Chapter XVII, (Q00-Q99) (version 23 June 2008)

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

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
Kleeblattschaedel 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 } of part of brain
Agenesis }
Aplasia }
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 } . 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 . disease or lesion } system

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

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

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 } of face and neck
. fistula }
. 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

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
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 } Atresia } of cricoid cartilage, epiglottis, glottis, 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

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

Added December 2007:

For facial clefts, codes Q35-37 the coding committee has decided to recommend use of WHO updates instead of the BPA extensions (BPA extensions are crossed through below with WHO update replacements shown). Only for Q369 the coding committee recommend to use the BPA version instead of the WHO update.

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~~
Q35.1 Cleft hard palate
Q35.3 Cleft soft palate
Q35.5 Cleft hard palate with cleft soft palate
~~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
~~Q35.8 Cleft palate, unspecified, bilateral~~

~~Q35.90 Cleft palate, unspecified, unilateral~~
~~Q35.99 Cleft palate, unspecified~~
Q35.9 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, median
Q36.90 Cleft lip, specified as unilateral
Q36.99 Cleft lip NOS

Q37 Cleft palate with cleft lip

Q37.0 Cleft hard palate with bilateral cleft lip
Q37.1 Cleft hard palate with unilateral cleft lip
~~Q37.10 Cleft hard palate with cleft lip, specified as unilateral~~
~~Q37.19 Cleft hard palate with cleft lip, NOS~~
Q37.2 Cleft soft palate with bilateral cleft lip
Q37.3 Cleft soft palate with unilateral cleft lip
Q37.4 Cleft hard and soft palate with bilateral cleft lip
Q37.5 Cleft hard and soft palate with unilateral cleft lip
~~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 bilateral cleft lip
Q37.9 Unspecified cleft palate with unilateral cleft lip
~~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 } (of) salivary gland or duct
Accessory }
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 } . insufficient } of caecum and colon

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: . epooephoron
. 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 } . cyst } of vulva . 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
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 } NOS of male genital organ
. deformity }

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

Q62.6 Malposition of ureter
Deviation } (of) ureter or
Displacement } ureteric orifice
Ectopic }
Implantation, anomalous }
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

Q62.7 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
. 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

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 }
Hemimelia NOS } of limb(s) NOS
Reduction defect }
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

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
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
Malformation of spine } with scoliosis
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-Sch”nberg 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
	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)
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: Sjoegren-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]
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)

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
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 Sjoegren-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
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 } spleen
Mis-shapen }
Accessory }
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
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