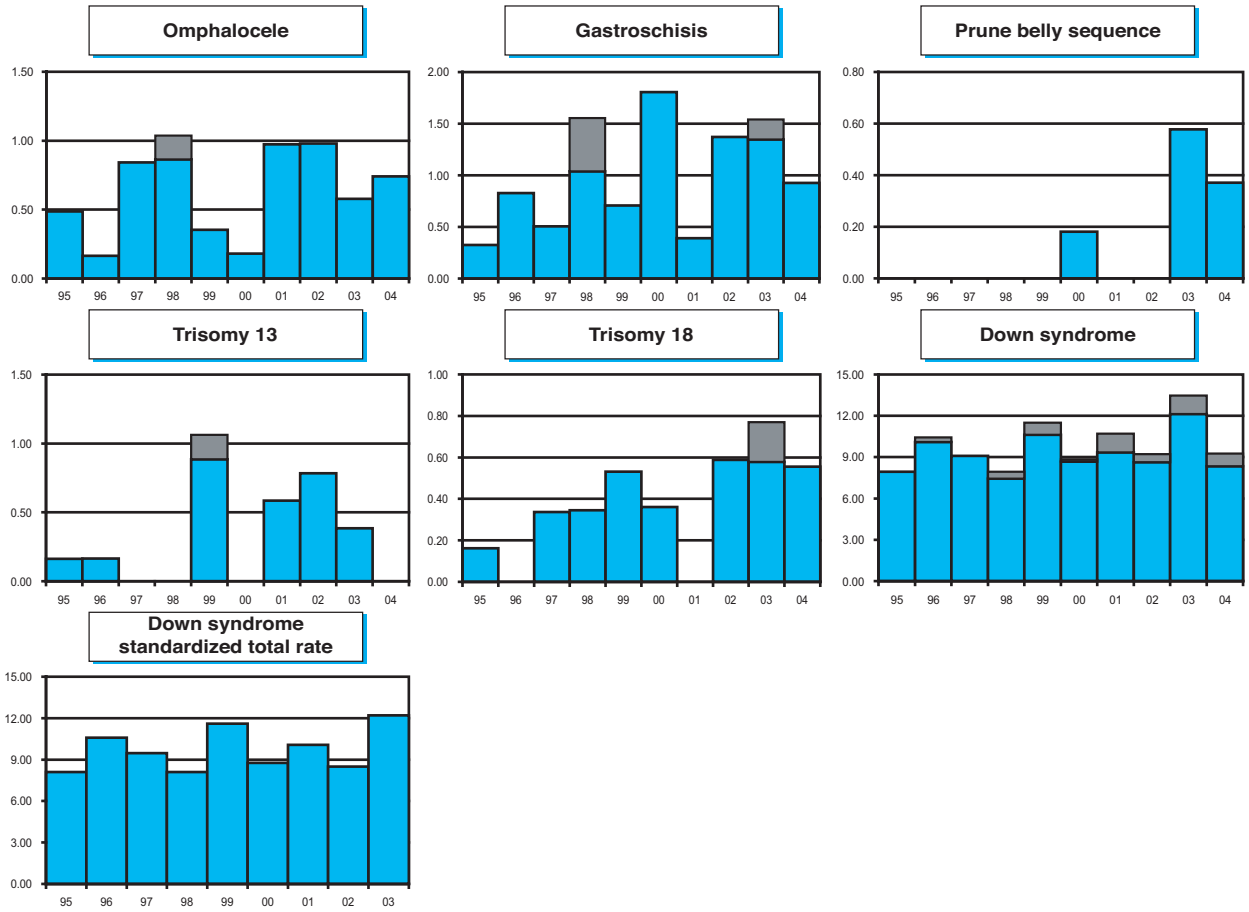
Slovak Republic

'Time trends 1995-2004 (Birth prevalence rates per 10,000)



Note: L+S rates, ToP rates

Note: L+S rates, ToP rates



Note: L+S rates, ToP rates

South America: ECLAMC

Latin American Collaborative Study of Congenital Malformations (ECLAMC)

History:
The Programme started in 1967 and has grown in size and coverage. The Programme became a full member of the International Clearinghouse in 1977.

Size and coverage:
The number of participating hospitals has grown from 20 in 1977 to 70 at the present time, distributed over most South Americans countries. The annual number of births covered is at present approximately 150,000, less than 1% of all births. Stillbirths of at least 500g birthweight have been included since 1978.

Legislation and funding:
The Programme is a research Programme with voluntary participation of hospitals and funded by research grants provided from several sources, mainly the national research councils of Argentina and Brazil.

Sources of ascertainment:
Reporting is made by collaborating pediatricians at the delivery units of participating hospitals.

Exposure information:
The mother of each reported infant and the mother of a control infant - the next non-malformed infant born at that hospital with the same sex as the proband - are interviewed on various exposures, including drug usage and parental occupation.

Background information:
Background information is obtained partly from summarising tables of births in each participating hospitals, partly from the matched control newborns.

Addresses and Staff:
Eduardo Castilla, MD, Programme Director ECLAMC/Dept.Genetica/FIOCRUZ C.P. 926 20010-970 Rio de Janeiro, Brazil

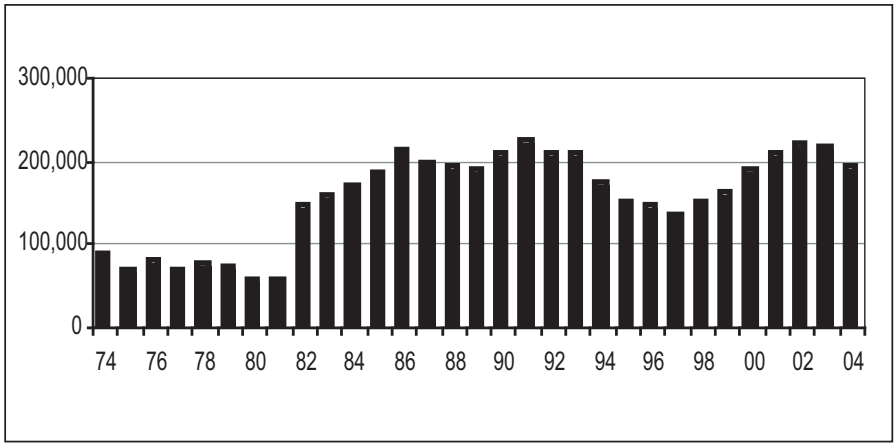
Phone: 55-21-25528952

Fax: 55-21-22604282(5521)

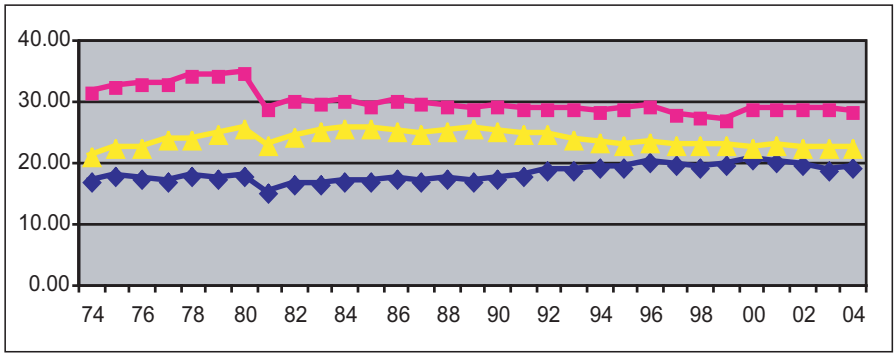
E-mail: castilla@centroin.com.br

South America: ECLAMC

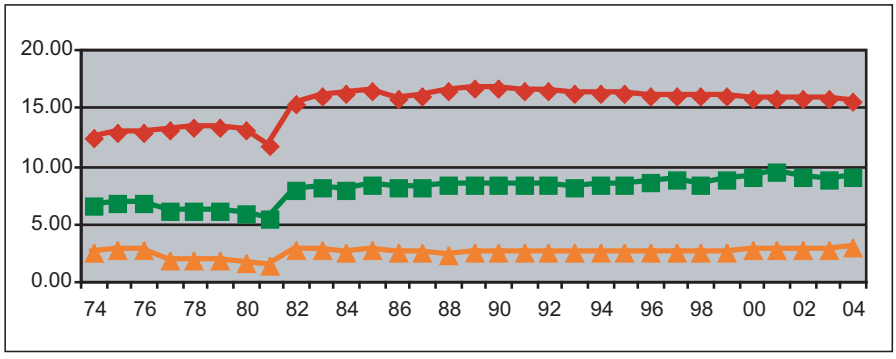
Total births by year



Percentage of births by maternal age



legenda: %births < 20 %births 20-24 %births 25-29



legenda: %births 30-34 %births 35-39 %births 40+

South America: 2004

Live births (LB)	188,884
Stillbirths (SB)	2317
Total births	191,201
Number of terminations of pregnancy (ToP) for birth defects	not permitted

Birth Defects	Number of cases			Rates*10,000
	LB	SB	ToP	Total rate
Anencephaly	59	69		6.69
Spina bifida	182	9		9.99
Encephalocele	54	10		3.35
Microcephaly	73	6		4.13
Arhinencephaly / Holoprosencephaly	23	9		1.67
Hydrocephaly	210	21		12.08
Anophthalmos	22	1		1.20
Microphthalmos	24	6		1.57
Unspecified Anophthalmos/ Microphthalmos	0	0		---
Anotia	4	0		0.21
Microtia	116	8		6.49
Unspecified Anotia/Microtia	4	1		---
Transposition of great vessels	25	0		1.31
Tetralogy of Fallot	30	1		1.62
Hypoplastic left heart syndrome	25	1		1.36
Coarctation of aorta	13	1		0.73
Choanal atresia, bilateral	5	0		0.26
Cleft palate without cleft lip	114	12		6.59
Cleft lip with or without cleft palate	263	19		14.75
Oesophageal atresia / stenosis with or without fistula	70	4		3.87
Small intestine atresia / stenosis	69	1		3.66
Anorectal atresia / stenosis	102	13		6.01
Undescended testis (36 weeks of gestation or later)	134	2		7.11
Hypospadias	80	0		4.18
Epispadias	2	0		0.10
Indeterminate sex	34	12		2.41
Renal agenesis	50	9		3.09
Cystic kidney	75	5		4.18
Bladder exstrophy	6	1		0.37
Polydactyly, preaxial	83	0		4.34
Total Limb reduction defects (include unspecified)	120	16		7.11
Transverse	60	5		3.40
Preaxial	18	5		1.20
Postaxial	10	0		0.52
Intercalary	14	0		0.73
Mixed	18	5		1.20
Unspecified	0	1		---
Diaphragmatic hernia	55	4		3.09
Omphalocele	60	14		3.87
Gastroschisis	77	5		4.29
Unspecified Omphalocele/Gastroschisis	8	2		---
Prune belly sequence	17	0		0.89
Trisomy 13	9	3		0.63
Trisomy 18	21	5		1.36
Down syndrome, all ages (include age unknown)	397	7		21.13
<20	28	0		7.61
20-24	61	2		11.77
25-29	38	1		9.20
30-34	45	0		15.11
35-39	105	2		62.35
40-44	110	1		217.52
45+	10	0		354.61
unknown	0	1		---

South America: Previous years rates 1974 - 2004

Prevalence rates: (LB+SB) * 10,000

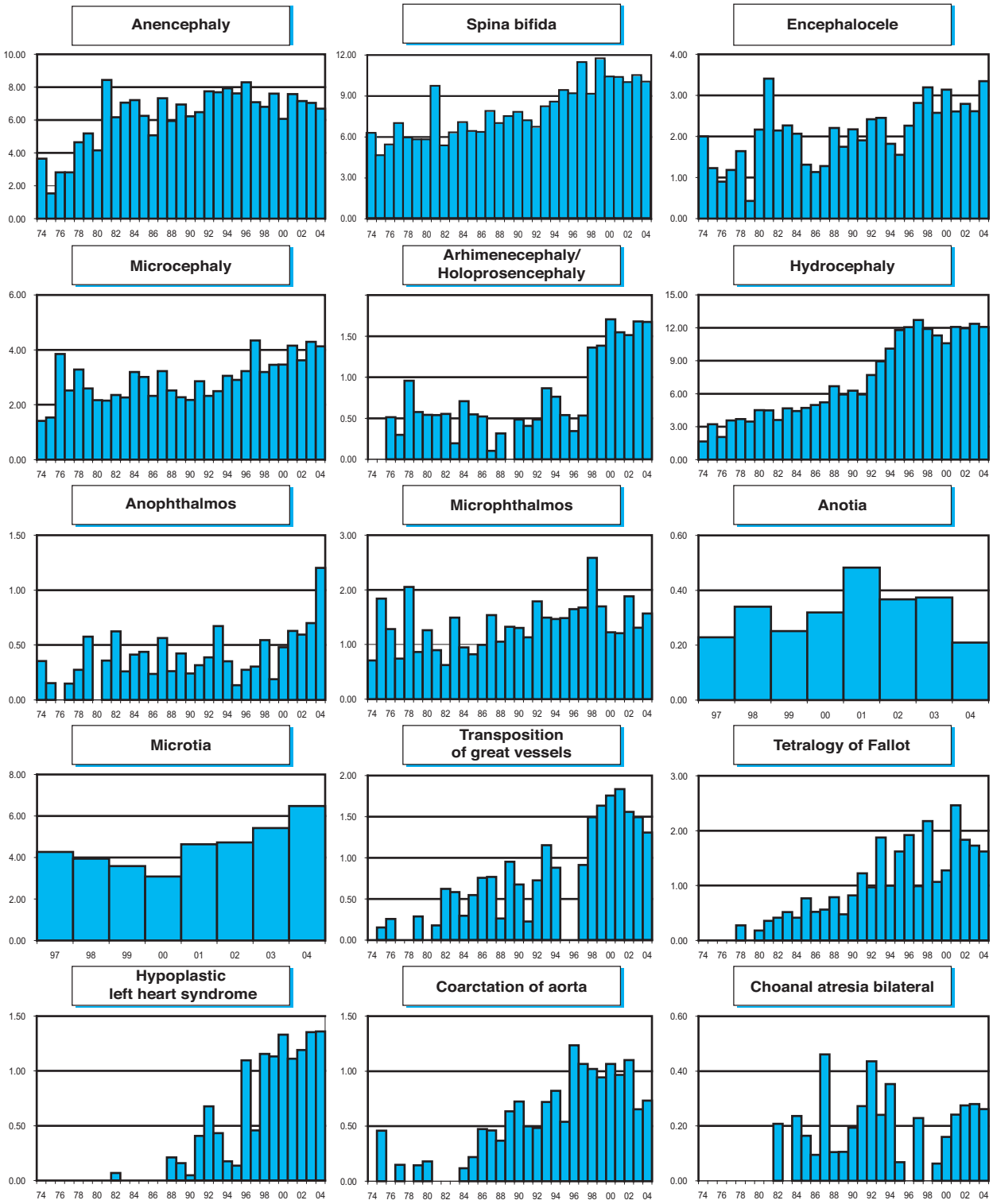
	1974-79	1980-84	1985-89	1990-94	1995-99	2000-04
Births	**438,055**	**579,119**	**968,001**	**1,012,539**	**731,513**	**1,018,471**
Anencephaly	3.47	6.73	6.28	7.18	7.49	6.93
Spina bifida	5.82	6.54	6.97	7.62	10.14	10.22
Encephalocele	1.26	2.28	1.53	2.16	2.47	2.89
Microcephaly	2.53	2.54	2.67	2.57	3.40	3.94
Arhinencephaly / Holoprosencephaly	0.39	0.50	0.30	0.59	0.85	1.62
Hydrocephaly	2.88	4.30	5.50	7.68	11.93	11.84
Anophthalmos	0.25	0.38	0.38	0.40	0.29	0.72
Microphthalmos	1.23	1.04	1.15	1.43	1.82	1.44
Unspecified Anophthalmos / Microphthalmos	---	---	---	---	---	---
Anotia	nr	nr	nr	nr	0.27*	0.35
Microtia	nr	nr	nr	nr	3.91*	4.88
Unspecified Anotia / Microtia	---	---	---	---	---	---
Transposition of great vessels	0.11	0.41	0.66	0.72	1.03	1.59
Tetralogy of Fallot	0.05	0.41	0.62	1.19	1.56	1.80
Hypoplastic left heart syndrome	0.00	0.02	0.07	0.36	0.81	1.27
Coarctation of aorta	0.11	0.05	0.43	0.64	0.96	0.90
Choanal atresia, bilateral	0.00	0.12	0.19	0.30	0.07	0.25
Cleft palate without cleft lip	3.20	3.35	3.31	3.87	3.73	5.03
Cleft lip with or without cleft palate	11.07	10.53	10.83	10.46	12.59	13.46
Oesophageal atresia / stenosis with or without fistula	2.01	2.68	2.58	2.83	3.40	3.67
Small intestine atresia / stenosis	0.48	1.64	1.44	1.82	2.01	3.06
Anorectal atresia / stenosis	2.76	3.82	3.64	4.46	4.85	5.70
Undescended testis (36 weeks of gestation or later)	1.58	3.35	4.58	4.80	5.21	6.74
Hypospadias	3.47	4.71	4.00	4.36	5.10	5.04
Epispadias	0.14	0.38	0.28	0.36	0.12	0.23
Indeterminate sex	1.00	2.33	1.90	1.88	1.72	2.29
Renal agenesis	0.43	0.66	0.85	1.61	2.24	2.50
Cystic kidney	0.55	1.11	1.47	2.06	3.70	4.28
Bladder exstrophy	0.14	0.21	0.28	0.25	0.33	0.39
Polydactyly, preaxial	2.97	2.28	2.52	2.64	2.95	3.92
Total Limb reduction defects (include unspecified)	4.09	5.49	4.83	5.52	6.14	6.77
Transverse	2.15	2.61	2.70	2.73	2.95	3.63
Preaxial	0.62	1.11	0.99	1.11	1.57	1.40
Postaxial	0.27	0.50	0.26	0.47	0.40	0.41
Intercalary	0.48	0.59	0.34	0.49	0.56	0.47
Mixed	0.48	0.60	0.42	0.58	0.55	0.63
Unspecified	---	---	---	---	---	---
Diaphragmatic hernia	0.73	1.31	1.73	2.13	3.43	3.89
Omphalocele	1.10	1.93	2.32	2.51	3.17	3.40
Gastroschisis	0.09	0.40	0.65	1.18	2.64	3.26
Unspecified Omphalocele / Gastroschisis	---	---	---	---	---	---
Prune belly sequence	0.02	0.54	0.74	0.74	1.18	1.03
Trisomy 13	0.18	0.50	0.43	0.55	0.85	0.89
Trisomy 18	0.23	0.71	0.99	0.98	1.78	2.02
Down syndrome, all ages (include age unknown)	14.63	14.78	14.94	15.84	18.00	19.75
<20	7.68	7.10	6.76	6.83	8.37	6.85
20-24	7.09	7.33	6.30	7.97	8.69	10.15
25-29	7.60	8.30	6.89	8.72	9.40	10.33
30-34	13.66	15.81	15.47	15.48	16.87	16.86
35-39	56.45	44.24	44.49	47.05	50.39	57.76
40-44	161.18	141.15	152.52	148.49	179.36	177.78
45+	269.28	283.57	270.52	244.78	361.34	374.62
unspecified	---	---	---	---	---	---

* data include less than 5 years

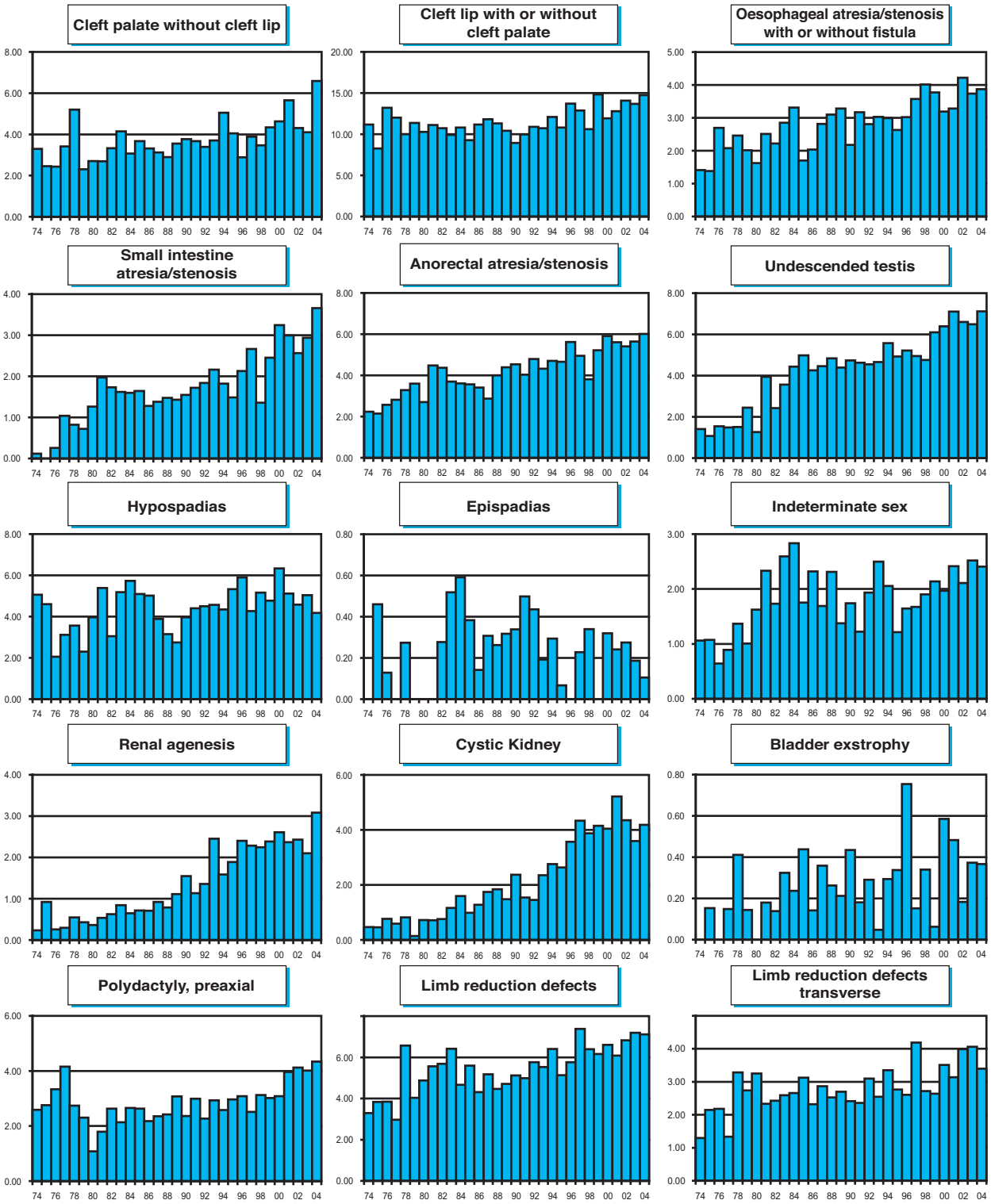
nr = not reported

South America: ECLAMC

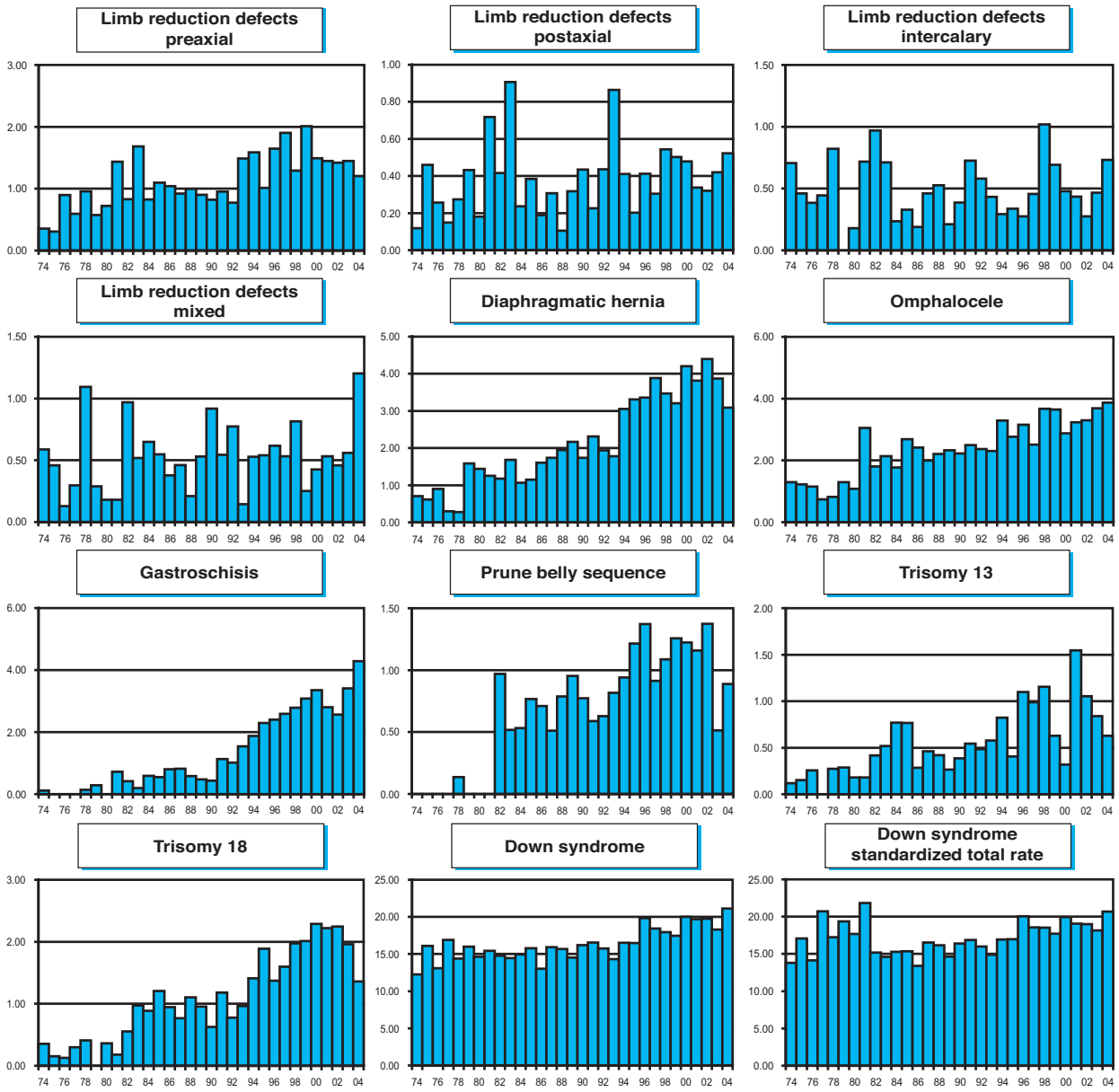
'Time trends 1974-2004 (Birth prevalence rates per 10,000)



Note: L+S rates



Note: L+S rates



Note: L+S rates

Spain: ECEMC

Spanish Collaborative Study of Congenital Malformations (ECEMC)

History:
The programme was created in 1976 by Prof. Dr. María Luisa Martínez-Frías, as a hospital-based case-control study and surveillance system. It became a full member of the ICBDSR in 1979. In January 2002 the ECEMC Programme became integrated into the CIAC (Research Center on Congenital Anomalies), of the Instituto de Salud Carlos III (ISCIII) from the Ministerio de Sanidad y Consumo of Spain, and is also directed by Prof. Martínez-Frías. Activity of the CIAC is coordinated in agreement with the IIER (Institute of Research on Rare Diseases), of the ISCIII too. The ECEMC has 2 Teratogen Information Services since 1991, one for the general population and another one for physicians.

Size and coverage:
Reports are obtained from hospitals distributed all over Spain. The annual number of births surpasses 100,000, representing near 25% of all Spanish births. Stillbirths of at least 24 weeks or 500 g. have been included since 1980.

Legislation and funding:
It is a research programme with voluntary participation of hospitals, and is financed mainly by the Spanish Administration and, partially, by non-governmental organisations.

Sources of ascertainment:
The detection period is the first 3 days of life, including major and/or minor/mild defects. Reports come from delivery units and paediatric departments of the participating hospitals. Mothers are interviewed directly to fill in the ECEMC standard protocols, which include more than 300 data for each child (family history, demographic and obstetrical data, prenatal exposures, etc), whether case or control. Controls are defined as the next non-malformed infant born at the same hospital that the case with the same sex as the malformed infant. In many instances, photographs, imaging studies, high-resolution bands karyotypes and molecular analysis when needed (which are performed at the central group of the ECEMC), and other complementary studies are available.

Exposure information:
The mother of each reported infant (case or control) is interviewed within the first three days after delivery on various exposures (parental occupation, maternal acute or chronic diseases, drug usage, exposure to other chemical or physical factors).

Background information:
Total number of births by sex and number of twin pairs in each participating hospital are gathered. Other background information is obtained from the control material.

Addresses and Staff:
Prof. María-Luisa Martínez-Frías, Programme Director ECEMC, Centro de Investigación sobre Anomalías Congénitas (CIAC) Instituto de Salud Carlos III, C/Sinesio Delgado nº 6, Pabellón 6, 28029-Madrid (Spain)

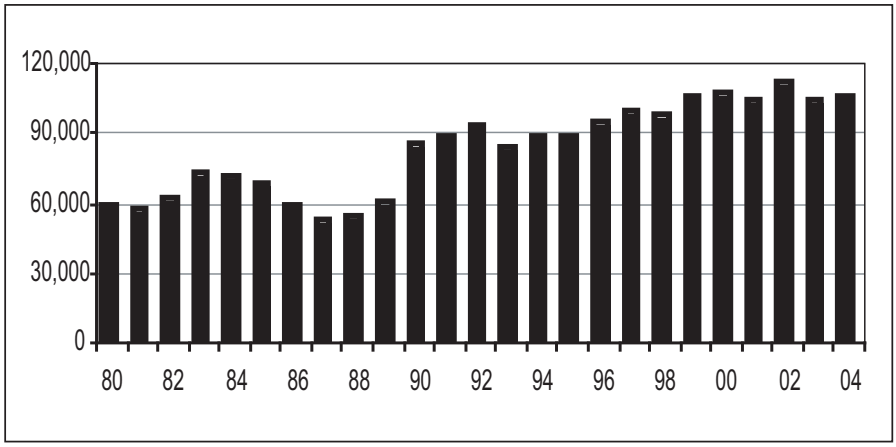
Phone: 34-91-3877538

Fax: 34-91-3877541

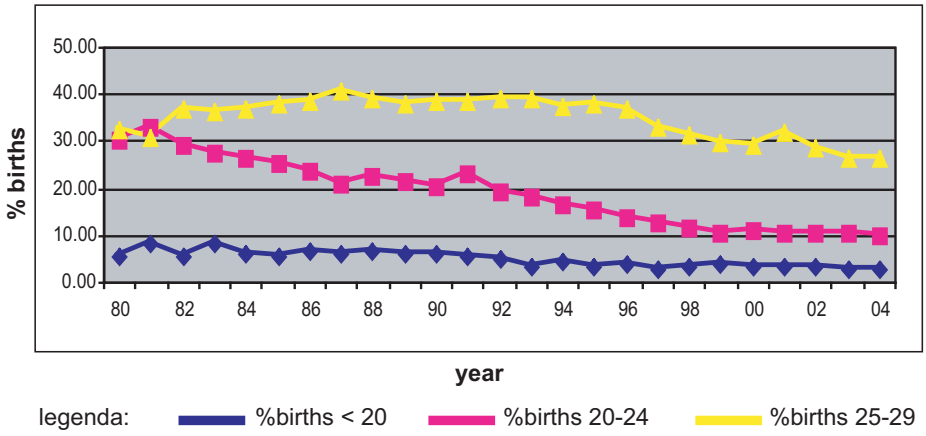
E-mail: mlmartinez.frias@isciii.es

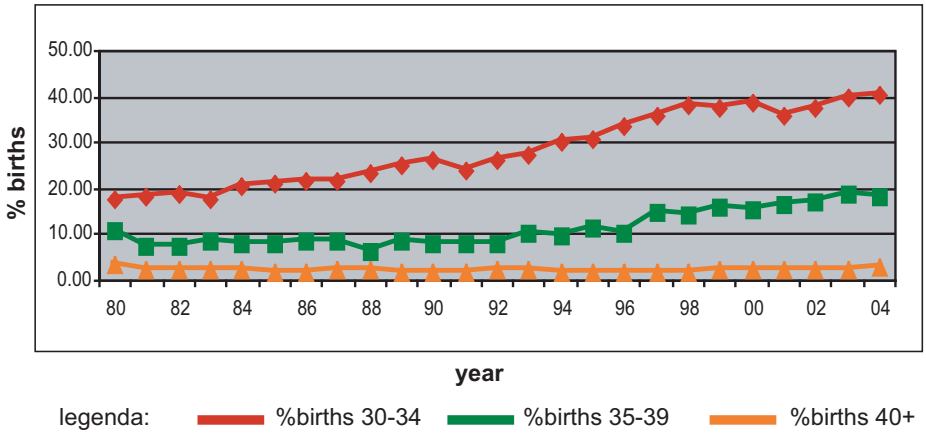
Spain: ECEMC

Total births by year



Percentage of births by maternal age





Spain: ECEMC, 2004

Live births (LB)	103,579
Stillbirths (SB)	430
Total births	104,009
Number of terminations of pregnancy (ToP) for birth defects	nr

Birth Defects	Number of cases			Rates*10,000
	LB	SB	ToP	Total rate
Anencephaly	2	0	nr	0.19
Spina bifida	12	1	nr	1.25
Encephalocele	3	0	nr	0.29
Microcephaly	6	0	nr	0.58
Arhinencephaly / Holoprosencephaly	3	0	nr	0.29
Hydrocephaly	18	0	nr	1.73
Anophthalmos	1	1	nr	0.19
Microphthalmos	6	0	nr	0.58
Unspecified Anophthalmos/ Microphthalmos	0	0	nr	---
Anotia	1	0	nr	0.10
Microtia	15	0	nr	1.44
Unspecified Anotia/Microtia	0	0	nr	---
Transposition of great vessels	24	1	nr	2.40
Tetralogy of Fallot	8	0	nr	0.77
Hypoplastic left heart syndrome	2	0	nr	0.19
Coarctation of aorta	10	0	nr	0.96
Choanal atresia, bilateral	2	0	nr	0.19
Cleft palate without cleft lip	48	0	nr	4.61
Cleft lip with or without cleft palate	48	1	nr	4.71
Oesophageal atresia / stenosis with or without fistula	21	1	nr	2.12
Small intestine atresia / stenosis	6	0	nr	0.58
Anorectal atresia / stenosis	21	2	nr	2.21
Undescended testis (36 weeks of gestation or later)	24	0	nr	2.31
Hypospadias	24	0	nr	2.31
Epispadias	0	0	nr	0.00
Indeterminate sex	4	0	nr	0.38
Renal agenesis	1	0	nr	0.10
Cystic kidney	18	0	nr	1.73
Bladder exstrophy	3	0	nr	0.29
Polydactyly, preaxial	20	0	nr	1.92
Total Limb reduction defects (include unspecified)	50	2	nr	5.00
Transverse	22	1	nr	2.21
Preaxial	6	1	nr	0.67
Postaxial	2	0	nr	0.19
Intercalary	3	0	nr	0.29
Mixed	9	0	nr	0.87
Unspecified	0	0	nr	---
Diaphragmatic hernia	6	0	nr	0.58
Omphalocele	3	0	nr	0.29
Gastroschisis	5	0	nr	0.48
Unspecified Omphalocele/Gastroschisis	0	0	nr	---
Prune belly sequence	2	1	nr	0.29
Trisomy 13	4	0	nr	0.38
Trisomy 18	4	0	nr	0.38
Down syndrome, all ages (include age unknown)	74	0	nr	7.11
<20	0	0	nr	0.00
20-24	5	0	nr	4.96
25-29	13	0	nr	4.76
30-34	27	0	nr	6.41
35-39	18	0	nr	9.58
40-44	9	0	nr	37.14
45+	0	0	nr	0.00
unknown	0	0	nr	---

nr = not reported

Spain: ECEMC, Previous years rates 1980 - 2004

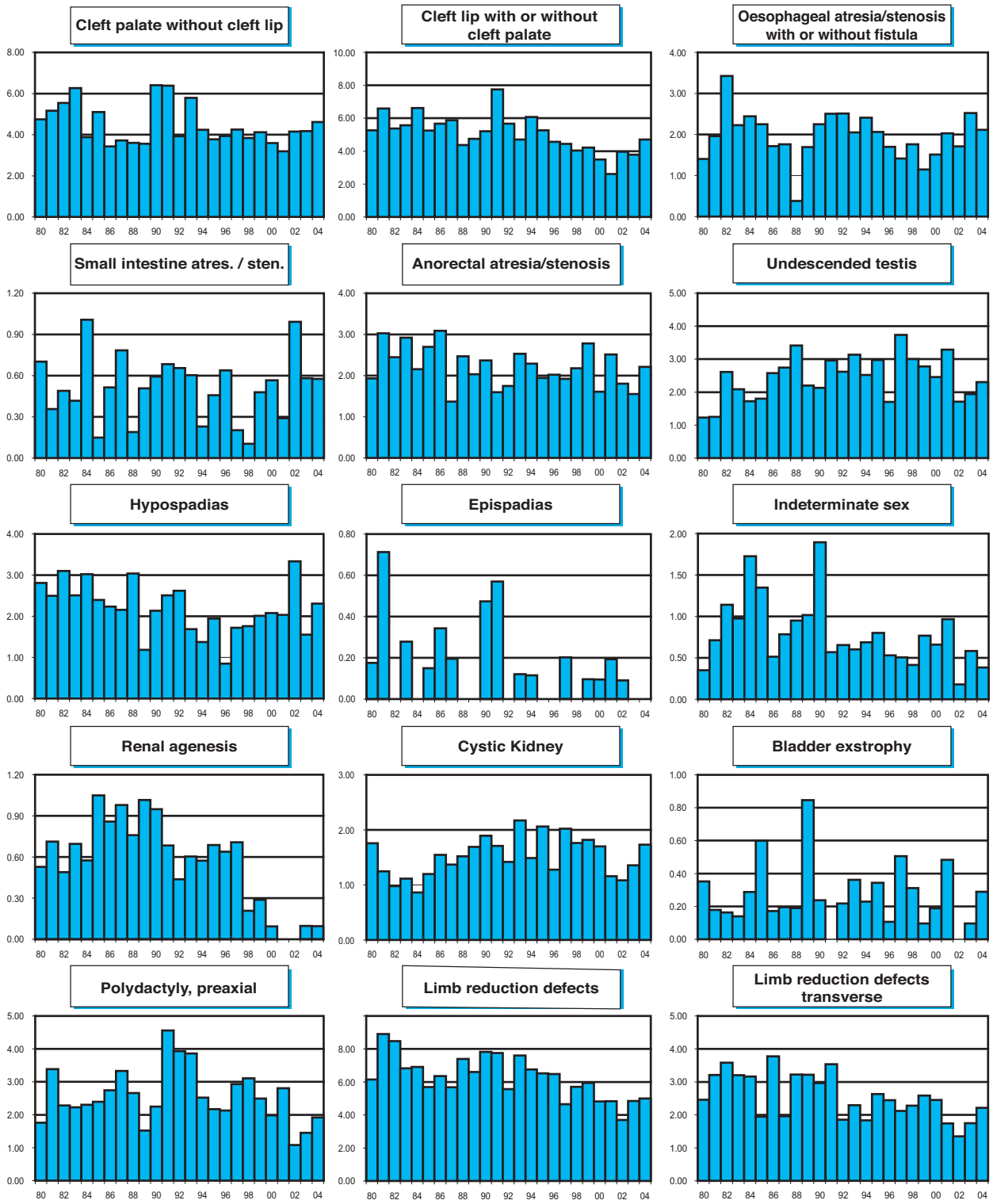
Birth prevalence rates: (LB+SB) * 10,000

	1974-79	1980-84	1985-89	1990-94	1995-99	2000-04
Births		**315,670**	**287,748**	**433,858**	**481,187**	**527,289**
Anencephaly		4.44	3.16	1.04	0.64	0.15
Spina bifida		4.44	4.66	3.62	2.24	1.29
Encephalocele		1.14	0.63	0.97	0.29	0.15
Microcephaly		2.15	1.91	2.35	1.72	1.02
Arhinencephaly / Holoprosencephaly		0.54	0.49	0.53	0.50	0.25
Hydrocephaly		2.47	2.81	2.86	2.49	1.82
Anophthalmos		0.79	0.38	0.25	0.21	0.09
Microphthalmos		2.03	1.74	1.75	1.35	0.83
Unspecified Anophthalmos / Microphthalmos		----	----	----	----	----
Anotia		0.00	0.03	0.16	0.15	0.09
Microtia		2.03	1.84	1.43	1.41	1.56
Unspecified Anotia / Microtia		----	----	----	----	----
Transposition of great vessels		0.70	0.90	1.36	1.43	1.29
Tetralogy of Fallot		0.19	0.42	1.11	1.16	0.95
Hypoplastic left heart syndrome		0.41	0.59	1.06	0.83	0.25
Coarctation of aorta		0.38	0.31	0.78	1.00	0.66
Choanal atresia, bilateral		0.13	0.35	0.28	0.21	0.13
Cleft palate without cleft lip		5.13	3.93	5.32	3.99	3.94
Cleft lip with or without cleft palate		5.89	5.18	5.90	4.49	3.72
Oesophageal atresia / stenosis with or without fistula		2.31	1.60	2.35	1.60	1.97
Small intestine atresia / stenosis		0.60	0.42	0.55	0.37	0.61
Anorectal atresia / stenosis		2.50	2.36	2.10	2.18	1.93
Undescended testis (36 weeks of gestation or later)		1.81	2.50	2.67	2.85	2.33
Hypospadias		2.79	2.19	2.07	1.66	2.28
Epispadias		0.22	0.14	0.25	0.06	0.08
Indeterminate sex		1.01	0.94	0.88	0.60	0.55
Renal agenesis		0.60	0.94	0.65	0.50	0.06
Cystic kidney		1.17	1.46	1.73	1.79	1.40
Bladder exstrophy		0.22	0.42	0.21	0.27	0.21
Polydactyly, preaxial		2.38	2.50	3.43	2.58	1.84
Total Limb reduction defects (include unspecified)		7.41	6.32	7.08	5.84	4.63
Transverse		3.14	2.81	2.49	2.41	1.90
Preaxial		1.17	1.04	0.92	0.73	0.59
Postaxial		0.13	0.17	0.18	0.25	0.11
Intercalary		0.57	0.24	0.65	0.15	0.34
Mixed		1.20	0.87	1.24	1.10	0.85
Unspecified		----	----	----	----	----
Diaphragmatic hernia		2.76	2.12	2.24	1.54	0.63
Omphalocele		1.81	1.32	1.11	0.91	0.53
Gastroschisis		0.60	0.38	0.35	0.46	0.36
Unspecified Omphalocele / Gastroschisis		----	----	----	----	----
Prune belly sequence		0.57	0.52	0.51	0.31	0.15
Trisomy 13		0.38	0.45	0.46	0.52	0.34
Trisomy 18		0.89	0.97	1.08	0.69	0.63
Down syndrome, all ages (include age unknown)		14.41	15.22	12.70	10.85	7.97
<20		7.35	8.07	8.57	1.81	1.79
20-24		6.98	6.37	4.50	5.09	5.07
25-29		6.10	8.18	7.74	5.78	5.01
30-34		9.77	15.09	14.26	10.92	6.93
35-39		46.67	41.61	35.92	23.77	12.68
40-44		147.44	186.36	64.92	51.94	45.00
45+		233.13	141.70	248.45	531.91	42.64
unknown		----	----	----	----	----

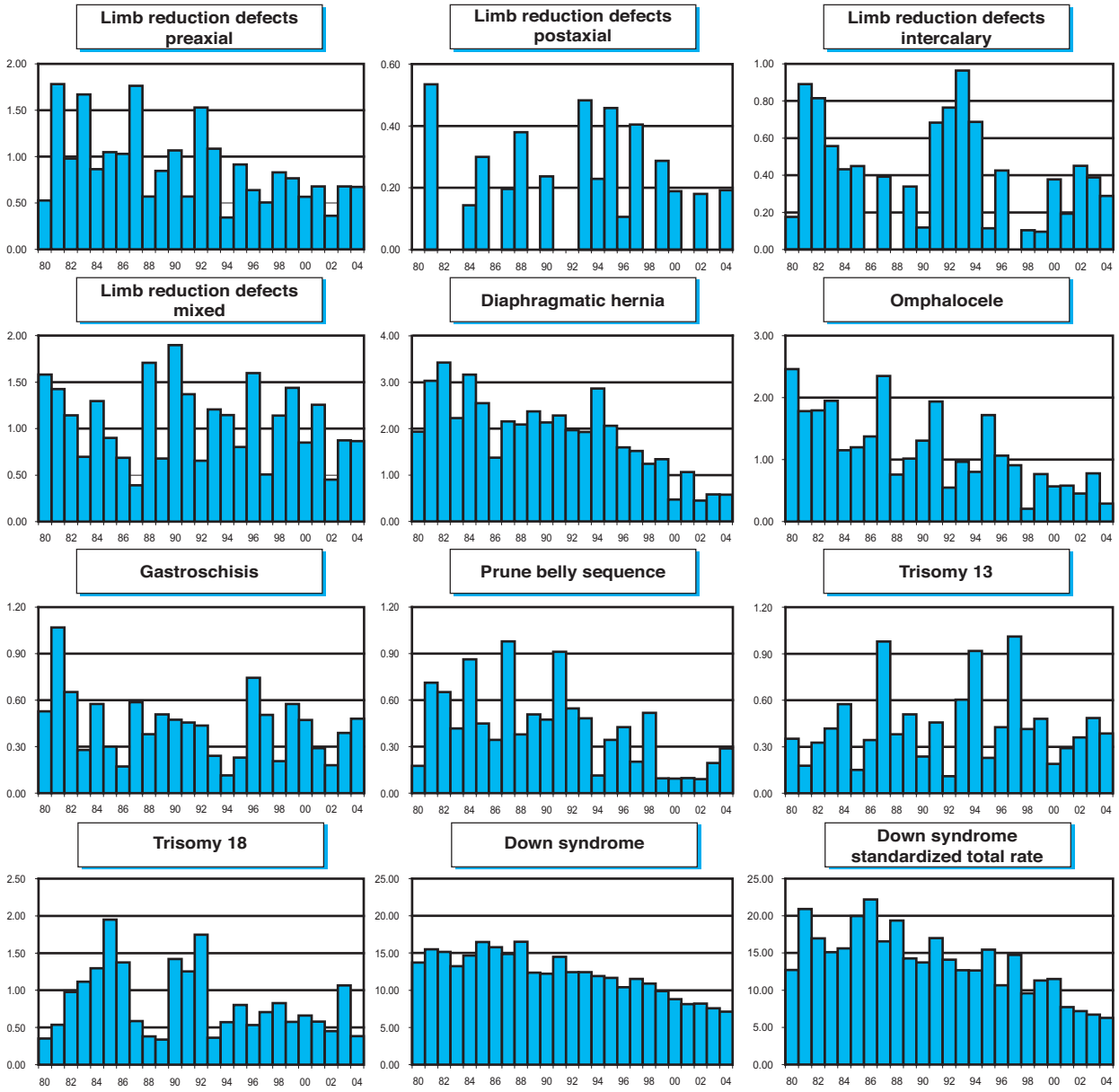
Spain: ECEMC

'Time trends 1980-2004 (Birth prevalence rates per 10,000)

Note: L+S rates



Note: L+S rates



Note: L+S rates

Sweden
The Swedish Birth Defects Registry

History:
The Registry of Congenital Malformations started in 1964 and the Medical Birth Registry in 1973. The Programme was a founding member of the ICBDSR and contributed with data until 1994. The registry has a new regime from 1999 and is since then again a full member of the ICBDSR.

Size and coverage:
All births in Sweden are included, approximately 100,000-120,000 annual births. The definition of stillbirth in Sweden is more than 28 weeks. Since 1999 all fetal deaths with congenital malformations more than 22 weeks are reported to the Swedish Registry of Congenital Malformations. In 1999 a special fetal congenital anomalies surveillance system was started to include those fetuses with congenital malformations who were terminated as a result of prenatal diagnosis.

Legislation and funding:
Reporting is compulsory for children with malformations, but not for terminated pregnancies with fetuses with congenital malformations.

Sources of ascertainment:
Reports are received from delivery units, paediatric clinics, pathology departments, child cardiology clinics, and cytogenetic laboratories.

Exposure information:
Some exposure information for all births is available in the Medical Birth Registry; maternal occupation, socio-economic factors, maternal smoking, drug use during pregnancy, contraceptive usage, maternal diseases.

Background information:
Epidemiological background data are available on all birth in the Medical Birth Registry.

Addresses and Staff:
Goran Anneren, Department of Clinical Genetics, Uppsala University Children's Hospital S-75185 UPPSALA, Sweden

Phone: +46-18-665942

Fax: +46-18-554025

E-mail: goran.anneren@akademiska.se

Birgitta Ollars, Department of Epidemiology, National Board of Health and Welfare, SE-106 30 Stockholm, Sweden.

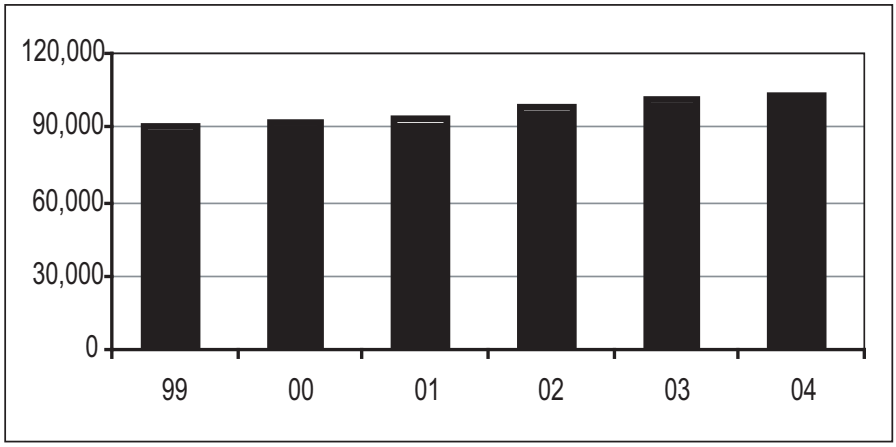
Phone: +46-8 55553123

Fax: +46-8-55553327

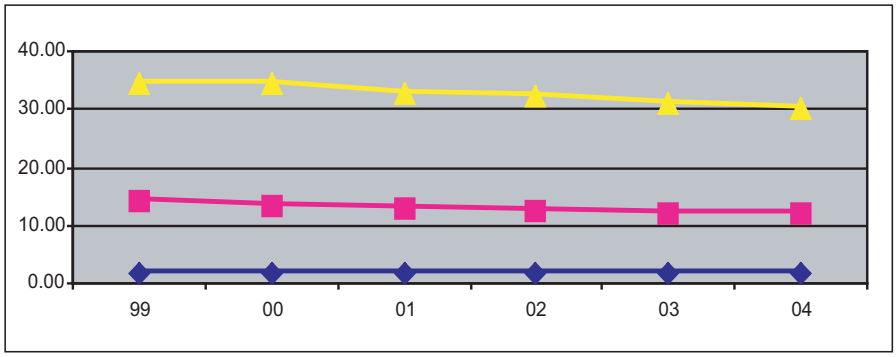
E-mail: birgitta.ollars@socialstyrelsen.se

Sweden

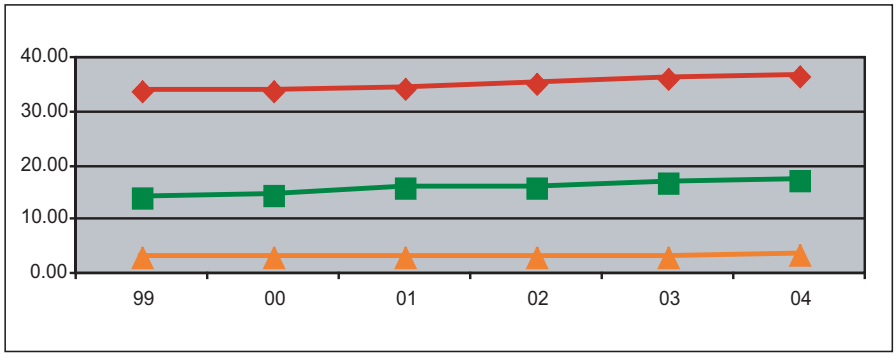
Total births by year



Percentage of births by maternal age



legenda: %births < 20 %births 20-24 %births 25-29



legenda: %births 30-34 %births 35-39 %births 40+

Sweden: 2004

Live births (LB)	100,928
Stillbirths (SB)	333
Total births	101,261
Number of terminations of pregnancy (ToP) for birth defects	455

Birth Defects	Number of cases			Rates*10,000
	LB	SB	ToP	Total rate
Anencephaly	2	0	32	3.34
Spina bifida	16	0	17	3.24
Encephalocele	1	0	13	1.38
Microcephaly	0	0	0	0.00
Arhinencephaly / Holoprosencephaly	3	0	6	0.88
Hydrocephaly	12	0	32	4.33
Anophthalmos	2	0	0	0.20
Microphthalmos	4	0	2	0.59
Unspecified Anophthalmos / Microphthalmos	0	0	0	---
Anotia	8	1	0	0.88
Microtia	2	0	0	0.20
Unspecified Anotia / Microtia	0	0	0	---
Transposition of great vessels	32	0	5	3.64
Tetralogy of Fallot	26	0	2	2.75
Hypoplastic left heart syndrome	14	0	11	2.46
Coarctation of aorta	43	0	3	4.52
Choanal atresia, bilateral	7	0	1	0.79
Cleft palate without cleft lip	45	2	4	5.01
Cleft lip with or without cleft palate	98	0	5	10.13
Oesophageal atresia / stenosis with or without fistula	27	0	2	2.85
Small intestine atresia / stenosis	33	0	2	3.44
Anorectal atresia / stenosis	29	1	7	3.64
Undescended testis (36 weeks of gestation or later)	nr	nr	nr	nr
Hypospadias	196	0	2	19.47
Epispadias	1	0	0	0.10
Indeterminate sex	1	0	1	0.20
Renal agenesis	3	0	8	1.08
Cystic kidney	20	0	15	3.44
Bladder exstrophy	3	0	1	0.39
Polydactyly, preaxial	53	0	6	5.80
Total Limb reduction defects (include unspecified)	44	0	9	5.21
Transverse	37	0	6	4.23
Preaxial	0	0	3	0.29
Postaxial	1	0	0	0.10
Intercalary	2	0	0	0.20
Mixed	4	0	0	0.39
Unspecified	0	0	0	---
Diaphragmatic hernia	21	0	7	2.75
Omphalocele	17	1	18	3.54
Gastroschisis	9	0	3	1.18
Unspecified Omphalocele / Gastroschisis	0	0	0	---
Prune belly sequence	0	0	2	0.20
Trisomy 13	12	0	20	3.15
Trisomy 18	10	0	54	6.29
Down syndrome, all ages (include age unknown)	106	1	138	24.09
<20	0	0	1	6.22
20-24	13	0	3	13.31
25-29	6	0	12	5.93
30-34	34	0	21	14.92
35-39	39	1	64	60.51
40-44	12	0	33	139.41
45+	2	0	4	555.56
unknown	0	0	0	---

nr = not reported

Sweden: Previous years rates 1999 - 2004

Prevalence rates: (LB+SB+TOP) * 10,000

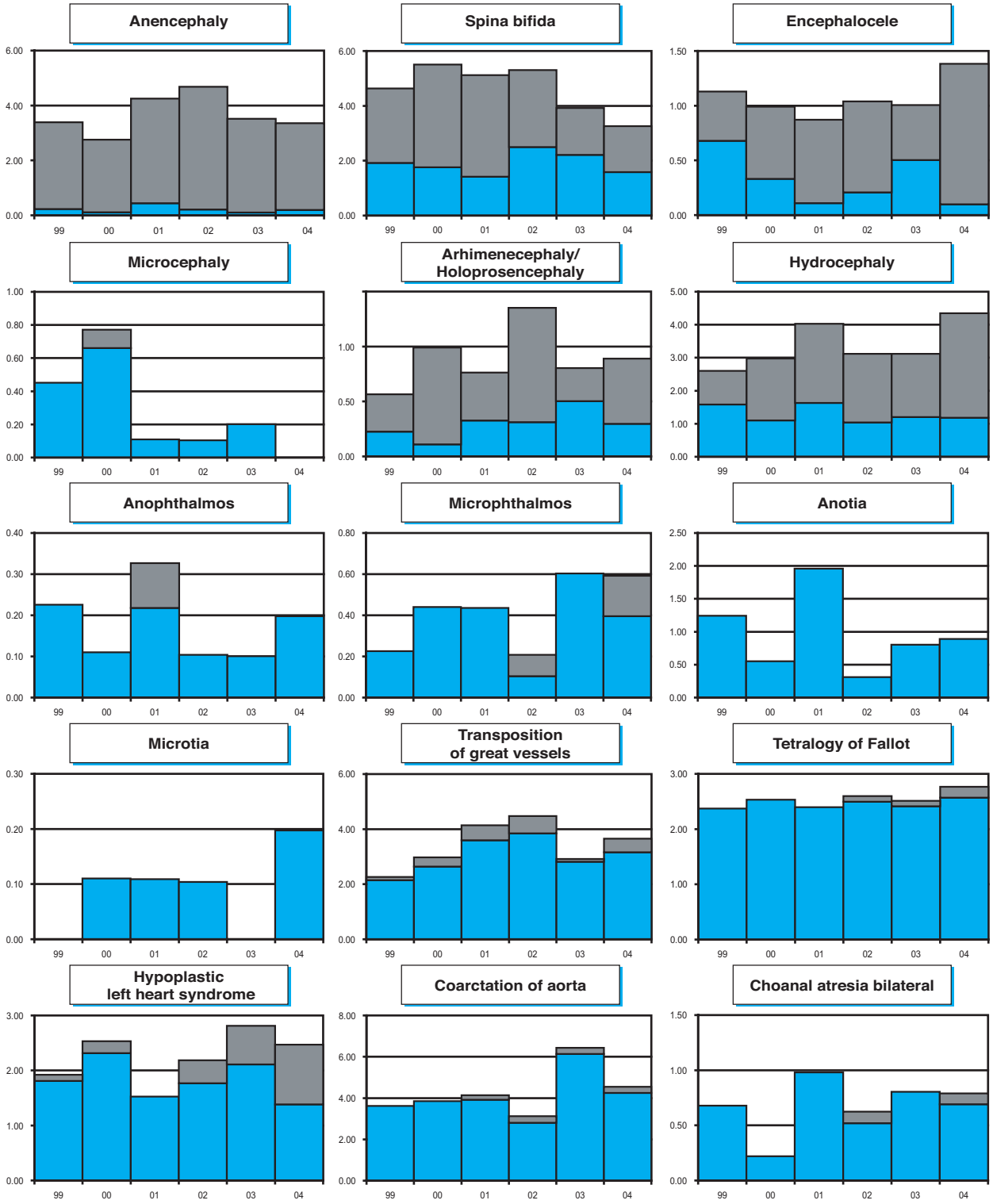
	1974-79	1980-84	1985-89	1990-94	1995-99*	2000-04
Births					**88,512**	**479,555**
Anencephaly					3.39	3.71
Spina bifida					4.63	4.59
Encephalocele					1.13	1.06
Microcephaly					0.45	0.23
Arhinencephaly / Holoprosencephaly					0.56	0.96
Hydrocephaly					2.60	3.52
Anophthalmos					0.23	0.17
Microphthalmos					0.23	0.46
Unspecified Anophthalmos / Microphthalmos					---	---
Anotia					1.24	0.90
Microtia					0.00	0.10
Unspecified Anotia / Microtia					---	---
Transposition of great vessels					2.26	3.63
Tetralogy of Fallot					2.37	2.56
Hypoplastic left heart syndrome					1.92	2.31
Coarctation of aorta					3.62	4.44
Choanal atresia, bilateral					0.68	0.69
Cleft palate without cleft lip					6.33	5.21
Cleft lip with or without cleft palate					11.64	9.78
Oesophageal atresia / stenosis with or without fistula					1.69	2.63
Small intestine atresia / stenosis					2.49	2.46
Anorectal atresia / stenosis					3.50	3.04
Undescended testis (36 weeks of gestation or later)					nr	nr
Hypospadias					20.79	20.35
Epispadias					0.11	0.21
Indeterminate sex					0.11	0.27
Renal agenesis					2.49	1.67
Cystic kidney					2.94	3.11
Bladder exstrophy					0.34	0.25
Polydactyly, preaxial					3.62	4.69
Total Limb reduction defects (include unspecified)					4.63	5.17
Transverse					2.71	3.86
Preaxial					0.23	0.25
Postaxial					0.11	0.19
Intercalary					0.11	0.23
Mixed					1.47	0.65
Unspecified					---	---
Diaphragmatic hernia					3.05	2.69
Omphalocele					2.26	2.61
Gastroschisis					2.37	1.77
Unspecified Omphalocele / Gastroschisis					---	---
Prune belly sequence					0.11	0.10
Trisomy 13					2.03	2.31
Trisomy 18					5.87	6.05
Down syndrome, all ages (include age unknown)					22.48	24.48
<20					5.97	9.62
20-24					7.22	9.32
25-29					6.26	9.12
30-34					15.57	16.89
35-39					58.93	55.36
40-44					143.39	170.42
45+					224.72	466.02
unspecified					---	---

* data include less than 5 years

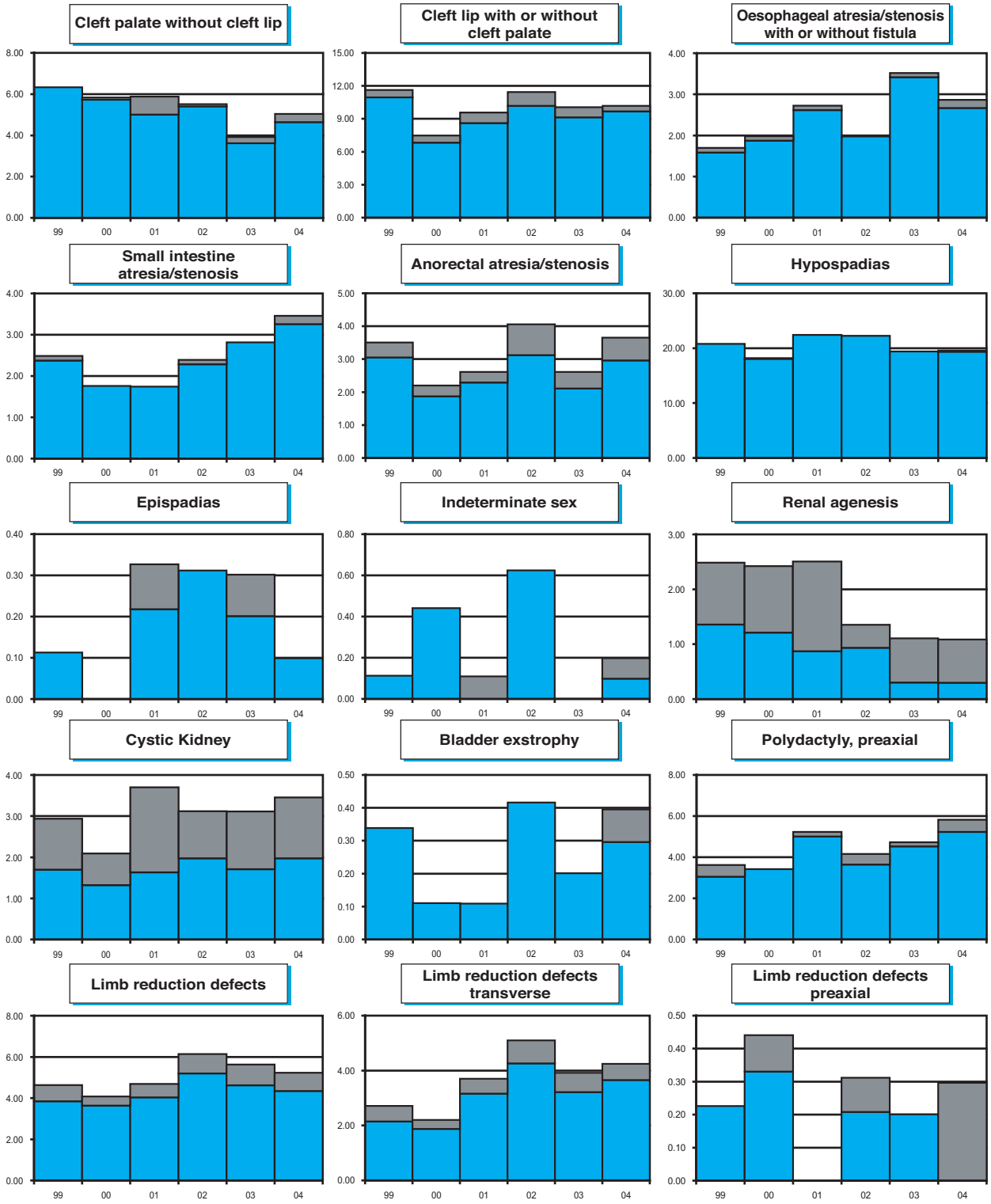
nr = not reported

Sweden

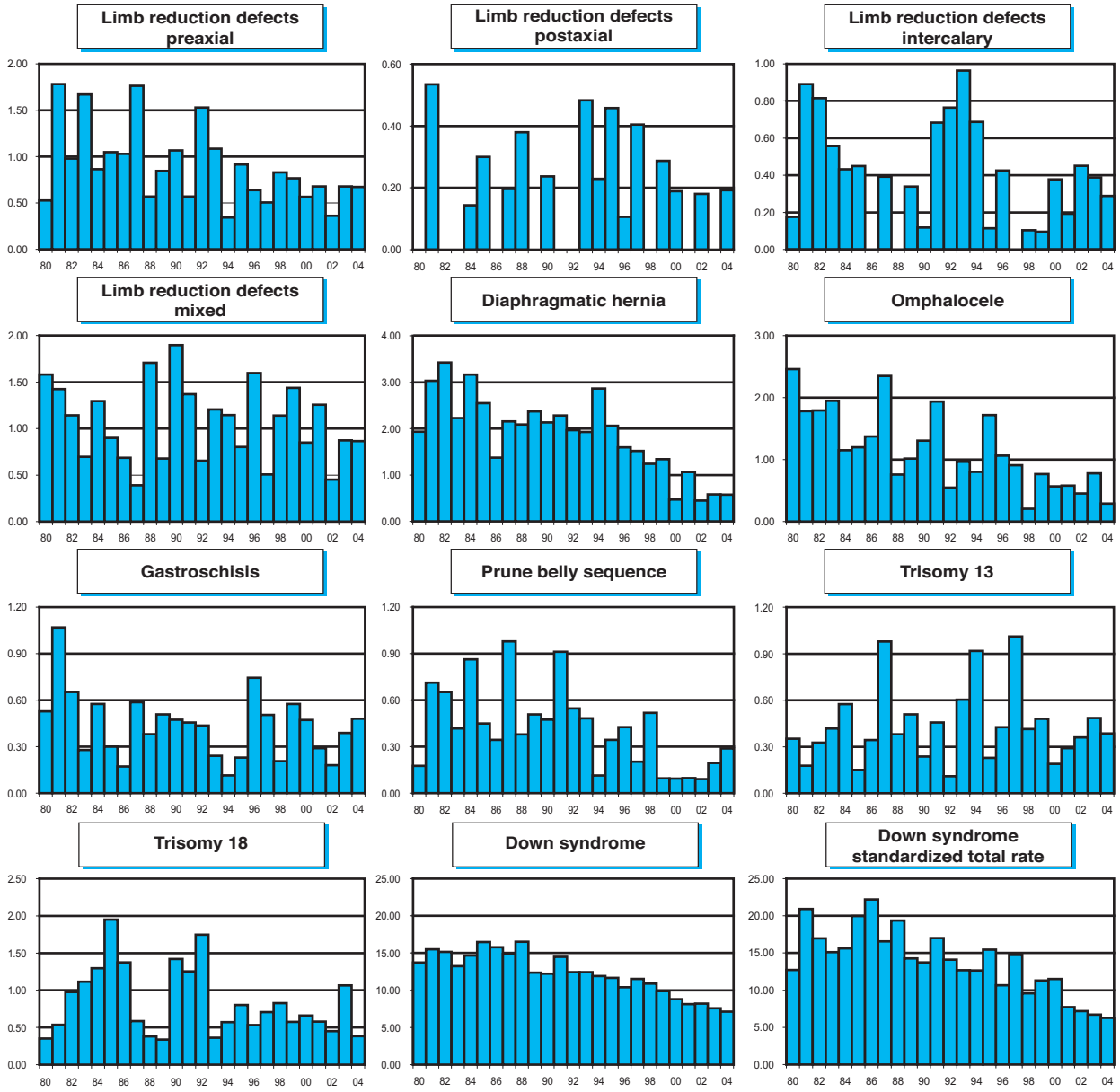
'Time trends 1999-2004 (Birth prevalence rates per 10,000)



Note: L+S rates, ToP rates



Note: L+S rates, ToP rates



Note: L+S rates, ToP rates

Ukraine: UABDP

Ukrainian-American Birth Defects Program

History:
In 1998, two birth defect (BD) teams, composed of neonatologists, medical geneticists and information officers were initiated. In 1999, two BD centers were established, one in the city of Lutsk to serve the Volyn State (Oblast) and in Rivne to serve the Rivne Oblast. Population based malformation monitoring was initiated in 2000 - about 20,000 births in both oblasts. In 2001, training of their BD teams was initiated and BD information centers were established (Kherson, Khmelniski and Crimea). Beyond monitoring, each BD team is also charged to enhance coordination of care, address recurrence prevention, supervise data coding and management by vital statistics partners and to nurture a Regional BD Information Center for health care providers, families and the public. In 1999, a Ukrainian Alliance for the Prevention of BD was formed to link patients, families and community organizations with health care specialists, academic and other state organizations. In 2001, an "OMNI" information and training center was established in Kyiv. The "OMNI" center is dedicated to promoting normal child development and the prevention of birth defects and offers its resources to health care providers, Alliance members as well as academic and other organizations. In 2001, the Programme became member of the World Alliance of Organizations for the Prevention of BD and an associate member of the ICBDSR.

Legislation and funding:
The planning of the Programme was funded by donations, mostly by USA and Canadian citizens. In 1988, the March of Dimes provided partial support and a model for the formation of the Ukrainian Alliance for the Prevention of Birth Defects. The Center for Disease Control Birth Defects Program, Greenwood Foundation, California BD Monitoring System, OMIM, European Alliance of Genetic Support Groups and the University of South Alabama provided resources for the OMNI Center. Training of Ukrainian BD teams and the establishment of BD centers in various oblasts were made possible by a cooperative agreement with and funds provided by the United States Agency of International Development (USAID). The resources listed are amply matched by the Ukrainian Ministry of Health, Hygiene Institute, Ukrainian Academy of Medicine and Mohyla Academy University who contribute facilities, dedicated personnel and other resources. Furthermore, the Legislative power enacted laws and the Ministry of Health issued decrees making BD reportable. Oblast BD data is submitted to the Oblast Vital Statistics Centers which forward aggregate data to the Ministry of Health. In addition, BD data devoid of identifiers is examined by Ukrainian and International experts to facilitate applications of international standards.

Sources of ascertainment:
Malformation monitoring is active - every neonatologist required to report every BD occurrence. Field trips to every birth site are made by BD team neonatologists who review compliance and search for additional data from obstetric, pediatric and other sources. Most patients are referred to centralized facilities for specialty care, which routinely calls for services by medical geneticists who also are members of BD teams. In addition, medical geneticists review death certificates and medical records generated by other specialists including family planning.

Exposure information:
Routine exposure of information collection is minimal except when ad hoc circumstances are noted. Recommendations for systematic collection of exposure data are under consideration.

Background information:
In terms of area and population size, Ukraine is similar to France. Natality in Ukraine has plummeted by at least 38% (in 1977-80 it stood at 15.5 birth per 1000 population compared to 9.7 in 1933-1994). The Chornobyl nuclear plant disaster contaminated 15% of the territory. In 1996, there were 45 physicians, 115 hospital beds per 10,000 population, 467,211 births, 664,156 induced abortions, 13% of children born to unwed mothers and 5.4% births under 2500 g. The rate of induced abortions has dropped from 83 per 10,000 women (1990) to 52 (1996) but remains nearly three folds greater than in Canada, France or United States. The overall Infant Mortality (under one year) for 1996 was 143.3 per 10,000 live births, of which 50.7 was attributable to "perinatal conditions" and 40.5 to "congenital defects", followed by "infectious and parasitic diseases" (11.6 each). Furthermore, "congenital anomalies" in Ukraine contribute nearly as much to child mortality (from birth to 14 years) as do "accidents, traumas and poisonings" (2.4 vs. 2.7 per 10,000 children in 1996, followed by "diseases of respiratory organs and nervous system" which contributed 0.9 each). The impact of

exposure to chronic low dose radiation related to the Chornobyl accident on child development remains undefined except for an epidemic of pediatric thyroid cancer. However, exposure of the unborn to alcohol, rubella, and a diet poor in foliates, iodine, iron and other micronutrients call for vigorous advocacy for birth defects prevention interventions due to these factors. Fortification of flour with folic acid is proposed to reduce the significant burden imposed by spina bifida, a frequent birth defect in regions under monitoring. Among other prevention interventions being advanced is a rubella immunization Programme to reduce rubella induced deafness, and avoidance of alcohol before and during pregnancy.

Addresses and Staff:
Wladimir Wertelecki, Programme International Coordinator Lyubov Yevtuskok: Programme Medical Coordinator

Phone: 251-460-7500

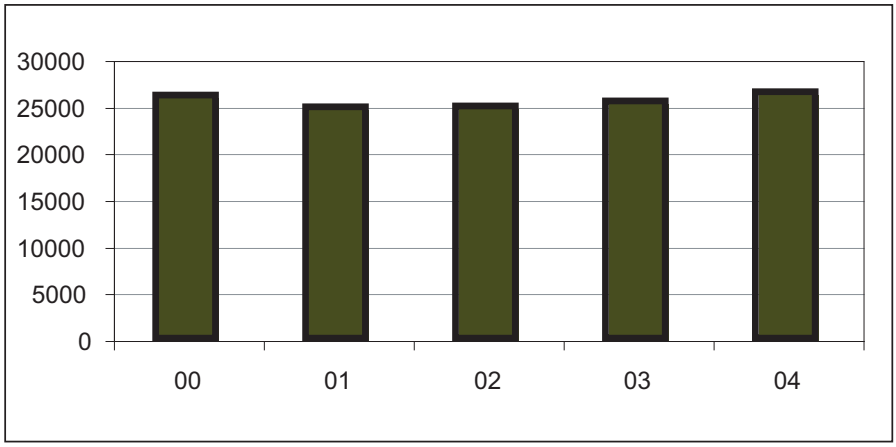
Fax: 251-461-1591

E-mail: genfir3@aol.com

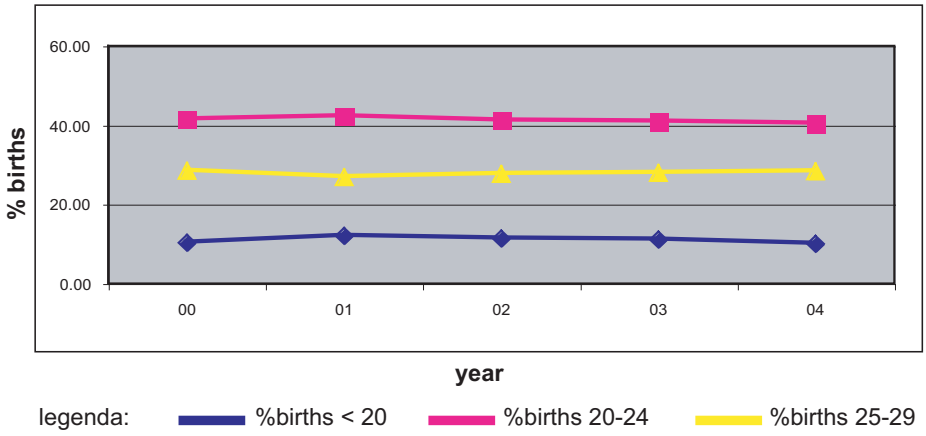
E-mail: wwertele@usamail.usouthal.edu
lyevtushok@bdp.rovno.ua

Ukraine: UABDP

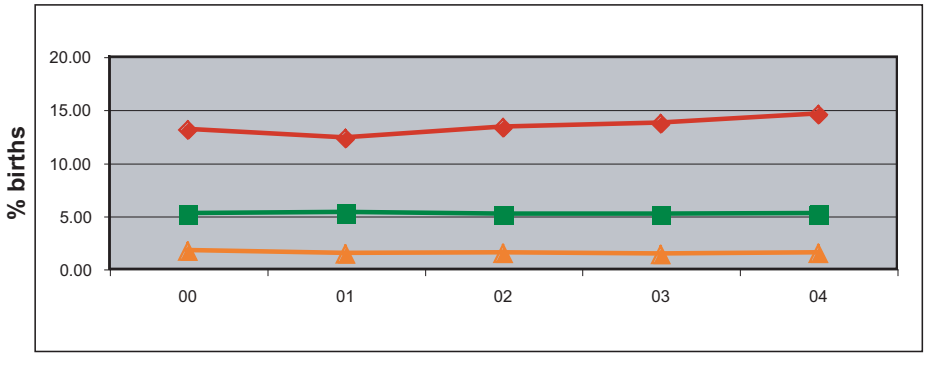
Total births by year



Percentage of births by maternal age



legenda: %births < 20 %births 20-24 %births 25-29



legenda: %births 30-34 %births 35-39 %births 40+

Ukraine: UABDP, 2004

Live births (LB)	26,315
Stillbirths (SB)	95
Total births	26,410
Number of terminations of pregnancy (ToP) for birth defects	nr

Birth Defects	Number of cases			Rates*10,000
	LB	SB	ToP	Total rate
Anencephaly	0	2	21	8.71
Spina bifida	16	2	18	13.63
Encephalocele	1	0	5	2.27
Microcephaly (1)	8	0	nr	3.03
Arhinencephaly / Holoprosencephaly	3	0	nr	1.14
Hydrocephaly	9	2	nr	4.17
Anophthalmos (1)	0	0	nr	0.00
Microphthalmos (1)	2	0	nr	0.76
Unspecified Anophthalmos/ Microphthalmos	0	0	nr	---
Anotia	0	0	nr	0.00
Microtia	3	0	nr	1.14
Unspecified Anotia/Microtia	0	0	nr	---
Transposition of great vessels	8	0	nr	3.03
Tetralogy of Fallot	4	0	nr	1.51
Hypoplastic left heart syndrome	5	0	nr	1.89
Coarctation of aorta	5	0	nr	1.89
Choanal atresia, bilateral	0	0	nr	0.00
Cleft palate without cleft lip	13	0	nr	4.92
Cleft lip with or without cleft palate	16	2	nr	6.82
Oesophageal atresia / stenosis with or without fistula	5	0	nr	1.89
Small intestine atresia / stenosis	4	0	nr	1.51
Anorectal atresia / stenosis	7	1	nr	3.03
Undescended testis (36 weeks of gestation or later)	98	0	nr	37.11
Hypospadias (2)	5	0	nr	1.89
Epispadias	0	0	nr	0.00
Indeterminate sex	0	0	nr	0.00
Renal agenesis	1	0	nr	0.38
Cystic kidney	8	0	nr	3.03
Bladder exstrophy	2	0	nr	0.76
Polydactyly, preaxial	5	0	nr	1.89
Total Limb reduction defects (include unspecified)	11	0	nr	4.17
Transverse	8	0	nr	3.03
Preaxial	1	0	nr	0.38
Postaxial	0	0	nr	0.00
Intercalary	1	0	nr	0.38
Mixed	1	0	nr	0.38
Unspecified	0	0	nr	---
Diaphragmatic hernia	5	2	nr	2.65
Omphalocele	3	0	nr	1.14
Gastroschisis	2	0	nr	0.76
Unspecified Omphalocele/Gastroschisis	0	0	nr	---
Prune belly sequence	0	0	nr	0.00
Trisomy 13 (1)	0	0	nr	0.00
Trisomy 18 (1)	1	0	nr	0.38
Down syndrome, all ages (include age unknown) (1)	33	1	nr	12.87
<20	1	0	nr	3.78
20-24	12	0	nr	11.25
25-29	4	1	nr	6.67
30-34	3	0	nr	7.80
35-39	5	0	nr	36.60
40-44	5	0	nr	140.45
45+	3	0	nr	1363.64
unknown	0	0	nr	---

(1) Clinical diagnosis only; with photodocumentation or measurements documented
(2) Includes penile, scrotal, and perineal hypospadias only

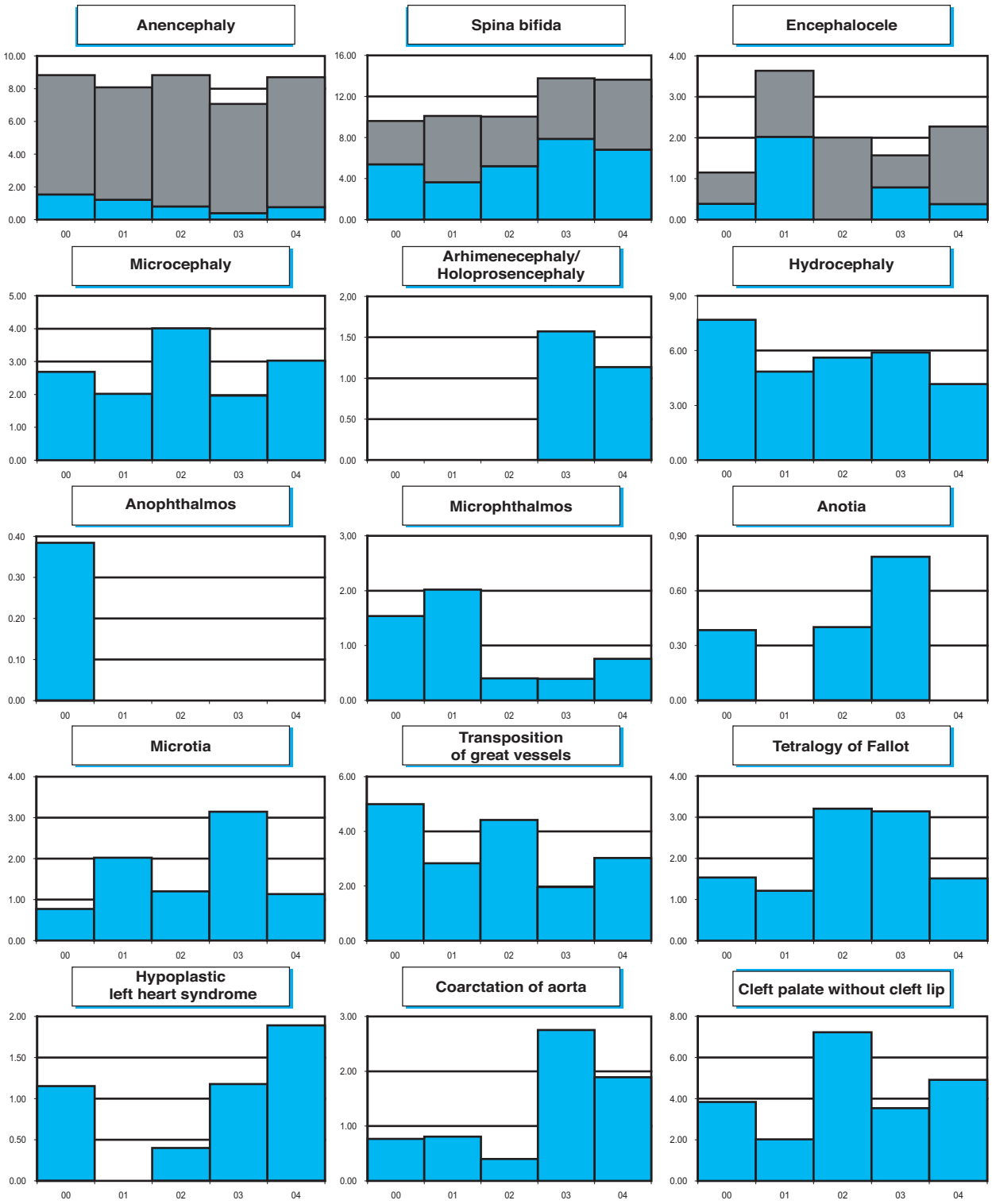
Ukraine: UABDP, Previous years rates 2000 - 2004

Birth prevalence rates: (LB+SB+TOP) * 10,000 for Anencephaly, Spina bifida and Encephalocele
Birth prevalence rates: (LB+SB) * 10,000 for all other malformations

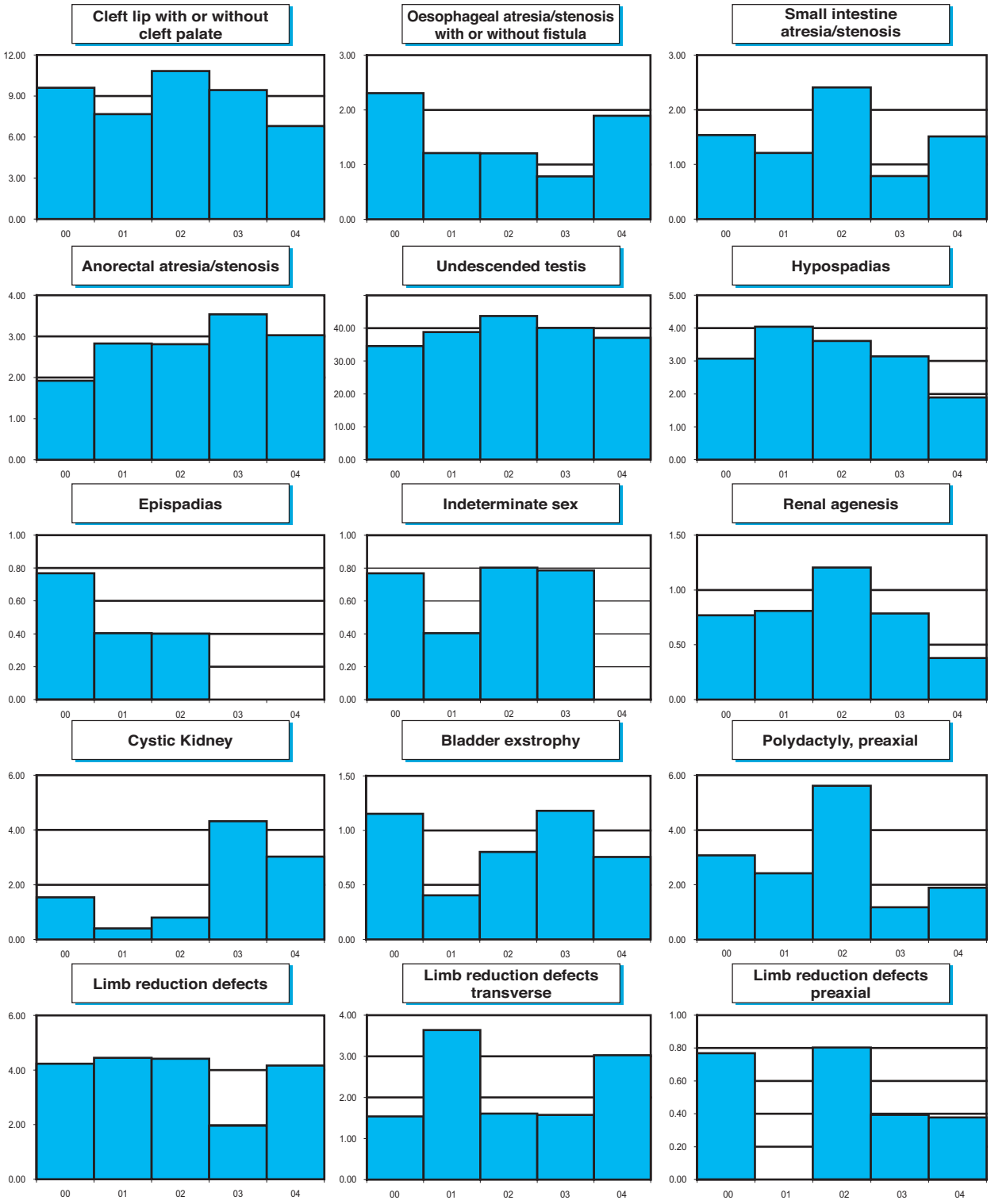
	1974-79	1980-84	1985-89	1990-94	1995-99	2000-04
Births						**127,547**
Anencephaly						8.31
Spina bifida						11.45
Encephalocele						2.12
Microcephaly						2.74
Arhinencephaly / Holoprosencephaly						0.55
Hydrocephaly						5.64
Anophthalmos						0.08
Microphthalmos						1.02
Unspecified Anophthalmos / Microphthalmos						---
Anotia						0.31
Microtia						1.65
Unspecified Anotia / Microtia						---
Transposition of great vessels						3.45
Tetralogy of Fallot						2.12
Hypoplastic left heart syndrome						0.94
Coarctation of aorta						1.33
Choanal atresia, bilateral						0.00
Cleft palate without cleft lip						4.31
Cleft lip with or without cleft palate						8.86
Oesophageal atresia / stenosis with or without fistula						1.49
Small intestine atresia / stenosis						1.49
Anorectal atresia / stenosis						2.82
Undescended testis (36 weeks of gestation or later)						38.80
Hypospadias						3.14
Epispadias						0.31
Indeterminate sex						0.55
Renal agenesis						0.78
Cystic kidney						2.04
Bladder exstrophy						0.86
Polydactyly, preaxial						2.82
Total Limb reduction defects (include unspecified)						3.84
Transverse						2.27
Preaxial						0.47
Postaxial						0.24
ntercalary						0.31
Mixed						0.24
Unspecified						---
Diaphragmatic hernia						1.88
Omphalocele						1.25
Gastroschisis						1.02
Unspecified Omphalocele / Gastroschisis						---
Prune belly sequence						0.00
Trisomy 13						0.16
Trisomy 18						0.31
Down syndrome, all ages (include age unknown)						12.30
<20						7.18
20-24						7.61
25-29						9.28
30-34						16.38
35-39						30.40
40-44						102.62
45+						816.33
unknown						---

Ukraine: UABDP

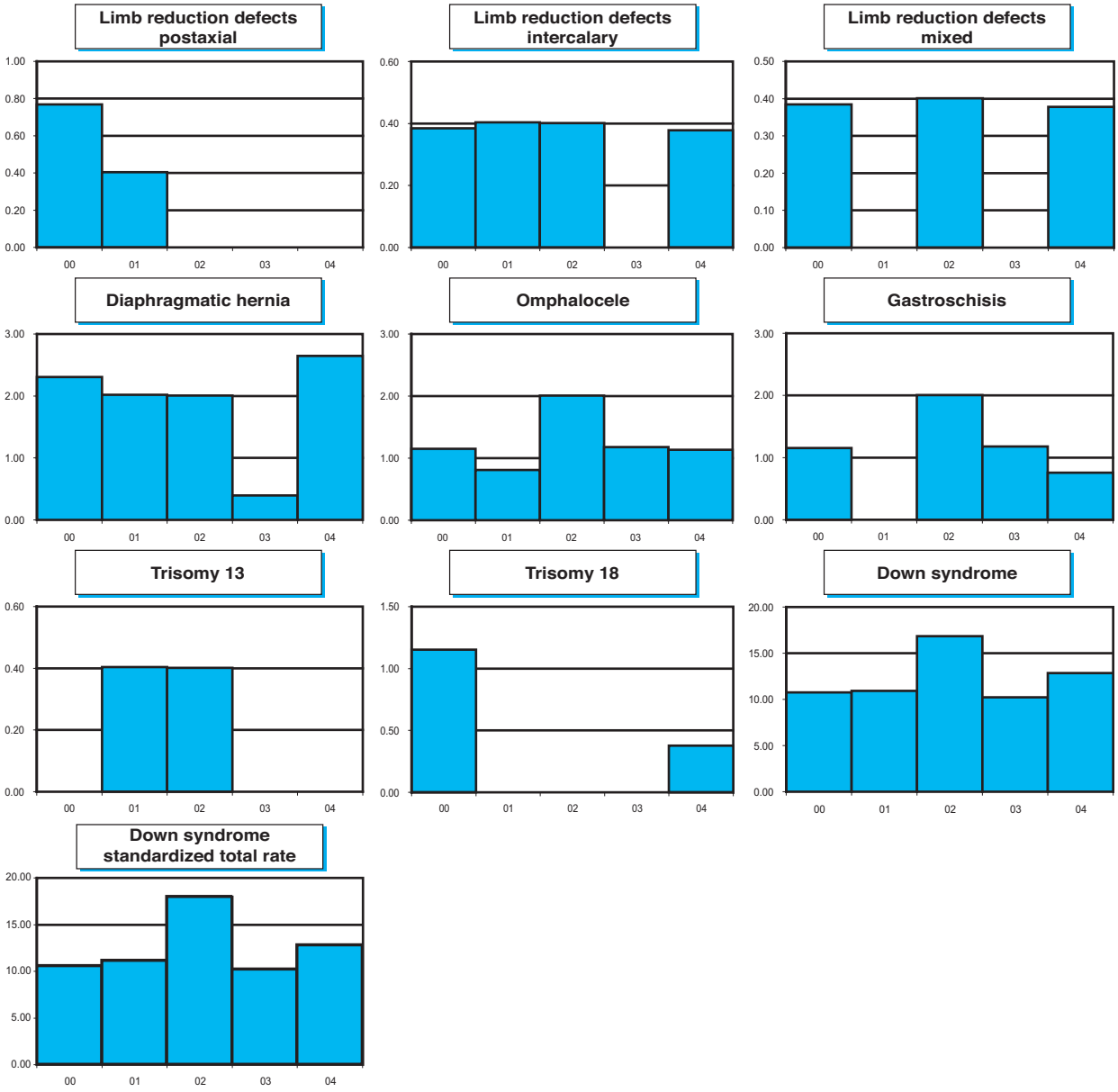
'Time trends 2000-2004 (Birth prevalence rates per 10,000)



Note: L+S rates, ToP rates



Note: L+S rates, ToP rates



Note: L+S rates, ToP rates

USA: Atlanta MACDP

Metropolitan Atlanta Congenital Defects Program (MACDP)

History:
The Programme started in 1967 and was a founding member of the ICBDSR. The Programme is a full member of the ICBDSR.

Size and coverage:
The Programme covers all births within a five-county area in metropolitan Atlanta, Georgia. The annual number of births in this area is approximately 50,000. Stillbirths and terminations of at least 20 weeks gestations are included. Elective terminations at any gestational age are included.

Legislation and funding:
In 1994 the Georgia Department of Human Resources (GDHR) added birth defects to the list of legally reportable conditions in Georgia. In 1997 the GDHR authorized the Birth Defects Branch at the Centers for Disease Control and Prevention (CDC) to act with and on its behalf to collect health information on children with birth defects. The Programme is funded by the Centers for Disease Control and Prevention.

Sources of ascertainment:
Multiple sources, such as delivery units, pediatric departments, neonatal intensive care units, laboratories, prenatal diagnostic centers, and tertiary care centers are used to ascertained malformed infants born in the defined area with a follow-up to age six years.

Exposure information:
Limited information on maternal illnesses, conditions, and maternal exposures such as medications.

Background information:
Demographic information, reproductive history, gestational age, birth weight, and pregnancy outcome information available from vital records for cases and live births and stillbirths.

Addresses and Staff:
Adolfo Correa, MD, PhD / Csaba Siffel, MD, PhD
National Center on Birth Defects and Developmental Disabilities Centers for Disease Control and Prevention, Mailstop E-86, 1600 Clifton Road Atlanta, GA 30333, USA

Phone: 1-404-498-4090

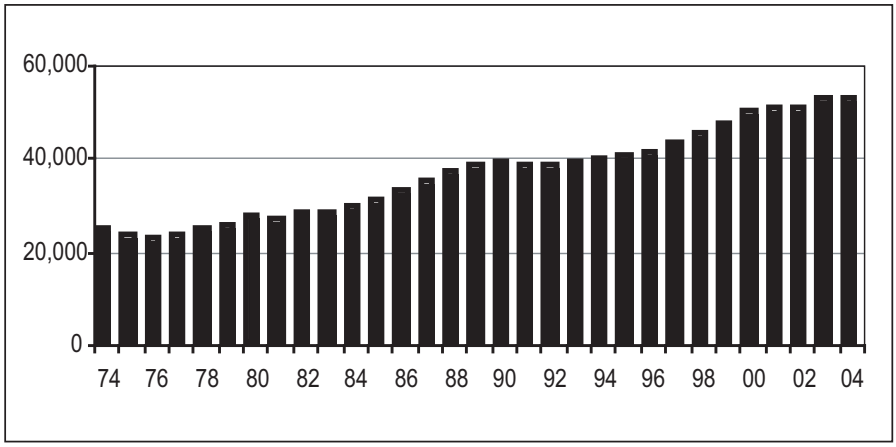
Fax: 1-404-498 3040

E-mail: ACorrea@cdc.gov; CSiffel@cdc.gov

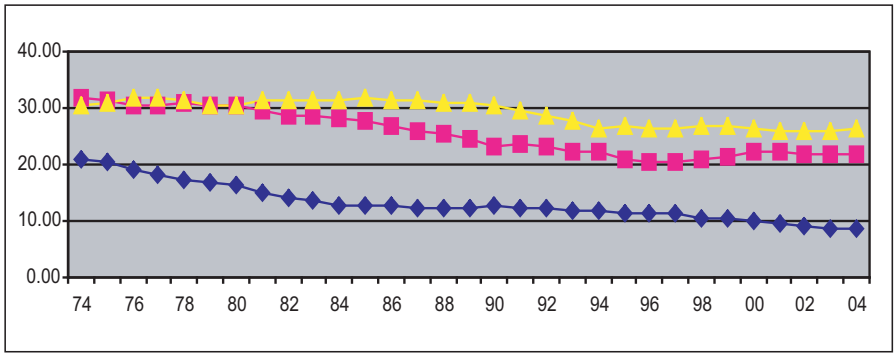
Website:
http://www.cdc.gov/ncbddd/bd/macdp.htm

USA: Atlanta MACDP

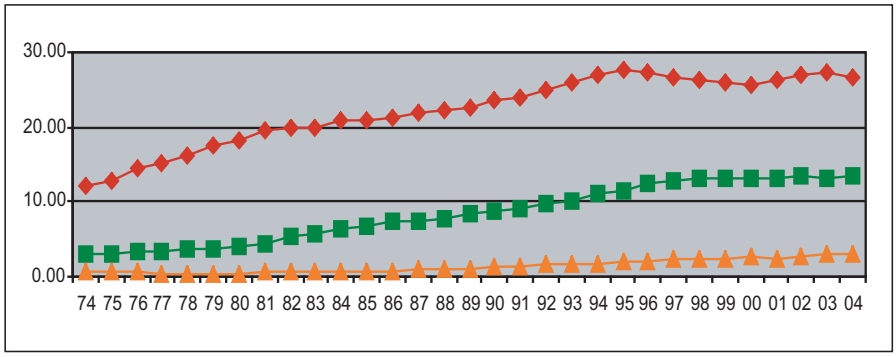
Total births by year



Percentage of births by maternal age



legenda: %births < 20 %births 20-24 %births 25-29



legenda: %births 30-34 %births 35-39 %births 40+

USA: Atlanta MACDP, 2004

Live births (LB)	51,808
Stillbirths (SB)	548
Total births	52,356
Number of terminations of pregnancy (ToP) for birth defects	nr

Birth Defects	Number of cases			Rates*10,000
	LB	SB	ToP	Total rate
Anencephaly	1	3	5	1.72
Spina bifida	6	1	6	2.48
Encephalocele	3	0	2	0.96
Microcephaly	22	0	0	4.20
Arhinencephaly / Holoprosencephaly	3	0	1	0.76
Hydrocephaly	24	4	2	5.73
Anophthalmos	2	0	1	0.57
Microphthalmos	9	0	0	1.72
Unspecified Anophthalmos/ Microphthalmos	0	0	0	---
Anotia	0	0	0	0.00
Microtia	7	0	0	1.34
Unspecified Anotia/Microtia	0	0	0	---
Transposition of great vessels	23	0	0	4.39
Tetralogy of Fallot	20	0	0	3.82
Hypoplastic left heart syndrome	8	1	0	1.72
Coarctation of aorta	27	0	0	5.16
Choanal atresia, bilateral	1	0	0	0.19
Cleft palate without cleft lip	28	0	0	5.35
Cleft lip with or without cleft palate	57	0	4	11.65
Oesophageal atresia / stenosis with or without fistula	9	0	0	1.72
Small intestine atresia / stenosis	7	0	0	1.34
Anorectal atresia / stenosis	15	0	0	2.87
Undescended testis (36 weeks of gestation or later)	24	0	0	4.58
Hypospadias	29	0	0	5.54
Epispadias	2	0	0	0.38
Indeterminate sex	7	0	0	1.34
Renal agenesis	3	0	1	0.76
Cystic kidney	29	0	1	5.73
Bladder exstrophy	0	0	0	0.00
Polydactyly, preaxial	11	0	0	2.10
Total Limb reduction defects (include unspecified)	14	0	1	2.87
Transverse	8	0	1	1.72
Preaxial	2	0	0	0.38
Postaxial	1	0	0	0.19
Intercalary	1	0	0	0.19
Mixed	2	0	0	0.38
Unspecified	0	0	0	---
Diaphragmatic hernia	10	0	0	1.91
Omphalocele	4	1	1	1.15
Gastroschisis	20	0	0	3.82
Unspecified Omphalocele/Gastroschisis	0	0	0	---
Prune belly sequence	3	0	1	0.76
Trisomy 13	4	1	5	1.91
Trisomy 18	7	5	7	3.63
Down syndrome, all ages (include age unknown)	68	0	16	16.04
<20	3	0	1	8.69
20-24	3	0	0	2.64
25-29	9	0	0	6.55
30-34	17	0	3	14.29
35-39	17	0	9	36.89
40-44	19	0	3	145.70
45+	0	0	0	0.00
unknown	0	0	0	---

nr = not reported

USA: Atlanta MACDP, Previous years rates 1974 - 2004

Birth prevalence rates: (LB+SB+TOP) * 10,000

	1974-79	1980-84	1985-89	1990-94	1995-99	2000-04
Births	**143,922**	**139,240**	**173,520**	**194,162**	**216,010**	**255,901**
Anencephaly	5.35	3.95	3.46	2.88	4.12	2.23
Spina bifida	7.23	6.61	5.94	4.48	4.77	3.44
Encephalocele	1.88	2.44	2.02	1.13	2.08	1.25
Microcephaly	5.00	6.03	6.11	4.84	8.38	5.74
Arhinencephaly / Holoprosencephaly	0.56	0.79	1.33	1.29	1.16	0.66
Hydrocephaly	9.59	9.26	7.03	5.67	7.45	6.68
Anophthalmos	0.56	0.65	0.58	0.82	0.23	0.35
Microphthalmos	3.89	3.81	3.11	2.88	2.96	2.42
Unspecified Anophthalmos / Microphthalmos	---	---	---	---	---	---
Anotia	0.21	0.22	0.06	0.21	0.19	0.27
Microtia	1.32	1.29	1.79	1.08	1.34	1.41
Unspecified Anotia / Microtia	---	---	---	---	---	---
Transposition of great vessels	4.86	5.82	4.73	4.84	5.42	5.55
Tetralogy of Fallot	2.92	3.73	4.15	3.61	4.54	4.14
Hypoplastic left heart syndrome	2.29	3.02	2.13	2.94	3.24	2.34
Coarctation of aorta	3.89	4.24	5.13	4.27	4.91	5.51
Choanal atresia, bilateral	0.42	0.07	0.46	0.21	0.46	0.43
Cleft palate without cleft lip	7.71	3.95	5.30	4.74	5.65	5.78
Cleft lip with or without cleft palate	11.46	11.35	9.11	9.37	9.44	8.68
Oesophageal atresia / stenosis with or without fistula	2.29	2.51	2.36	2.16	2.04	1.99
Small intestine atresia / stenosis	1.46	1.65	1.67	1.80	1.67	1.88
Anorectal atresia / stenosis	4.66	3.52	4.26	3.71	3.47	3.13
Undescended testis (36 weeks of gestation or later)	nr	nr	nr	nr	21.27*	14.93
Hypospadias	1.25	1.58	4.61	4.79	8.29	8.01
Epispadias	0.97	0.86	0.63	0.62	0.42	0.43
Indeterminate sex	2.29	1.80	1.15	1.13	1.34	1.60
Renal agenesis	2.08	1.87	1.21	1.34	1.16	1.02
Cystic kidney	2.43	2.73	4.21	5.10	5.83	5.94
Bladder exstrophy	0.49	0.22	0.23	0.36	0.09	0.12
Polydactyly, preaxial	1.95	1.87	2.59	3.35	2.64	2.07
Total Limb reduction defects (include unspecified)	6.18	4.45	4.61	4.79	6.34	5.35
Transverse	3.68	3.09	2.77	3.45	3.38	3.28
Preaxial	1.18	0.57	0.81	0.82	1.20	0.94
Postaxial	0.21	0.22	0.35	0.26	0.32	0.16
ntercalary	0.63	0.22	0.29	0.05	0.28	0.20
Mixed	0.07	0.36	0.29	0.15	0.74	0.66
Unspecified	---	---	---	---	---	---
Diaphragmatic hernia	2.57	2.30	2.82	2.37	2.18	2.81
Omphalocele	3.89	3.16	3.11	2.58	2.45	1.91
Gastroschisis	1.67	1.87	2.31	2.73	1.71	3.17
Unspecified Omphalocele / Gastroschisis	---	---	---	---	---	---
Prune belly sequence	0.63	0.50	0.46	0.36	0.32	0.47
Trisomy 13	1.18	1.08	1.67	1.18	1.94	1.91
Trisomy 18	0.63	1.94	2.07	2.47	4.44	4.45
Down syndrome, all ages (include age unknown)	8.82	10.99	10.37	12.67	17.68	17.16
<20	nr	7.01	6.48	7.23	10.48	6.35
20-24	nr	6.68	8.65	6.95	10.21	4.97
25-29	nr	8.29	6.82	7.22	8.01	7.93
30-34	nr	16.81	12.67	11.70	13.55	14.57
35-39	nr	20.65	22.97	31.73	43.33	47.82
40-44	nr	80.32	66.62	70.80	137.67	113.82
45+	nr	0.00	0.00	487.80	251.26	119.76
unknown	---	---	---	---	---	---

* data include less than 5 years

nr = not reported

USA: Atlanta MACDP

'Time trends 1974-2004 (Birth prevalence rates per 10,000)

Note: L+S rates, ToP rates

Note: L+S rates, ToP rates

Note: L+S rates, ToP rates

USA: Texas
Texas Center for Birth Defects Research

History:
BDES (Birth Defects Epidemiology and Surveillance) was established after an unusual cluster of anencephaly cases that occurred in Brownsville, Texas in 1991. Epidemiologic investigations revealed a higher than expected rate of neural tube defects among children born to Hispanic mothers living in South Texas. In recognition that epidemiologic resources are routinely needed to investigate birth defects clusters, the Texas State Legislature passed the Texas Birth Defects Act in 1993, which authorized the establishment of BDES. Since 1994, BDES has maintained the Texas Birth Defects Registry, an active population-based birth defects surveillance system, which has been statewide since 1999. Through multiple sources of information, the Registry monitors all births in Texas and identifies cases of birth defects. Children identified through the Registry are referred to appropriate medical and community services. In 1996, the CDC-funded Texas Center for Birth Defects Research and Prevention was established under the auspices of BDES . The Programme is a full member of the ICBDSR.

Size and coverage
The Programme covers all deliveries to mothers residing in Texas (approximately 380,000 annually). Stillbirths and terminations of any gestational age are included. Cases diagnosed up to age one are included (up to any age for fetal alcohol syndrome). As of 2006, there were over 100,000 birth defect cases in the Registry.

Legislation and funding
Birth defects surveillance was mandated by the Texas Birth Defects Act in 1993, and is codified in the Texas Health and Safety Code Chapter 87. About one-half of funding for the birth defects registry is from state general revenue with the remainder from federal block grants .

Sources of ascertainment
Birth hospitals, birthing centres, lay midwives, hospitals where affected children are treated.

Exposure information
Limited information on maternal illnesses and conditions, limited information on maternal exposures such as medications.

Background information
Basic demographics, reproductive history, gestational age, delivery information.

Addresses and Staff:
Mark A. Canfield, Ph.D., Branch Manager Texas Birth Defects Epidemiology and Surveillance 1100 W. 49th Street, Suite T-707, Austin, Texas 78756-3180, USA

Branch phone: 512-458-7232

Direct line/voicemail: 512-458-7111 ext.1-6158

Fax: 512-458-7330

E-mail: mark.canfield@dshs.state.tx.us

Lisa K. Robinson (Marengo), M.S. Epidemiologist Texas Department of State Health Services Birth Defects Epidemiology and Surveillance Branch, Tower Bldg. Suite 704.11

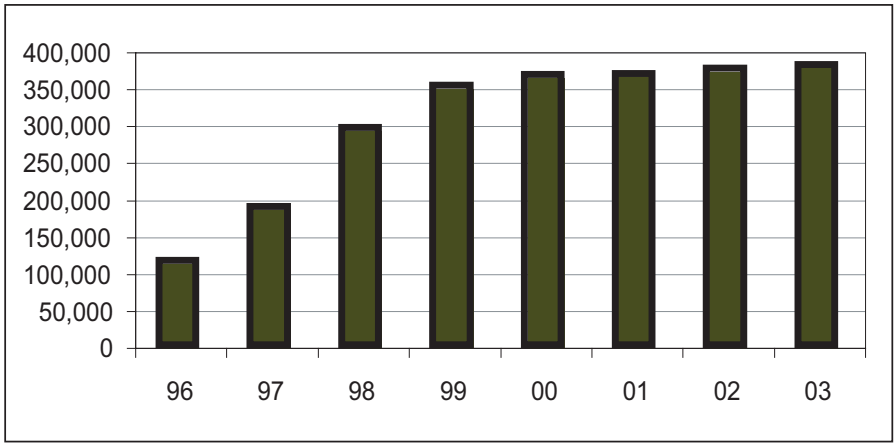
phone: 512.458.7111 ext. 6657

E-mail: Lisa.Marengo@dshs.state.tx.us

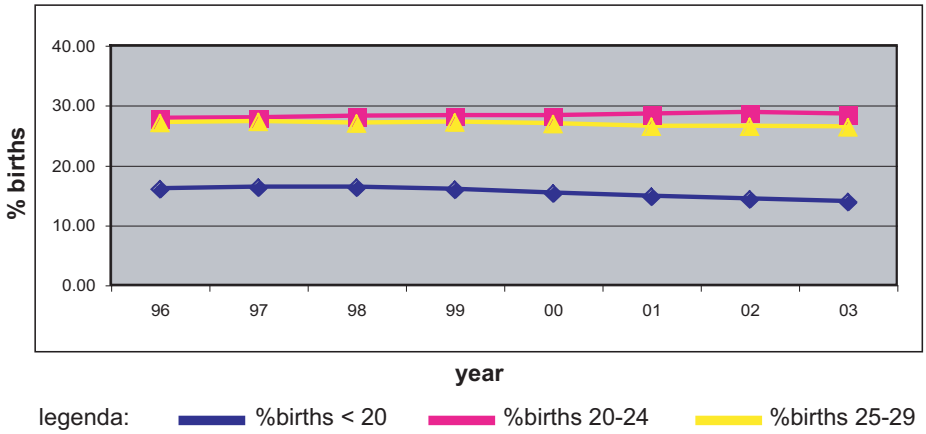
Website: www.dshs.state.tx.us/birthdefects

USA: Texas

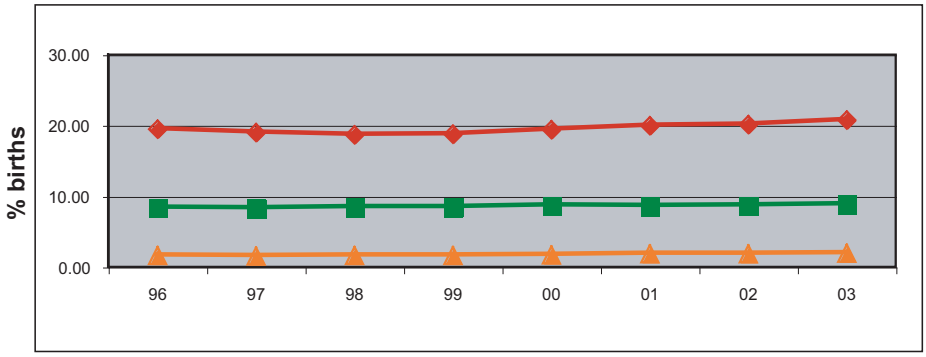
Total births by year



Percentage of births by maternal age



legenda: %births < 20 %births 20-24 %births 25-29



legenda: %births 30-34 %births 35-39 %births 40+

USA: Texas, 2003

Live births (LB)	377,374
Stillbirths (SB)	2,265
Total births	379,639
Number of terminations of pregnancy (ToP) for birth defects	179

Birth Defects	Number of cases			Rates*10,000
	LB	SB	ToP	Total rate
Anencephaly	38	25	29	2.42
Spina bifida	103	7	8	3.11
Encephalocele	24	2	4	0.79
Microcephaly	245	0	1	6.48
Arhinencephaly / Holoprosencephaly	35	3	2	1.05
Hydrocephaly	185	9	6	5.27
Anophthalmos	9	1	1	0.29
Microphthalmos	83	0	2	2.24
Unspecified Anophthalmos/ Microphthalmos	0	0	0	---
Anotia	7	2	0	0.24
Microtia	81	1	0	2.16
Unspecified Anotia/Microtia	0	0	0	---
Transposition of great vessels	168	1	1	4.48
Tetralogy of Fallot	131	1	1	3.50
Hypoplastic left heart syndrome	65	1	2	1.79
Coarctation of aorta	176	1	0	4.66
Choanal atresia, bilateral	31	0	0	0.82
Cleft palate without cleft lip	170	2	4	4.63
Cleft lip with or without cleft palate	327	20	9	9.37
Oesophageal atresia / stenosis with or without fistula	69	0	0	1.82
Small intestine atresia / stenosis	48	0	0	1.26
Anorectal atresia / stenosis	179	4	3	4.90
Undescended testis (36 weeks of gestation or later)	343	1	0	9.06
Hypospadias	544	1	0	14.35
Epispadias	30	0	0	0.79
Indeterminate sex	22	7	7	0.95
Renal agenesis	58	11	8	2.03
Cystic kidney	161	2	5	4.42
Bladder exstrophy	9	0	0	0.24
Polydactyly, preaxial	133	3	0	3.58
Total Limb reduction defects (include unspecified)	163	11	6	4.74
Transverse	84	6	4	2.47
Preaxial	31	0	2	0.87
Postaxial	6	0	0	0.16
Intercalary	3	1	0	0.11
Mixed	29	3	0	0.84
Unspecified	10	1	0	---
Diaphragmatic hernia	89	3	2	2.47
Omphalocele	53	4	6	1.66
Gastroschisis	156	9	4	4.45
Unspecified Omphalocele/Gastroschisis	17	2	2	---
Prune belly sequence	13	1	1	0.39
Trisomy 13	19	5	9	0.87
Trisomy 18	38	18	16	1.90
Down syndrome, all ages (include age unknown)	378	15	23	10.95
<20	27	0	0	5.16
20-24	51	4	0	5.09
25-29	54	1	1	5.61
30-34	54	1	3	7.37
35-39	69	2	8	23.59
40-44	51	1	6	84.91
45+	5	1	0	165.75
unknown	67	5	5	---

USA: Texas, Previous years rates 1996 - 2003

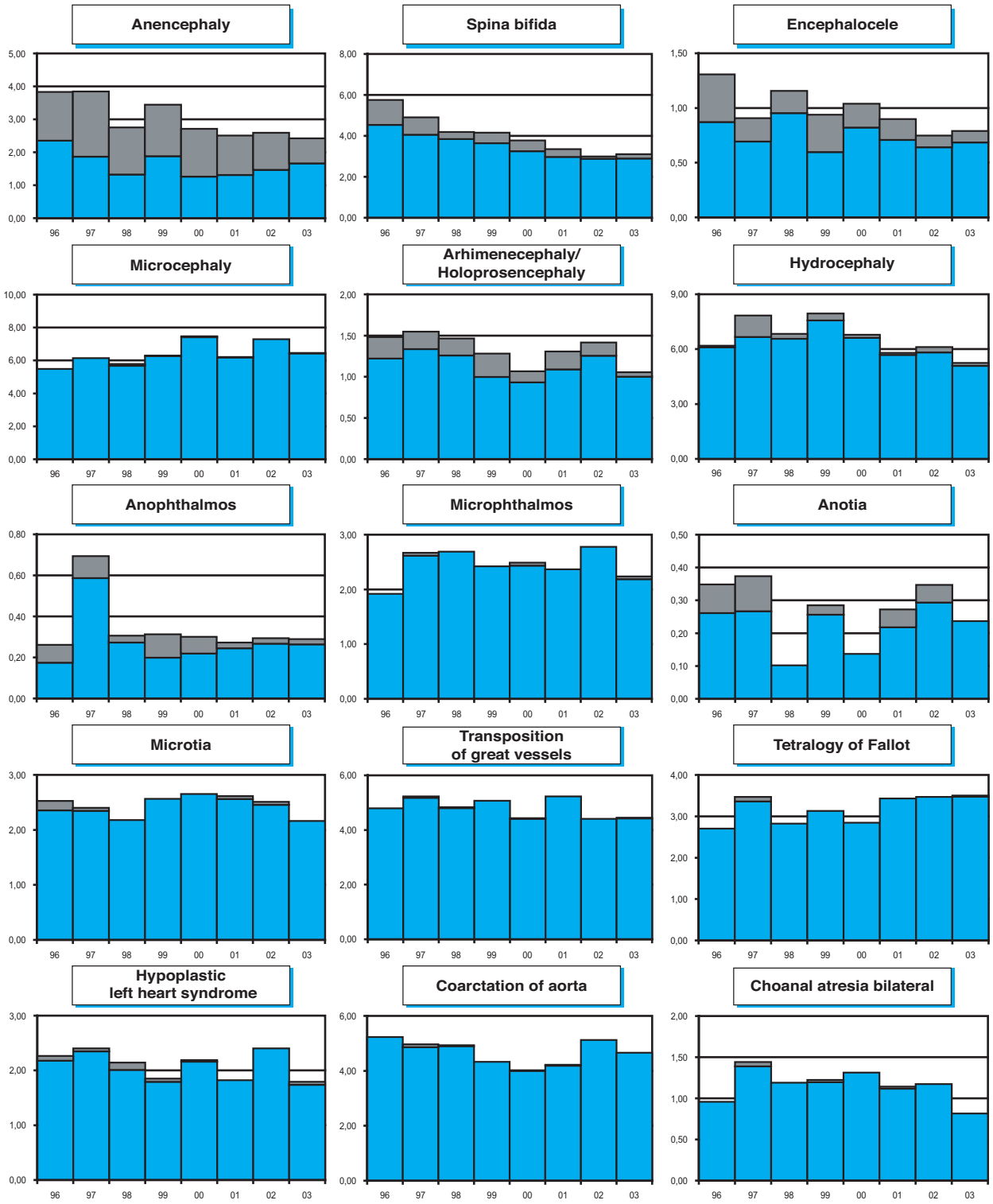
Birth prevalence rates: (LB+SB+TOP) * 10,000

	1974-79	1980-84	1985-89	1990-94	1995-99*	2000-03
Births					**947,232**	**1,487,174**
Anencephaly					3.36	2.56
Spina bifida					4.51	3.30
Encephalocele					1.05	0.87
Microcephaly					6.01	6.85
Arhinencephaly / Holoprosencephaly					1.41	1.21
Hydrocephaly					7.37	5.98
Anophthalmos					0.38	0.29
Microphthalmos					2.49	2.47
Unspecified Anophthalmos / Microphthalmos					---	---
Anotia					0.25	0.25
Microtia					2.41	2.48
Unspecified Anotia / Microtia					---	---
Transposition of great vessels					4.99	4.63
Tetralogy of Fallot					3.05	3.32
Hypoplastic left heart syndrome					2.10	2.05
Coarctation of aorta					4.75	4.51
Choanal atresia, bilateral					1.22	1.11
Cleft palate without cleft lip					5.81	5.33
Cleft lip with or without cleft palate					10.80	10.26
Oesophageal atresia / stenosis with or without fistula					2.24	2.00
Small intestine atresia / stenosis					1.71	1.60
Anorectal atresia / stenosis					4.42	4.73
Undescended testis (36 weeks of gestation or later)					7.82	8.49
Hypospadias					18.25	16.24
Epispadias					0.58	0.71
Indeterminate sex					1.66	1.01
Renal agenesis					2.11	1.92
Cystic kidney					4.30	4.43
Bladder exstrophy					0.18	0.24
Polydactyly, preaxial					2.82	3.19
Total Limb reduction defects (include unspecified)					5.48	5.23
Transverse					2.58	2.65
Preaxial					1.14	1.04
Postaxial					0.26	0.24
Intercalary					0.08	0.15
Mixed					1.26	0.93
Unspecified					---	---
Diaphragmatic hernia					2.68	2.52
Omphalocele					2.27	2.06
Gastroschisis					3.78	4.12
Unspecified Omphalocele / Gastroschisis					---	---
Prune belly sequence					0.26	0.28
Trisomy 13					1.16	1.13
Trisomy 18					2.44	2.06
Down syndrome, all ages (include age unknown)					11.88	12.33
<20					7.63	6.40
20-24					5.69	5.04
25-29					4.84	5.57
30-34					9.45	8.88
35-39					27.11	28.49
40-44					97.18	109.67
45+					117.45	205.37
unknown					---	---

* data include less than 5 years

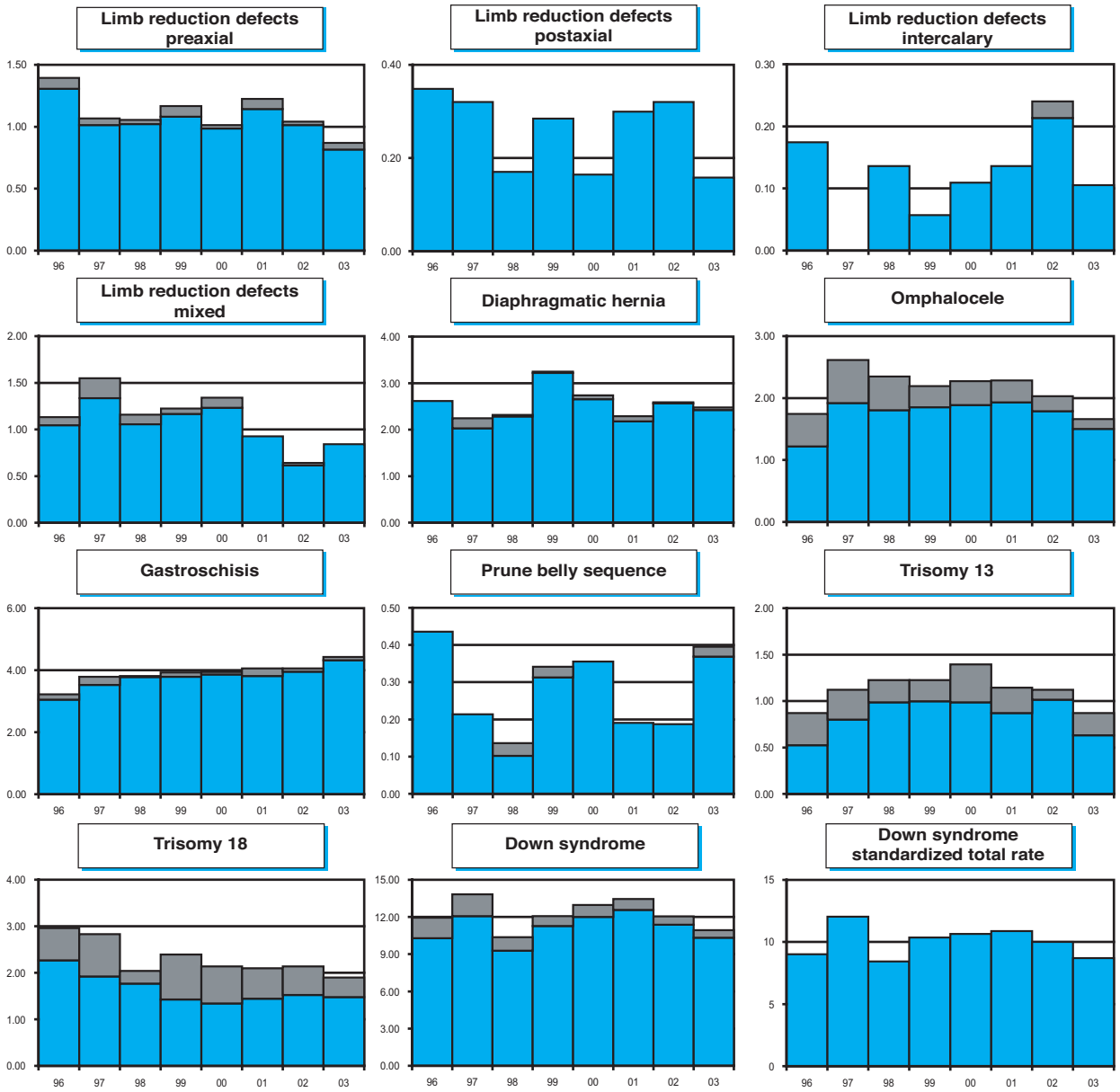
USA: Texas

'Time trends 1996-2003 (Birth prevalence rates per 10,000)



Note: L+S rates, ToP rates

Note: L+S rates, ToP rates



Note: L+S rates, ToP rates

USA: Utah UBDN

Utah Birth Defects Network (UBDN)

History:
The Utah Birth Defect Network (UBDN) evolved from a population-based pilot project on neural tube defects surveillance in 1994 to become a full, statewide, population-based surveillance program between 1994 and 1999. The UBDN currently monitors virtually all major structural birth defects occurring in Utah. Grants from the Centers for Disease Control and Prevention, the Utah Chapter of the March of Dimes, and Maternal Child Health Block Grants provided the funding for the UBDN to expand to its full surveillance system from the initial focus on neural tube defects (NTDs). The UBDN has ongoing legislative funding from the Utah Department of Health. In 2003 the UBDN became one of the ten Centers funded by the CDC to study causes of birth defects through the National Birth Defects Prevention Study. The UBDN is a full member of the Clearinghouse.

Size and coverage
The UBDN is a state-wide population-based surveillance system that monitors approximately 50,000 births annually. All pregnancy outcomes are monitored, including stillbirths and pregnancy terminations of at least 20 weeks' gestation. Terminations less than 20 weeks with a diagnosis of a major birth defect are also included.

Legislation and funding
In 1999, an Administrative Rule was enacted under the Utah Health Code Statute which mandates all delivery hospitals and laboratories to report any pregnancy or infant diagnosed with a birth defect. This administrative rule also covers health care providers and other agencies that voluntarily report a birth defect case to the UBDN. The UBDN has secured ongoing funding through the State Legislature for surveillance activities.

Sources of ascertainment
The UBDN uses multiple sources of ascertainment, including delivery units, birth hospitals, pediatric departments, site champions, laboratories, prenatal diagnostic centers, hospital discharge data, and specialty clinics.

Exposure information
Exposure information is obtained through the medical record abstraction process.

Background information
General epidemiological data for all births are available through the Utah Department of Health's Office of Vital Records and Statistics. Background information for the Utah Birth Defect Network is available on the UBDN website at www.health.utah.gov/birthdefect

Addresses and Staff:
Marcia Feldkamp, Director

E-mail: Marcia.Feldkamp@hsc.utah.edu

Lorenzo Botto, Medical Epidemiologist

E-mail: Lorenzo.Botto@hsc.utah.edu

Miland Palmer, Surveillance Coordinator

E-mail: mpalmer@utah.gov

Utah Birth Defect Network, Utah Department of Health PO Box 144697, Salt Lake City, Utah 84114-4697 USA

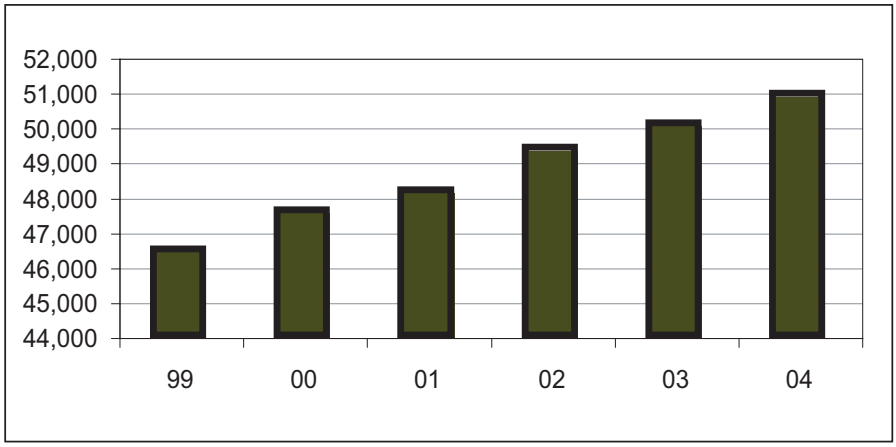
phone: 801 257 0566

Fax: 801 257 0572

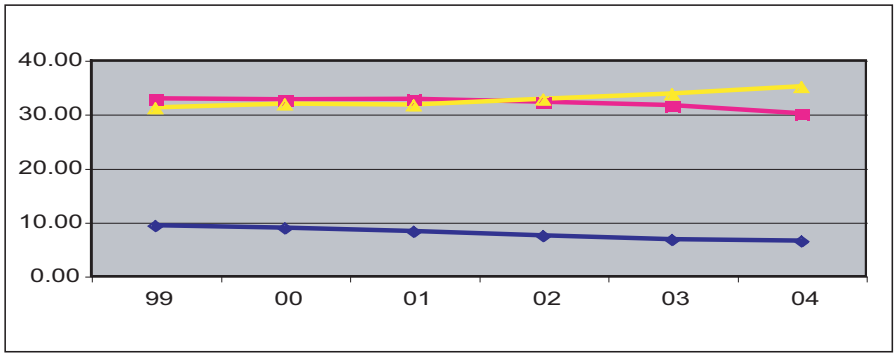
Website: www.health.utah.gov/birthdefect

USA: Utah UBDN

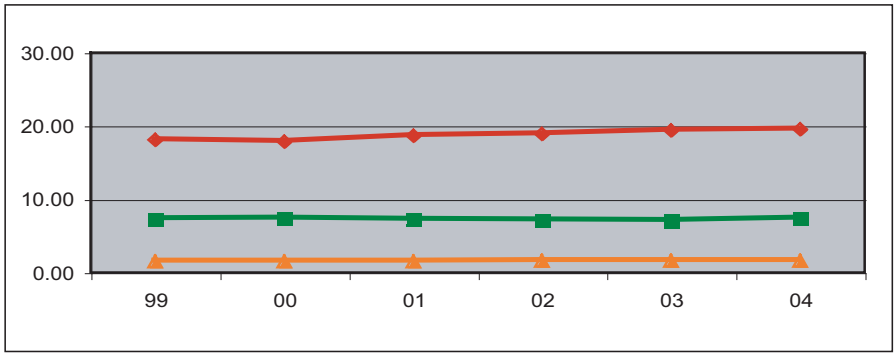
Total births by year



Percentage of births by maternal age



legenda: %births < 20 %births 20-24 %births 25-29



legenda: %births 30-34 %births 35-39 %births 40+

USA: Utah UBDN, 2004

Live births (LB)	50,653
Stillbirths (SB)	265
Total births	50,918
Number of terminations of pregnancy (ToP) for birth defects	56

Birth Defects	Number of cases			Rates*10,000
	LB	SB	ToP	Total rate
Anencephaly	2	1	5	0.59
Spina bifida	24	1	3	4.90
Encephalocele	2	0	2	0.39
Microcephaly	21	0	1	4.12
Arhinencephaly / Holoprosencephaly	1	0	3	0.20
Hydrocephaly	4	2	1	1.18
Anophthalmos	2	0	0	0.39
Microphthalmos	4	0	1	0.78
Unspecified Anophthalmos/ Microphthalmos	0	0	0	---
Anotia	0	0	0	0.00
Microtia	22	0	0	4.32
Unspecified Anotia/Microtia	0	0	0	---
Transposition of great vessels	30	1	0	6.08
Tetralogy of Fallot	12	0	0	2.35
Hypoplastic left heart syndrome	16	2	0	3.53
Coarctation of aorta	37	1	0	7.45
Choanal atresia, bilateral	0	0	0	0.00
Cleft palate without cleft lip	36	0	1	7.06
Cleft lip with or without cleft palate	52	2	5	10.59
Oesophageal atresia / stenosis with or without fistula	8	0	1	1.57
Small intestine atresia / stenosis	9	0	0	1.77
Anorectal atresia / stenosis	11	0	1	2.16
Undescended testis (36 weeks of gestation or later)	nr	nr	nr	nr
Hypospadias	147	0	0	28.84
Epispadias	2	0	0	0.39
Indeterminate sex	nr	nr	nr	nr
Renal agenesis	12	1	2	2.55
Cystic kidney	29	0	3	5.69
Bladder exstrophy	1	0	0	0.20
Polydactyly, preaxial	nr	nr	nr	nr
Total Limb reduction defects (include unspecified)	20	2	4	4.32
Transverse	7	0	2	1.37
Preaxial	5	1	1	1.18
Postaxial	1	0	0	0.20
Intercalary	0	0	0	0.00
Mixed	5	1	1	1.18
Unspecified	2	0	0	---
Diaphragmatic hernia	17	0	1	3.34
Omphalocele	8	2	3	1.96
Gastroschisis	23	2	4	4.90
Unspecified Omphalocele/Gastroschisis	0	0	0	---
Prune belly sequence	1	0	1	0.20
Trisomy 13	1	1	2	0.39
Trisomy 18	10	4	7	2.75
Down syndrome, all ages (include age unknown)	68	3	5	13.93
<20	3	0	0	9.21
20-24	18	0	0	11.77
25-29	11	2	0	7.30
30-34	12	0	1	12.03
35-39	12	1	2	34.33
40-44	11	0	2	143.79
45+	1	0	0	243.90
unknown	0	0	0	---

nr = not reported

USA: Utah UBDN, Previous years rates 1999 - 2004

Birth prevalence rates: (LB+SB+TOP) * 10,000

	1974-79	1980-84	1985-89	1990-94	1995-99*	2000-04
Births					**46,458**	**246,111**
Anencephaly					1.94	2.07
Spina bifida					3.44	4.06
Encephalocele					0.22	0.93
Microcephaly					3.01	3.62
Arhinencephaly / Holoprosencephaly					1.08	1.30
Hydrocephaly					2.37	2.32
Anophthalmos					0.00	0.24
Microphthalmos					1.94	1.46
Unspecified Anophthalmos / Microphthalmos					---	---
Anotia					0.22	0.08
Microtia					1.08	2.72
Unspecified Anotia / Microtia					---	---
Transposition of great vessels					6.46	4.96
Tetralogy of Fallot					4.52	2.76
Hypoplastic left heart syndrome					3.87	3.53
Coarctation of aorta					8.61	7.27
Choanal atresia, bilateral					0.00	0.16
Cleft palate without cleft lip					6.03	7.76
Cleft lip with or without cleft palate					12.70	14.38
Oesophageal atresia / stenosis with or without fistula					4.30	2.19
Small intestine atresia / stenosis					3.66	2.60
Anorectal atresia / stenosis					4.52	3.13
Undescended testis (36 weeks of gestation or later)					nr	nr
Hypospadias					27.34	29.26
Epispadias					0.65	0.28
Indeterminate sex					nr	nr
Renal agenesis					4.30	3.37
Cystic kidney					3.44	5.97
Bladder exstrophy					0.22	0.24
Polydactyly, preaxial					nr	nr
Total Limb reduction defects (include unspecified)					6.03	5.81
Transverse					4.09	2.64
Preaxial					0.86	1.71
Postaxial					0.00	0.12
Intercalary					0.00	0.08
Mixed					0.65	1.06
Unspecified					---	---
Diaphragmatic hernia					2.58	3.66
Omphalocele					2.15	2.84
Gastroschisis					4.30	4.84
Unspecified Omphalocele / Gastroschisis					---	---
Prune belly sequence					0.00	0.20
Trisomy 13					0.65	1.71
Trisomy 18					3.66	3.33
Down syndrome, all ages (include age unknown)					15.93	15.56
<20					6.96	11.41
20-24					7.89	8.82
25-29					11.08	8.50
30-34					9.54	15.49
35-39					76.54	46.37
40-44					109.38	158.47
45+					185.19	526.32
unspecified					---	---

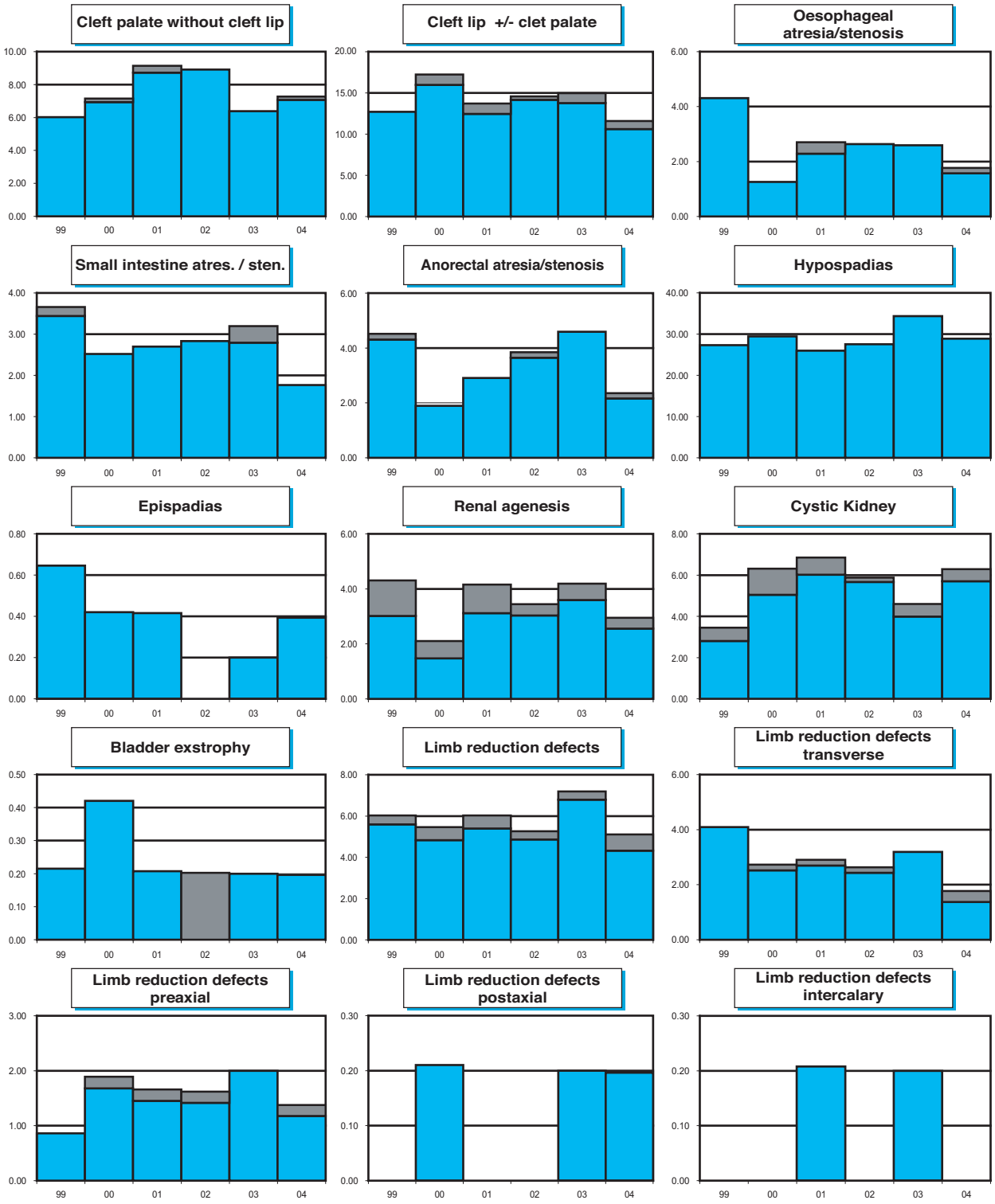
* = data available for less than 5 years

nr= not reported

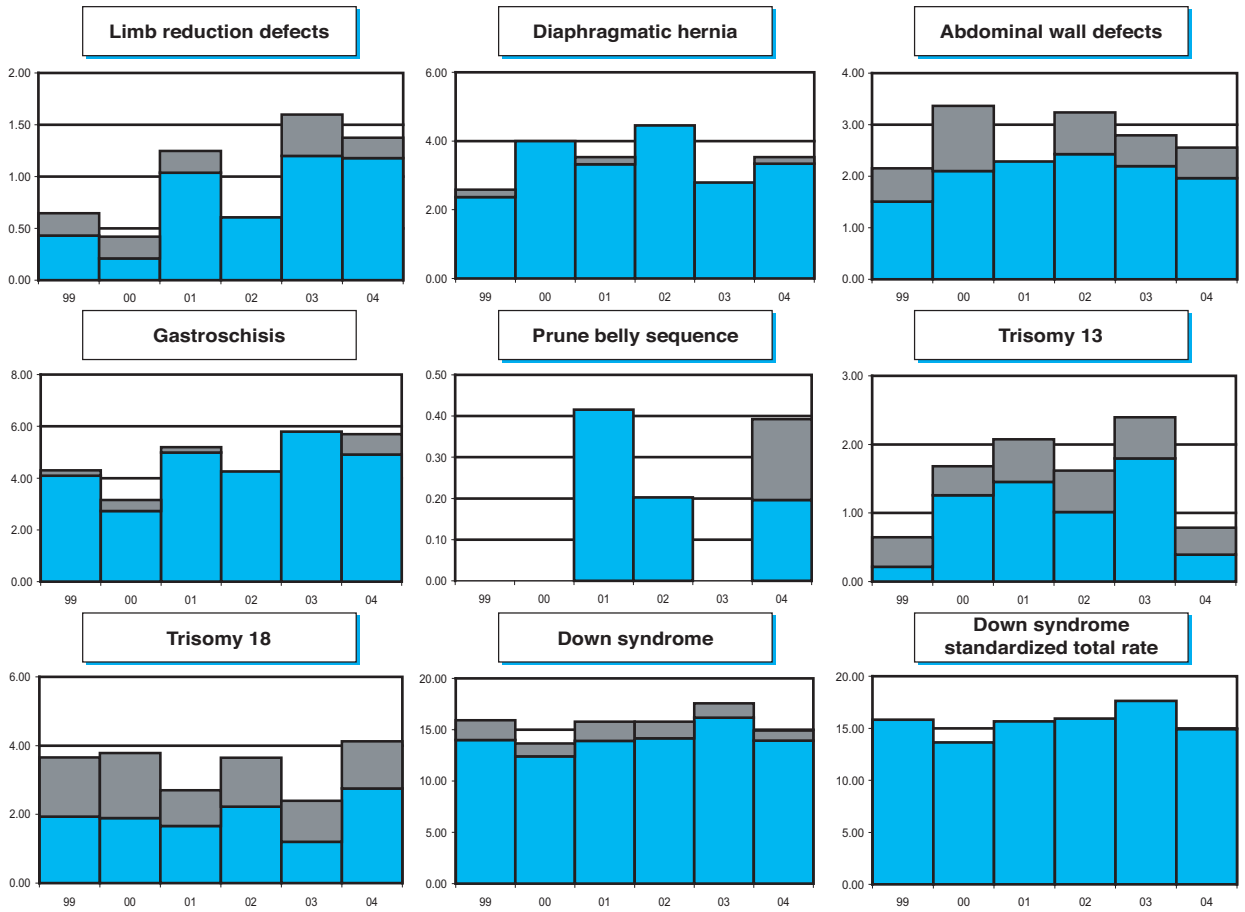
USA: Utah UBDN

'Time trends 1999-2004 (Birth prevalence rates per 10,000)

Note: L+S rates, ToP rates



Note: L+S rates, ToP rates



Note: L+S rates, ToP rates

Wales: CARIS

Congenital Anomaly Register and Information Service (CARIS)

History and Funding:
Data collection commenced on 1st January 1998 and includes any baby where pregnancy ended after this date. CARIS joined EUROCAT in 1998 and ICBDSR in 2004. CARIS is based at Singleton Hospital, Swansea and is funded by the National Assembly for Wales. CARIS aims to collect data which can be used to describe the pattern of congenital anomalies across Wales. This should help:

- Build up and monitor the picture of congenital anomalies in Wales
- Assess interventions intended to help prevent or detect congenital anomalies
- Plan and co-ordinate provision of health services for affected babies and children
- Assess possible clusters of birth defects and their causes

Population Coverage:
The Registry covers the entire country of Wales (population-based = All mothers resident in defined geographic area) with an annual number of births of around 32,000.

Sources of Ascertainment:
Reporting is voluntary. The Register relies upon multi-source reporting including: antenatal clinics, delivery units, pediatric departments, ophthalmology, cytogenetics, pathology, orthopaedics, maxillo-facial and regional centres of pediatric surgery. Each delivery unit has a nominated co-ordinator to help ensure good reporting and chase for further details. CARIS staff also visit units to help with data collection. Registration covers all fetuses with prenatally diagnosed anomalies. There is no lower age of cut off, so the fetal losses and early terminations with anomalies are registered. All liveborn babies with structural anomalies are registered if diagnosed before their 1st birthday, but all chromosomal anomalies are registered, even if diagnosed later. Data exchange with the Mersey Register is also important as babies needing specialist services in North Wales are referred to Liverpool

Termination of Pregnancy:
Termination of pregnancy is legal up to 24 weeks of gestation. Terminations of pregnancy are registered. If congenital anomaly is diagnosed, there is no upper gestational age limit for termination in cases of major anomaly.

Stillbirth Definition and Early Fetal Deaths:
Stillbirth definition: 24 weeks gestation (late fetal death after 23 completed weeks of gestation). Stillbirths of 24 weeks or more gestation are registered. Early fetal deaths/spontaneous abortions have no lower limit for inclusion on the register (earliest recorded go down to 8 weeks gestation. Autopsy rates were not given.

Exposure Data Availability:
Exposure information: information on maternal drug use, maternal and paternal diseases and occupations, outcomes of previous pregnancies is available. Folic acid supplementation before and during pregnancy is also collected.

Denominators and Controls Information:
Denominator date is obtained from the Office for National Statistics.

Addresses and Staff:
Mr David Tucker, Congenital Anomaly Register and Information Service (CARIS) Congenital Anomaly Register & Information Service, Level 3 West Wing, Singleton Hospital, Sketty Lane
Swansea, Wales, SA2 8QA

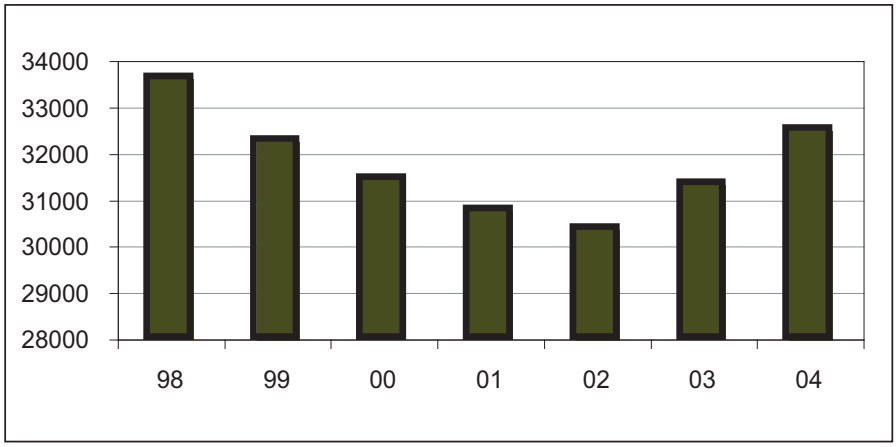
phone: 801 257 0566

Fax: 801 257 0572

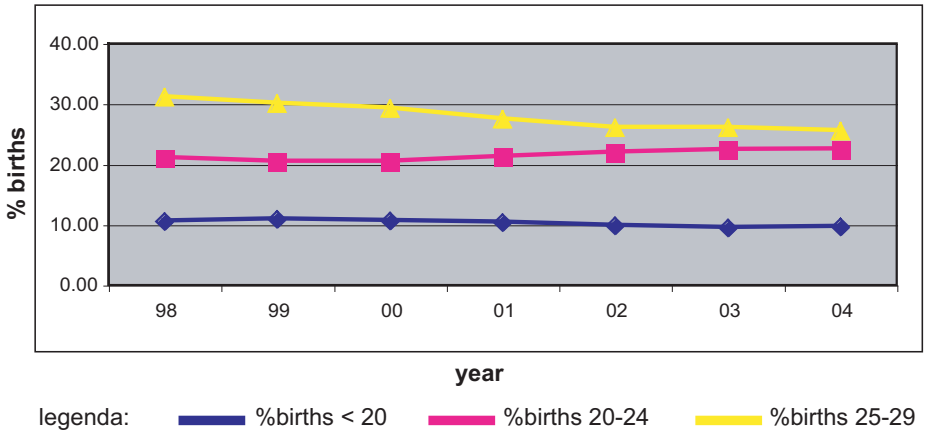
E-mail: mpalmer@utah.gov

Wales: CARIS

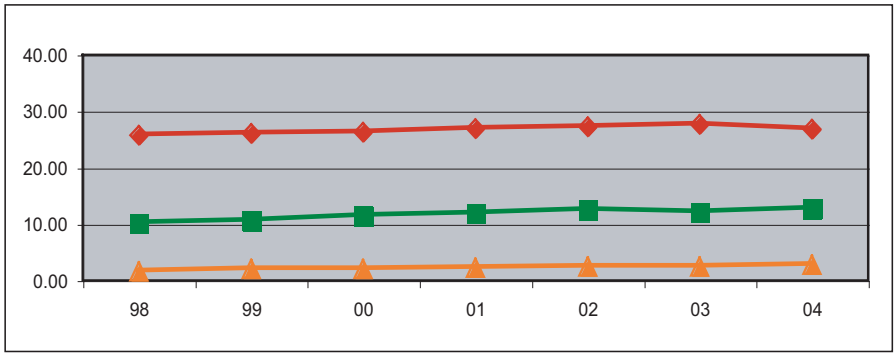
Total births by year



Percentage of births by maternal age



legenda: %births < 20 %births 20-24 %births 25-29



legenda: %births 30-34 %births 35-39 %births 40+

Wales: CARIS, 2004

Live births (LB)	32,325
Stillbirths (SB)	179
Total births	32,504
Number of terminations of pregnancy (ToP) for birth defects	179

Birth Defects	Number of cases			Rates*10,000
	LB	SB	ToP	Total rate
Anencephaly	0	0	21	6.43
Spina bifida	5	0	20	7.65
Encephalocele	1	0	2	0.92
Microcephaly	16	0	0	4.90
Arhinencephaly / Holoprosencephaly	1	0	5	1.84
Hydrocephaly	12	0	15	8.26
Anophthalmos	1	0	0	0.31
Microphthalmos	3	0	0	0.92
Unspecified Anophthalmos/ Microphthalmos	0	0	0	---
Anotia	0	0	0	0.00
Microtia	4	0	0	1.22
Unspecified Anotia/Microtia	0	0	0	---
Transposition of great vessels	10	0	1	3.37
Tetralogy of Fallot	10	0	0	3.06
Hypoplastic left heart syndrome	2	1	2	1.53
Coarctation of aorta	18	0	3	6.43
Choanal atresia, bilateral	2	0	0	0.61
Cleft palate without cleft lip	24	0	4	8.57
Cleft lip with or without cleft palate	31	0	4	10.71
Oesophageal atresia / stenosis with or without fistula	6	0	1	2.14
Small intestine atresia / stenosis	9	0	0	2.75
Anorectal atresia / stenosis	8	0	3	3.37
Undescended testis (36 weeks of gestation or later)	4	0	0	1.22
Hypospadias	54	0	0	16.52
Epispadias	0	0	0	0.00
Indeterminate sex	0	0	0	0.00
Renal agenesis	0	0	3	0.92
Cystic kidney	24	2	11	11.32
Bladder exstrophy	0	0	0	0.00
Polydactyly, preaxial	2	0	1	0.92
Total Limb reduction defects (include unspecified)	12	1	5	5.51
Transverse	6	0	2	2.45
Preaxial	0	0	2	0.61
Postaxial	0	0	0	0.00
Intercalary	1	0	0	0.31
Mixed	2	1	1	1.22
Unspecified	3	0	0	---
Diaphragmatic hernia	7	0	0	2.14
Omphalocele	5	1	5	3.37
Gastroschisis	29	0	2	9.49
Unspecified Omphalocele/Gastroschisis	0	0	1	---
Prune belly sequence	1	0	0	0.31
Trisomy 13	2	0	6	2.45
Trisomy 18	4	0	17	6.43
Down syndrome, all ages (include age unknown)	37	1	37	22.95
<20	3	0	1	12.76
20-24	4	0	1	6.84
25-29	9	1	1	13.28
30-34	6	0	10	18.33
35-39	10	0	14	57.46
40-44	5	0	9	160.55
45+	0	0	1	312.50
unknown	0	0	0	---

Wales: CARIS, Previous years rates 1998 - 2004

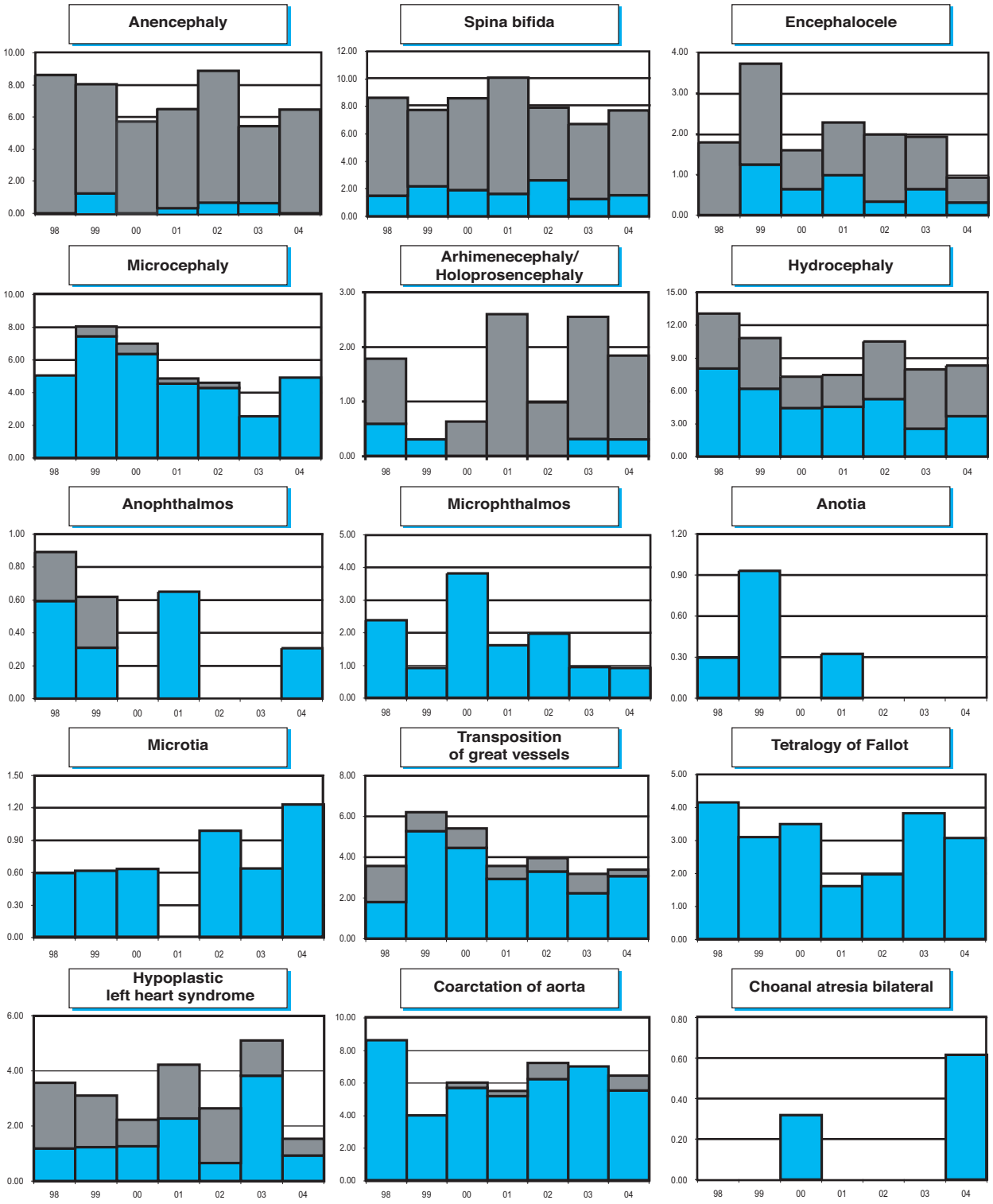
Birth prevalence rates: (LB+SB+TOP) * 10,000

	1974-79	1980-84	1985-89	1990-94	1995-99*	2000-04
Births					**65,886**	**156,423**
Anencephaly					8.35	6.58
Spina bifida					8.20	8.18
Encephalocele					2.73	1.73
Microcephaly					6.53	4.79
Arhinencephaly / Holoprosencephaly					1.06	1.73
Hydrocephaly					11.99	8.31
Anophthalmos					0.76	0.19
Microphthalmos					1.67	1.85
Unspecified Anophthalmos / Microphthalmos					---	---
Anotia					0.61	0.06
Microtia					0.61	0.70
Unspecified Anotia / Microtia					---	---
Transposition of great vessels					4.86	3.90
Tetralogy of Fallot					3.64	2.81
Hypoplastic left heart syndrome					3.34	3.13
Coarctation of aorta					6.37	6.46
Choanal atresia, bilateral					0.00	0.19
Cleft palate without cleft lip					9.87	8.18
Cleft lip with or without cleft palate					11.23	9.85
Oesophageal atresia / stenosis with or without fistula					0.76	1.34
Small intestine atresia / stenosis					3.95	2.37
Anorectal atresia / stenosis					4.71	4.48
Undescended testis (36 weeks of gestation or later)					3.19	2.69
Hypospadias					26.26	23.72
Epispadias					0.46	0.58
Indeterminate sex					0.15	0.64
Renal agenesis					6.83	4.86
Cystic kidney					10.93	10.23
Bladder exstrophy					0.61	0.19
Polydactyly, preaxial					0.46	0.70
Total Limb reduction defects (include unspecified)					10.93	8.69
Transverse					5.62	3.96
Preaxial					2.12	1.02
Postaxial					0.30	0.26
Intercalary					0.91	1.47
Mixed					0.61	1.15
Unspecified					---	---
Diaphragmatic hernia					3.49	3.84
Omphalocele					3.04	3.96
Gastroschisis					5.31	5.82
Unspecified Omphalocele / Gastroschisis					---	---
Prune belly sequence					0.15	0.06
Trisomy 13					1.97	2.49
Trisomy 18					5.01	4.54
Down syndrome, all ages (include age unknown)					18.67	19.82
<20					11.40	7.70
20-24					8.06	5.90
25-29					11.93	10.73
30-34					15.23	17.06
35-39					54.01	49.92
40-44					149.65	171.40
45+					0.00	277.78
unknown					---	---

* data include less than 5 years

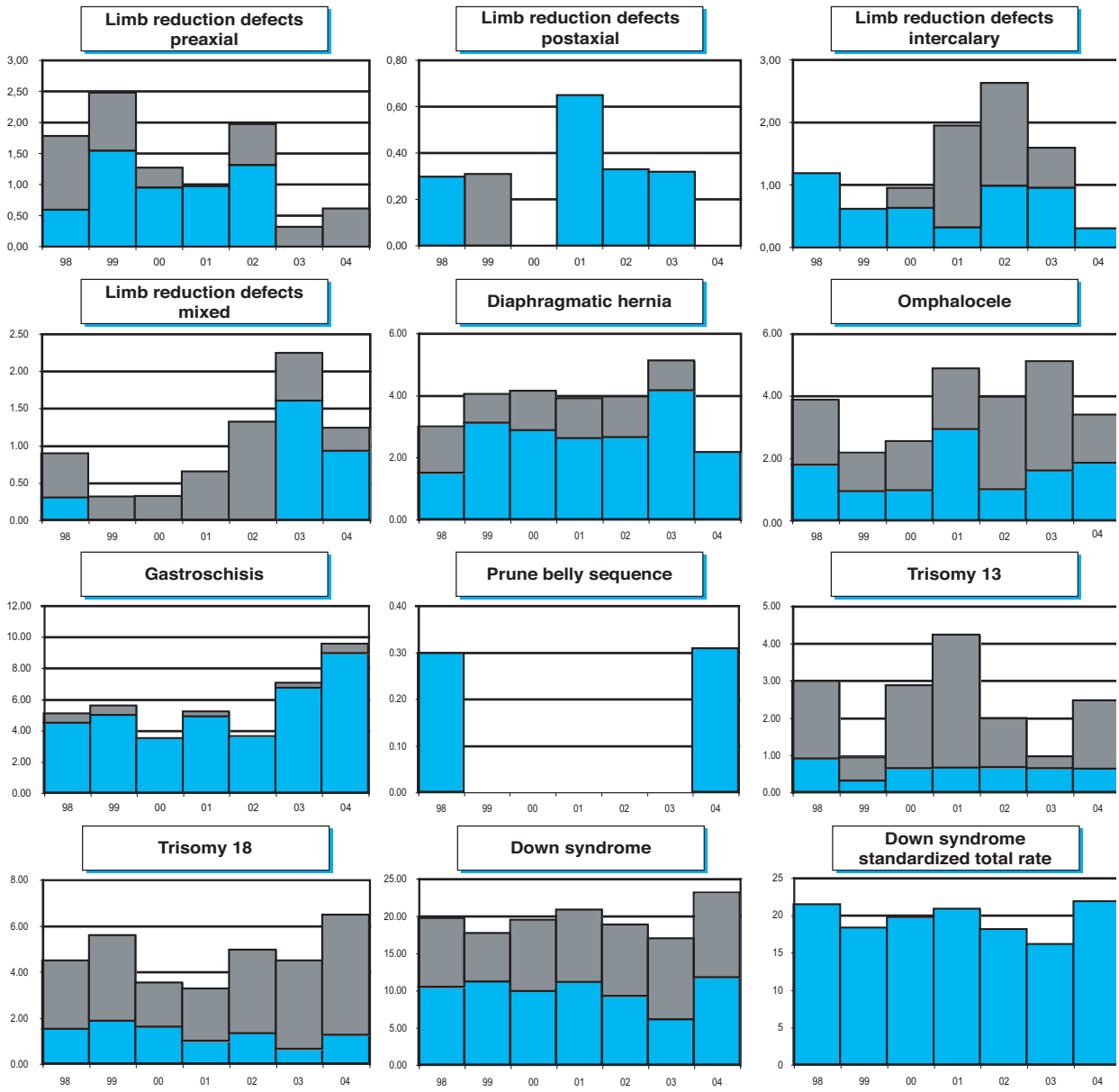
Wales: CARIS

'Time trends 1998-2004 (Birth prevalence rates per 10,000)



Note: L+S rates, ToP rates

Note: L+S rates, ToP rates



Note: L+S rates, ToP rates

Monitoring Systems, not contributing with Annual Data : description of the registry

Australia: National

Australian Congenital Malformation Monitoring System

History:
The mechanism for national monitoring of birth defects was established in 1981. However not all States and Territories collected birth defect data at this time. All the States and Territories now hold Perinatal Data Collections and birth defect information is collected as additional information. The national Programme became an associate member of the Clearinghouse in 1982 and full member in 1984. Australia has not contributed national data to the Clearinghouse for the last 2 years. Australian is currently under taking a national review of birth defect data collections and anticipates the resumption of data contribution to the ICBDSR in the foreseeable future.

Size and coverage:
All births in Australia are covered. Births have remained stable at approximately 250,000 annual births for the last 10 years. All births of 20 weeks or 400 grams gestation are registered.

Legislation and funding:
There is no national legislation requiring the reporting of birth defects at the national level. In some States, notification to their birth defect registry is required as part of their respective Public Health Acts. In most States and Territories, birth defect data is collected as part of another collection, and funding, if any is determined by the jurisdiction. The State and Territory Health Departments report to the central data custardian which receives funding from the Australian Institute of Health and Welfare.

Sources of ascertainment:
Birth defects are first notified to State and Territory birth defect registries from the perinatal data collection. The State and Territory birth defect registries operate independently and there is enormous variation in the breadth of notification sources and level of ascertainment. Other sources of notification may include death certificates, autopsies, hospital morbidity databases, notification from health professionals, cytogenetic and prenatal screening. At the minimum, State and Territory birth defect registries send electronic notification to the central data custardian annually.

Exposure information:
Currently not available.

Background information:
In the absence of national legislation, there is enormous variation is the scope, quality of data and ascertainment between the States and Territories. Australia is currently under taking a national data review with participation from all key stakeholders and jurisdictions. One of the stated out-comes of the data rewiew is to develop an agreed minimum data set, and, to expand the scope, improve the quality and consistency of data including standardized coding and agreed timeframes for reporting at the national and international levels.

Address for further information:
Elisabeth Sullivan, MD, Programme Director AIHW National Perinatal Statistics Unit, Sydney Children's Hospital, 2nd Floor, McNevin Dickson Building, Randwick Hospital Campus Randwick NSW 2031 Australia

Phone: 61-3-96162729

Fax: 61-2-93821025

E-mail: e.sullivan@unsw.edu.au

Website: www.npsu.unsw.edu.au

South Africa: SABDSS

South Africa Birth Defects Surveillance Systems (SABDSS).

History:
The Programme started in 1988 and became a full member of the Clearinghouse in 1992.

Size and coverage:
The Programme is hospital based covering 15 sentinel sites over the country with approximately 50,000 annual birth or 5% of all births in South Africa.

Legislation and Funding:
The Programme is funded by the Department of National Health. Participation in the Programme is voluntary.

Sources of ascertainment:
Reports are obtained from delivery units and paediatric units of the participating hospitals.

Exposure information:
No exposure information is routinely available.

Background information:
Total births for some participating hospitals are not accurately known.

Address for further information:
David Bourne-Rauf Sayed, Programme Director
Dept. of Public Health and Primary Health Care
University of Cape Town-Medical School
Observatoy 7925, Cape Town, South Africa

Phone: 27-21-4066482

Fax: 27-21-4066163

Email: db@phfm.vet.ac.za
rauf@phfm.vet.ac.za

United Arab Emirates
Congenital Abnormality Study Group

History:
Although started 1992, the Programme started continuos monitoring only in 1994. It is now an Associate Member of the Clearinghouse.

Size and coverage:
The Programme covers about 8,000 births a year occurring in three major hospitals of the Al Ain Medical District, situated in the eastern part of the Abu Dhabi Emirate. It has a population of about 270,000. Still births with a weight of only more than 500 gm are included.

Legislation and funding:
The Programme is funded by the Faculty of Medicine and Health Sciences of the UAE University.

Sources of ascertainment:
In each hospital, there is a neonatologist who examines, identifies abnormalities and records at birth in a form provided. The diagnosis is further assisted by a clinical geneticist/dysmorphologist and pediatricians.

Exposure information:
Some basic information on exposure such as maternal disease is collected in all cases.

Background information:
General epidemiological data for all births are available.

Addresses and Staff:
Lihadh Al-Gazali, MD, Programme Director Dept. of Paediatrics Faculty of Medicine & Health Sciences, United Arab Emirates University, PO Box 17666

Phone: 971-3-672000

Fax: 971-3-672022

E-mail: algazali@hotmail.com

USA: California

California Birth Defects Monitoring Program

History:
The California Birth Defects Monitoring Programme was established in 1983 to monitor rates and trends and conduct epidemiological investigations to find causes of birth defects. The Programme is funded through the California Department of Health Services and jointly operated with the March of Dimes Birth Defects Foundation. In 1997 the Centers for Disease Control designated the Programme one of eight Centers of Excellence in Birth Defects Research. The Programme is an associate member of the Clearinghouse.

Size and coverage:
The Programme operates a population-based registry among 56,000 births. The registry includes 8 counties whose birth defects rates and trends are representative of California and who reflect the state's racial/ethnic diversity.

Legislation and funding:
The Programme operates under statutory authority: Health and Safety Code, Division 102, Part 2, Chapter 1, Sections 103825-103855. State funding is appropriated each year through the state budget. The Programme also receives research grants from the National Institutes of Health and the Centers for Disease Control.

Sources of ascertainment:
Staff actively ascertain data at hospitals and genetic centers by reviewing logs and identifying children with structural birth defects (BPA 740-759) diagnosed prenatally through age 1. All diagnostic information is abstracted directly from medical records; registry files are cross-linked with vital statistics data to verify demographic information.

Exposure information:
Bilingual interviewers collect environmental exposure information through large, case-control interview studies. Exposures under investigation include nutrition, health status and family history, medications, lifestyle, and chemical exposures through hobbies and occupation. Study participants also submit biological samples for analysis of genetic factors that might be contributing. The Programme has published more than 200 articles reporting research and registry findings in medical and scientific journals.

Background information:
Registry data, research findings, publications, and a description of Programme activities are available on their website.

Addresses and Staff:
John A. Harris, MD, Program Director California Birth Defects Monitoring Program, 1917 Fifth Street Berkeley, CA 94710, USA

Phone: 1-510-849 5841

Fax: 1-510-849 5865

E-mail: jha@cbdmp.org

Website: www.cbdmp.org

Selection of papers by Programme Directors and their collaborators are reported as following. The details are sent from the Programme Directors only for the listed Monitoring Systems. The collaborative publications, made by two or more ICBDSR members in any context, are first shown and not repeated in the specific registry section.
Papers can be obtained contacting authors.

Collaborative Publications

Botto LD, Lisi A, Robert-Gnansia E, Erickson JD, Vollset SE, Mastroiacovo P, Botting B, Cocchi G, de Vigan C, de Walle H, Feijoo M, Irgens LM, McDonnell B, Merlob P, Ritvanen A, Scarano G, Siffel C, Metneki J, Stoll C, Smithells R, Goujard J. International retrospective cohort study of neural tube defects in relation to folic acid recommendations: are the recommendations working? BMJ. 2005; 330(7491):571.

Lisi A, Botto LD, Rittler M, Castilla E, Bianchi F, Botting B, De Walle H, Erickson Jd, Gatt M, De Vigan C, Irgens LM, Johnson W, Lancaster P, Merlob P, Mutchinick OM, Ritvanen A, Robert E, Scarano G, Stoll C, Mastroiacovo P. Sex and congenital malformations: An international perspective. American Journal of Medical Genetics 2005;134a:49–57. (Erratum in: American Journal of Medical Genetics 2005;134a:463.)

Botto LD, Robert-Gnansia E, Siffel C, Harris J, Borman B, Mastroiacovo P. Fostering international collaboration in birth defects research and prevention: a perspective from the International Clearinghouse for Birth Defects Surveillance and Research. Am J Public Health. 2006 May;96(5):774-80. Epub 2006 Mar 29.

Botto LD, Lisi A, Bower C, Canfield MA, Dattani N, De Vigan C, De Walle H, Erickson DJ, Halliday J, Irgens LM, Lowry RB, McDonnell R, Metneki J, Poetzsch S, Ritvanen A, Robert-Gnansia E, Siffel C, Stoll C, Mastroiacovo P. Trends of selected malformations in relation to folic acid recommendations and fortification: an international assessment. Birth Defects Res A Clin Mol Teratol. 2006; 76(10):693-705.

Australia: WABDR

Bourke J, Bower C, Blair E, Charles A, Knuiman M. The effect of terminations of pregnancy for fetal abnormalities on trends in mortality to one year of age in Western Australia. Paediatric and Perinatal Epidemiology 2005;19:284-93.

Bower C, Hansen M. Assisted reproductive technologies and birth outcomes: overview of recent systematic reviews. Reproduction, Fertility, and Development 2005;17:329-33.

Carey M, Mylvaganam A, Rouse I, Bower C. Risk factors for isolated talipes equinovarus in Western Australia, 1980-1994. Paediatric and Perinatal Epidemiology 2005;19:238-45.

Hansen M, Bower C, Milne E, de Klerk N, Kurinczuk JJ. Assisted reproductive technologies and the risk of birth defects-a systematic review. Human Reproduction 2005;20:328-38.

Jablensky AV, Morgan V, Zubrick SR, Bower C, Yellachich LA. Pregnancy, delivery, and neonatal complications in a population cohort of women with schizophrenia and major affective disorders. American Journal of Psychiatry 2005;162:79-91.

Colvin J, Sokol J, Dickinson JE, Bower C. Outcome of diaphragmatic hernia: a population based study in Western Australia. Pediatrics 2005;116:356-363.

Hadlow NC, Hewitt BG, I Dickinson JE, Jacoby P, Bower C. Community based screening for Down syndrome in the first trimester using ultrasound and maternal serum biochemistry. Brit J Obstet Gynaecol 2005;112:1561-4.

Payne J, Elliott EJ, D'Antoine H, O'Leary C, Mahony A, Haan E, Bower C. Health professionals' knowledge, practice and opinions about fetal alcohol syndrome and alcohol consumption in pregnancy. ANZ J Public Health 2005; 29:558-64.

Bower C, de Klerk N, Hickling S, Ambrosini G, Flicker L, Geelhoed E, Milne E. Assessment of the potential effect of incremental increases in folic acid intake on neural tube defects in Australia and New Zealand. ANZ J Public Health 2006; 30:369-74.

O'Leary P, Breheny N, Dickinson JE, Bower C, Goldblatt J, Hewitt B, Murch A, Stock R. First-trimester combined screening for Down syndrome and other fetal anomalies. Obstet Gynecol 2006;107:869-876.

Bower C, Miller M, Payne J, Serna P. Folate intake and the primary prevention of non-neural birth defects. Aust NZ J Public Health 2006;30:258-261.

Bittles AH, Bower C, Hussain R, Glasson EJ. The four ages of Down syndrome. European Journal of Public Health European J Public Health 2006; epub July 19.

Bower C, de Klerk N, Milne E, Bailey H, Ambrosini G, Hickling S, Geelhoed E, Flicker L, O'Leary P. Plenty

of evidence on mandatory folate fortification (letter). Aust NZ J Public Health 2006;30:81-81 (and Erratum, Aust NZ J Public Health 2006; 30:188).

Hadlow NC, Hewitt BG, Dickinson JE, Jacoby P, Bower C. Community-based screening for Downs Syndrome in the first trimester using ultrasound and maternal serum biochemistry. (letter). Brit J Obstet Gynaecol 2006; 113: 363-364.

O'Leary C. Bower C. Payne J. Elliott E. Fetal alcohol syndrome. [Letter] Australian Family Physician. 35(4):184, 2006

Bower C. Primary prevention of neural tube defects with folate in Western Australia: the value of the Western Australian Birth Defects Registry. Congenital Anomalies 2006; 46(2):118-21.

Elliott EJ, Payne J, Haan E, Bower C. Diagnosis of fetal alcohol syndrome and alcohol use in pregnancy: a survey of paediatricians' and trainees' knowledge, attitudes and practice. J Paed Child Health 2006;42:698-703

Hansen M., Sullivan E., Jequier A.M., Burton P., Junk S., Yovich J., Bower C. Practitioner reporting of birth defects in children born following ART: does it still have a role in surveillance of birth defects? Human Reproduction Hum Reprod. 2007 Feb;22(2):516-20. Epub 2006 Oct 4.

France: Paris

De Vigan C, Khoshnood B, Lhomme A, Vodovar V, Goujard J, Goffinet F. Prévalence et diagnostic prénatal des malformations en population parisienne: vingt ans de surveillance par le Registre des malformations congénitales de Paris. J Gynecol Obstet Biol Reprod 2005; 34: 8-16

Khoshnood B, De Vigan C, Vodovar V, Goujard J, Lhomme A, Bonnet D, Goffinet F. Trends in prenatal diagnosis, pregnancy termination, and perinatal mortality of newborns with congenital heart disease in France, 1983-2000: a population- based evaluation. Pediatrics 2005; 115: 95-101

Morris JK, De Vigan C, Mutton DE, Alberman E. Risk of a Down syndrome live birth in women 45 years of age and older. Prenat Diagn 2005; 25: 275-78

Garne E, Loane M, Dolk H, De Vigan C, Scxarano G, Tucker D, Stoll C, Gener B, Pierini A, Nelen V, Rosch C, Gillerot Y, Feijoo M, Tincheva R, Queisser-Luft A, Addor MC, Mosquera C, Gatt M, Barisic I. Prenatal diagnosis of severe structural congenital malformations in Europe. Ultrasound Obstet Gynecol 2005; 25: 6-11

Dolk H, Loane M, Garne E, De Walle H, Queisser-Luft A, De Vigan C, Addor MC, Gener B, Haeusler M, Jordan H, Tucker D, Stoll C, Feijoo M, Lillis D, Bianchi F. Trends and geographic inequalities in the prevalence of Down syndrome in Europe, 1980-1999. Rev Epidemiol Sante Pub 2005, 53 :2S887-2S95

Khoshnood B, De Vigan C, Vodovar V, Bréart B, Goffinet F, Blondel B. Advances in medical technology and creation of disparities: the case of Down syndrome. Am J Public Health 2006; 96: 2139-44

Khoshnood B, De Vigan C, Blondel B, Vodovar V, Garel M, Goffinet F. Women's interpretation of an abnormal result on measurement of nuchal tanslucency and maternal serum screening for prenatal testing of Down syndrome. Ultrasound Obstet Gynecol 2006; 28: 242-8

De Vigan C, Khoshnood B, Vodovar V, Chausson M, Mahamadaly S, Cadio E, Goffinet F. Epidémiologie des malformations cardiaques congénitales. Pourrquoi réaliser une étude prospective en population (EPICARD)? Médecine Foetale et Echographie en Gynécologie 2006; 68: 20-25.

Germany: Saxony-Anhalt

Poetzsch, Simone; Hoyer-Schuschke, Jana; Koehn, Andrea; Vogt, Cornelia; Goetz, Dorit; Haase, Marion Jahresbericht des Bundeslandes Sachsen-Anhalt zur Häufigkeit von congenitalen Fehlbildungen und Anomalien sowie genetisch bedingten Erkrankungen 2005. ISSN 1861-3535, 87 S.

Ludwig, A. K.; Katalinic, A.; Steinbicker, Volker; Diedrich, K.; Ludwig, M.Antenatal care in singleton pregnancies after ICSI as compared to spontaneous conception : data from a prospective controlled cohort study in Germany. In: Human reproduction, ISSN 0268-1161, Bd. 21 (2006), 3, S. 713-720

Poetzsch, Simone; Hoyer-Schuschke, Jana; Seelig, Manuela; Steinbicker, Volker. Knowledge among young people about folic acid and its importance during pregnancy : a survey in the Federal State of Saxony-Anhalt (Germany). In: Journal of applied genetics. ISSN 1234-1983, Bd. 47 (2006), 2, S. 187-190

Stadler, Sonja C.; Polanetz, Roman; Maier, Esther M.; Heidenreich, Sylvia C.; Niederer, Birgit; Mayerhofer, Peter U.; Lagler, Florian; Koch, Hans-Georg; Santer, René; Fletcher, Janice M.; Ranieri, Enzo; Das, Anibh M.; Spiekerkötter, Ute; Schwab, Karl O.; Poetzsch, Simone; Marquardt, Iris; Hennermann, Julia B.; Knerr, Ina; Mercimek-

Mahmutoglu, Saadet; Kohlschmidt, Nicolai; Liebl, Bernhard; Fingerhut, Ralph; Olgemöller, Bernhard; Muntau, Ania C.; Roscher, Adelbert A.; Röschinger, Wulf. Newborn screening for 3-methylcrotonyl-CoA carboxylase deficiency : population heterogeneity of MCCA and MCCB mutations and impact on risk assessment In: Human mutation : variation, databases, and disease. ISSN 1059-7794, Bd. 27 (2006), 8, S. 748-759

Hoyer-Schuschke, Jana; Poetzsch, Simone; Gerloff, Claudia; Krause, Hardy; Kawa, Susan; Goetz, Dorit; Haase, Marion; Vogt, Cornelia; Koehn, Andrea. Gastroschisis - eine Fehlbildung mit steigender Prävalenz? In: Ärzteblatt Sachsen-Anhalt: ISSN 0938-9261, Bd. 17 (2006), 5, S. 64-67

Koehn, Andrea; Poetzsch, Simone; Hoyer-Schuschke, Jana. Kenntnisse über Mikronährstoffe : Ergebnisse einer repräsentativen Befragung unter Schülern in Sachsen-Anhalt In: Ernährungs-Umschau : ISSN 0174-0008, Bd. 53 (2006), 4, S. 130-134

Seelig, Manuela; Hoyer-Schuschke, Jana; Koehn, Andrea; Poetzsch, Simone. Kenntnisstand von SchülerInnen in Sachsen-Anhalt zum Thema "Folsäure und Schwangerschaft". In: Päd : praktische Pädiatrie, ISSN 0949-7641, Bd. 12 (2006), 3, S. 197-203

Koehn, Andrea. Mikronährstoffe : Kenntnisstand von Schülerinnen und Schülern In: Praxis der Naturwissenschaften - Biologie in der Schule. ISSN 1617-5697, Bd. 55 (2006), 5, S. 44-46

Heinz, Judith; Kästner, Steffi; Seewald, M.; Poetzsch, Simone. Unzureichende Umsetzung der perikonzeptionellen Folsäureeinnahme zur Prävention von Neuralrohrdefekten. In: Geburtshilfe und Frauenheilkunde : German journal of obstetrics and gynecology. ISSN 0016-5751, Bd. 66 (2006), 2, S. 156-162

Hungary

Acs N, Banhidy F, Puho E, Czeizel AE. Maternal influenza during pregnancy and risk of congenital abnormalities in offspring. Birth Defects Res A Clin Mol Teratol. 2005; 73(12):989-96.

Acs N, Banhidy F, Puho E, Czeizel AE. Population-based case-control study of mebendazole in pregnant women for birth outcomes. Congenit Anom (Kyoto). 2005; 45(3):85-8.

Acs N, Puho E, Banhidy F, Czeizel AE. Association between bronchial asthma in pregnancy and shorter gestational age in a population-based study. J Matern Fetal Neonatal Med. 2005; 18(2):107-12.

Czeizel AE, Dudas I, Puho E. Short-term paracetamol therapy during pregnancy and a lower rate of preterm birth. Paediatr Perinat Epidemiol. 2005; 19(2):106-11.

Kazy Z, Puho E, Czeizel AE. Effect of vaginal metronidazole + miconazole treatment during pregnancy for gestational age and birth weight in a population-based study. Arch Gynecol Obstet. 2005; 272(4):294-7.

Kazy Z, Puho E, Czeizel AE. Parenteral polymyxin B treatment during pregnancy. Reprod Toxicol. 2005; 20(2):181-2.

Kazy Z, Puho E, Czeizel AE. Population-based case-control study of oral ketoconazole treatment for birth outcomes. Congenit Anom (Kyoto). 2005; 45(1):5-8.

Kazy Z, Puho E, Czeizel AE. Teratogenic potential of vaginal metronidazole treatment during pregnancy. Eur J Obstet Gynecol Reprod Biol. 2005; 123(2):174-8.

Kazy Z, Puho E, Czeizel AE. The possible association between the combination of vaginal metronidazole and miconazole treatment and poly-syndactyly Population-based case-control teratologic study. Reprod Toxicol. 2005; 20(1):89-94.

Metneki J, Czeizel AE. Increasing total prevalence rate of cases with Down syndrome in Hungary. Eur J Epidemiol. 2005; 20(6):525-35.

Metneki J, Puho E, Czeizel AE. Maternal diseases and isolated orofacial clefts in Hungary. Birth Defects Res A Clin Mol Teratol. 2005; 73(9):617-23.

Nielsen GL, Norgard B, Puho E, Rothman KJ, Sorensen HT, Czeizel AE. Risk of specific congenital abnormalities in offspring of women with diabetes. Diabet Med. 2005; 22(6):693-6.

Norgard B, Puho E, Czeizel AE, Skriver MV, Sorensen HT. Aspirin use during early pregnancy and the risk of congenital abnormalities: a population-based case-control study. Am J Obstet Gynecol. 2005; 192(3):922-3.

Petik D, Puho E, Czeizel AE. Evaluation of maternal infusion therapy during pregnancy for fetal development. Int J Med Sci. 2005; 2(4):137-42.

Puho E, Metneki J, Czeizel AE. Maternal employment status and isolated orofacial clefts in Hungary. Cent Eur J Public Health. 2005; 13(3):144-8.

Vogt G, Puho E, Czeizel AE. A population-based case-control study of isolated anophthalmia and

microphthalmia. Eur J Epidemiol. 2005; 20(11):939-46.
Vogt G, Puho E, Czeizel AE. A population-based case-control study of isolated ocular coloboma. Ophthalmic Epidemiol. 2005; 12(3):191-7.

Vogt G, Puho E, Czeizel AE. Population-based case-control study of isolated congenital cataract. Birth Defects Res A Clin Mol Teratol. 2005; 73(12):997-1005.

Acs N, Banhidy F, Horvath-Puho E, Czeizel AE. Maternal panic disorder and congenital abnormalities: a population-based case-control study. Birth Defects Res A Clin Mol Teratol. 2006; 76(4):253-61.

Acs N, Banhidy F, Horvath-Puho E, Czeizel AE. Population-based case-control study of the common cold during pregnancy and congenital abnormalities. Eur J Epidemiol. 2006; 21(1):65-75.

Acs N, Banhidy F, Puho E, Czeizel AE. A possible dose-dependent teratogenic effect of ergotamine. Reprod Toxicol. 2006; 22(3):551-2.

Acs N, Banhidy F, Puho E, Czeizel AE. Pregnancy complications and delivery outcomes of pregnant women with influenza. J Matern Fetal Neonatal Med. 2006; 19(3):135-40.

Acs N, Banhidy F, Puho E, Czeizel AE. Teratogenic effects of vaginal boric acid treatment during pregnancy. Int J Gynaecol Obstet. 2006; 93(1):55-6.

Acs N, Banhidy F, Puho EH, Czeizel AE. Acute respiratory infections during pregnancy and congenital abnormalities: a population-based case-control study. Congenit Anom (Kyoto). 2006; 46(2):86-96.

Banhidy F, Acs N, Horvath-Puho E, Czeizel AE. Maternal severe migraine and risk of congenital limb deficiencies. Birth Defects Res A Clin Mol Teratol. 2006; 76(8):592-601.

Banhidy F, Acs N, Horvath-Puho E, Czeizel AE. Pregnancy complications and delivery outcomes in pregnant women with severe migraine. Eur J Obstet Gynecol Reprod Biol. 2006 Nov 8; [Epub ahead of print]

Banhidy F, Acs N, Puho E, Czeizel AE. Association between maternal panic disorders and pregnancy complications and delivery outcomes. Eur J Obstet Gynecol Reprod Biol. 2006; 124(1):47-52.

Banhidy F, Acs N, Puho E, Czeizel AE. Association between maternal panic disorders and pregnancy complications and delivery outcomes. Eur J Obstet Gynecol Reprod Biol. 2006; 124(1):47-52.

Banhidy F, Acs N, Puho E, Czeizel AE. Pregnancy complications and delivery outcomes of pregnant women with common cold. Cent Eur J Public Health. 2006; 14(1):10-4.

Banhidy F, Acs N, Puho EH, Czeizel AE. Maternal kidney stones during pregnancy and adverse birth outcomes, particularly congenital abnormalities in the offspring. Arch Gynecol Obstet. 2006 Nov 10; [Epub ahead of print]

Banhidy F, Acs N, Puho EH, Czeizel AE. Maternal urinary tract infection and related drug treatments during pregnancy and risk of congenital abnormalities in the offspring. BJOG. 2006; 113(12):1465-71.

Banhidy F, Puho E, Acs N, Czeizel AE. Possible association between maternal recurrent orofacial herpes in pregnancy and a lower rate of preterm birth. J Matern Fetal Neonatal Med. 2006; 19(9):537-42.

Czeizel AE, Puho E, Acs N, Banhidy F. Inverse association between severe nausea and vomiting in pregnancy and some congenital abnormalities. Am J Med Genet A. 2006; 140(5):453-62.

Czeizel AE, Puho E. Maternal use of nutritional supplements during the first month of pregnancy and decreased risk of Down's syndrome: case-control study. Nutrition. 2005; 21(6):698-704; discussion 774.

Czeizel AE, Puho EH, Banhidy F. No association between periconceptional multivitamin supplementation and risk of multiple congenital abnormalities: a population-based case-control study. Am J Med Genet A. 2006; 140(22):2469-77.

Dudas I, Puho E, Czeizel AE. Population-based case-control study of oxoline acid use during pregnancy for birth outcomes. Congenit Anom (Kyoto). 2006; 46(1):39-42.

Kazy Z, Puho EH, Czeizel AE. The possible preterm birth preventive effect of ampicillin during pregnancy. Arch Gynecol Obstet. 2006; 274(4):215-21.

Norgard B, Norgaard M, Czeizel AE, Puho E, Sorensen HT. Maternal herpes labialis in pregnancy and neural tube defects. Dev Med Child Neurol. 2006; 48(8):674-6.

Somoskovi A, Bartfai Z, Tamasi L, Kocsis J, Puho E, Czeizel AE. Population-based case-control study of allergic rhinitis during pregnancy for birth outcomes. Eur J Obstet Gynecol Reprod Biol. 2007; 131(1):21-7. Epub 2006 Jan 25.

Tamasi L, Somoskovi A, Muller V, Bartfai Z, Acs N, Puho E, Czeizel AE. A population-based case-control study on the effect of bronchial asthma during pregnancy for congenital abnormalities of the offspring. J Asthma. 2006; 43(1):81-6.

Vogt G, Horvath-Puho E, Czeizel AE. A population-based case-control study of isolated primary congenital glaucoma. Am J Med Genet A. 2006; 140(11):1148-55.

Vogt G, Horvath-Puho E, Czeizel E. [A population-based case-control study of isolated congenital cataract] Orv Hetil. 2006; 147(23):1077-84. Hungarian.

Vogt G, Szunyogh M, Czeizel AE. Birth characteristics of different ocular congenital abnormalities in Hungary. Ophthalmic Epidemiol. 2006; 13(3):159-66.

Wogelius P, Horvath-Puho E, Pedersen L, Norgaard M, Czeizel AE, Sorensen HT. Maternal use of oral contraceptives and risk of hypospadias – a population-based case-control study. Eur J Epidemiol. 2006; 21(10):777-81.

Iran: TROCA

Dastgiri S, Imani S, Klankesh L, Barzegar M, Heidarzaeh M. Congenital anomalies in Iran: a cross-sectional study on 1574 cases in the North-West country. Child: care, health and development, 2007, 33(3):257-261

Ireland: Dublin

Busby A, Armstrong B, Dolk H, Armstrong N, Haeusler M, Berghold A, Gillerot Y, Baguette A, Gjergja R, Barisic I, Christiansen M, Goujard J, Steinbicker V, Rosch C, McDonnell R, Scarano G, Calzolari E, Neville A, Cocchi G, Bianca S, Gatt M, Walle HD, Braz P, Latos-Bielenska A, Gener B, Portillo I, Addor MC, Abramsky L, Ritvanen A, Robert-Gnansia E, Daltveit AK, Aneren G, Olars B, Edwards G. Preventing neural tube defects in Europe: a missed opportunity. Reprod Toxicol. 2005: 20(3):393-40

Busby A, Ritvanen A, Dolk H, Armstrong N, De Walle H, Riaño-Galán I, Gatt M, McDonnell R, Nelen V, Stone D. Survey of informed consent for registration of congenital anomalies in Europe. Br Med J 2005:331;140-141

Busby A, Abramsky L, Helen Dolk H, Ben Armstrong B, a EUROCAT Folic Acid Working Group. Preventing neural tube defects in Europe: a population based study. Br Med J 2005: 330; 574-575.

Israel : IBDMS

P. Merlob, O. Sapir, J. Sulkes, B. Fisch.The prevalence of major congenital malformations during two periods of time, 1986 - 1994 and 1995 - 2002 in newborns conceived by assisted reproduction technology Eur J Med Genet, 48:5-11, 2005.

M. Osovsky, P. Merlob.Trichomegaly in two sisters with synophrys in the older sibling. Am J Med Genet, 136A:398, 2005.

B. Bar-Oz, M. Clementi, E. Di Giantonio, R. Greenberg, M. Beer, P. Merlob, J. Arnon, A. Ornoy, D.M. Zimmerman, M. Berkovitch.Metamizol (dipyrone, optalgin) in pregnancy, is it safe? A prospective comparative study. Eur J Obstet Gynecol Reprod Biol, 119:176-179, 2005.

I. Gull, I. Wolman, P. Merlob, A.J. Jaffa, J.B. Lessing, Y. Yaron. Nomograms for the sonographic measurement of the fetal philtrum and chin. Fetal Diag Ther, 20:127-131, 2005.

R. Mashiach, M. Davidovits, B. Eisenstein, D. Kidron, M. Kovo, J. Shalev, M. Merlob, D. Vardimon, Z. Efrat, I. Meizner. Fetal hyperechogenic kidney with normal amniotic fluid volume: a diagnostic dilemma. Prenatal Diagn, 25:533-558, 2005.

G. Dubnov, R. Fogelman, P. Merlob.Prolonged QT interval in a newborn to a fluoxetine-treated mother.Arch Dis Child, 90:972-973, 2005.

R. Levinson-Castiel, P. Merlob, N. Linder, L. Sirota, G. Klinger.Neonatal abstinence syndrome following in utero exposure to selective serotonin reuptake inhibitors in term infants.Arch Pediatr Adolesc Med, 160; 173-176, 2006.

D. Bader, M. Grun, P. Merlob.Correspondence concerning Hunter and Yotsuyangi's "the external ear: more attention to detail may aid syndrome and contribute answers to embryological questions".Am J Med Genet, 140:798; 2006 (letter).

Mexico: RYVEMCE

OM Mutchinick, JJ Morales, JC Zenteno, C Fernández del Castillo. "A rare case of gonadal agenesis with paramesonephric derivaties in a patient with normal female karyotype. Fertility & Sterility, 83: 201-204, 2005

JC Zenteno, V Verdón-Zapata, S Kofman-Alfaro, OM Mutchinick. "Isolated ectrodactyly caused by an heterozygous missense mutation in the transactivation domain of *TP63*" Am J Med Genet, 134:74-76, 2005

AJ González, JJ Morales, L Luna, J Arteaga, OM Mutchinick. "Beguinner's guide to genetics: cancer genetics", StudentBMJ,13(2):52-53, 2005

OM Mutchinick, H Arrieta, J Morales, J Arteaga, R Macías, L Luna, N Monroy, AJ González. "Beguinner's guide to genetics: past, present and future" StudentBMJ, 13(3):96-97, 2005

JL Guéant, RM Guéant-Rodriguez, R Debard, S Thirion, JP Bronowicki, NW Chabi, A Sanni, G Anello, P Bosco, C Romano, E Amouzou, HR Arrieta, BE Sánchez, A Romano, B Herbeth, JC Guilland, OM Mutchinick. "Prevalence of methylenetetrahydrofolate reductase 677T and 1298C alleles and folate status: a comparative study among Mexican, West African and West European populations. Am J Clin Nutr, 83:701-707, 2006

Northern Netherlands

Leegte B, Van der Hout AH, Deffenbaugh AM, Bakker MK, Mulder IM, Ten Berge A, Leenders EP, Wesseling J, De Hullu J, Hoogerbrugge N, Ligtenberg MJL, Ardern-Jones A, Bancroft E, Salmon A, Barwell J, Eeles R, Oosterwijk JC. Phenotypic expression of double heterozygosity for BRCA1 and BRCA2 germline mutations. J. Med Genet 2005; 42:e20.

Busby A, Abramsky L, Dolk H, Armstrong B, a Eurocat Folic Acid Working Group. Preventing neural tube defects in Europe: population based study. BMJ 2005; Volume 330, 12 March 2005.

Meijer WM, De Walle HEK, Kerstjens-Frederikse WS, De Jong-van den Berg LTW. Folic acid sensitive birth defects in association with intrauterine exposure to folic acid antagonists. Reproductive Toxicology 2005; 20: p. 203-207.

Busby A, Ritvanen A, Dolk H, Armstrong N, De Walle H, Riano-Galán I, Gatt M, McDonnell R, Nelen V, Stone D. Survey of informed consent for registration of congenital anomalies in Europe. BMJ 2005; Volume 331: 140-1.

Busby A, Armstrong B, Dolk H, Armstrong N, Haeusler M, Berghold A, Gillerot Y, Baguette A, Gjergja R, Basiric I, Christiansen M, Goujard J, Steinbicker V, Rösch C, McDonnell R, Scarano G, Calzolari E, Neville A, Cocchi G, Bianca S, Gatt M, De Walle H, Braz P, Latos-Bielenska A, Gener B, Portillo I, Addor M, Abramsky L, Ritvanen A, Robert-Gnansia E, Kjersti Daltveit A, Aneren G, Olars B, Edwards G. Preventing neural tube defects in Europe: A missed opportunity. Reproductive Toxicology 2005; Volume 20: 393-402.

Wiesel A, Queisser-Luft A, Clementi M, Bianca S, Stoll C, the EUROSCAN Study Group. Prenatal Detection of Congenital Renal Malformations by Fetal Ultrasonographic Examination: An Analysis of 709,030 Births in 12 European Countries. European Journal of Medical Genetics 48 (2005) 131-144.

Meijer WM, De Walle HEK. Verschillen in foliumzuurbeleid en prevalentie van neuralebuisdefecten in Europa; aanbevelingen voor voedselverrijking in een EUROCAT-rapport. Ned. Tijdschr. Geneeskunde 2005; 12 november 149(46) p 2561-2564.

Dolk H, Loane M, Garne E, De Walle H, Queisser-Luft A, De Vigan C, Addor MC, Gener B, Haeusler M, Jordan H, Tucker D, Stoll C, Feijoo M, Lillis D, Bianchi F. Trends and geographic inequalities in the prevalence of Down syndrome in Europe, 1980-1999. Rev Epidemiol Sante Publique, 2005. 53: 2887-2895.

Bakker MK, Jentink J, Vroom F, Van den Berg PB, De Walle HEK, De Jong-Van Den Berg LTW. Drug prescription patterns before, during and after pregnancy for chronic, occasional and pregnancy-related drugs in the Netherlands. BJOG 2006; 113: 559-568.

Meijer WM, De Jong-Van Den Berg LTW, Van Den Berg MD, Verheij JBGM, De Walle HEK. Clomiphene and Hypospadias on a Detailed Level: Signal or Chance? Birth Defects Research (Part A) 2006; 76:249-252.

Meijer WM, Cornel MC, Dolk H, De Walle HEK, Armstrong NC, De Jong-van den Berg LTW. EUROCAT Working Group. The potential of the European network of congenital anomaly registers (EUROCAT) for drug safety surveillance: a descriptive study. Pharmacoepidemiology and Drug Safety 2006; 15: 675-682.

De Jong-Van den Berg LTW, Bakker MK, De Walle HEK. Duidelijk verhoogd risico op congenitale afwijkingen door het gebruik van angiotensineconverterend-enzym (ACE)-remmers in de zwangerschap. Ned. Tijdschr. Geneeskd. 2006; 7 oktober;150(40): p. 2222. (ingezonden brief).

Calzolari E, Pierini A, Astolfi G, Bianchi F, Neville A, Riveri F and a EUROCAT Working Group, Ässociated Anomalies in Multi-Malformed Infants with Cleft Lip and Palate: An Epidemiological Study Based on Nearly 6 Million Births in 23 EUROCAT Registries", American Journal of Medical Genetics. (in press)

Vroom F, De Walle HEK, Van de Laar MAJF, Brouwers RBJ, De Jong-Van den Berg LTW. Disease-Modifying Antirheumatic Drugs in Pregnancy. Drug Safety 2006;29(10).

Niessen RC, Berends MJW, Wu Y, Sijmons RH, Hollema H, Ligtenberg MJL, De Walle HEK, De Vries EGE, Karrenbeld A, Buys CHC, Van der Zee AGJ, Hofstra RMW, Kleibeuker JH. Identification of mismatch repair gene mutations in young patients with colorectal cancer and in patients with multiple tumours associated with hereditary non-polyposis colorectal cancer. 2006 Gut; 55: 1781-1788.

Norway

Ahlberg N, Vangen S. Pregnancy and birth in multicultural Norway. (In Norwegian) Tidsskr Nor Laegeforen 2005;125:586-8.

Berle JO, Mykletun A, Daltveit AK, Rasmussen S, Holsten F, Dahl AA. Neonatal outcomes in offspring of women with anxiety and depression during pregnancy. A linkage study from The Nord-Trondelag Health Study (HUNT) and Medical Birth Registry of Norway. Arch Women Ment Health 2005;8(3):181-9.

Berle JO. The Pregnancy and Post Partum Periods Studies of Fetal Growth Anxiety and Depression. Thesis. University of Bergen 2005. ISBN: 82-308-0052-9.

Busby A, Armstrong B, Dolk H, Armstrong N, Haeusler M, Berghold A, Gillerot Y, Baguette A, Gjergia R, Barisic I, Christiansen M, Goujard J, Steinbicker V, Rosch C, McDonnell R, Scarano G, Calzolari E, Neville A, Cocchi G, Bianca S, Gatt M, Walle HD, Braz P, Latos-Bielenska A, Gener B, Portillo I, Addor MC, Abramsky L, Ritvanen A, Robert-Gnansia E, Daltveit AK, Aneren G, Olars B, Edwards G. Preventing neural tube defects in Europe: A missed opportunity. Reprod Toxicol 2005;20(3):393-402.

Chen KT, Eskild A, Bresnahan M, Stray-Pedersen B, Sher A, Jenum PA. Previous maternal infection with Toxoplasma gondii and the risk of fetal death. Am J Obstet Gynecol 2005;193:443-9.

Christiansen M, Tonder N, Larsen LA, Andersen PS, Simonsen H, Oyen N, Kanters JK, Jacobsen JR, Fosdal I, Wettrell G, Kjeldsen K. Mutations in the HERG K+-ion channel: a novel link between long QT syndrome and sudden infant death syndrome. Am J Cardiol 2005;95(3):433-4.

Dahl J, Myhr KM, Daltveit AK, Hoff JM, Gilhus NE. Pregnancy, delivery, and birth outcome in women with multiple sclerosis. Neurology. 2005;65(12):1961-3.

Eide MG, Oyen N, Skjaerven R, Irgens LM, Bjerkedal T, Nilsen ST. Breech Delivery and Intelligence: A Population-Based Study of 8,738 Breech Infants. Obstet Gynecol 2005;105:4-11.

Eide MG, Oyen N, Skjaerven R, Nilsen ST, Bjerkedal T, Tell GS. Size at Birth and Gestational Age as Predictors of Adult Height and Weight. Epidemiology 2005;16:175-181.

Eide MG. Associations of Perinatal Conditions with Adult Body Size and intelligence. A register-based Cohort Study in Norway 1967-1999. Thesis. University of Bergen 2005. ISBN: 82-308-0086-3.

Eide MG. Influences of perinatal conditions on adult body size and intellectual performance: a register-based cohort study. Norsk epidemiologi 2005;15:29-40.

Eskild A, Jenum PA, Bruu AL. Maternal antibodies against cytomegalovirus in pregnancy and the risk of fetal death and low birth weight. Acta Obstet Gynecol Scand 2005;84(11):1035-41.

Eskild A, Bruu AL, Stray-Pedersen B, Jenum P. Epstein-Barr virus infection during pregnancy and the risk of adverse pregnancy outcome. BJOG 2005;112(12):1620-4.

Fossa SD, Magelssen H, Melve K, Jacobsen AB, Langmark F, Skjaerven R. Parenthood in Survivors After Adulthood Cancer and Perinatal Health in Their Offspring: A Preliminary Report. J Natl Cancer Inst, Monographs 2005;No.34:77-82.

Haga H-J, Gram Gjesdal C, Irgens LM, Ostensen M. Reproduction and gynaecological manifestations in women with primary Sjögren's syndrome: a case-control study. Scand J Rheumatol 2005;34:45–48.

Haga H-J, Gjesdal CG, Koksvik HS, Skomsvoll JF, Irgens LM, Ostensen M. Pregnancy Outcome in Patients with Primary Sjögren's Syndrome. A Case-Control Study. J Rheumatol 2005;32(9):1734-1736.

Harville EW, Wilcox AJ, Lie RT, Vindenes H, Abyholm F. Cleft Lip and Palate versus Cleft Lip Only: Are They Distinct Defects? Am J Epidemiol 2005;162(5):448-53.

Hoff JM, Gilhus NE, Daltveit AK. Pregnancies and deliveries in patients with Charcot-Marie-Tooth disease. Neurology. 2005;64:459-62

Irgens LM, Kristensen P, Vatten LJ. Early exposure and later disease. Preface. (In Norwegian). Norsk Epidemiologi 2005;15:3-4.

Johnsen SL, Rasmussen S, Sollien R, Kiserud T. Fetal age assessment based on femur length at 10 25 weeks of gestation, and reference ranges for femur length to head circumference ratios. Acta Obstet Gynecol Scand 2005:84:725–33.

Kristensen P, Bjerkedal T, Irgens LM, Gravseth HM, Brevik JI. Impact of life course determinants on work participation among young Norwegian men. Norsk Epidemiologi 2005;15:65-74.

Kvestad E, Kvaerner KJ, Roysamb E, Tambs K, Harris JR, Magnus P. Heritability of recurrent tonsillitis. Arch Otolaryngol Head Neck Surg 2005;131:383-7.
Lie RT, Lyngstadaas A, Orstavik KH, Bakketeig LS, Jacobsen G, Tanbo T. Birth defects in children conceived by ICSI compared with children conceived by other IVF-methods; a meta-analysis. Int J Epidemiol 2005;34:696-701.

Lie RT, Skjaerven R. Follow-up studies of children with birth defects in the Medical Birth Registry of Norway: A review. Norsk Epidemiologi 2005;15:21-24.

Lippert T, Skjaerven R, Salvesen KA. Why do some women only give birth to boys or to girls? (In Norwegian) Tidsskr Nor Laegeforen 2005;125:3414-7.

Magelssen H, Haugen TB, von During V, Melve KK, Sandstad B, Fossa SD. Twenty Years Experience with Semen Cryopreservation in Testicular Cancer Patients: Who Needs It? European Urology 2005;48:779-85.

Markestad T, Kaaresen PI, Ronnestad A, Reigstad H, Lossius K, Medbo S, Zanussi G, Engelund IE, Skjaerven R, Irgens LM. Norwegian Extreme Prematurity Study Group. Early death, morbidity, and need of treatment among extremely premature infants. Pediatrics. 2005;115:1289-98.

Moster D, Lie RT, Markestad T. Sense and sensibility in delivery care. (In Norwegian). Tidsskr Nor Laegeforen 2005;125(20):2818-20.

Nafstad P, Samuelsen SO, Irgens LM, Bjerkedal T. Complications during pregnancy and risk of asthma among Norwegians born 1967-1993. (In Norwegian) Norsk Epidemiologi 2005;15:47-54.

Nordby K-C, Andersen Aa, Irgens LM, Kristensen P. Indicators of mancozeb exposure in relation to thyroid cancer and neural tube defects in farmers' families. Scand J Work Environ Health 2005;31(2):89–96.

Nystad W, Roysamb E, Magnus P, Tambs K, Harris JR. A comparison of genetic and environmental variance structures for asthma, hay fever and eczema with symptoms of the same diseases: a study of Norwegian twins. Int J Epidemiol 2005;34:1302-9.

Olsen AO, Grjibovski A, Magnus P, Tambs K, Harris JR. Psoriasis in Norway as observed in a population-based Norwegian twin panel. Br J Dermatol 2005;153:346-51.

Rasmussen S, Bergsjo P, Jacobsen G, Haram K, Bakketeig LS. Haemoglobin and serum ferritin in pregnancy – correlation with smoking and body mass index. European J Obst Gyn Reprod Biol 2005;123:27-34.

Ronnestad A, Abrahamsen TG, Medbo S, Reigstad H, Lossius K, Kaaresen PI, Engelund IE, Irgens LM, Markestad T. Septicemia in the First Week of Life in a Norwegian National Cohort of Extremely Premature Infants. Pediatrics 2005;115:e262-68.

Ronnestad A, Abrahamsen TG, Medbo S, Reigstad H, Lossius K, Kaaresen PI, Egeland T, Engelund IE, Irgens LM, Markestad T. Late-Onset Septicemia in a Norwegian National Cohort of Extremely Premature Infants Receiving Very Early Full Human Milk Feeding. Pediatrics 2005;115:e269-76.

Singhammer J, Mittelmark MM, Daltveit AK, Tell GS. The influence on birthweight of maternal living conditions a decade prior to giving birth. Norsk epidemiologi 2005;15:91-40.

Skjaerven R, Vatten LJ, Wilcox AJ, Ronning T, Irgens LM, Lie RT. Recurrence of pre-eclampsia across generations: exploring fetal and maternal genetic components in a population based cohort. BMJ 2005;331:877-79.

Skulstad S, Rasmussen S, Seglem S, Svanaes R, Aareskjold H, Kiserud T. The effect of umbilical venous constriction on placental development, cord length and perinatal outcome. Early Human Development 2005;81:325-31.

Spydslaug A, Trogstad L, Skrondal A, Eskild A. Recurrent risk of anal sphinchter laseration in women with vaginal deliveries. Obstetrics and Gynecology 2005;105:307-313.

Staff AC, Holven K, Loken EB, Sygnestveit K, Vollset SE, Smeland S. Effects of public initiatives aimed at reducing neural tube defects with folic acid supplementation. (In Norwegian) Tidsskr Nor Laegeforen 2005;125:435-37.

Staff AC, Holven K, Loken EB, Sygnestveit K, Vollset SE, Smeland S. Does folic acid have effects on other health problems than neural tube defects? (In Norwegian) Tidsskr Nor Laegeforen 2005;125:438-41.

Trogstad LIS, Stoltenberg C, Magnus P, Skjaerven R, Irgens LM. Recurrence risk in hyperemesis gravidarum. BJOG 2005;112:1641-45.

Vatten LJ, Skjaerven R, Wilcox AJ, Ronning T, Irgens LM, Lie RT. Recurrence of pre-eclampsia across generations: exploring fetal and maternal genetic components in a popluation based cohort. BMJ. 2005;331:877-880.

Vollset SE, Gjessing HK, Tandberg A, Ronning T, Irgens LM, Baste V, Nilsen RM, Daltveit AK. Folate Supplementation and Twin Pregnancies. Epidemiology. 2005;16(2):201-205.

Arntzen A, Samuelsen SO, Daltveit AK, Stoltenberg C. Post-neonatal mortality in Norway 1969–95: A cause-specific analysis. Int J Epidemiol 2006;35:1083-89.

Aschim EL, Haugen TB, Tretli S, Daltveit AK, Grotmol T. Risk factors for testicular cancer - differences between pure non-seminoma and mixed seminoma/non-seminoma? Int J Androl 2006;29(4):458-67.

Bakketeig LS, Jacobsen G, Skjaerven R, Carneiro IG, Knudsen LB. Low birthweight and mortality: the tendency to repeat low birthweight and its association with early neonatal and infant morbidity and mortality. Paediatr Perinat Epidemiol 2006;20:507-11.

Basso O, Rasmussen S, Weinberg CR, Wilcox AJ, Irgens LM, Skjaerven R. Trends in fetal and infant survival following preeclampsia. JAMA 2006;296(11):1357-62.

Berle JO, Mykletun A, Daltveit AK, Rasmussen S, Dahl AA. Outcomes in adulthood for children with foetal growth retardation. A linkage study from the Nord-Trondelag Health Study (HUNT) and the Medical Birth Registry of Norway. Acta Psychiatr Scand 2006;113(6):501-9.

Dahl J, Myhr KM, Daltveit AK, Gilhus NE. Planned vaginal births in women with multiple sclerosis: delivery and birth outcome. Acta Neurol Scand Suppl 2006;183:51-4.

Eide MG, Skjaerven R, Irgens LM, Bjerkedal T, Oyen N. Associations of birth defects with adult intellectual performance, disability and mortality: population-based cohort study. Pediatr Res 2006;59(6):848-53.

Gasemyr K, Irgens LM, Aavitsland P. Abortion - no longer need for research? / P. Aavitsland replies. Letter to the editor. (In Norwegian). Tidsskr Nor Laegeforen 2006;126:2832-33.

Hoff JM, Daltveit AK, Gilhus NE. Artrogryposis multiplex congenita - a rare fetal condition caused by maternal myasthenia gravis. Acta Neurol Scand Suppl 2006;183:26-7.

Irgens LM (editor). Annual report 2003/2004, Medical Birth Registry of Norway. Norwegian Institute of Public Health, Bergen 2006. ISBN – ISSN 1504-3320.

Johnsen SL, Wilsgaard T, Rasmussen S, Sollien R, Kiserud T. Longitudinal reference charts for growth of the fetal head, abdomen and femur. Eur J Obstet Gynecol Reprod Biol 2006;127(2):172-85.

Johnsen SL, Rasmussen S, Wilsgaard T, Sollien R, Kiserud T. Longitudinal reference ranges for estimated fetal weight. Acta Obstet Gynecol Scand 2006;85:286-97.

Johnsen SL, Rasmussen S, Sollien R, Kiserud T. Accuracy of second trimester fetal head circumference and biparietal diameter for predicting the time of spontaneous birth. J Perinat Med. 2006;34(5):367-70.

Kazaura MR, Lie RT, Skjaerven R. Grandparents' age and the risk of Down syndrome in Norway. Acta Obstet Gynecol Scand 2006;85:236-40.

Kolas T, Saugstad OD, Daltveit AK, Nilsen ST, Oian P. Planned cesarean versus planned vaginal delivery at term: Comparison of newborn infant outcomes. Am J Obstet Gynecol 2006;195:1538-43.

Lie RT, Wilcox AJ, Skjaerven R. Maternal and Paternal Influences on Length of Pregnancy. Obstet Gynecol 2006;107:880-5.

Lie RT. An International Perspective on Anencephaly and Spina Bifida: Prevalences by the Turn of the Century. In Wyszynski DF ed. Neural tube defects: From origin to treatment. Oxford University Press. New York, 2006;117-32.

Lunde A. Mixed-effects models with application to insurance and genetic epidemiology. Thesis. University of Bergen 2006. ISBN 82-308-0289-0.

Magnus P, Haug K, Nystad W, Skjaerven R. The Mother and Child Cohort Study will give new answers. (In Norwegian). Tidsskr Nor Laegeforen 2006;126(13):1747-1749.

Magnus P, Irgens LM, Haug K, Nystad W, Skjaerven R, Stoltenberg C. Cohort profile: The Norwegian Mother and Child Cohort Study (MoBa). Int J Epidemiol 2006;35:1146-50.

Mjoen G, Saetre DO, Lie RT, Tynes T, Blaasaas KG, Hannevik M, Irgens LM. Paternal occupational exposure to radiofrequency electromagnetic fields and risk of adverse pregnancy outcome. Eur J Epidemiol 2006;21(7):529-535.

Nordby KC, Irgens LM, Kristensen P. Immunological exposures in Norwegian agriculture and pre-eclampsia. Paediatr Perinat Epidemiol 2006;20(6):462-70.

Nordby K-C. Exposure to selected natural and man-made toxic substances in agriculture and their associations with cancers and reproductive outcomes in farmer's families. Thesis. University of Oslo 2006.

Nurk E, Tell GS, Refsum H, Ueland PM, Vollset SE. Factor V Leiden, pregnancy complications and adverse outcomes: the Hordaland Homocysteine Study. QJM 2006;99:289-98.

Rasmussen S, Irgens LM. The effects of smoking and hypertensive disorders on fetal growth. BioMed Central Pregnancy and Childbirth 2006;6:16. http://www.biomedcentral.com/content/pdf/1471-2393-6-16.pdf

Rasmussen S, Kiserud T, Albrechtsen S. Foetal size and body proportion at 17-19 weeks of gestation and neonatal size, proportion, and outcome. Early Hum Dev 2006;82(10):683-90.

Refsum H, Nurk E, Smith AD, Ueland PM, Gjesdal CG, Bjelland I, Tverdal A, Tell GS, Nygard O, Vollset SE. The Hordaland Homocysteine Study: a community-based study of homocysteine, its determinants, and associations with disease. Review. J Nutr 2006;136(6 Suppl):1731S-1740S.

Romundstad LB, Romundstad PR, Sunde A, von Düring V, Skjaerven R, Vatten LJ. Increased risk of placenta previa in pregnancies following IVF/ICSI; a comparison of ART and non-ART pregnancies in the same mother. Hum Reprod 2006;21(9):2353-2358.

Ronningen KS, Paltiel L, Meltzer HL, Nordhagen R, Lie KK, Hovengen R, Haugen M, Nystad W, Hoppin JA, Magnus P. The biobank of the Norwegian mother and child cohort Study. Norsk Epidemiologi 2006;16:59-62.

Ronningen KS, Paltiel L, Meltzer HL, Nordhagen R, Lie KK, Hovengen R, Haugen M, Nystad W, Magnus P, Hoppin JA. The biobank of the Norwegian mother and child cohort Study: A resource for the next 100 years. European Journal of Epidemiology 2006;21(8):619-25.

Samuelsen SO, Bakketeig LS, Tretli S, Johannesen TB, Magnus P. Head circumference at birth and risk of brain cancer in childhood: a population-based study. Lancet Oncol. 2006;7(1):39-42.

Stene LC, Thorsby PM, Berg JP, Ronningen KS, Undlien DE, Joner G and Norwegian Childhood Diabetes Study Group. The relation between size at birth and risk of type 1 diabetes is not influenced by adjustment for the insulin gene (-23HphI) polymorphism or HLA-DQ genotype. Diabetologia 2006;49(9):2068-73.

Thompson JM, Irgens LM, Rasmussen S, Daltveit AK. Secular trends in socio-economic status and the implications for preterm birth. Paediatr Perinat Epidemiol. 2006;20(3):182-7.

Vikse BE, Irgens LM, Bostad L, Iversen BM. Adverse perinatal outcome and later kidney biopsy in the mother.J Am Soc Nephrol 2006;17:837-45.

Slovak Republic

Szabová, E., Nesãáková, E., Zeljenková, D., Kudláãková, M., Varga, I., Ginter, E.:Biological-genetic profile of Romany population in western Slovakia. Reproductive Toxicology, 20, 2005, 483-484.

Zeljenková, D., Szabová, E., Kovrïlnych, J.: Banking of biological and environmental samples. Reproductive Toxicology, 20, 2005, 490-491.

Szabová, E., Zeljenková, D., Kudláãková, M., E-N.Emmanouil-Nikoloussi, Valachoviãová, M., Nikoloussis, E., Navarová, J., Ujházy, E., Ginter, E.: Study on the life style of Romany population. Slov.Anthropol.8(1), 2005, 158-161,.

Zeljenková, D., Szabová, E., Emmanouil-Nikoloussi, E-N., Navarová, J.,Ujházy, E., Nikoloussis, M.: Banking of biological and environmental samples. Slov. Anhtropol., 8(1), 2005, 195-197.

Emmanouil-Nikoloussi, E.-N., Frangou-Massourides, H., Nikoloussis, M., Massouridou, S., Szabová, E., Zeljenková, D., Navarová, J.: Apoptic effect of 13-Cis retinoic acid and hydroxyurea on CASPASE-3 activity in mice placenta : an early marker of congenital malformations. Slov.Anthropol. 8(1), 2005, 129-132.

Szabová, E., Zeljenková, D., Malová, J.: Evaluation of fecundability in Slovak population by „Time-to-pregnancy" assay. Slov.antropol., 8, (2), 2005, 137-141.

Zeljenková, D., Kovrïlnycj, J., Szabová, E.:Acute toxicity of selected chemicals in adult zebrafish (Danio rerio) and its early life stages-the comparative study.ALTEX, 2, 2006, 137.

Szabová, E., Ne‰ãáková, E., Zeljenková, D., Kudláãková, M., Varga, I., Ginter, E.: Overview of biological and health profile of the Romanies in

western Slovakia. In Monitoring health status of vulnerable groups in Europe:Past and present, L.Abreau and J.Sandor (Eds), 2006, p.189-203.

Szabová, E., Zeljenková, D., KovriÏnych, J.: Reproductive effects of chemicals with potential estrogenic effect on human population. Reproductive Toxicology, 22, 2006, 284

Zeljenková, D., KovriÏnych, J., Szabová, E.:Acute toxicity of selected chemicals in adult Zebrafish (Danio rerio) and its early life stages-The comparative study. Reproductive Toxicology, 22, 2006, 285

Szabová, E., Zeljenková, D., and ORFCOM collaborative group.: Study on birth defects. Analysis of socioeconomic parameters. Abstract book.11th Interdisciplinary Slovak-Czech Toxicology Conference, 5-7 June 2006, 60.

Zeljenková, D., Szabová, E.: New EU chemical policy and REACH. Abstract book.11th Interdisciplinary Slovak-Czech Toxicology Conference, 5-7 June 2006, 67.

Szabová, E., Zeljenková, D., Malová, J.: Evaluation of fecundability in some Slovakian regions. Slov.antropol., 9, (1), 2006, 38-41.

Spain: ECEMC

Ali M, Highet LJ, Lacombe D, Goizet C, King MD, Tacke U, van der Knaap MS, Lagae L, Rittey C, Brunner HG, von Bokhoven H, Hamel B, Oade YA, Sanchis A, Desguerre I, Cau D, Mathieu N, Moutard ML, Lebon P, Kumar D, Jackson AP, Crow YJ. A second locus for Aicardi-Goutieres syndrome at chromosome 13q14-21. J Med Genet. Published online 20 May: doi:10.1136/mg.2005.031880.

Bermejo E, Cuevas L, Mendioroz J, Martínez-Frías ML. Anomalías congénitas en España: Vigilancia epidemiológica en el último cuarto de siglo (1980-2004). Bol ECEMC: Rev Dismor Epidemiol 2005; V,4:62-85. (Spanish. Abstract in English. Access through http://bvs.isciii.es/mono/pdf/CIAC_04.pdf)

Bermejo E, Félix V, Lapunzina P, Galán E, Soler V, Delicado A, Pantoja A, Márquez MD, García M, Mora E, Cuevas L, Ureta A, López-Pajares I, Martínez-Frias ML. Craniofacial dyssynostosis: Description of the first four Spanish cases and review. Am J Med Genet 2005;132A:41-48.

Bermejo E, Lapunzina P, Galán E, Félix V, Soler V, Martínez-Frías ML. Correspondence: New findings in craniofacial dyssynostosis. Am J Med Genet 2005;134A:344-345.

Bermejo E, Marco JJ, Paisán L, Félix V, Marugán V, Huertas H, Aparicio P, Sanchis A, Centeno F, Ayala A, Pérez JL, Peñas A, Gomar JL, Lertxundi MM, Burón E, Vázquez MS, Gómez H, Barcia JM, Hernández F, Martínez-Frías ML. Epidermolisis bullosa (EB): Patogénesis, aspectos clínicos, diagnósticos y genéticos, base molecular, aspectos epidemiológicos, manejo del paciente con EB e implicaciones translacionales del análisis de mutaciones. Bol ECEMC: Rev Dismor Epidemiol 2005; V,4:2-13. (Spanish. Abstract in English. Access through http://bvs.isciii.es/mono/pdf/CIAC_04.pdf)

Bermejo E, Mendioroz J, Cuevas L, Mansilla E, Martínez-Frías ML. Análisis de los principales aspectos clínico-epidemiológicos de los recién nacidos con defectos congénitos. Bol ECEMC: Rev Dismor Epidemiol 2005; V,4:19-35. (Spanish. Abstract in English. Access through http://bvs.isciii.es/mono/pdf/CIAC_04.pdf)

Fernández Martín P, Mejías Pavón C, Rodríguez-Pinilla E, Acle M, Martínez-Frías ML. a vacuna de la rubéola y el embarazo. Bol ECEMC: Rev Dismor Epidemiol 2005; 48-51. (Spanish. Abstract in English. Access through http://bvs.isciii.es/mono/pdf/CIAC_04.pdf)

González de Dios J. Aproximación al concepto de prevención cuaternaria en Genética y Dismorfología Clínica. Bol ECEMC: Rev Dismor Epidemiol 2005; V,4:40-46. (Spanish. Abstract in English. Access through http://bvs.isciii.es/mono/pdf/CIAC_04.pdf)

Martínez-Frías ML. The real earliest historical evidence of Down syndrome. Am J Med Genet 2005; 132A:231 (letter).

Martínez-Frías ML. Editoriales: Esterilidad masculina y microdeleciones del cromosoma Y. Med Clin (Barc) 2005; 125(19):736-739.

Martínez-Frías ML, Bermejo E, Rodríguez-Pinilla E, Dequino G y Grupo Periférico del ECEMC. Evolución secular y por autonomías de la frecuencia de tratamientos de fertilidad, partos múltiples y cesáreas en España. Med Clin (Barc) 2005;124(4): 132-139. (Spanish. Abstract in English).

Martínez-Frías ML, Bermejo E, Rodríguez-Pinilla E, Prieto D, Prieto L. Correspondence: MTHFR 677C-T Polymorphism is not excluded as maternal risk for Down syndrome among Turkish women. Am J Med Genet 2005;134A:461.

Martínez-Frías ML, Fernández Toral J, López-Grondona F, Mendioroz J, Bermejo E. Clinical Report: Growth deficiency, facial anomalies, and

brachydactyly (Frías syndrome): A second family. Am J Med Genet 2005; 137A:288-291.

Martínez-Frías ML, Frías JP, Bermejo E, Rodríguez-Pinilla E, Prieto L, Frías JL. Pre-gestational maternal body mass index predicts an increased risk of congenital malformations in infants of mothers with gestational diabetes. Diabetic Medicine 2005;22:775-781.

Martínez-Frías ML, Rodríguez-Pinilla E, Bermejo E, y Grupo periférico del ECEMC. Consumo de tabaco durante el embarazo en España: Análisis por años, comunidades autónomas y características maternas. Med Clin (Barc) 2005;124(3): 86-92. (Spanish. Abstract in English).

Martínez-Frías ML, Rodríguez-Pinilla E, Bermejo E. Respuesta de los autores a los comentarios de Mateos y Aranda sobre 'Consumo de tabaco durante el embarazo en España: Análisis por años, comunidades autónomas y características maternas'. Med Clin (Barc) 2005; 125(5):598-599 (letter).

Mendioroz J, Bermejo E, Martínez Santana S, De La Serna E, Gómez-Ullate J, Alcaraz M, Ayala A, Félix V, García San Miguel M, Lara A, Sanchis A, Vega M, Mansilla E, Cuevas L, Martínez-Frías ML. Síndromes muy poco frecuentes. . Bol ECEMC: Rev Dismor Epidemiol 2005; V,4:36-39. (Spanish. Abstract in English. Access through http://bvs.isciii.es/mono/pdf/CIAC_04.pdf)

Mendioroz J, Fernández-Toral J, Suárez E, López-Grondona F, Kjaer KW, Bermejo E, Martínez-Frías ML. Clinical Report: Sensorineural deafness, abnormal genitalia, synostosis of metacarpals and metatarsals 4 and 5, and mental retardation: Description of a second patient and exclusion of HOXD13. Am J Med Genet 2005;135A:211-213.

Ochoa Sangrador C. Luces y sombras de los estudios casos y controles. Impacto sobre el conocimiento de las malformaciones congénitas. Bol ECEMC: Rev Dismor Epidemiol 2005; V,4:52-60. (Spanish. Abstract in English. Access through http://bvs.isciii.es/mono/pdf/CIAC_04.pdf)

Pogue R, Ehtesham N, Repetto GM, Carrero-Valenzuela R, Bazán de Casella C, Pintos de Pons S, Martínez-Frías ML, Heuertz S, Cormier-Daire V, Cohn DH . Research letter: Probable identity-by-descent for a mutation in the Dyggve-Melchior-Clausen/Smith-McCort dysplasia (Dymeclin) gene among patients from Guam, Chile, Argentina and Spain. Am J Med Genet 2005; 138A:75-78.

Rodríguez L, Martínez-Fernández ML, Mansilla E, Blanco P, Martín Sanz F, Martínez-Frías ML. Trisomía parcial 7q y monosomía subtelomérica 20p. Presentación clínica de un caso y revisión. Bol ECEMC: Rev Dismor Epidemiol 2005; V,4:14-18. (Spanish. Abstract in English. Access through http://bvs.isciii.es/mono/pdf/CIAC_04.pdf)

Rodríguez L, Starke H, Martínez Guardia N, Tönnies H, Neitzel H, Kozlowski P, Mazauric ML, Heller A, López Grondona F, Mansilla E, Santos Muñoz MJ, Liehr T, Martínez-Frías ML. Three new cases with a supernumerary ring chromosome 1. Clin Dysmorphol 2005; 14:169-175.

Rodríguez L, Zollino M, Climent S, Mansilla E, López-Grondona F, Martínez-Fernández ML, Murdolo M, Martínez-Frías ML. Clinical Report: The new Wolf-Hirschhorn syndrome critical region (WHSCR-2): A description of a second case. Am J Med Genet 2005; 136A:175-178.

Rodríguez-Pinilla E, Mejías Pavón C, Fernández Martín P, Acle M, Martínez-Frías ML. Resultados de la actividad de los Servicios de Información Telefónica sobre Teratógenos (SITTE y SITE) durante el año 2004 y análisis del nivel cultural de la población usuaria. Bol ECEMC: Rev Dismor Epidemiol 2005; V,4:106-112. (Spanish. Abstract in English. Access through http://bvs.isciii.es/mono/pdf/CIAC_04.pdf)

Sanchis A, Cerveró L, Bataller A, Tortajada JL, Huguet J, Crow YJ, Au M, Higuet LJ, Martínez-Frías ML. Genetics syndromes mimic congenital infections. J Pediatr 2005;146:701-705.

Bermejo E, Cuevas L, Mendioroz J, Martínez-Frías ML y Grupo Periférico del ECEMC. Vigilancia epidemiológica de anomalías congénitas en España: treinta años de existencia del registro del ECEMC. Bol. ECEMC: Rev Dismor Epidemiol 2006;V,5:58-81. (Spanish. Abstract in English. Access through http://bvs.isciii.es/mono/pdf/CIAC_05.pdf)

Bermejo E, Mendioroz J, Cuevas L, Martínez-Frías ML. Integración de los aspectos clínicos en el análisis epidemiológico de los recién nacidos con defectos congénitos registrados en el ECEMC: 30 años preparándonos para el futuro. Bol. ECEMC: Rev Dismor Epidemiol 2006;V,5:26-43. (Spanish. Abstract in English. Access through http://bvs.isciii.es/mono/pdf/CIAC_05.pdf)

Bermejo E, Mendioroz J, Cuevas L, Martínez-Frías ML. The incidence of gastroschisis: Is also increasing in Spain particularly among babies of young mothers. BMJ (18 Feb) 2006; 332:424. doi:10.1136/bmj.332.7538.424. Access through: http://bmj.bmjjournals.com/cgi/content/full/332/7538/424

Centeno Malfaz F, Bello Martínez B, Beltrán Pérez AI, Alcalde Martín C, López García C. Síndrome de intervalo QT largo congénito. Bol. ECEMC: Rev Dismor Epidemiol 2006;V,5:84-90. (Spanish. Abstract in English. Access through http://bvs.isciii.es/mono/pdf/CIAC_05.pdf)

Crow YJ, Leitch A, Hayward BE, Garner A, Parmar R, Griffith E, Ali M, Semple C, Aicardi J, Babul-Hirji R, Baumann C, Baxter P, Bertini E, Chandler KE, Chitayat D, Cau D, Déry C, Fazzi E, Goizet C, King MD, Klepper J, Lacombe D, Lanzi G, Lyall H, Martínez-Frías ML, Mathieu M, McKeown C, Monier A, Oade Y, Quarrell OW, Rittey CD, Curtis Rogers R, Sanchis A, Stephenson JBP, Tacke U, Till M, Tolmie JL, Tomlin P, Voit T, Weschke B, Geoffrey Woods C, Lebon P, Bonthron DT, Ponting CP, Jackson AP. Mutations in genes enconding ribonuclease H2 subunits cause Aicardi-Goutières syndrome and mimic congenital viral brain infection. Nature Genetics 2006;38:910-916.

Cuevas L, Barcia Ruiz JM, López Soler JA, Félix Rodríguez V, Sanchis Calvo A, Aparicio Lozano P, Arroyo Carrera I, Ayala Garcés A, Conde Nieto MC, Egüés Jimeno J, García González MM, Rosal Roig J, Vázquez García S, Zuazo Zamalloa E, Mendioroz J, Bermejo E, Martínez-Frías ML. Síndromes muy poco frecuentes. Bol. ECEMC: Rev Dismor Epidemiol 2006;V,5:44-48. (Spanish. Abstract in English. Access through http://bvs.isciii.es/mono/pdf/CIAC_05.pdf)

Liehr T, Mrasek K, Weise A, Dufke A, Rodríguez L, Martínez Guardia N, Sanchís A, Vermeesch JR, Ramel C, Polityko A, Haas OA, Anderson J, Claussen U, Von Eggeling F, Starke H. Small supernumerary marker chromosomes-progress towards a genotype-phenotype correlation. Cytogenet Genome Res 2006; 112:23-34.

Mansilla E, Rodríguez L, Martínez-Fernández ML, Rodríguez de Cía J, García Vicent C, Martínez-Frías ML. Monosomía parcial 10p en un caso con fenotipo similar al síndrome de Ritscher-Schinzel. Bol. ECEMC: Rev Dismor Epidemiol 2006;V,5:18-25. (Spanish. Abstract in English. Access through http://bvs.isciii.es/mono/pdf/CIAC_05.pdf)

Martínez-Frías ML. Folic acid: a public-health challenge.Lancet 2006 Jun 24;367(9528):2057 (letter).

Martínez-Frías ML. Técnicas de reproducción asistida y defectos congénitos: ¿riesgo "teratogénico" o genético?. Evid Pediatr 2006;2:66. (Spanish).
Martínez-Frías ML, Bermejo E. Do we have enough evidences to consider that infertility treatments may not be causally related with congenital anomalies in newborn infants?. BMJ 2006; (17 Noviembre). http://www.bmj.com/cgi/eletters/333/7570/665 (letter).

Martínez-Frías ML, Bermejo E, Mendioroz J, Rodríguez-Pinilla E, Grupo Periférico del ECEMC y Grupo de Trabajo de REpIER. Análisis comparativo de las frecuencias de ciertos defectos congénitos y su evolución secular en 11 comunidades autónomas. Prog Obstet Ginecol 2006; 49,5:221-237. (Spanish. Abstract in English).

Martínez-Frías ML, Bermejo-Sánchez E, Rodríguez-Pinilla E, Prieto-Merino D y Grupo Periférico del ECEMC. Características de los neonatos con y sin arteria umbilical única. Análisis de dos series consecutivas de recién nacidos con y sin defectos congénitos. An Pediatr (Barc) 2006;65,6:541-550. (Spanish. Abstract in English).

Martínez-Frías ML, Pérez B, Desviat LR, Castro M, Leal F, Rodríguez L, Mansilla E, Martínez-Fernández ML, Bermejo E, Rodríguez-Pinilla E, Prieto D, Ugarte M and ECEMC Working Group. Maternal polymorphisms 677C-T and 1298A-C of MTHFR, and 66A-G MTRR genes: Is there any relationship between polymorphisms of the folate pathway, maternal homocysteine levels, and the risk for having a child with Down syndrome?. Am J Med Genet Part A 2006;140A:987-997.

Mendioroz J, Cuevas L, Bermejo E, Martínez-Frías ML. Revisión: Aspectos clínicos y genéticos de las hamartoneoplasias que pueden ser diagnosticadas en los tres primeros días de vida. Bol. ECEMC: Rev Dismor Epidemiol 2006;V,5:2-17. (Spanish. Abstract in English. Access through http://bvs.isciii.es/mono/pdf/CIAC_05.pdf)

Rodríguez-Pinilla E, Fernández Martín P, Mejías Pavón C, Lucas V, Martínez-Frías ML. Resultados de la actividad de los Servicios SITTE y SITE durante el año 2005 y análisis de las llamadas por etnia materna. Bol. ECEMC: Rev Dismor Epidemiol 2006;V,5:92-107. (Spanish. Abstract in English. Access through http://bvs.isciii.es/mono/pdf/CIAC_05.pdf)

Rodríguez-Pinilla E, Mejías C, Fernández P, Lucas V, Martínez-Frías ML y Grupo de Trabajo del ECEMC. Análisis de la utilización de medicamentos y otros datos demográficos en la población inmigrante (período 2000-2004). Bol. ECEMC: Rev Dismor Epidemiol 2006;V,5:50-55. (Spanish. Abstract in English. Access through http://bvs.isciii.es/mono/pdf/CIAC_05.pdf)

Rodríguez-Pinilla E, Prieto-Merino D, Dequino G, Mejías C, Fernández P, Martínez-Frías ML. Exposición prenatal a glucocorticoides para acelerar la maduración pulmonar fetal y su repercusión sobre

el peso, la talla y el perímetro cefálico del recién nacido. Med Clin (Barc) 2006;127,10:361-367 (Spanish. Abstract in English).

Zurriaga Lloréns O, Martínez García C, Arizo Luque V, Sánchez Pérez MJ, Ramos Aceitero JM, García Blasco J, Ferrari Arroyo MJ, Perestelo Pérez L, Ramalle Gómara E, Martínez-Frías ML, Posada de la Paz. Los Registros de Enfermedades en la Investigación Epidemiológica de las Enfermedades Raras en España. Rev Esp Salud Pública 2006;80:249-257. (Spanish. Abstract in English).

Sweden

Hansson T, Dahlbom I, Rogberg S, Nyberg BI, Dahlstrom J, Anneren G, Klareskog L, Dannaeus A. Antitissue transglutaminase and antithyroid autoantibodies in children with Down syndrome and celiac disease. J Pediatr Gastroenterol Nutr. 2005 Feb;40(2):170-4; discussion 125-7.

Le Blanc K, Gotherstrom C, Ringden O, Hassan M, McMahon R, Horwitz E, Anneren G, Axelsson O, Nunn J, Ewald U, Norden-Lindeberg S, Jansson M, Dalton A, Astrom E, Westgren M. Fetal mesenchymal stem-cell engraftment in bone after in utero transplantation in a patient with severe osteogenesis imperfecta. Transplantation. 2005 Jun 15;79(11):1607-14.

Ljunger E, Cnattingius S, Lundin C, Anneren G. Chromosomal anomalies in first-trimester miscarriages. Acta Obstet Gynecol Scand. 2005 Nov;84(11):1103-7.

Schoumans J, Nordgren A, Ruivenkamp C, Brondum-Nielsen K, Teh BT, Anneren G, Holmberg E, Nordenskjold M, Anderlid BM. Genome-wide screening using array-CGH does not reveal microdeletions/microduplications in children with Kabuki syndrome. Eur J Hum Genet. 2005 Feb;13(2):260-3

Busby A, Abramsky L, Dolk H, Armstrong B, Addor MC, Anneren G, Armstrong N, Baguette A, Barisic I, Berghold A, Bianca S, Braz P, Calzolari E, Christiansen M, Cocchi G, Daltveit AK, De Walle H, Edwards G, Gatt M, Gener B, Gillerot Y, Gjergja R, Goujard J, Haeusler M, Latos-Bielenska A, McDonnell R, Neville A, Ollars B, Portillo I, Ritvanen A, Robert-Gnansia E, Rosch C, Scarano G, Steinbicker V. Preventing neural tube defects in Europe: a missed opportunity.Reprod Toxicol. 2005 Sep-Oct;20(3):393-402. Erratum in: Reprod Toxicol. 2006 Jan;21(1):116.

George L, Granath F, Johansson AL, Anneren G, Cnattingius S. Environmental tobacco smoke and risk of spontaneous abortion. Epidemiology. 2006 Sep;17(5):500-5.

Hedov G, Wikblad K, Annerén G. Sickness absence in Swedish parents of children with Down syndrome: Relation to self-perceived health and stress. J Intell Disabil Res 2006; 50:546-52.

Mansouri MR, Carlsson B, Davey E, Nordenskjold A, Wester T, Anneren G, Lackgren G, Dahl N. Molecular genetic analysis of a de novo balanced translocation t(6;17)(p21.31;q11.2) associated with hypospadias and anorectal malformation. Hum Genet. 2006 Mar;119(1-2):162-8. Epub 2006 Jan 3.

Ollars B, Annerén G. Missbildningsregistrering 2005 (Registration of congenital malformations 2005) (In Swedish with English summary). National Board of Health and Welfare, Centre for Epidemiology, ISBN 91-85482-78-1 Article No. 2006-42-10.

Soderbergh A, Gustafsson J, Ekwall O, Hallgren A, Nilsson T, Kampe O, Rorsman F, Anneren G. Autoantibodies linked to autoimmune polyendocrine syndrome type I are prevalent in Down syndrome. Acta Paediatr. 2006 Dec;95(12):1657-60.

Wester U, Bondeson ML, Edeby C, Anneren G. Clinical and molecular characterization of individuals with 18p deletion: a genotype-phenotype correlation. Am J Med Genet A. 2006 Jun 1;140(11):1164-71.

USA: Atlanta

Bailey LB, Berry RJ. Folic acid supplementation and the occurrence of congenital heart defects, orofacial clefts, multiple births, and miscarriage. Am J Clin Nutr 81(5):1213S-1217S, 2005.

Berry RJ, Kihlberg R. Folic acid supplementation is associated with an increase in dizygotic twinning. Early Hum Dev 81(5):465-7, 2005.

Berry RJ, Kihlberg R, Devine O. Impact of misclassification of in vitro fertilisation in studies of folic acid and twinning: modelling using population based Swedish vital records. BMJ 330(7495):815, 2005.

Berry RJ. Impact of ovarian stimulation on studies of twinning. Am J Obstet Gynecol 193(3 Pt 2):1287-8, 2005.

Canfield CA, Collins JS, Botto LD, Williams LJ, Mai CT, Kirby RS, Pearson K, Devine O, Mulinare, J. Changes in the birth prevalence of selected birth defects after grain fortification with folic acid in the United States: findings from a multi-state pop-

ulation-based study. Birth Defects Research (Part A) 73:679-89. 2005.

Centers for Disease Control and Prevention. Improved national prevalence estimates for 18 selected major birth defects - United States, 1999-2001. MMWR 54(51 & 52):1301-1305, 2005.

Centers for Disease Control and Prevention. Use of dietary supplements containing folic acid among women of childbearing age - United States, 2005. MMWR 54(38):955-958, 2005.

Lagoy CT, Joshi N, Cragan JD, Rasmussen SA. Medication use during pregnancy and lactation: an urgent call for public health action. Journal of Women's Health 14(2):104-109, 2005

Rasmussen SA, Hayes EB. Public health approach to emerging infections among pregnant women. Am J Public Health 95(11):1942-4, 2005.

Schieve LA, Rasmussen SA, Reefhuis J. Risk of birth defects among children conceived with assisted reproductive technology: providing an epidemiologic context to the data. Fertil Steril 84(5):1320-4, 2005.

Siffel C, Alverson CJ, Correa A. Analysis of seasonal variation of birth defects in Atlanta. Birth Defects Res A Clin Mol Teratol 73(10):655-62, 2005.

Than LC, Honein MA, Watkins ML, Yoon PW, Daniel KL, Correa A. Intent to become pregnant as a predictor of exposures during pregnancy: is there a relation? J Reprod Med 50(6):389-96, 2005

Williams LJ, Kucik JE, Alverson CJ, Olney RS, Correa A. Epidemiology of gastroschisis in metropolitan Atlanta, 1968 through 2000. Birth Defects Res A Clin Mol Teratol 73(3):177-83, 2005

Williams LJ, Rasmussen SA, Flores A, Kirby RS, Edmonds LD. Decline in the prevalence of spina bifida and anencephaly by race/ethnicity: 1995-2002. Pediatrics 116(3):580-6, 2005

Werler MM, Mitchell AA, Hernandez-Diaz S, Honein MA, for the National Birth Defects Prevention Study. Use of over-the-counter medications during pregnancy. Am J Obstet Gynecol 193(3):771-7, 2005

Yuskiv N, Honein MA, Moore CA. Reported multivitamin consumption and the occurrence of multiple congenital anomalies. Am J Med Genet A 136(1):1-7, 2005.

Biernath KR, Reefhuis J, Whitney CG, Mann EA, Costa P, Eichwald J, Boyle C. Bacterial meningitis among children with cochlear implants beyond 24 months after implantation. Pediatrics 117(2):284-9, 2006.

Callaghan WM, MacDorman MF, Rasmussen SA, Qin C, Lackritz EM. The contribution of preterm birth to infant mortality rates in the United States. Pediatrics 118(4):1566-73, 2006.

Canfield MA, Honein MA, Yuskiv N, Xing J, Mai CT, Collins JS, Devine O, Petrini J, Ramadhani TA, Hobbs CA, Kirby RS. National estimates and race/ethnic-specific variation of selected birth defects in the United States, 1999-2001. Birth Defects Res A Clin Mol Teratol 76(11):747-56, 2006.

Cono J, Cragan JD, Jamieson DJ, Rasmussen SA. Prophylaxis and treatment of pregnant women for emerging infections and bioterrorism emergencies. Emerg Infect Dis 12(11):1631-7, 2006.

Cragan JD, Friedman JM, Holmes LB, Uhl K, Green NS, Riley L. Ensuring the safe and effective use of medications during pregnancy: planning and prevention through preconception care. Matern Child Health J 10(5 Suppl):129-35, 2006.

Crider KS, Reefhuis J, Woomert A, Honein MA. Racial and ethnic disparity in participation in DNA collection at the Atlanta site of the National Birth Defects Prevention Study. Am J Epidemiol 164(8):805-12, 2006.

Dott M, Chace D, Fierro M, Kalas TA, Hannon WH, Williams J, Rasmussen SA. Metabolic disorders detectable by tandem mass spectrometry and unexpected early childhood mortality: a population-based study. Am J Med Genet A 140(8):837-42, 2006.

Jamieson DJ, Kourtis AP, Bell M, Rasmussen SA. Lymphocytic choriomeningitis virus: an emerging obstetric pathogen? Am J Obstet Gynecol 194(6):1532-6, 2006.

Jamieson DJ, Theiler RN, Rasmussen SA. Emerging infections and pregnancy. Emerg Infect Dis 12(11):1638-43, 2006.

Kenneson A, Kolor K, Yang Q, Olney RS, Rasmussen SA, Friedman JM. Trends and racial disparities in muscular dystrophy deaths in the United States, 1983-1998: an analysis of multiple cause mortality data. Am J Med Genet A 140(21):2289-97, 2006.

Lawrence JM, Watkins ML, Chiu V, Erickson JD, & Petitti DB. Do racial and ethnic differences in serum folate values exist after food fortification with folic acid? Am J Obstet Gynecol 194:520-6, 2006.

O'Leary DR, Kuhn S, Kniss KL, Hinckley AF, Rasmussen SA, Pape WJ, Kightlinger LK, Beecham BD, Miller TK, Neitzel DF, Michaels SR, Campbell GL, Lanciotti RS, Hayes EB. Birth outcomes following West Nile Virus infection of pregnant women in the United States: 2003-2004. Pediatrics 117(3):e537-45, 2006.

Rasmussen SA, Wong LY, Correa A, Gambrell D, Friedman JM. Survival in infants with Down syndrome, Metropolitan Atlanta, 1979-1998. J Pediatr 148(6):806-12, 2006.

Reefhuis J, Rasmussen SA, Friedman JM. Selective serotonin-reuptake inhibitors and persistent pulmonary hypertension of the newborn. N Engl J Med 354(20):2188-90, 2006.

Rowland CA, Correa A, Cragan JD, Alverson CJ. Are encephaloceles neural tube defects? Pediatrics 118(3):916-23, 2006.

Shaw GM, Carmichael SL, Laurent C, Rasmussen SA. Maternal nutrient intakes and risk of orofacial clefts. Epidemiology 17(3):285-91, 2006.

Siffel C, Strickland MJ, Gardner BR, Kirby RS, Correa A. Role of geographic information systems in birth defects surveillance and research. Birth Defects Res A Clin Mol Teratol 76(11):825-33, 2006.

Whitehead NS, Rasmussen SA, Cox S, Posner SF. Prevalence and predictors of receipt of prenatal information about genetic screening. Prenat Diagn 26(10):944-50, 2006.

Yang Q, Botto LD, Erickson JD, Berry RJ, Sambell C, Johansen H, Friedman JM. Improvement in stroke mortality in Canada and the United States, 1990 to 2002. Circulation 113:1335-43, 2006.

Yang Q, Chen H, Correa A, Devine O, Mathews TJ, Honein MA. Racial differences in infant mortality attributable to birth defects in the United States, 1989-2002. Birth Defects Res A Clin Mol Teratol 76(10):706-13, 2006.

USA: Texas

Anderson JL, Waller DK, Canfield MA, Shaw GM, Watkins ML, Werler MM. Maternal obesity, gestational diabetes and central nervous system birth defects: a Texas case-control study. Epidemiology 2005; 16:87-92.

Blanton SH, Cortez A, Stal S, Mulliken JB, Finnell RH, Hecht JT: Variation in IRF6 contributes to nonsyndromic cleft lip and palate. Am J Med Genet, 2005;137:259-62.

Canfield MA, Collins JS, Botto LD, Williams LJ, Mai CT, Kirby RS, Pearson K, Devine O, Mulinare J, for the National Birth Defects Prevention Network. Changes in the birth prevalence of selected birth defects after grain fortification with folic acid in the US: findings from a multi-state, population-based study. Birth Defects Research Part A 2005;73:678-688.

Etheredge AJ, Christensen K, del Junco D, Murray JC, Mitchell LE. Evaluation of two methods for assessing gene-environment interactions using data from the Danish case-control study of facial clefts. Birth Defects Research (A) 73:541-546, 2005.

Felkner M, Suarez L, Brender JD, Scaife B, Hendricks K. Iron status indicators in women with prior neural tube defect-affected pregnancies. Maternal and Child Health J 2005; 9:421-428.

Felkner M, Suarez L, Hendricks K, Larsen R. Implementation and outcomes of recommended folic acid supplementation in Mexican American women with prior neural tube defect-affected pregnancies. Prev Med 2005:40:867-871.

Gilboa SM, Mendola P, Olshan AF, Langlois PH, Savitz DA, Loomis D. Relation between ambient air quality and selected birth defects, seven county study, Texas, 1997-2000. Am J Epidemiol 2005;162:238-52.

Hashmi SS, Waller K, Langlois P, Canfield M, Hecht JT. Prevalence of nonsyndromic oral clefts in Texas: 1995-1999. Am J Med Genet A, 2005 May 1;134(4):368-372.

Heck AL, Bray M, Scott A, Blanton SH, Hecht JT: Variation in CASP10 gene is associated with idiopathic talipes equinovarus. J Pediatr Orthop, 2005; 25:598-602.

Jensen LE, Hoess K, Whitehead AS, Mitchell LE. The NAT1 C1095A polymorphism, maternal multivitamin use and smoking, and the risk of spina bifida. Birth Defects Research Part A 73:512-516, 2005.

Mason CA, Kirby RS, Sever LE, Langlois PH. Prevalence is the preferred measure of frequency of birth defects. Birth Defects Research Part A 2005;73:690-692.

McBride KL, Marengo L, Canfield M, Langlois P, Fixler D, Belmont JW. Epidemiology of left ventricle outflow tract obstruction malformations (aortic valve stenosis, coarctation of the aorta, hypoplastic left heart syndrome) in Texas, 1999-2001. Birth Def Res Part A 2005;73:555-6.

Mitchell LE. Epidemiology of neural tube defects.

American Journal of Medical Genetics 135:88-94, 2005.

Mitchell LE, Weinberg CR. Evaluation of offspring and maternal genetic effects on disease risk using a family-based approach: the "Pent" design. American Journal of Epidemiology. 162:676-685, 2005.

Moorthi, RN, Hashmi SS, Langlois P, Canfield M, Waller K, Hecht JT. Idiopathic talipes equinovarus (ITEV) (clubfeet) in Texas. Am J Med Genet, 2005;132(4):376-80.

Scheuerle A, Heller K, Elder F. Complete trisomy 1q with mosaic Y;1 translocation: A recurrent aneuploidy presenting diagnostic dilemmas. M J Med Genet A. 2005;138A:166-170.

Suarez L, Gilani Z, Felkner M, Brender J, Henry J, Hendricks K. Exposure to polychlorinated biphenyls and risk of neural tube defects in a Mexican American population. Int J Occup Environ Health 2005;11:234-38.

Waller DK, Dawson T. Relation between maternal obesity and adverse pregnancy outcomes. In: Hornstra G. Uauy R, Yang X (eds): The impact of maternal nutrition on the offspring. Nestle Nutrition Workshop Series Pediatric Program, Volume 55, pp 197-211, Nestec LTD., Vevey/S. Karger AG, Basel, 2005.

Brender, JD, Zhan FB, Suarez L, Langlois P, Gilani Z, DeLima I, Moody K. Linking environmental hazards and birth defects data: methods and issues. Int J Occup Environ Health 2006; 12:126 – 133.

Brender JD, Suarez L, Felkner M, Gilani Z, Stinchcomb D, Moody K, Henry J, Hendricks K. Maternal exposure to arsenic, cadmium, lead, and mercury and neural tube defects in offspring. Environ Res 2006; 101:132-139.

Brender JD, Zhan FB, Suarez L, Langlois PH, Moody K. Maternal residential proximity to waste sites and industrial facilities and oral clefts in offspring. J Occup Environ Med 2006; 48:565-572.

Canfield MA, Honein MA, Yuskiv N, Xing J, Mai CT, Collins JS, Devine O, Petrini J, Ramadhani TA, Hobbs CA, Kirby RS, for the National Birth Defects Prevention Network. National estimates and race/ethnic-specific variation of selected birth defects in the United States, 1999-2001. Birth Defects Res Part A 2006; 76:747-756.

Canfield MA, Przybyla, Case AP, Ramadhani T, Suarez L, Dyer J. Folic acid awareness and supplementation among women of childbearing age. Prev Med 2006; 43:27-30.

Canfield MA, Ramadhani T, Langlois P, Waller DK. Residential mobility patterns and exposure misclassification in epidemiologic studies of birth defects. Journal of Exposure Science and Environmental Epidemiology 2006 (published online).

CDC. Improved national prevalence estimates for 18 selected major birth defects – United States, 1999-2001. MWMR 2006; 54:1301-1305.

Gilboa SM, Mendola O, Olshan AF, Savitz DA, Herring AH, Loomis D, Langlois PH, Keating K. Characteristics that predict locating and interviewing mother identified by a state birth defects registry and vital records. Birth Defects Research Part A 76: 60-65, 2006.

Jensen LE, Etheredge AJ, Brown KS, Mitchell LE, Whitehead AS. Maternal genotype for the monocyte chemoattractant protein 1 A(-2518)G promoter polymorphism is associated with the risk of spina bifida in offspring. American Journal of Medical Genetics A, 140:1114-1118, 2006.

Jensen LE, Hoess K, Mitchell LE, Whitehead AS. Loss of function polymorphisms in NAT1 protect against spina bifida. Human Genetics 120:52-57, 2006.

Langlois P, Scheuerle A, Winter A. Severity of birth defects as a tool for dealing with detection bias in cluster investigations. Birth Defects Monitor 12(1): 2-3, 2006.

Missmer SA, Suarez L, Felkner M, Wang E, Merrill AH, Rothman KJ, Hendricks KA. Exposure to fumonisins and the occurrence of neural tube defects along the Texas-Mexico border. Environ Health Perspectives 114:237-241, 2006.

Mitchell LE, Finnell RH, Whitehead AS. Aetiology and prevention of spina bifida. In: David TJ ed. Recent Advances in Pediatrics. Royal Society of Medicine Press Ltd. 2006.

Zhan FB, Brender JD, Han Y, Suarez L, Langlois PH. GIS-EpiLink: a spatial search tool for linking environmental and health data. J Med Syst 2006; 30: 405-412.

Zhan FB, Brender JD, De Lima I, Suarez L, Langlois PH. Match rate and positional accuracy of two geocoding methods for epidemiologic research. Ann Epidemiol 2006; 16:842-849.

Archer NP, Langlois PH, Case AP, Wolfe LJ. Linking

teratogen service and birth defects registry databases to improve knowledge of birth defect status. Birth Defects Research Part A (in press)

Archer N, Langlois P, Suarez L, Brender J, Shanmugam R. Association of paternal age with prevalence of selected birth defects. Birth Defects Res A Clin Mol Teratol (in press).

Finnell RH, Mitchell LE. Neural tube defects. Rimoin's Principals and Practices of Medical Genetics, 5th Edition, in press.

Gilboa AM, Mendola P, Olshan AF, Harness C, Loomis D, Langlois P, Savitz DA, Herring AH. Comparison of residential geocoding methods in a population-based study of air quality and birth defects. Environ Res (In Press).

Mitchell LE. Epidemiology of Cleft Lip and Palate. Comprehensive Cleft Care, in press.

Mitchell LE, Etheredge AJ, Hill DS, Finnell RH. Spina Bifida. Encyclopedia of Molecular Mechanisms of Disease, in press.

Zhan FB, Brender JD, De Lima I, Suarez L, Langlois PH. Improving match rate and positional accuracy of two geocoding methods for epidemiological research. Ann Epidemiol 2006 (In Press)

Usa: Utah UBDN

Canfield MA, Collins JS, Botto LD, Williams LJ, Mai CT, Kirby RS, Pearson K, Devine O, Mulinare J; National Birth Defects Preventin Network.

Changes in the birth prevalence of selected birth defects after grain fortification with folic acid in the United States: findings from a multi-state population-based study. Birth Defects Res A Clin Mol Teratol. 2005 Oct;73(10):679-89.

Yang Q, Botto LD, Erickson JD, Berry RJ, Sambell C, Johansen H, Friedman JM. Improvement in stroke mortality in Canada and the United States, 1990 to 2002. Circulation. 2006 Mar 14;113(10):1335-43.

Bleyl SB, Botto LD, Carey JC, Young LT, Bamshad MJ, Leppert MF, Ward K. Analysis of a Scottish founder effect narrows the TAPVR-1 gene interval to chromosome 4q12. Am J Med Genet A. 2006 Nov 1;140(21):2368-73.

ANNUAL REPORT

Order form

q Please send me _____copies of 2006 Annual Report (15 US $)/12 Euro each
q Please send me _____copies of 200__ Annual Report (5 US $)/4 Euro each
q Please send me _____copies of 199__ Annual Report (5 US $)

I enclose / will send a check of _____ US$*/Euros
Payable to International Centre on Birth Defects

**contribution for printing and shipping cost*

Name
_____

Shipping Address
_____
_____
_____

Please fax/e-mail this form to:
ICBDSR CENTRE
Fax: +39-06-3701904
E-mail: centre@icbdsr.org
Website: www.icbdsr.org

Editing, design, graphics and printing:

Tel.: 39.06.86216255
E mail: info@betmultimedia.it

Annual Report 2006

with data for 2004

Published by
International Clearinghouse
for Birth Defects
Surveillance and Research