Chapter XVII, (Q00-Q99)

Congenital malformations, deformations and chromosomal abnormalities

Excludes: inborn errors of metabolism (E70-E90)

This chapter contains the following blocks:

Q00-Q07	Congenital malformations of the nervous system
Q10-Q18	Congenital malformations of eye, ear, face and neck
Q20-Q28	Congenital malformations of the circulatory system
Q30-Q34	Congenital malformations of the respiratory system
Q35-Q37	Cleft lip and palate
Q38-Q45	Other congenital malformations of the digestive system
Q50-Q56	Congenital malformations of genital organs
Q60-Q64	Congenital malformations of the urinary system
Q65-Q79	Congenital malformations and deformations of the musculoskeletal system
Q80-Q89	Other congenital malformations
Q90-Q99	Chromosomal abnormalities, not elsewhere classified

Q00-Q07 Congenital malformations of the nervous system

Q00 Anencephaly and similar malformations

Q00.00 Anencephaly, NOS
Acephaly
Acrania
Amyelencephaly
Excludes: hydranencephaly (Q04.35)

Q00.01 Incomplete anencephaly
Hemianencephaly
Hemicephaly

Q00.1 Craniorachischisis
Rachischisis: . craniospinal
. complete
. total

Q00.2 Iniencephaly

Q00.20 Iniencephaly, open

Q00.21 Iniencephaly, closed

Q01 Encephalocele
Includes: encephalomyelocele
hydroencephalocele
hydromeningocele, cranial
meningocele, cerebral
meningoencephalocele
Note: cranial hydromeningocele and cerebral meningocele are not considered to be encephaloceles as they do not contain brain tissue but have been included here in ICD-10
Excludes: Meckel-Gruber syndrome (Q61.9)

Q01.0 Frontal encephalocele

Q01.1 Nasofrontal encephalocele

Q01.2 Occipital encephalocele

Q01.8 Encephalocele of other sites

Q01.80 Parietal encephalocele

Q01.81 Orbital encephalocele

Q01.82 Nasal encephalocele

Q01.83 Nasopharyngeal encephalocele

Q01.9 Encephalocele, unspecified

Q02 # Microcephaly

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           Hydromicrocephaly
           Micrencephalon
          Excludes: Meckel-Gruber syndrome (Q61.9)
                    microcephaly due to:
                     . congenital infection (P35-P37)
                     . exposure to ionising radiation (Q86.85)

Q03       Congenital hydrocephalus
           Includes: hydrocephalus in newborn
            Excludes: Arnold-Chiari syndrome (Q07.0)
                      hydrocephalus:
                      .acquired (G91.-)
                      .due to congenital toxoplasmosis (P37.1)
                      .with spina bifida (Q05.0-Q05.4)
Q03.0     Malformations of aqueduct of Sylvius
           Aqueduct of Sylvius: .anomaly
                                .obstruction, congenital
                                .stenosis
Q03.1     Atresia of foramina of Magendie and Luschka
           Dandy-Walker syndrome
Q03.8     Other congenital hydrocephalus
Q03.80    Clover leaf skull
           Kleeblattsch„del deformity syndrome

Q03.9     Congenital hydrocephalus, unspecified

Q04       Other congenital malformations of brain
           Excludes: cyclopia (Q87.03)
                     macrocephaly (Q75.3)
Q04.0     Congenital malformations of corpus callosum
Q04.00    Agenesis of corpus callosum
Q04.1     Arhinencephaly
Q04.2     Holoprosencephaly
Q04.3     Other reduction deformities of brain
@           Absence   }
            Agenesis  }
            Aplasia   } of part of brain
            Hypoplasia}
           Excludes: congenital malformations of corpus callosum (Q04.0)
Q04.30    Reduction anomalies of cerebrum
Q04.31    Reduction anomalies of hypothalamus
Q04.32    Reduction anomalies of cerebellum
Q04.33    Agyria or lissencephaly
Q04.34    Microgyria or pachygyria
           Polygyria
           Micropolygyria
Q04.35    Hydranencephaly
Q04.4     Septo-optic dysplasia
Q04.5     Megalencephaly
Q04.6     Congenital cerebral cysts
            Porencephaly
            Schizencephaly
           Excludes: acquired porencephalic cysts (G93.0)
Q04.60    Multiple congenital cerebral cysts
Q04.61    Single congenital cerebral cyst
Q04.8     Other specified congenital malformations of brain
           Macrogyria
           Walnut brain
           Congenital haematocephalus
           Congenital malformation of cerebral meninges
Q04.9     Congenital malformation of brain, unspecified
           Congenital: .anomaly          }
                       .deformity        }
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.disease or lesion } NOS of brain
.multiple anomalies}

Q05 Spina bifida
Includes: hydromeningocele (spinal)
meningocele (spinal)
meningomyelocele
myelocele
myelomeningocele
spinal rachischisis
spina bifida (aperta)(cystica)
syringomyelocele
Excludes: Arnold-Chiari syndrome (Q07.0)
spina bifida occulta (Q76.0)
rachischisis (Q00.1): . cranial
. craniospinal

Note: For Spina bifida Q05.0-Q05.8 the following fifth-character subdivision can be used if desired-
.....1 open, aperta, not covered with skin or membrane
.....2 closed, cystica, covered with skin or membrane
.....9 if not known whether lesion is open or closed

Q05.0 Cervical spina bifida with hydrocephalus
Q05.1 Thoracic spina bifida with hydrocephalus
Spina bifida: .dorsal }
.thoracolumbar } with hydrocephalus
.dorsolumbar }
Q05.2 Lumbar spina bifida with hydrocephalus
Lumbosacral spina bifida with hydrocephalus
Q05.3 Sacral spina bifida with hydrocephalus
Q05.4 Unspecified spina bifida with hydrocephalus
Site unspecified
Q05.5 Cervical spina bifida without hydrocephalus
Q05.6 Thoracic spina bifida without hydrocephalus
Spina bifida: .dorsal NOS
.thoracolumbar NOS
.dorsolumbar NOS
Q05.7 Lumbar spina bifida without hydrocephalus
Lumbosacral spina bifida NOS
Q05.8 Sacral spina bifida without hydrocephalus
Q05.9 Spina bifida, unspecified

Q06 Other congenital malformations of spinal cord
Excludes: syringomyelia and syringobulbia (G95.0)
Q06.0 Amyelia
Q06.1 Hypoplasia and dysplasia of spinal cord
Atelomyelia
Myelatelia
Myelodysplasia of spinal cord
Q06.2 Diastematomyelia
Q06.3 Other congenital cauda equina malformations
Q06.4 Hydromyelia
Hydrorachis
Q06.8 Other specified congenital malformations of spinal cord
Q06.9 Congenital malformations of spinal cord, unspecified
Congenital: .anomaly }
.deformity } NOS of spinal cord
.disease or lesion} or meninges

Q07 Other congenital malformations of nervous system
Excludes: familial dysautonomia [Riley-Day] (G90.1)
neurofibromatosis (nonmalignant) (Q85.0)

Q07.0 Arnold-Chiari syndrome

Q07.8 Other specified congenital malformations of nervous system
Agenesis of nerve, NOS
Cayler syndrome
Congenital facial diplegia
Displacement of brachial plexus
Nuclear agenesis
Excludes: Moebius syndrome (Q87.06)
Duane syndrome (H50.8)

Q07.80 Jaw-winking syndrome
Marcus Gunn's syndrome

Q07.81 Optic nerve hypoplasia
Congenital optic atrophy

Q07.82 Crocodile tears

Q07.9 Congenital malformations of nervous system, unspecified
Congenital malformation of meninges, unspecified
Congenital: .anomaly }
.deformity } NOS of nervous system
.disease or lesion }

Q10-Q18 Congenital malformations of eye, ear, face and neck
Excludes: cleft lip and cleft palate (Q35-37)
congenital malformations of:
.cervical spine (Q05.0, Q05.5, Q67.5, Q76.0-Q76.4)
.larynx (Q31.-)
.lip NEC (Q38.0)
.nose (Q30.-)
.parathyroid gland (Q89.2)
.thyroid gland (Q89.2)
retinoblastoma (C69.2)

Q10 Congenital malformations of eyelid, lacrimal apparatus and orbit
Excludes: cryptophthalmos:
.NOS (Q11.2)
.syndrome (Q87.02)
Goldenhar syndrome [oculo-auriculo-vertebral syndrome] (Q87.04)

Q10.0 Congenital ptosis
Blepharophimosis-ptosis syndrome

Q10.1 Congenital ectropion

Q10.2 Congenital entropion

Q10.3 Other congenital malformations of eyelid
Ablepharon (absence of eyelids)
Absence or agenesis of: .cilia (eyelashes)
.eyelid
Accessory: .eyelid
.eye muscle
Blepharophimosis, congenital [fused eyelids]
Congenital symblepharon
Coloboma of eyelid
Mongoloid slant (of palpebral fissure)
Antimongoloid slant (of palpebral fissure)
Congenital malformation of eyelid NOS

Q10.4 Absence and agenesis of lacrimal apparatus
Absence of punctum lacrimale

Q10.5 Congenital stenosis and stricture of lacrimal duct

Q10.6 Other congenital malformations of lacrimal apparatus
Congenital malformations of lacrimal apparatus NOS

Q10.7 Congenital malformations of orbit

Q11 Anophthalmos, microphthalmos and macrophthalmos

Q11.0 Cystic eyeball

Q11.1 Other anophthalmos
Agenesis }
Aplasia } of eye
Excludes: cryptophthalmos syndrome (Q87.02)
Q11.2 Microphthalmos
Cryptophthalmos NOS
Dysplasia of eye
Fraser syndrome
Hypoplasia of eye
Lenz' microphthalmus syndrome
Rudimentary eye
Excludes: cryptophthalmos syndrome (Q87.02)
Q11.3 Macrophthalmos
Excludes: macrophthalmos in congenital glaucoma (Q15.0)

Q12 Congenital lens malformations
Q12.0 Congenital cataract
Q12.1 Congenital displaced lens
Q12.2 Coloboma of lens
Q12.3 Congenital aphakia
Q12.4 Spherophakia
Q12.8 Other congenital lens malformations
Q12.80 Microphakia
Q12.9 Congenital lens malformation, unspecified

Q13 Congenital malformations of anterior segment of eye
Q13.0 Coloboma of iris
Coloboma NOS
Q13.1 Absence of iris
Aniridia
See also nephroblastoma [Wilms' tumour] (C64)
Q13.2 Other congenital malformations of iris
Anisocoria, congenital
Atresia of pupil
Congenital malformation of iris NOS
Corectopia
Polycoria
Excludes: ectopic pupil (H21.5)
Q13.3 Congenital corneal opacity
Q13.4 Other congenital corneal malformations
Congenital malformation of cornea NOS
Microcornea
Peter's anomaly
Q13.5 Blue sclera
Q13.8 Other congenital malformations of anterior segment of eye
Rieger's anomaly
Iridogoniodysgenesis with somatic anomalies
Q13.9 Congenital malformations of anterior segment of eye, unspecified

Q14 Congenital malformations of posterior segment of eye
Q14.0 Congenital malformation of vitreous humour
Congenital vitreous opacity
Q14.1 Congenital malformation of retina
Congenital retinal aneurysm
Coloboma of retina
Q14.10 Congenital retinoschisis
Q14.2 Congenital malformation of optic disc
Coloboma of optic disc
Q14.3 Congenital malformation of choroid
Q14.8 Other congenital malformations of posterior segment of eye
Coloboma of the fundus
Q14.9 Congenital malformation of posterior segment of eye, unspecified

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Q15      Other congenital malformations of eye
          Excludes: congenital nystagmus (H55)
                    ocular albinism (E70.3)
                    retinitis pigmentosa (H35.5)
Q15.0    Congenital glaucoma
          Buphthalmos
          Glaucoma of newborn Hydrophthalmos
          Macrophthalmos in congenital glaucoma
Q15.00   Congenital keratoglobus
          Enlarged cornea
          Megalocornea
Q15.8    Other specified congenital malformations of eye
Q15.9    Congenital malformation of eye, unspecified
          Congenital: .anomaly  }
                      .deformity} NOS of eye

Q16      Congenital malformations of ear causing hearing impairment
          Excludes: congenital deafness (H90.-)
Q16.0    Congenital absence of (ear) auricle
          Anotia
          Congenital absence of ear lobe
Q16.1    Congenital absence, atresia and stricture of auditory canal
          (external)
            Atresia, stenosis or stricture of osseous meatus
Q16.2    Absence of Eustachian tube
Q16.3    Congenital malformation of ear ossicles
          Fusion of ear ossicles
Q16.4    Other congenital malformations of middle ear
          Congenital malformations of middle ear NOS
Q16.5    Congenital malformation of inner ear
          Anomaly of: .membranous labyrinth
                      .organ of Corti
Q16.9    Congenital malformation of ear causing impairment of hearing,
          unspecified
           Congenital absence of ear NOS

Q17      Other congenital malformations of ear
          Excludes: preauricular sinus (Q18.1)
Q17.0    Accessory auricle
           Accessory tragus
           Polyotia
           Preauricular appendage or tag
           Supernumary: .ear
                        .lobule
          Excludes: Goldenhar syndrome
                     [oculo-auriculo-vertebral syndrome] (Q87.04)
Q17.1    Macrotia
Q17.2    Microtia
Q17.3    Other misshapen ear
          Pointed ear
          Vulcan ear
          Simple ear
Q17.4    Misplaced ear
           Low set ears
          Excludes: cervical auricle (Q18.2)
Q17.5    Prominent ear
          Bat ear
Q17.8    Other specified congenital malformations of ear
@         Darwin's tubercle
          Branchio-oro-renal syndrome
          Melnick-Fraser syndrome
Q17.9    Congenital malformation of ear, unspecified
          Congenital anomaly of ear NOS
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Q18 Other congenital malformations of face and neck
Excludes: cleft lip and cleft palate (Q35-37)
conditions classified to Q67.0-Q67.4
congenital malformations of skull and face bones (Q75.-)
cyclopia (Q87.03)
dentofacial anomalies [including malocclusion] (K07.-)
malformation syndromes affecting facial appearance (Q87.0-)
persistent thyroglossal duct (Q89.2)

Q18.0 Sinus, fistula and cyst of branchial cleft
Branchial vestige

Q18.1 Preauricular sinus and cyst
Fistula : .of auricle, congenital
.cervicoaural

Q18.2 Other branchial cleft malformations
Branchial cleft malformations NOS
Cervical auricle
Otocephaly

Q18.3 Webbing of neck
Pterygium colli

Q18.4 Macrostomia

Q18.5 Microstomia

Q18.6 Macrocheilia
Hypertrophy of lip, congenital

Q18.7 Microcheilia

Q18.8 Other specified congenital malformations of face and neck
Medial: .cyst }
.fistula} of face and neck
.sinus }

Q18.80 Synophrys

Q18.9 Congenital malformation of face and neck, unspecified
Congenital anomaly NOS of face and neck

Q20-Q28 Congenital malformations of the circulatory system

Q20 Congenital malformations of cardiac chambers and connections
Excludes: dextrocardia with situs inversus (Q89.3)
mirror image atrial arrangement with situs inversus (Q89.3)

Q20.0 Common arterial trunk
Persistent truncus arteriosus

Q20.1 Double outlet right ventricle
Taussig-Bing syndrome

Q20.2 Double outlet left ventricle

Q20.3 Discordant ventriculoarterial connection
Dextrotransposition of aorta
Transposition of great vessels (complete)

Q20.4 Double inlet ventricle
Common ventricle
Cor triloculare biatriatum
Single ventricle

Q20.5 Discordant atrioventricular connection
Corrected transposition
Levotransposition
Ventricular inversion

Q20.6 Isomerism of atrial appendages
Isomerism of atrial appendages with asplenia or polysplenia
Ivemark syndrome

Q20.8 Other congenital malformations of cardiac chambers and connections
Cor biloculare

Q20.9 Congenital malformation of cardiac chambers and connections, unspecified

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Q21      Congenital malformations of cardiac septa
           Excludes: acquired cardiac septal defect (I51.0)
Q21.0    Ventricular septal defect
          Roger's disease [Maladie de Roger]
          Small VSD with no significant haemodynamic effects
Q21.1    Atrial septal defect
          ASD
Q21.10   Ostium secundum atrial septal defect (type II)
Q21.11   Patent or persistent foramen ovale
Q21.12   Sinus venosus defect
Q21.13   Coronary sinus defect
Q21.14   Lutembacher's syndrome (ASD plus mitral stenosis)
Q21.15   Common atrium
          Cor triloculare biventriculare
Q21.18   Other specified atrial septal defect
          Excludes: ostium primum atrial septal defect (type I) Q21.20
Q21.2    Atrioventricular septal defect
Q21.20   Ostium primum atrial septal defect (type I)
Q21.21   Common atrioventricular canal
Q21.28   Other specified atrioventricular septal defect
          Endocardial cushion defect NOS
Q21.3    Tetralogy of Fallot
          Ventricular septal defect with pulmonary stenosis or
           atresia, dextroposition of aorta and hypertrophy of
           right ventricle.
Q21.4    Aortopulmonary septal defect
          Aortic septal defect
          Aortopulmonary window
Q21.8    Other congenital malformations of cardiac septa
Q21.80   Left ventricle to right atrial communication
          Gerbode defect
Q21.81   Eisenmenger's syndrome
Q21.82   Pentalogy of Fallot
          Fallot's tetralogy plus atrial septal defect
Q21.9    Congenital malformation of cardiac septum, unspecified
          Septal heart defect, NOS

Q22      Congenital malformations of pulmonary and tricuspid valves
Q22.0    Pulmonary valve atresia
Q22.1    Congenital pulmonary valve stenosis
Q22.2    Congenital pulmonary valve insufficiency
          Congenital pulmonary valve regurgitation
Q22.3    Other congenital malformations of pulmonary valve
          Congenital malformation of pulmonary valve NOS
Q22.4    Congenital tricuspid stenosis
          Tricuspid atresia
Q22.5    Ebstein's anomaly
Q22.6    Hypoplastic right heart syndrome
Q22.8    Other congenital malformations of tricuspid valve
Q22.9    Congenital malformation of tricuspid valve, unspecified

Q23      Congenital malformations of aortic and mitral valves
Q23.0    Congenital stenosis of aortic valve
           Congenital aortic: .atresia
                              .stenosis
           Excludes: congenital subaortic stenosis (Q24.4)
                     that in hypoplastic left heart syndrome (Q23.4)
Q23.1    Congenital insufficiency of aortic valve
          Congenital aortic insufficiency
Q23.10   Bicuspid aortic valve
Q23.2    Congenital mitral stenosis
          Congenital mitral atresia
Q23.3    Congenital mitral insufficiency
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Q23.4 Hypoplastic left heart syndrome
Atresia, or marked hypoplasia of aortic orifice or valve, with hypoplasia of ascending aorta and defective development of left ventricle (with mitral valve stenosis or atresia)
Q23.8 Other congenital malformations of aortic and mitral valves
Q23.9 Congenital malformation of aortic and mitral valves, unspecified

Q24 Other congenital malformations of heart
Excludes: endocardial fibroelastosis (I42.4)
Q24.0 Dextrocardia
Excludes: dextrocardia with situs inversus (Q89.3)
isomerism of atrial appendages (with asplenia or polysplenia) (Q20.6)
mirror image atrial arrangement with situs inversus (Q89.3)
Q24.1 Laevocardia
Q24.2 Cor triatriatum
Q24.3 Pulmonary infundibular stenosis
Q24.4 Congenital subaortic stenosis
Q24.5 Malformation of coronary vessels
Congenital coronary (artery) aneurysm
Q24.6 Congenital heart block
Q24.8 Other specified congenital malformations of the heart
Congenital malformation of: .myocardium
.pericardium
Malposition of heart
Uhl's disease
Congenital cardiomegaly
Fallot's trilogy
Ectopia cordis
Q24.80 Congenital diverticulum of left ventricle
Q24.9 Congenital malformations of the heart, unspecified
Congenital: .anomaly
.disease NOS of heart

Q25 Congenital malformations of great arteries
Q25.0 Patent ductus arteriosus
PDA
Patent ductus Botallo
Persistent ductus arteriosus
Q25.1 Coarctation of aorta
Q25.10 Preductal coarctation of aorta
Q25.11 Postductal coarctation of aorta
Q25.19 Coarctation of aorta unspecified
Q25.2 Atresia of aorta
Interrupted aortic arch
Q25.3 Stenosis of aorta
Supravalvular aortic stenosis
Excludes: congenital aortic stenosis (valvular) (Q23.0)
Q25.4 Other congenital malformations of aorta
Absence}
Aplasia} of aorta
Persistent convolutions of aortic arch
Excludes: hypoplasia of aorta in hypoplastic left heart syndrome (Q23.4)
Q25.40 Hypoplasia of aorta
Tubular hypoplasia of aorta
Q25.41 Persistent right aortic arch
Q25.42 Overriding aorta
Q25.43 Aneurysm of sinus of Valsalva (ruptured)
Q25.44 Double aortic arch
Vascular ring due to double aortic arch
Q25.45 Congenital aneurysm of aorta

Congenital dilatation of aorta
Q25.5 Atresia of pulmonary artery
Q25.6 Stenosis of pulmonary artery
Q25.7 Other congenital malformations of pulmonary artery
Agenesis }
Anomaly } of pulmonary artery
Hypoplasia }
Q25.70 Pulmonary arteriovenous aneurysm
Q25.71 Aberrant pulmonary artery
Q25.72 Congenital aneurysm of pulmonary artery
Congenital dilatation of pulmonary artery
Q25.8 Other congenital malformations of great arteries
Q25.80 Vascular ring due to anomalous right subclavian artery
Q25.81 Vascular ring, other and unspecified
Excludes: vascular ring due to double aortic arch (Q25.44)
Q25.9 Congenital malformations of great arteries, unspecified

Q26 Congenital malformations of great veins
Q26.0 Congenital stenosis of vena cava
Q26.00 Congenital stenosis of inferior vena cava
Q26.01 Congenital stenosis of superior vena cava
Q26.1 Persistent left superior vena cava
Q26.2 Total anomalous pulmonary venous connection
Total anomalous pulmonary venous drainage
TAPVD
Q26.20 Total anomalous pulmonary venous connection-subdiaphragmatic
Q26.21 Total anomalous pulmonary venous connection-supradiaphragmatic
Q26.3 Partial anomalous pulmonary venous connection
Q26.4 Anomalous pulmonary venous connection, unspecified
Q26.5 Anomalous portal venous connection
Q26.6 Portal vein-hepatic artery fistula
Q26.8 Other congenital malformations of great veins
Absence of vena cava (inferior) (superior)
Azygos continuation of inferior vena cava
Persistent left posterior cardinal vein
Scimitar syndrome
Q26.9 Congenital malformation of great vein, unspecified
Anomaly of vena cava (inferior) (superior) NOS

Q27 Other congenital malformations of peripheral vascular system
Excludes: anomalies of: .cerebral and precerebral vessels (Q28.0-Q28.3)
.coronary vessels (Q24.5)
.pulmonary artery (Q25.5-Q25.7)
congenital retinal aneurysm (Q14.1)
haemangioma and lymphangioma (D18.-)
congenital naevi (Q82.5-)
Q27.0 Congenital absence and hypoplasia of umbilical artery
Single umbilical artery
Q27.1 Congenital renal artery stenosis
Q27.2 Other congenital malformations of renal artery
Congenital malformation of renal artery NOS
Multiple renal arteries
Q27.3 Peripheral arteriovenous malformation
Arteriovenous aneurysm
Excludes: acquired arteriovenous aneurysm (I77.0)
Q27.4 Congenital phlebectasia
Q27.8 Other specified congenital malformations of peripheral vascular system
Absence, atresia of artery or vein NEC Congenital:
.aneurysm (peripheral)
.stricture, artery
.varix

Q27.80 Aberrant subclavian artery
Anomalous right subclavian artery
Excludes: vascular ring due to anomalous right subclavian artery (Q25.80)

Q27.9 Congenital malformation of peripheral vascular system, unspecified
Anomaly of artery or vein NOS

Q28 Other congenital malformations of circulatory system
Excludes: congenital aneurysm: .NOS (Q27.8)
.coronary (Q24.5)
.peripheral (Q27.8)
.pulmonary (Q25.7)
.retinal (Q14.1)
.aneurysm of sinus of Valsalva (ruptured) (Q25.43)
ruptured: .cerebral arteriovenous malformation (I60.8)
.malformation of precerebral vessels (I72.-)
Von Hippel-Lindau syndrome (Q85.82)

Q28.0 Arteriovenous malformation of precerebral vessels
Congenital arteriovenous precerebral aneurysm (nonruptured)

Q28.1 Other malformations of precerebral vessels
Congenital: .malformation of precerebral vessels NOS
.precerebral aneurysm (nonruptured)

Q28.2 Arteriovenous malformation of cerebral vessels
Arteriovenous malformation of brain NOS
Congenital arteriovenous cerebral aneurysm (nonruptured)
See also Sturge-Weber(-Dimitri) syndrome (Q85.81)

Q28.3 Other malformations of cerebral vessels
Congenital: .cerebral aneurysm (nonruptured)
.malformation of cerebral vessels NOS

Q28.8 Other specified congenital malformations of circulatory system
Congenital aneurysm, specified site NEC
Congenital lymphatic abnormalities

Q28.9 Congenital malformation of circulatory system, unspecified

Q30-Q34 Congenital malformations of the respiratory system

Q30 Congenital malformations of nose
Excludes: congenital deviation of nasal septum (Q67.4)

Q30.0 Choanal atresia
Atresia }
Congenital stenosis } of nares (anterior)(posterior)
CHARGE association

Q30.1 Agenesis and underdevelopment of nose
Congenital absence of nose

Q30.2 Fissured, notched and cleft nose

Q30.3 Congenital perforated nasal septum

Q30.8 Other congenital malformations of nose
Accessory nose
Congenital anomaly of nasal sinus wall

Q30.9 Congenital malformation of nose, unspecified

Q31 Congenital malformations of larynx

Q31.0 Web of larynx
Web of larynx: .NOS
.glottic
.subglottic

Q31.1 Congenital subglottic stenosis

Q31.2 Laryngeal hypoplasia

Q31.3 Laryngocele

Q31.4 Congenital laryngeal stridor

Congenital stridor (larynx) NOS
Q31.40 Congenital laryngomalacia
Q31.48 Other congenital laryngeal stridor
Q31.8 Other congenital malformations of larynx
Absence)
Agenesis) of cricoid cartilage, epiglottis, glottis,
Atresia) larynx or thyroid cartilage
Cleft thyroid cartilage
Congenital stenosis of larynx NEC
Fissure of epiglottis
Posterior cleft of cricoid cartilage
Q31.80 Congenital laryngeal cleft
Q31.9 Congenital malformation of larynx, unspecified

Q32 Congenital malformations of trachea and bronchus
Excludes: congenital bronchiectasis (Q33.4)
Q32.0 Congenital tracheomalacia
Q32.1 Other congenital malformations of trachea
Anomaly of tracheal cartilage
Atresia of trachea
Congenital: . dilatation }
. malformation } of trachea
. tracheocele
Q32.10 Congenital tracheal stenosis
Complete (cartilaginous) tracheal ring [stovepipe trachea]
Q32.11 Congenital tracheo-oesophageal cleft
Excludes: congenital tracheo-oesophageal fistula (Q39.1, Q39.2)
Q32.2 Congenital bronchomalacia
Q32.20 Primary congenital bronchomalacia
Q32.21 Secondary congenital bronchomalacia
Congenital bronchomalacia associated with vascular ring
Q32.3 Congenital stenosis of bronchus
Q32.4 Other congenital malformations of bronchus
Congenital malformation of bronchus NOS
Q32.40 Tracheal bronchus
Q32.41 Bronchus picus
Q32.42 Congenital diverticulum of bronchus
Q32.43 Absence of bronchus
Agenesis}
Atresia } of bronchus

Q33 Congenital malformations of lung
Q33.0 Congenital cystic lung
Congenital: .cystic lung disease
.bronchogenic cyst
Excludes: cystic lung disease, acquired or unspecified (J98.4)
Q33.00 Congenital single lung cyst
Q33.01 Congenital polycystic lung
Congenital multiple lung cysts
Q33.02 Congenital honeycomb lung
Q33.1 Accessory lobe of lung
Q33.10 Azygos lobe of lung
Q33.2 Sequestration of lung
Q33.3 Agenesis of lung
Absence of lung (lobe)
Q33.4 Congenital bronchiectasis
Q33.5 Ectopic tissue in lung
Q33.6 Hypoplasia and dysplasia of lung
Excludes: pulmonary hypoplasia associated with:
.short gestation (P28.0)
.prolonged rupture of membranes (P01.1)
Q33.8 Other congenital malformations of lung
Q33.80 Congenital (cystic) adenomatoid malformation of the lung

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Q33.81   Broncho-pulmonary isomerism
Q33.9    Congenital malformation of lung, unspecified

Q34      Other congenital malformations of respiratory system
Q34.0    Anomaly of pleura
Q34.1    Congenital cyst of mediastinum
Q34.8    Other specified congenital malformations of respiratory system
          Atresia of nasopharynx
Q34.80   Congenital pulmonary lymphangiectasis
Q34.9    Congenital malformation of respiratory system, unspecified
          Congenital: .absence    }
                      .anomaly NOS} of respiratory organ
Q35-Q37  Cleft lip and cleft palate
          Excludes: Robin's syndrome (Q87.08)

Q35   $$ Cleft palate
          Includes: fissure of palate
                    palatoschisis
          Excludes: cleft palate with cleft lip (Q37.-)
Q35.0    Cleft hard palate, bilateral
Q35.10   Cleft hard palate, unilateral
Q35.19   Cleft hard palate, unspecified
Q35.2    Cleft soft palate, bilateral
Q35.30   Cleft soft palate, unilateral
Q35.39   Cleft soft palate, unspecified
Q35.4    Cleft hard palate with cleft soft palate, bilateral
          Bilateral complete cleft palate
Q35.50   Cleft hard palate with cleft soft palate, unilateral
          Unilateral complete cleft palate
Q35.59   Cleft hard palate with cleft soft palate, unspecified
          Complete cleft palate, unspecified
Q35.6    Cleft palate, medial
          Median cleft of soft and/or hard palate
Q35.60   Central complete cleft palate
Q35.61   Central incomplete cleft palate
Q35.7    Cleft uvula
          Bifid uvula
Q35.8    Cleft palate, unspecified, bilateral
Q35.90   Cleft palate, unspecified, unilateral
Q35.99   Cleft palate, unspecified

Q36      Cleft lip
          Includes: cheiloschisis
                    congenital fissure of lip
                    harelip
                    labium leporinum
          Excludes: cleft lip with cleft palate (Q37.-)
Q36.0    Cleft lip, bilateral
Q36.1    Cleft lip, medial
Q36.90   Cleft lip, specified as unilateral
Q36.99   Cleft lip NOS

Q37   $$ Cleft palate with cleft lip
Q37.0    Cleft hard palate with cleft lip, bilateral
Q37.10   Cleft hard palate with cleft lip, specified as unilateral
Q37.19   Cleft hard palate with cleft lip, NOS
Q37.4    Cleft hard and soft palate with cleft lip, bilateral
Q37.50   Cleft hard and soft palate with cleft lip, specified as unilateral
Q37.59   Cleft hard and soft palate with cleft lip NOS

Q37.8    Unspecified, cleft palate with cleft lip, bilateral

Q37.90   Unspecified, cleft palate with cleft lip, specified as unilateral
```

Q37.99 Cleft palate with cleft lip NOS

Q38-Q45 Other congenital malformations of the digestive system
Excludes: hernia: . inguinal (K40)
. femoral (K41)
. umbilical (K42)
. ventral (K43)

Q38 Other congenital malformations of tongue, mouth and pharynx
Excludes: macrostomia (Q18.4)
microstomia (Q18.5)
Q38.0 Congenital malformations of lips, not elsewhere classified
Congenital malformation of lip NOS
Labial pit
Van der Woude's syndrome
Excludes: cleft lip (Q36.-)
.with cleft palate (Q37.-)
macrocheilia (Q18.6)
microcheilia (Q18.7)
Q38.00 Congenital fistula of lip
Q38.08 Other congenital malformations of lips, not elsewhere classified
Q38.1 Ankyloglossia
Tongue tie
Q38.2 Macroglossia
Q38.3 Other congenital malformations of tongue
Bifid tongue
Congenital: .adhesion of tongue
.fissure of tongue
.dislocation or displacement of tongue
Hypoglossia
Hypoplasia of tongue
Microglossia
Lobulated tongue
Hamartomata of tongue
Q38.30 Aglossia
Q38.39 Congenital malformation of tongue NOS
Q38.4 Congenital malformations of salivary glands and ducts
Absence)
Accessory) (of) salivary gland or duct
Atresia)
Congenital fistula of salivary gland
Q38.5 Congenital malformations of palate, not elsewhere classified
Absence of uvula
Congenital malformation of palate NOS
Excludes: cleft palate (Q35.-)
.with cleft lip (Q37.-)
Q38.50 High arched palate
Q38.58 Other congenital malformations of palate, not elsewhere classified
Q38.6 Other congenital malformations of mouth
Congenital malformation of mouth NOS
Q38.7 Pharyngeal pouch
Diverticulum of pharynx
Excludes: pharyngeal pouch syndrome (D82.1)
Q38.8 Other congenital malformations of pharynx
Congenital malformation of pharynx NOS
Q38.80 Congenital palato-oesophageal incoordination
Naso-pharyngeal dysmotility

Q39 Congenital malformations of oesophagus
Excludes: congenital tracheo-oesophageal cleft (Q32.11)
Q39.0 Atresia of oesophagus without fistula
Atresia of oesophagus NOS

Q39.1 Atresia of oesophagus with tracheo-oesophageal fistula
Atresia of oesophagus with broncho-oesophageal fistula
Q39.10 Atresia of oesophagus with fistula between trachea and upper oesophageal pouch
Q39.11 Atresia of oesophagus with fistula between trachea and lower oesophageal pouch
Q39.2 Congenital tracheo-oesophageal fistula without atresia
Congenital tracheo-oesophageal fistula NOS
TOF
Q39.20 Congenital broncho-oesophageal fistula without atresia
Q39.3 Congenital stenosis and stricture of oesophagus
Q39.4 Oesophageal web
Q39.5 Congenital dilatation of oesophagus
Q39.50 Congenital cardiospasm
Achalasia of cardia, congenital
Q39.6 Diverticulum of oesophagus
Oesophageal pouch
Q39.8 Other congenital malformations of oesophagus
Absent oesophagus
Congenital displacement of oesophagus
Q39.80 Congenital duplication of oesophagus
Q39.81 Oesophageal dysmotility
Pseudo-obstruction of oesophagus
Q39.9 Congenital malformation of oesophagus, unspecified

Q40 Other congenital malformations of upper alimentary tract
Q40.0 Congenital hypertrophic pyloric stenosis
Congenital or infantile: . constriction)
. hypertrophy)
. spasm) of pylorus
. stenosis)
. stricture)
Pyloric stenosis, NOS, in infant less than three months old
Infantile hypertrophic pyloric stenosis
Q40.1 Congenital hiatus hernia
Displacement of cardia through oesophageal hiatus
Partial thoracic stomach
Excludes: congenital diaphragmatic hernia (Q79.0)
Q40.2 Other specified congenital malformations of stomach
Megalogastria
Microgastria
Congenital: .displacement of stomach
.diverticulum of stomach
.hourglass stomach
Prepyloric diaphragm
Q40.21 Dysmotility of stomach
Pseudo-obstruction of stomach
Q40.22 Duplication of stomach
Q40.3 Congenital malformation of stomach, unspecified
Q40.8 Other specified congenital malformations of upper alimentary tract
Pyloric atresia
Q40.9 Congenital malformation of upper alimentary tract,unspecified
Congenital: . anomaly)
. deformity) NOS of upper alimentary tract

Q41 Congenital absence, atresia and stenosis of small intestine
Includes: congenital obstruction, occlusion and stricture of small intestine or intestine NOS
Excludes: meconium ileus (E84.1)
Q41.0 Congenital absence, atresia and stenosis of duodenum
Q41.1 Congenital absence, atresia and stenosis of jejunum
Apple peel syndrome
Imperforate jejunum

Q41.2 Congenital absence, atresia and stenosis of ileum
Q41.8 Congenital absence, atresia and stenosis of other specified parts of small intestine
Congenital absence, atresia and stenosis of multiple regions of small intestine
Q41.9 Congenital absence, atresia and stenosis of small intestine, part unspecified
Congenital absence, atresia and stenosis of intestine NOS

Q42 Congenital absence, atresia and stenosis of large intestine
Includes: congenital obstruction, occlusion and stricture of large intestine
Q42.0 Congenital absence, atresia and stenosis of rectum with fistula
For Q42.0 the following optional fifth character codes may be used if desired to indicate the type of fistula:

.....0 rectourethral
.....1 rectovesical
.....2 rectovulval
.....3 rectocutaneous
.....4 rectocloacal
.....8 other specified (see below)

N.B. For Congenital absence, atresia and stenosis of rectum with rectovaginal fistula, use Q42.0 and Q52.2

For Congenital gastrointestinal-urinary tract fistula without rectal absence, atresia or stenosis, use Q64.74

Q42.1 Congenital absence, atresia and stenosis of rectum without fistula
Imperforate rectum
Q42.2 Congenital absence,atresia and stenosis of anus with fistula
For Q42.2 the following optional fifth character codes may be used if desired to indicate the type of fistula:

.....0 anocutaneous
.....1 anovestibular
.....8 other

Q42.3 Congenital absence,atresia,stenosis of anus without fistula
Imperforate anus
Congenital anal stenosis
Q42.8 Congenital absence, atresia and stenosis of other parts of large intestine
Congenital absence, atresia and stenosis of appendix
Q42.9 Congenital absence, atresia and stenosis of large intestine, part unspecified
Q42.90 Colonic atresia

Q43 Other congenital malformations of intestine
Q43.0 Meckel's diverticulum
Q43.00 Persistent omphalomesenteric duct
Persistent vitelline duct
Q43.01 Omphalomesenteric band
Q43.02 Omphalomesenteric cyst
Q43.1 Hirschsprung's disease
Aganglionosis
Congenital (aganglionic) megacolon
Hirschsprung's disease NOS
Q43.10 Short segment Hirschsprung's disease
Q43.11 Long segment Hirschsprung's disease
Q43.12 Total colonic aganglionosis
Q43.13 Total intestinal aganglionosis

Q43.2 Other congenital functional disorders of colon
Congenital dilatation of colon
Congenital macrocolon, not aganglionic
Small left colon syndrome
Megacystis, microcolon, hypoperistalsis syndrome
Neuronal intestinal dysplasia
Hyperganglionosis
Q43.20 Large intestinal dysmotility
Pseudo-obstruction of large intestine
Q43.3 Congenital malformations of intestinal fixation
Jackson's membrane
Universal mesentery
Other anomalies of mesentery
Q43.30 Malrotation of colon
Rotation:
. failure of }
. incomplete } of caecum and colon
. insufficient }
Q43.31 Congenital intraabdominal adhesions [bands]
Congenital adhesions [bands]: .omental, anomalous
.peritoneal
Ladd's bands
Q43.38 Other congenital malformations of intestinal fixation
Q43.4 Duplication of intestine
Duplication of anus, appendix, caecum and intestine
Enterogenous cyst
Q43.5 Ectopic anus
Misplaced anus
Q43.6 Congenital fistula of rectum and anus
Excludes: congenital fistula: .rectovaginal (Q52.2
.urethrorectal (Q64.7)
pilonidal fistula or sinus (L05.-)
congenital fistula of rectum and anus with absence, atresia and stenosis (Q42.0, Q42.2)
Q43.7 Persistent cloaca
Cloaca NOS
Q43.8 Other specified congenital malformations of intestine
Congenital: .blind loop syndrome
.diverticulitis, colon
.diverticulum, intestine
Dolichocolon
Megaloappendix
Megaloduodenum
Transposition of: .appendix
.colon
.intestine
Persistent inversion of appendix
Q43.80 Microcolon

Q43.81 Small intestinal dysmotility
Pseudo-obstruction of small intestine
Q43.82 Generalised intestinal dysmotility
Q43.83 Congenital intestinal blind loop
Q43.9 Congenital malformation of intestine, unspecified

Q44 Congenital malformations of gallbladder, bile ducts and liver
Q44.0 Agenesis, aplasia and hypoplasia of gallbladder
Congenital absence of gallbladder
Q44.1 Other congenital malformations of gallbladder
Congenital malformation of gallbladder NOS
Intrahepatic gallbladder
Duplication of gallbladder
Q44.2 Atresia of bile ducts

Biliary atresia NOS
Q44.20 Intrahepatic biliary atresia
Q44.21 Extrahepatic biliary atresia
Q44.3 Congenital stenosis and stricture of bile ducts
Q44.4 Choledochal cyst
Q44.5 Other congenital malformations of bile ducts
Accessory hepatic duct
Congenital malformation of bile duct NOS
Duplication: .biliary duct
.cystic duct
Q44.6 Cystic disease of liver
Fibrocystic disease of liver
Q44.7 Other congenital malformations of liver
Accessory liver
Congenital: .hepatomegaly
.malformation of liver NOS
Q44.70 Absence or agenesis of liver, total or lobe
Q44.71 Alagille's syndrome
Q44.72 Congenital atrophy of left lobe of liver
Q44.73 Riedel's lobe of liver
Q44.74 Ectopic liver
Q44.75 Focal nodular hypoplasia of liver

Q45 Other congenital malformations of digestive system
Excludes: congenital: .diaphragmatic hernia (Q79.0)
.hiatus hernia (Q40.1)
Q45.0 Agenesis, aplasia and hypoplasia of pancreas
Congenital absence of pancreas
Q45.1 Annular pancreas
Q45.2 Congenital pancreatic cyst
Q45.3 Other congenital malformations of pancreas and pancreatic duct
Accessory pancreas
Congenital malformation of pancreas or pancreatic duct NOS
Excludes: diabetes mellitus: .congenital (E10.-)
.neonatal (P70.2)
fibrocystic disease of pancreas (E84.-)
Q45.30 Ectopic pancreas
Q45.8 Other specified congenital malformations of digestive system
Q45.80 Absence (complete)(partial) of alimentary tract NOS
Q45.81 Duplication of digestive organs NOS
Q45.82 Malposition, congenital of digestive organs NOS
Q45.83 Congenital mesenteric cyst
Q45.9 Congenital malformation of digestive system, unspecified
Congenital: .anomaly }
.deformity NOS } of digestive system

Q50-Q56 Congenital malformations of genital organs
Excludes: androgen resistance syndrome [testicular feminisation syndrome] (E34.5)
syndromes associated with anomalies in the number and form of chromosomes (Q90-Q99)

Q50 Congenital malformations of ovaries, fallopian tubes and broad ligaments
Q50.0 Congenital absence of ovary
Excludes: Turner's syndrome (Q96.-)
Q50.00 Congenital absence of ovary, unilateral
Q50.01 Congenital absence of ovary, bilateral
Q50.1 Developmental ovarian cyst
Q50.10 Developmental ovarian cyst, single
Q50.11 Developmental ovarian cyst, multiple
Q50.2 Congenital torsion of ovary

Q50.3 Other congenital malformations of ovary
Accessory ovary
Dysplastic ovary
Congenital malformation of ovary NOS
Q50.30 Ovarian streak
Q50.4 Embryonic cyst of fallopian tube
Fimbrial cyst
Q50.5 Embryonic cyst of broad ligament
Cyst: . epo”phoron
. Gartner's duct
. parovarian
. of mesenteric remnant
Q50.6 Other congenital malformations of fallopian tube and broad ligament
Accessory) (of) fallopian tube or broad ligament
Atresia)
Congenital malformation of fallopian tube or broad ligament NOS
Q50.60 Absence of fallopian tube or broad ligament

Q51 Congenital malformations of uterus and cervix
Q51.0 Agenesis and aplasia of uterus
Congenital absence of uterus
Q51.1 Doubling of uterus with doubling of cervix and vagina
Q51.2 Other doubling of uterus
Doubling of uterus NOS
Q51.3 Bicornate uterus
Bicornuate uterus
Q51.4 Unicornate uterus
Unicornuate uterus
Q51.5 Agenesis and aplasia of cervix
Congenital absence of cervix
Q51.6 Embryonic cyst of cervix
Q51.7 Congenital fistula between uterus and digestive and urinary tracts
Uterointestinal fistula
Uterovesical fistula
Q51.8 Other congenital malformations of uterus and cervix
Displaced uterus
Hydrometrocolpos with post-axial polysyndactyly syndrome
Hypoplasia of uterus and cervix
Kaufman-McKusick syndrome
MURCS syndrome
Rudimentary cervix
Q51.9 Congenital malformation of uterus and cervix, unspecified

Q52 Other congenital malformations of female genitalia
Q52.0 Congenital absence of vagina
Q52.1 Doubling of vagina
Septate vagina
Excludes: doubling of vagina with doubling of uterus and cervix (Q51.1)
Q52.2 Congenital rectovaginal fistula
Excludes: cloaca (Q43.7)
Q52.3 Imperforate hymen
Q52.4 Other congenital malformations of vagina
Congenital malformation of vagina NOS
Congenital cyst of canal of Nuck
Q52.40 Embryonic vaginal cyst
Q52.5 Fusion of labia
Excludes: acquired labial adhesions (N90.8)
fused labia secondary to inflammation (N76.80)
Q52.6 Congenital malformation of clitoris
Q52.7 Other congenital malformations of vulva
Congenital: . absence } of vulva
. cyst }

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                    . malformation NOS }
Q52.8    Other specified congenital malformations of female genitalia
          Congenital cyst of hydatid of Morgagni in female
Q52.80   Congenital cyst of Wolffian duct in female
Q52.81   Female hypospadias
Q52.9    Congenital malformation of female genitalia, unspecified

Q53      Undescended testicle
           Excludes: retractile testicle (Q55.20)
          For Q53.0-.2 the following optional fifth character
           subdivisions denoting abnormal site of testis may be used if
           desired:
                   .....0 inguinal
                   .....1 canalicular
                   .....2 intraabdominal
                   .....8 other

Q53.0    Ectopic testis
          Unilateral or bilateral ectopic testis
Q53.1    Undescended testicle, unilateral
Q53.2    Undescended testicle, bilateral
Q53.9    Undescended testicle, unspecified
          Cryptorchidism NOS

Q54      Hypospadias
          Excludes: epispadias (Q64.0)
Q54.0    Hypospadias, balanic
          Hypospadias: .coronal
                       .glandular
Q54.1    Hypospadias, penile
Q54.2    Hypospadias, penoscrotal
Q54.3    Hypospadias, perineal
Q54.4    Congenital chordee
Q54.8    Other hypospadias
          Excludes: female hypospadias (Q52.81)
Q54.9    Hypospadias, unspecified

Q55      Other congenital malformations of male genital organs
          Excludes: congenital hydrocele (P83.5)
                    hypospadias (Q54.-)
Q55.0    Absence and aplasia of testis
Q55.00   Absence and aplasia of testis, unilateral
          Monorchism
Q55.01   Absence and aplasia of testis, bilateral
          Anorchism
Q55.1    Hypoplasia  of testis and scrotum
          Fusion of testes
Q55.2    Other congenital malformations of testis and scrotum
@         Congenital malformation of testis or scrotum NOS
          Polyorchism
Q55.20   Retractile testis
Q55.21   Bifid scrotum
Q55.3    Atresia of vas deferens
Q55.4    Other congenital malformations of vas deferens, epididymis,
          seminal vesicles and prostate
           Absence or aplasia of: .prostate
                                  .spermatic cord
           Congenital malformation of vas deferens, epididymis,
            seminal vesicles or prostate NOS
           Cysts of embryonal remnants [persistent Wolffian duct]
Q55.40   Congenital cyst of hydatid of Morgagni in male
Q55.5    Congenital absence and aplasia of penis
Q55.6    Other congenital malformations of penis
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Congenital malformation of penis NOS
Curvature of penis lateral
Hypoplasia of penis
Micropenis
Penile duplication
Penoscrotal transposition

Q55.8 Other specified congenital malformations of male genital organs

Q55.9 Congenital malformation of male genital organ, unspecified
Congenital: .anomaly }
.deformity } NOS of male genital organ

Q56 Indeterminate sex and pseudohermaphroditism
Excludes: pseudohermaphroditism:
. female, with adrenocortical disorder (E25.-)
. male, with androgen resistance (E34.5)
. with specified chromosomal anomaly (Q96-Q99)

Q56.0 Hermaphroditism, not elsewhere classified
Ovotestis
Excludes: Chimera 46,XX/46,XY true hermaphrodite (Q99.0)

Q56.1 Male pseudohermaphroditism, not elsewhere classified
Male pseudohermaphroditism NOS

Q56.2 Female pseudohermaphroditism, not elsewhere classified
Female pseudohermaphroditism NOS

Q56.3 Pseudohermaphroditism, unspecified

Q56.4 Indeterminate sex, unspecified
Ambiguous genitalia

Q60-Q64 Congenital malformations of the urinary system

Q60 Renal agenesis and other reduction defects of kidney
Includes: atrophy of kidney: . congenital
. infantile
congenital absence of kidney

Q60.0 Renal agenesis, unilateral

Q60.1 Renal agenesis, bilateral

Q60.2 Renal agenesis, unspecified

Q60.3 Renal hypoplasia, unilateral

Q60.4 Renal hypoplasia, bilateral

Q60.5 Renal hypoplasia, unspecified

Q60.6 Potter's syndrome
Potter's sequence
Oligohydramnios sequence

Q61 Cystic kidney disease
Excludes: acquired cyst of kidney (N28.1)

Q61.0 Congenital single renal cyst
Cyst of kidney (congenital)(single)

Q61.1 Polycystic kidney, infantile type

Q61.2 Polycystic kidney, adult type

Q61.3 Polycystic kidney, unspecified

Q61.4 Renal dysplasia

Q61.40 Multicystic dysplastic kidney, unilateral
Cystic renal dysplasia, unilateral

Q61.41 Multicystic dysplastic kidney, bilateral
Cystic renal dysplasia, bilateral

Q61.48 Other specified renal dysplasia

Q61.5 Medullary cystic kidney
Sponge kidney NOS

Q61.50 Juvenile medullary cystic kidney
Nephronophthisis

Q61.51 Adult type medullary cystic kidney

Q61.52 Medullary sponge kidney

Q61.8 Other cystic kidney disease
@ Fibrocystic renal degeneration or disease
Cystic kidney disease associated with:
. tuberous sclerosis (Q85.1)
. Zellweger's syndrome (Q87.83)
Glomerular cystic disease
Q61.9 Cystic kidney disease, unspecified
Q61.90 Meckel-Gruber syndrome
Microcephalus with cystic kidney disease

Q62 Congenital obstructive defects of renal pelvis and congenital malformations of ureter
Q62.0 Congenital hydronephrosis
Ante-natally diagnosed hydronephrosis
Q62.1 Atresia and stenosis of ureter
Congenital occlusion of ureter
Impervious ureter
Q62.10 Congenital pelviureteric junction obstruction, unilateral
Q62.11 Congenital pelviureteric junction obstruction, bilateral
Q62.12 Congenital vesicoureteric junction obstruction, unilateral
Q62.13 Congenital vesicoureteric junction obstruction, bilateral
Q62.18 Other specified atresia and stenosis of ureter
Q62.2 Congenital megaloureter
Congenital dilatation of ureter
Q62.3 Other obstructive defects of renal pelvis and ureter
Congenital ureterocele
Q62.30 Ectopic ureterocele
Q62.31 Orthotopic ureterocele
Q62.32 Congenital polyp of ureter
Q62.33 Congenital hydroureter
Q62.4 Agenesis of ureter
Absent ureter
Q62.5 Duplication of ureter
Accessory ureter
Q62.50 Double ureter
Duplex ureter
Complete duplication of ureter
Q62.51 Triple ureter

For Q62.6 the following optional fifth character subdivision can be used if desired, to indicate the site of ureteric drainage:

.....0 bladder neck
.....1 urethra
.....2 vagina
.....3 vulva
.....4 vas deferens
.....5 seminal vesicles
.....8 other

Q62.6 Malposition of ureter
Deviation }
Displacement }
Ectopic } (of) ureter or ureteric orifice
Implantation, anomalous }
Q62.7 Congenital vesico-uretero-renal reflux
Congenital vesicoureteric reflux
VUR
Excludes: vesicoureteral-reflux-associated nephropathy (N13.7)
Q62.70 Congenital vesico-uretero-renal reflux, unilateral
Q62.71 Congenital vesico-uretero-renal reflux, bilateral
Q62.8 Other congenital malformations of ureter

```
          Anomaly of ureter NOS

Q63       Other congenital malformations of kidney
          Excludes: congenital nephrotic syndrome (N04.-)
Q63.0     Accessory kidney
Q63.00    Double or triple kidney
          Duplex or triplex kidney
Q63.1     Lobulated, fused and horseshoe kidney
            Renal fusion anomalies without ectopia
          Excludes: crossed ectopia of kidney with fusion anomaly (Q63.22)
Q63.10    Horseshoe kidney
Q63.18    Other specified renal fusion anomaly
Q63.19    Renal fusion anomaly, unspecified
Q63.2     Ectopic kidney
          Renal ectopia
          Congenital displaced kidney
          Malrotation of kidney
Q63.20    Pelvic kidney
Q63.21    Crossed ectopia of kidney (without fusion)
Q63.22    Crossed ectopia of kidney with fusion anomaly
Q63.28    Other specified renal ectopia
Q63.29    Renal ectopia, unspecified
Q63.3     Hyperplastic and giant kidney
Q63.8   @ Other specified congenital malformations of kidney
Q63.81    Congenital calyceal diverticulum
Q63.9     Congenital malformation of kidney, unspecified

Q64       Other congenital malformations of urinary system
Q64.0     Epispadias
          Excludes: hypospadias (Q54.-)
Q64.1     Exstrophy of urinary bladder
          Ectopia vesicae
          Extroversion of bladder
Q64.10    Cloacal exstrophy
          Ectopia cloacae
Q64.20    Congenital posterior urethral valves
Q64.21    Congenital anterior urethral valves
Q64.3     Other atresia and stenosis of urethra and bladder neck
@          Impervious urethra
Q64.30    Congenital bladder neck obstruction
Q64.31    Congenital stricture of urethra
          Congenital stricture of anterior urethra
Q64.32    Congenital stricture of urethral meatus
Q64.33    Hypoplasia of urethra
          Atresia of urethra
Q64.4     Malformation of urachus
Q64.40    Cyst of urachus
Q64.41    Patent urachus
Q64.42    Urachal diverticulum
Q64.48    Other specified malformation of urachus
          Prolapse of urachus
Q64.5     Congenital absence of bladder and urethra
Q64.6     Congenital diverticulum of bladder
          Congenital paraureteric diverticulum
Q64.7     Other congenital malformations of bladder and urethra
          Accessory: .bladder
                     .urethra
          Congenital: .hernia of bladder
                      .malformation of bladder or urethra NOS
                      .prolapse of: . urethra
                                    . urinary meatus
Q64.70    Anterior urethral diverticulum
Q64.71    Congenital prolapse of bladder (mucosa)
```

Q64.72 Double urethra
Double urinary meatus
Q64.73 Ectopic urethra or urethral orifice
Q64.74 Congenital gastrointestinal-urinary tract fistula
Congenital: . urethrorectal fistula
. rectovesical fistula
Q64.75 Congenital megalourethra
Q64.76 Megacystis-megaureter syndrome
Q64.78 Congenital urethral syringocele
Q64.8 Other specified congenital malformations of urinary system
Q64.9 Congenital malformation of urinary system, unspecified
Congenital: .anomaly }
.deformity} NOS of urinary system

Q65-Q79 Congenital malformations and deformations of musculoskeletal system

Q65 Congenital deformities of hip
CDH
Excludes: clicking hip (R29.4)
Q65.0 Congenital dislocation of hip, unilateral
Q65.1 Congenital dislocation of hip, bilateral
Q65.2 Congenital dislocation of hip, unspecified
Q65.3 Congenital subluxation of hip, unilateral
Q65.4 Congenital subluxation of hip, bilateral
Q65.5 Congenital subluxation of hip, unspecified
Q65.6 Unstable hip
Dislocatable hip
Subluxatable hip
Q65.60 Unstable hip, unilateral
Q65.61 Unstable hip, bilateral
Q65.8 Other congenital deformities of hip
Q65.80 Dysplastic hip, unilateral
Congenital acetabular dysplasia, unilateral
Q65.81 Dysplastic hip, bilateral
Congenital acetabular dysplasia, bilateral
Q65.82 Anteversion of femoral neck
Anteversion of femur
Q65.83 Congenital coxa valga
Q65.84 Congenital coxa vara
Q65.9 Congenital deformity of hip, unspecified

Q66 Congenital deformities of feet
Excludes: reduction defects of feet (Q72.-)
valgus deformities (acquired) (M21.0)
varus deformities (acquired) (M21.1)
Q66.0 Talipes equinovarus
Q66.1 Talipes calcaneovarus
Q66.2 Metatarsus varus
Metatarsus adductus
Q66.3 Other congenital varus deformities of feet
Hallux varus, congenital
Q66.4 Talipes calcaneovalgus
Q66.5 Congenital pes planus
Flat foot: .congenital
.rigid
.spastic (everted)
Excludes: pes planus acquired (M21.4)
Q66.6 Other congenital valgus deformities of feet
Metatarsus valgus
Q66.7 Pes cavus
Q66.8 Other congenital deformities of feet
Clubfoot NOS

Hammer toe, congenital
Talipes: . NOS
. asymmetric
Tarsal coalition
Vertical talus

Q66.80 Rocker bottom foot
Q66.81 Congenital short Achilles tendon
Q66.9 Congenital deformity of feet, unspecified

Q67 Congenital musculoskeletal deformities of head, face, spine and chest
Excludes: congenital malformation syndromes classified to Q87.-
Potter's sequence [syndrome] (Q60.6)
Q67.0 Facial asymmetry
Q67.1 Compression facies
Excludes: Potter's facies (Q60.6)
Q67.2 Dolichocephaly
Q67.3 Plagiocephaly
Asymmetric head
Q67.4 Other congenital deformities of skull, face and jaw
Hemifacial atrophy or hypertrophy
Squashed or bent nose, congenital
Excludes: dentofacial anomalies [including malocclusion] (K07.-)
syphilitic saddle nose (A50.5)
Goldenhar syndrome [oculo-auriculo-vertebral syndrome] (Q87.04)
Q67.40 Depressions in skull
Q67.41 Deviation of nasal septum, congenital
Q67.5 Congenital deformity of spine
@ Excludes: infantile idiopathic scoliosis (M41.0)
scoliosis due to congenital bony malformation (Q76.3)
Q67.50 Congenital scoliosis, postural
Q67.52 Congenital postural curvature of spine, NOS
Q67.58 Other specified congenital deformity of spine
Q67.6 Pectus excavatum
Congenital funnel chest
Q67.7 Pectus carinatum
Congenital pigeon chest
Q67.8 Other congenital deformities of chest
Congenital deformity of chest wall NOS

Q68 Other congenital musculoskeletal deformities
Excludes: reduction defects of limb(s) (Q71-Q73)
Q68.0 Congenital deformity of sternocleidomastoid muscle
Congenital (sternomastoid) torticollis
Contracture of sternocleidomastoid (muscle)
Sternomastoid tumour (congenital)
Excludes: sternomastoid swelling due to birth trauma (P15.2)
Q68.1 Congenital deformity of hand
@ Congenital clubfinger
Camptodactyly
Q68.10 Clinodactyly
Q68.2 Congenital deformity of knee
Q68.20 Congenital dislocation of knee
Q68.21 Congenital genu recurvatum
Q68.28 Other specified congenital deformity of knee
Q68.3 Congenital bowing of femur
Excludes: anteversion of femur (neck) (Q65.8)
Q68.4 Congenital bowing of tibia and fibula
Q68.5 Congenital bowing of long bones of leg, unspecified
Q68.8 Other specified congenital musculoskeletal deformities
@ Congenital deformity of: .clavicle
.elbow

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                                .forearm
                                .scapula
          Congenital dislocation of shoulder
          Arthrogryposis NOS
         Excludes: arthrogryposis multiplex congenita (Q74.3)
Q68.80   Congenital dislocation of radial head

Q69      Polydactyly
         Excludes: acrocephalopolysyndactyly (Q87.01)
          For Q69.0-Q69.2 the following RCPCH fifth-character
           extensions can be used if desired:

              .....0 Preaxial
              .....1 Mesoaxial
              .....2 Postaxial
              .....9 unspecified

Q69.0    Accessory finger(s)
         Supernumerary finger(s)
Q69.1    Accessory thumb(s)
         Supernumerary thumb(s)
Q69.2    Accessory toe(s)
         Supernumerary toe(s)
         Accessory [supernumerary] hallux
Q69.9    Polydactyly, unspecified
         Supernumerary digit(s) NOS

Q70      Syndactyly
         Excludes: acrocephalopolysyndactyly (Q87.00)
                   acrocephalosyndactyly (Q87.01)
Q70.0    Fused fingers
         Complex syndactyly of fingers with synostosis
Q70.1    Webbed fingers
         Simple syndactyly of fingers without synostosis
Q70.2    Fused toes
         Complex syndactyly of toes with synostosis
Q70.3    Webbed toes
         Simple syndactyly of toes without synostosis
Q70.4    Polysyndactyly
Q70.9    Syndactyly, unspecified
Q70.90   Symphalangism
         Symphalangy NOS

Q71      Reduction defects of upper limb
Q71.0    Congenital complete absence of upper limb(s)
         Amelia of upper limb
Q71.1    Congenital absence of upper arm and forearm with hand present
         Phocomelia of upper limb
Q71.2    Congenital absence of both forearm and hand
Q71.3    Congenital absence of hand and finger(s)
Q71.30   Congenital absence of finger(s)
         [Remainder of hand intact]
Q71.31   Absence or hypoplasia of thumb
         [Other digits intact]
Q71.4    Longitudinal reduction defect of radius
          Clubhand (congenital)
          Radial clubhand
          Absence of radius
         Excludes: thrombocytopenia with absent radius syndrome (Q87.25)
                   Fanconi's anaemia with absent radius (D61.0)
Q71.5    Longitudinal reduction defect of ulna
Q71.6    Lobster-claw hand
         Congenital cleft hand
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Q71.8 Other reduction defects of upper limb(s)
Congenital shortening of upper limb(s)
Q71.9 Reduction defect of upper limb, unspecified
Congenital amputation of upper limb NOS
Constriction ring syndrome of upper limb NOS

Q72 Reduction defects of lower limb
Q72.0 Congenital complete absence of lower limb(s)
Amelia of lower limb
Q72.1 Congenital absence of thigh and lower leg with foot present
Phocomelia of lower limb
Q72.2 Congenital absence of both lower leg and foot
Q72.3 Congenital absence of foot and toe(s)
Q72.30 Congenital absence or hypoplasia of toe(s) with remainder of foot intact
Q72.31 Absence or hypoplasia of first toe with other digits present
Q72.4 Longitudinal reduction defect of femur
Proximal femoral focal deficiency
Q72.5 Longitudinal reduction defect of tibia
Absence of tibia
Q72.6 Longitudinal reduction defect of fibula
Absence of fibula
Q72.7 Split foot
Q72.8 Other reduction defects of lower limb(s)
Congenital shortening of lower limb(s)
Q72.9 Reduction defect of lower limb, unspecified
Congenital amputation of lower limb NOS
Constriction ring syndrome of lower limb NOS

Q73 Reduction defects of unspecified limb
Q73.0 Congenital absence of unspecified limb(s)
Amelia NOS
Q73.1 Phocomelia, unspecified limb(s)
Phocomelia NOS
Q73.8 Other reduction defects of unspecified limb(s)
Longitudinal reduction deformity of unspecified limb(s)
Ectromelia NOS } of limb(s) NOS
Hemimelia NOS } of limb(s) NOS
Reduction defect } of limb(s) NOS
Amputation of unspecified limb(s)
Constriction ring syndrome of unspecified limb(s)
Q73.80 Absent digits NOS
Excludes: congenital absence of all fingers (Q71.80)
congenital absence of all toes (Q72.80)

Q74 Other congenital malformations of limb(s)
Excludes: polydactyly (Q69.-)
reduction defect of limb (Q71-Q73)
syndactyly (Q70.-)
Q74.0 Other congenital malformations of upper limb(s), including shoulder girdle
Congenital pseudoarthrosis of clavicle
Congenital cubitus valgus or varus
Q74.00 Accessory carpal bones
Q74.01 Madelung's deformity
Q74.02 Cleidocranial dysostosis
Q74.03 Sprengel's deformity
Congenital elevation of the scapula
Q74.04 Macrodactylia (fingers)
Q74.05 Triphalangeal thumb
Q74.06 Radioulnar synostosis
Radioulnar dysostosis
Q74.07 Humeroulnar synostosis

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Q74.08   Humeroradial synostosis
Q74.09   Bifid digit(s) of upper limb
Q74.1    Congenital malformation of knee
           Congenital: .absence of patella
                       .dislocation of patella
                       .genu: . valgum
                              . varum
           Rudimentary patella
          Excludes: congenital: . dislocation of knee(Q68.2)
                                . genu recurvatum(Q68.2)
                    nail patella syndrome(Q87.2)
Q74.2   Other congenital malformations of lower limb(s), including
         pelvic girdle
           Congenital malformation (of): .ankle (joint)
                                         .sacroiliac (joint)
            Excludes: anteversion of femur (neck) (Q65.8)
Q74.20   Congenital fusion of sacroiliac joint
Q74.21   Astragaloscaphoid synostosis
Q74.22   Congenital angulation of tibia
Q74.23   Bifid digit(s) of lower limb
Q74.3    Arthrogryposis multiplex congenita
          Excludes: primary disorders of muscles (G71.-)
                    congenital viral myositis (P35.8)
                    infantile spinal muscular atrophy (G12.0)
Q74.8    Other specified congenital malformations of limb(s)
Q74.80   Brachydactyly
Q74.81   Congenital overgrowth of limb(s)
          Congenital hemihypertrophy
Q74.82   Congenital undergrowth of limb(s)
          Excludes: hemiatrophy NOS (R68.82)
Q74.83   Congenital limb asymmetry, unspecified
Q74.84   Larsen's syndrome
Q74.9    Unspecified congenital malformation of limb(s)
          Congenital anomaly of limb(s) NOS

Q75      Other congenital malformations of skull and face bones
          Excludes: congenital malformation of face NOS (Q18.-)
                    congenital malformation syndromes classified to Q87.-
                    dentofacial anomalies [including malocclusion] (K07.-)
                    musculoskeletal deformities of head and face (Q67.0-Q67.4)
                    skull defects associated with congenital anomalies of
                     brain such as: . anencephaly (Q00.0)
                                    . encephalocele (Q01.-)
                                    . hydrocephalus (Q03.-)
                                    . microcephaly (Q02)
Q75.0    Craniosynostosis
           Imperfect fusion of skull
           Pfeiffer syndrome
          Excludes: thanatophoric dwarfism/trigonocephaly association (Q77.1)
                    acrocephalo(poly)syndactyly (Q87.0-)
                    clover leaf skull (Kleeblattsch„del
                     deformity syndrome) (Q03.80)
Q75.00   Coronal craniosynostosis
          Brachycephaly
Q75.01   Sagittal craniosynostosis
          Scaphocephaly
Q75.02   Trigonocephaly
          Excludes: thanatophoric dwarfism (Q77.1)
Q75.03   Craniosynostosis of other multiple sutures
          Acrocephaly
          Oxycephaly
          Turricephaly
Q75.1    Craniofacial dysostosis
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Crouzon's disease
Q75.2 Hypertelorism
Q75.3 Macrocephaly
Q75.30 Familial (benign) macrocephaly
Q75.4 Mandibulofacial dysostosis
Note: Code Q75.4 is to be used for the isolated anomaly of skull and face bones. When this condition occurs as part of Treacher Collins [-Franceschetti] [-Klein] syndrome use (Q87.0A).
Q75.5 Oculomandibular dysostosis
Note: Code Q75.5 is to be used for the isolated anomaly of skull and face bones. When this condition occurs as part of Hallerman-Streiff syndrome use (Q87.05).
Q75.8 Other specified congenital malformations of skull and face bones
Absence of skull bone, congenital
Congenital deformity of forehead
Platybasia
Q75.80 Localised skull defects
Q75.81 Frontonasal dysplasia
Median cleft facial syndrome
Q75.9 Congenital malformation of skull and face bones, unspecified
Congenital anomaly of: . face bones NOS
. skull NOS

Q76 Congenital malformations of spine and bony thorax
Excludes: congenital musculoskeletal deformities of spine and chest (Q67.5-Q67.8)
Q76.0 Spina bifida occulta
Excludes: meningocele (spinal) (Q05.-)
spina bifida (aperta)(cystica) (Q05.-)
Q76.1 Klippel-Feil syndrome
Cervical fusion syndrome
Q76.2 Congenital spondylolisthesis
Congenital spondylolysis
Excludes: spondylolisthesis (acquired) (M43.1)
spondylolysis (acquired) (M43.0)
Q76.3 Congenital scoliosis due to congenital bony malformation
@ Kyphoscoliosis due to congenital bony malformation
Fusion or failure of segmentation with scoliosis
Q76.30 Single hemivertebra with congenital scoliosis
Q76.38 Congenital scoliosis due to other specified congenital bony malformation
Q76.4 Other congenital malformations of spine, not associated with
@ scoliosis
Congenital:
. fusion of spine }
. gibbus }
. kyphosis }
. lordosis } unspecified or
. malformation of lumbosacral (joint) (region) } not associated with scoliosis
Malformation of spine }
Platyspondylisis }
Supernumerary vertebra }
Q76.40 Congenital absence of vertebra(e)
Q76.41 Congenital anomalies of sacral vertebrae
Sacral agenesis
Q76.42 Congenital anomalies of other vertebrae
Q76.43 Congenital lordosis, postural
Q76.5 Cervical rib
Supernumerary rib in cervical region
Q76.6 Other congenital malformations of ribs
Congenital malformation of ribs NOS

Excludes: short rib syndrome (Q77.2)
Q76.60 Congenital absence of rib
Q76.61 Congenital fusion of ribs
Q76.62 Accessory rib
Excludes: cervical rib (Q76.5)
Q76.7 Congenital malformation of sternum
Misshapen sternum
Excludes: pectus excavatum (Q67.6)
pectus carinatum (Q67.7)
Q76.70 Congenital absence of sternum
Q76.71 Sternum bifidum
Q76.78 Other specified congenital malformation of sternum
Q76.8 Other congenital malformations of bony thorax
Q76.9 Congenital malformation of bony thorax, unspecified

Q77 Osteochondrodysplasia with defects of growth of tubular bones and spine
Excludes: mucopolysaccharidosis (E76.0-E76.3)
Q77.0 Achondrogenesis
Q77.00 Achondrogenesis, type I
Q77.01 Achondrogenesis, type II
Q77.02 Hypochondrogenesis
Q77.1 Thanatophoric short stature
Thanatophoric dwarfism/trigonocephaly association
Thanatophoric dysplasia (with clover leaf skull)
Q77.2 Short rib syndrome
Asphyxiating thoracic dysplasia [Jeune]
Jeune's syndrome
Q77.3 Chondrodysplasia punctata
Chondrodystrophia calcificans congenita
Conradi (-Hunerman) syndrome
Congenital multiple epiphyseal dysplasia
Rhizomelic syndrome
Excludes: warfarin embryopathy (Q86.2)
Q77.4 Achondroplasia
Achondroplastic dwarfism
Hypochondroplasia
Q77.5 Diastrophic dysplasia
Diastrophic dwarfism
Q77.6 Chondroectodermal dysplasia
Ellis-van Creveld syndrome
Q77.7 Spondyloepiphyseal dysplasia
Q77.8 Other osteochondrodysplasia with defects of growth of tubular bones and spine
Acrodysostosis
Kniest dysplasia
Q77.80 Metatropic dwarfism
Metatropic dysplasia
Q77.81 Metaphyseal chondrodysplasia
Metaphyseal dysostosis
Q77.9 Osteochondrodysplasia with defects of growth of tubular bones and spine,unspecified

Q78 Other osteochondrodysplasias
Q78.0 Osteogenesis imperfecta
Fragilitas ossium
Osteopsathyrosis
Q78.00 Osteogenesis imperfecta congenita
Q78.08 Other osteogenesis imperfecta
Osteogenesis imperfecta tarda
Q78.1 Polyostotic fibrous dysplasia
McCune-Albright(-Sternberg) syndrome
Q78.2 Osteopetrosis

Albers-Schönberg syndrome
Marble bone disease

Q78.3 Progressive diaphyseal dysplasia
Camurati-Engelmann syndrome

Q78.4 Enchondromatosis

Q78.40 Enchondromatosis with haemangiomata
Maffuci's syndrome [Kast's syndrome]

Q78.48 Other specified enchondromatosis
Dyschondroplasia
Ollier's disease
Osteochondromatosis syndrome
Excludes: osteochondromatosis, NOS (D48.0)

Q78.5 Metaphyseal dysplasia
Pyle's syndrome

Q78.6 Multiple congenital exostoses
Diaphyseal aclasis

Q78.8 Other specified osteochondrodysplasias
Excludes: chondrodystrophic myotonia [Schwartz-Jampel] (G71.16)

Q78.80 Osteopoikilosis

Q78.9 Osteochondrodysplasia, unspecified
Chondrodystrophy NOS
Osteodystrophy NOS

Q79 Congenital malformations of the musculoskeletal system, not elsewhere classified
Excludes: congenital (sternomastoid) torticollis (Q68.0)

Q79.0 Congenital diaphragmatic hernia
Excludes: congenital hiatus hernia (Q40.1)

Q79.00 Congenital anterior (foramen of Morgagni) hernia

Q79.01 Congenital posterolateral (foramen of Bochdalek) hernia

Q79.1 Other congenital malformations of diaphragm
Congenital malformation of diaphragm NOS

Q79.10 Congenital eventration of diaphragm

Q79.11 Congenital absent hemidiaphragm, (unilateral)

Q79.12 Congenital absent diaphragm
Congenital absent hemidiaphragm, bilateral

Q79.2 Exomphalos
Omphalocele
Excludes: umbilical hernia (K42.-)

Q79.3 Gastroschisis

Q79.4 Prune belly syndrome

Q79.5 Other congenital malformations of abdominal wall
Excludes: umbilical hernia (K42.-)

Q79.6 Ehlers-Danlos syndrome

Q79.8 Other congenital malformations of the musculoskeletal system
Accessory muscle
Popliteal web syndrome
Congenital shortening of tendon
Excludes: achilles tendon (Q66.81)

Q79.80 Congenital constriction bands

Q79.81 Absence of muscle and/or tendon

Q79.82 Poland's anomaly [syndrome]

Q79.9 Congenital malformation of musculoskeletal system, unspecified
Congenital: .anomaly NOS }
.deformity NOS} of musculoskeletal system NOS
Unspecified anomalies of muscle, tendon, bones, cartilage or connective tissue

Q80-Q89 Other congenital malformations

Q80 Congenital ichthyosis
Excludes: Refsum's disease (G60.1)

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Q80.0    Ichthyosis vulgaris
Q80.1    X-linked ichthyosis
Q80.2    Lamellar ichthyosis
          (Non-bullous ichthyosiform erythroderma)
          Severe form known as - Collodion baby
Q80.3    Congenital bullous ichthyosiform erythroderma
          (Epidermolytic hyperkeratosis)
Q80.4    Harlequin fetus
Q80.8    Other congenital ichthyosis
          Excludes: Sj”gren-Larsson syndrome (Q87.1A)
Q80.9    Congenital ichthyosis unspecified

Q81      Epidermolysis bullosa
Q81.0    Epidermolysis bullosa simplex
          Excludes: Cockayne's syndrome (Q87.1)
Q81.1    Epidermolysis bullosa letalis
          Herlitz' syndrome
Q81.2    Epidermolysis bullosa dystrophica
Q81.8    Other epidermolysis bullosa
Q81.9    Epidermolysis bullosa, unspecified

Q82      Other congenital malformations of skin
          Excludes: acrodermatitis enteropathica (E83.2)
                    congenital erythropoietic porphyria (E80.0)
                    pilonidal cyst or sinus (L05.-)
                    Sturge-Weber(-Dimitri) syndrome (Q85.8)
Q82.0    Hereditary lymphoedema
Q82.1    Xeroderma pigmentosum
Q82.2    Mastocytosis
           Urticaria pigmentosa
          Excludes: malignant mastocytosis (C96.2)
Q82.3    Incontinentia pigmenti
Q82.4    Ectodermal dysplasia (anhidrotic)
          Excludes: Ellis-van Creveld syndrome (Q77.6)
                    ectodermal dysplasia, hidrotic (Q82.82)
Q82.5    Congenital non-neoplastic naevus
           Birthmark NOS
           Naevus: .sanguineous
                   .vascular NOS
                   .verrucous
          Excludes: caf, au lait spots (L81.3)
                    lentigo (L81.4)
                    naevus: . NOS (D22.-)
                            . araneus (I78.1)
                            . melanocytic (D22.-)
                            . pigmented (D22.-)
                            . spider (I78.1)
                            . stellar (I78.1)
                    capillary haemangioma (D18.00)
                    cavernous haemangioma (D18.01)
                    mixed haemangioma (D18.02)
Q82.50   Naevus flammeus [Portwine stain]
Q82.51   Strawberry naevus
              Note: This term should be used for typical strawberry
                     naevi. Massive, non-superficial or otherwise
                     atypical lesions should be coded to D18.0-.
Q82.52   Mongolian blue spot
Q82.58   Other specified congenital non-neoplastic naevus
Q82.8    Other specified congenital malformations of skin
           Benign familiar pemphigus [Hailey-Hailey]
           Cutis laxa (hyperelastica)
           Dermatoglyphic anomalies [excludes: abnormal palmar
            creases - Q82.80]
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          Inherited keratosis palmaris et plantaris
          Keratosis follicularis [Darier-White]
         Excludes: Ehlers-Danlos syndrome (Q79.6)
Q82.80   Abnormal palmar creases
Q82.81   Accessory skin tags
Q82.82   Ectodermal dysplasia, hidrotic
         Excludes: ectodermal dysplasia, anhidrotic (Q82.4)
Q82.83   Hypomelanosis of Ito
Q82.9    Congenital malformation of skin, unspecified

Q83      Congenital malformations of breast
         Excludes: absence of pectoral muscle (Q79.81)
Q83.0    Congenital absence of breast with absent nipple
Q83.1    Accessory breast
         Supernumerary breast
Q83.2    Absent nipple
Q83.3    Accessory nipple
         Supernumerary nipple
Q83.8    Other congenital malformations of breast
         Hypoplasia of breast
Q83.9    Congenital malformation of breast, unspecified

Q84      Other congenital malformations of integument
Q84.0    Congenital alopecia
         Congenital atrichosis
Q84.1    Congenital morphological disturbances of hair, not elsewhere
         classified
          Beaded hair
          Monilethrix
          Pili annulati
          Pili torti
         Excludes: Menkes' kinky hair syndrome (E83.0)
Q84.2    Other congenital malformations of hair
         Congenital malformation of hair NOS
         Persistent lanugo
Q84.20   Congenital hypertrichosis
Q84.3    Anonychia
          Congenital absent nails
         Excludes: nail patella syndrome (Q87.2)
Q84.4    Congenital leukonychia
Q84.5    Enlarged and hypertrophic nails
         Congenital onychauxis
         Pachyonychia
Q84.6    Other congenital malformations of nails
         Congenital: .clubnail
                     .koilonychia
                     .malformation of nail NOS
Q84.8    Other specified congenital malformations of integument
Q84.80   Aplasia cutis congenita
Q84.9    Congenital malformation of integument, unspecified
         Congenital: .anomaly NOS  }
                     .deformity NOS} of integument NOS

Q85      Phakomatoses, not elsewhere classified
         Excludes: ataxia-telangiectasia [Louis-Bar] (G11.30)
                   familial dysautonomia [Riley-Day] (G90.1)
Q85.0    Neurofibromatosis (nonmalignant)
         Von Recklinghausen's disease
Q85.1    Tuberous sclerosis
         Bourneville's disease
         Epiloia
Q85.8    Other phakomatoses, not elsewhere classified
         Excludes: Meckel-Gruber syndrome (Q61.9)
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Q85.80   Peutz-Jeghers syndrome
Q85.81   Sturge-Weber(-Dimitri) syndrome
Q85.82   Von Hippel-Lindau syndrome
Q85.83   Gardner's syndrome
          Osteomatosis-intestinal polyposis syndrome
Q85.9    Phakomatosis, unspecified
          Hamartosis NOS

Q86      Congenital malformation syndromes due to known exogenous
           causes, not elsewhere classified
          Excludes: iodine-deficiency-related hypothyroidism (E00-E02)
                    nonteratogenic effects of substances transmitted
                     via placenta or breast milk (P04.-)
Q86.0    Fetal alcohol syndrome (dysmorphic)
Q86.1  @ Fetal hydantoin syndrome
Q86.2    Dysmorphism due to warfarin
Q86.8    Other congenital malformation syndromes due to known
          exogenous causes
           Congenital malformations due to methylmercury
Q86.80   Congenital malformations due to valproate
Q86.81   Congenital malformations due to Vitamin A
Q86.82   Congenital malformations due to thalidomide
Q86.83   Congenital malformations due to cytotoxic agents
Q86.84   Congenital malformations due to other drugs
Q86.85   Congenital malformations due to ionising radiation

Q87      Other specified congenital malformation syndromes affecting
          multiple systems
Q87.0    Congenital malformation syndromes predominantly affecting
          facial appearance
           Excludes: cherubism (K10.80)
                     Waardenburg's syndrome (E70.30)
Q87.00   Acrocephalopolysyndactyly
          Acrocephalopolysyndactyly type I, Noack syndrome
          Acrocephalopolysyndactyly type II, Carpenter syndrome
Q87.01   Acrocephalosyndactyly
          Apert's syndrome
          Vogt cephalodactyly
Q87.02   Cryptophthalmos syndrome
Q87.03   Cyclopia [cyclops][cyclopism][synophthalmia]
Q87.04   Goldenhar syndrome
          Oculo-auriculo-vertebral syndrome [Hemifacial microsomia syndrome]
Q87.05   Hallerman-Streiff syndrome
          Excludes: (isolated) oculomandibular dysostosis (Q75.5)
Q87.06   Moebius syndrome
Q87.07   Oro-facial-digital syndrome
          Oro-facial-digital syndrome types I and II
          Mohr syndrome
Q87.08   Pierre Robin sequence
          Robin syndrome/sequence
Q87.09   Stickler syndrome
          Hereditary progressive arthro-ophthalmopathy
Q87.0A   Treacher Collins [-Franceschetti] [-Klein] syndrome
          Excludes: (isolated) mandibulofacial dysostosis (Q75.4)
Q87.0B   Trico-rhino-phalangeal syndrome
          Type I
          Type II [Langer-Giedion]
Q87.0C   Whistling face syndrome
Q87.0D   Ullrich-Feichtiger's syndrome
          Dyscraniopygophalangism
Q87.0E   Pena-Shokeir syndrome
          Camptodactyly-ankyloses-facial anomalies-pulmonary
           hypoplasia syndrome
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Q87.0F Other specified congenital malformation syndromes predominantly affecting facial appearance

Q87.1 Congenital malformation syndromes predominantly associated with short stature
Excludes: Ellis-van Creveld syndrome (Q77.6)
Q87.10 Aarskog syndrome
Q87.11 Cockayne syndrome
Q87.12 Cornelia de Lange syndrome
Amsterdam dwarf [Brachmann-de Lange syndrome]
Q87.13 Dubowitz syndrome
Q87.14 Noonan syndrome
Q87.15 Prader-Willi syndrome
Q87.16 Robinow-Silverman-Smith syndrome
Q87.17 Russell-Silver syndrome
Q87.18 Seckel syndrome
Bird-headed dwarfism
Microcephalic primordial dwarfism
Q87.19 Smith-Lemli-Opitz syndrome
7-dehydrocholesterol reductase deficiency
Q87.1A Sj”gren-Larsson syndrome
Fatty alcohol:nicotinamide adenine dinucleotide oxido-reductase deficiency
Q87.1B Other specified congenital malformation syndromes predominantly associated with short stature

Q87.2 Congenital malformation syndromes predominantly involving limbs
Excludes: Fanconi's anaemia with absent radius (D61.0)
Q87.20 Holt-Oram syndrome
Q87.21 Klippel-Tr,naunay-Weber syndrome
Q87.22 Nail patella syndrome
Q87.23 Rubinstein-Taybi syndrome
Q87.24 Sirenomelia syndrome
Q87.25 Thrombocytopenia with absent radius syndrome
TAR syndrome
Q87.26 VATER association
VACTERL association
Q87.28 Other specified congenital malformation syndromes predominantly involving limbs

Q87.3 Congenital malformation syndromes involving early overgrowth
Q87.30 Beckwith-Wiedemann syndrome
Beckwith's syndrome
Q87.31 Sotos syndrome
Cerebral gigantism
Q87.32 Weaver syndrome
Q87.38 Other specified congenital malformation syndromes involving early overgrowth

Q87.4 Marfan's syndrome
Arachnodactyly NOS
Q87.5 Other congenital malformation syndromes with other skeletal changes

Q87.8 Other specified congenital malformation syndromes, not elsewhere classified
Q87.80 Alport's syndrome
Q87.81 Laurence-Moon-Biedl syndrome
Laurence-Moon(-Bardet)-Biedl syndrome

Q87.83 Zellweger syndrome
Note: this is a peroxisomal disorder

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          Excludes: Zellweger-like syndrome (E88.8F)
                    pseudo-Zellweger syndrome (E88.8J)

Q87.84   William's syndrome
Q87.85   Angelman's syndrome
          [Happy puppet syndrome]

Q89      Other congenital malformations, not elsewhere classified
Q89.0    Congenital malformations of spleen
           Congenital splenomegaly [hyperplasia of spleen]
           Hypoplasia of }
           Mis-shapen    }
           Accessory     }  spleen
           Ectopic       }
          Excludes: isomerism of atrial appendages (with
                     asplenia or polysplenia) (Q20.6)
Q89.00   Congenital asplenia
          Congenital absence of spleen
Q89.08   Other specified congenital malformation of spleen
Q89.1    Congenital malformations of adrenal gland
           Accessory    } adrenal gland
           Ectopic      }
          Excludes: congenital adrenal hyperplasia (E25.0)
Q89.10   Congenital absence of adrenal gland
Q89.11   Congenital adrenal hypoplasia
Q89.18   Other specified congenital malformation of adrenal gland
Q89.2    Congenital malformations of other endocrine glands
Q89.20   Congenital malformations of pituitary gland
Q89.21   Congenital malformations of thyroid gland
Q89.22   Persistent thyroglossal duct
Q89.23   Thyroglossal cyst
Q89.24   Congenital malformations of parathyroid gland
Q89.25   Congenital malformations of thymus
Q89.3    Situs inversus
          Excludes: dextrocardia NOS (Q24.0)
Q89.30   Dextrocardia with situs inversus
Q89.31   Mirror-image atrial arrangement with situs inversus
Q89.32   Situs inversus abdominalis
          Situs transversus abdominalis
          Transposition of abdominal viscera
Q89.33   Situs inversus thoracis
          Situs transversus thoracis
          Transposition of thoracic viscera
Q89.34   Kartagener's syndrome
           Kartagener's triad
          Excludes: other immotile cilia syndromes (J98.80)
Q89.38   Other specified situs inversus
Q89.4    Conjoined twins
Q89.40   Dicephaly
          Two heads
Q89.41   Craniopagus
          Head-joined twins
Q89.42   Thoracopagus
          Thorax-joined twins
Q89.43   Xiphopagus
          Xiphoid and pelvis-joined twins
Q89.44   Pygopagus
          Buttock-joined twins
Q89.45   Double monster
Q89.48   Other specified conjoined twins
Q89.7    Multiple congenital malformations, not elsewhere classified
@          Multiple congenital: .anomalies NOS
                                .deformities NOS
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Excludes: congenital malformation syndromes affecting multiple systems (Q87.-)

Q89.8 Other specified congenital malformations

Q89.80 Caudal dysplasia sequence

Q89.9 Congenital malformation, unspecified

Congenital: .anomaly NOS

.deformity NOS

Q90-Q99 Chromosomal abnormalities, not elsewhere classified

Q90 Down's syndrome

Q90.0 Trisomy 21, meiotic nondisjunction

Q90.1 Trisomy 21, mosaicism (mitotic nondisjunction)

Q90.2 Trisomy 21, translocation

Q90.9 Down's syndrome, unspecified

Trisomy 21 NOS

Q91 Edward's syndrome and Patau's syndrome

Q91.0 Trisomy 18, meiotic nondisjunction

Q91.1 Trisomy 18, mosaicism (mitotic nondisjunction)

Q91.2 Trisomy 18, translocation

Q91.3 Edward's syndrome, unspecified

Q91.4 Trisomy 13, meiotic nondisjunction

Q91.5 Trisomy 13, mosaicism (mitotic nondisjunction)

Q91.6 Trisomy 13, translocation

Q91.7 Patau's syndrome, unspecified

Q92 Other trisomies and partial trisomies of the autosomes, not elsewhere classified

Includes: unbalanced translocations and insertions

Excludes: trisomies of chromosomes 13, 18, 21 (Q90-Q91)

Q92.0 Whole chromosome trisomy, meiotic nondisjunction

Q92.1 Whole chromosome trisomy, mosaicism (mitotic nondisjunction)

Q92.2 Major partial trisomy

Whole arm or more duplicated

Q92.3 Minor partial trisomy

Less than whole arm duplicated

Q92.4 Duplications seen only at prometaphase

Q92.5 Duplications with other complex rearrangements

Q92.6 Extra marker chromosomes

Q92.7 Triploidy and polyploidy

Q92.8 Other specified trisomies and partial trisomies of autosomes

Q92.9 Trisomy and partial trisomy of autosomes, unspecified

Q93 Monosomies and deletions from the autosomes, not elsewhere classified

Q93.0 Whole chromosome monosomy, meiotic nondisjunction

Q93.1 Whole chromosome monosomy, mosaicism (mitotic nondisjunction)

Q93.2 Chromosome replaced with ring or dicentric

Q93.3 Deletion of short arm of chromosome 4

Wolff-Hirschorn syndrome

Q93.4 Deletion of short arm of chromosome 5

Cri du chat syndrome

Q93.5 Other deletions of part of a chromosome

Deletion of long arm of chromosome 13

Deletion of long or short arm of chromosome 18 [18p- or 18q syndrome]

Q93.50 Deletion of long arm of chromosome 21

Anti-mongolism syndrome

Q93.6 Deletions seen only at prometaphase

Q93.7 Deletions with other complex rearrangements

Q93.8 Other deletions from the autosomes

Q93.9 Deletion from autosomes, unspecified

Q95 Balanced rearrangements and structural markers, not elsewhere classified
Includes: Robertsonian and balanced reciprocal translocations and insertions
Q95.0 Balanced translocation and insertion in normal individual
Q95.1 Chromosome inversion in normal individual
Q95.2 Balanced autosomal rearrangement in abnormal individual
Q95.3 Balanced sex/autosomal rearrangement in abnormal individual
Q95.4 Individuals with marker heterochromatin
Q95.5 Individuals with autosomal fragile site
Q95.8 Other balanced rearrangements and structural markers
Q95.9 Balanced rearrangement and structural marker, unspecified

Q96 Turner's syndrome
Excludes: Noonan's syndrome (Q87.14)
Q96.0 Karyotype 45,X
Q96.1 Karyotype 46,X iso (Xq)
Q96.2 Karyotype 46,X with abnormal sex chromosome, except iso (Xq)
Q96.3 Mosaicism, 45,X/46,XX or XY
Q96.4 Mosaicism, 45,X/other cell line(s) with abnormal sex chromosome
Q96.8 Other variants of Turner's syndrome
Q96.9 Turner's syndrome, unspecified

Q97 Other sex chromosome abnormalities, female phenotype, not
@ elsewhere classified
Q97.0 Karyotype 47,XXX
Q97.1 Female with more than three X chromosomes
Q97.2 Mosaicism, lines with various numbers of X chromosomes
Q97.3 Female with 46,XY karyotype
Excludes: Drash syndrome (N07)
Q97.8 Other specified sex chromosome abnormalities, female phenotype
Q97.9 Sex chromosome abnormality, female phenotype, unspecified

Q98 Other sex chromosome abnormalities, male phenotype, not elsewhere classified
Q98.0 Klinefelter's syndrome karyotype 47,XXY
Q98.1 Klinefelter's syndrome,male with more than two X chromosomes
Q98.2 Klinefelter's syndrome, male with 46,XX karyotype
Q98.3 Other male with 46,XX karyotype
Q98.4 Klinefelter's syndrome, unspecified
Q98.5 Karyotype 47,XYY
Q98.6 Male with structurally abnormal sex chromosome
Q98.7 Male with sex chromosome mosaicism
Q98.8 Other specified sex chromosome abnormalities, male phenotype
Q98.9 Sex chromosome abnormality, male phenotype, unspecified

Q99 Other chromosome abnormalities, not elsewhere classified
Q99.0 Chimera 46,XX/46,XY
Chimera 46,XX/46,XY true hermaphrodite
Q99.1 46,XX true hermaphrodite
46,XX with streak gonads
46,XY with streak gonads
Pure gonadal dysgenesis
Q99.2 Fragile X chromosome
Fragile X syndrome
Q99.8 Other specified chromosome abnormalities
Q99.9 Chromosomal abnormality, unspecified