Chapter 17

Congenital malformations, deformations and chromosomal abnormalities (Q00-Q99)

Note: Codes from this chapter are not for use on maternal or fetal records

Excludes2: inborn errors of metabolism (E70-E88)

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 cleft 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

Congenital malformations of the nervous system (Q00-Q07)

Q00 Anencephaly and similar malformations

Q00.0 Anencephaly
Acephaly
Acrania
Amyelencephaly
Hemianencephaly
Hemicephaly

Q00.1 Craniorachischisis

Q00.2 Iniencephaly

Q01 Encephalocele

Includes: Arnold-Chiari syndrome, type III
encephalocystocele
encephalomyelocele
hydroencephalocele
hydromeningocele, cranial
meningocele, cerebral
meningoencephalocele

Excludes1: Meckel-Gruber syndrome (Q61.9)

Q01.0 Frontal encephalocele

Q01.1 Nasofrontal encephalocele

Q01.2 Occipital encephalocele

Q01.8 Encephalocele of other sites

Q01.9 Encephalocele, unspecified

Q02 Microcephaly

Includes: hydromicrocephaly

micrencephalon

Excludes1: Meckel-Gruber syndrome (Q61.9)

Q03 Congenital hydrocephalus

Includes: hydrocephalus in newborn

Excludes1: Arnold-Chiari syndrome, type II (Q07.0-)
acquired hydrocephalus (G91.-)
hydrocephalus due to congenital toxoplasmosis (P37.1)
hydrocephalus with spina bifida (Q05.0-Q05.4)

Q03.0 Malformations of aqueduct of Sylvius
Anomaly of aqueduct of Sylvius
Obstruction of aqueduct of Sylvius, congenital
Stenosis of aqueduct of Sylvius

Q03.1 Atresia of foramina of Magendie and Luschka
Dandy-Walker syndrome

Q03.8 Other congenital hydrocephalus

Q03.9 Congenital hydrocephalus, unspecified

Q04 Other congenital malformations of brain

Excludes1: cyclopia (Q87.0)
macrocephaly (Q75.3)

Q04.0 Congenital malformations of corpus callosum
Agenesis of corpus callosum

Q04.1 Arhinencephaly

Q04.2 Holoprosencephaly

Q04.3 Other reduction deformities of brain
Absence of part of brain
Agenesis of part of brain
Agyria
Aplasia of part of brain
Hydranencephaly
Hypoplasia of part of brain
Lissencephaly
Microgyria
Pachygyria

Excludes1: congenital malformations of corpus callosum (Q04.0)

Q04.4 Septo-optic dysplasia of brain

Q04.5 Megalencephaly

Q04.6 Congenital cerebral cysts
Porencephaly
Schizencephaly

Excludes1: acquired porencephalic cyst (G93.0)

Q04.8 Other specified congenital malformations of brain
Arnold-Chiari syndrome, type IV
Macrogyria

Q04.9 Congenital malformation of brain, unspecified
Congenital anomaly NOS of brain
Congenital deformity NOS of brain
Congenital disease or lesion NOS of brain
Multiple anomalies NOS of brain, congenital

Q05 Spina bifida

Includes: hydromeningocele (spinal)
meningocele (spinal)
meningomyelocele
myelocele
myelomeningocele
rachischisis
spina bifida (aperta)(cystica)
syringomyelocele

Use additional code for any associated paraplegia (paraparesis) (G82.2-)

Excludes1: Arnold-Chiari syndrome, type II (Q07.0-)
spina bifida occulta (Q76.0)

Q05.0 Cervical spina bifida with hydrocephalus

Q05.1 Thoracic spina bifida with hydrocephalus
Dorsal spina bifida with hydrocephalus
Thoracolumbar spina bifida with hydrocephalus

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

Q05.5 Cervical spina bifida without hydrocephalus

Q05.6 Thoracic spina bifida without hydrocephalus
Dorsal spina bifida NOS
Thoracolumbar spina bifida 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

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 malformation of spinal cord, unspecified
Congenital anomaly NOS of spinal cord
Congenital deformity NOS of spinal cord
Congenital disease or lesion NOS of spinal cord

Q07 Other congenital malformations of nervous system

Excludes2: congenital central alveolar hypoventilation syndrome (G47.35)
familial dysautonomia [Riley-Day] (G90.1)
neurofibromatosis (nonmalignant) (Q85.0-)

Q07.0 Arnold-Chiari syndrome
Arnold-Chiari syndrome, type II

Excludes1: Arnold-Chiari syndrome, type III (Q01.-)
Arnold-Chiari syndrome, type IV (Q04.8)

Q07.00 Arnold-Chiari syndrome without spina bifida or hydrocephalus

Q07.01 Arnold-Chiari syndrome with spina bifida

Q07.02 Arnold-Chiari syndrome with hydrocephalus

Q07.03 Arnold-Chiari syndrome with spina bifida and hydrocephalus

Q07.8 Other specified congenital malformations of nervous system
Agenesis of nerve
Displacement of brachial plexus
Jaw-winking syndrome
Marcus Gunn's syndrome

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

Congenital malformations of eye, ear, face and neck (Q10-Q18)

Excludes2: cleft lip and cleft palate (Q35-Q37)
congenital malformation of cervical spine (Q05.0, Q05.5, Q67.5, Q76.0-Q76.4)
congenital malformation of larynx (Q31.-)
congenital malformation of lip NEC (Q38.0)
congenital malformation of nose (Q30.-)
congenital malformation of parathyroid gland (Q89.2)
congenital malformation of thyroid gland (Q89.2)

Q10 Congenital malformations of eyelid, lacrimal apparatus and orbit

Excludes1: cryptophthalmos NOS (Q11.2)
cryptophthalmos syndrome (Q87.0)

Q10.0 Congenital ptosis

Q10.1 Congenital ectropion

Q10.2 Congenital entropion

Q10.3 Other congenital malformations of eyelid
Ablepharon
Blepharophimosis, congenital

Coloboma of eyelid
Congenital absence or agenesis of cilia
Congenital absence or agenesis of eyelid
Congenital accessory eyelid
Congenital accessory eye muscle
Congenital malformation of eyelid NOS

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

Q10.5 Congenital stenosis and stricture of lacrimal duct

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

Q10.7 Congenital malformation of orbit

Q11 Anophthalmos, microphthalmos and macrophthalmos

Q11.0 Cystic eyeball

Q11.1 Other anophthalmos
Anophthalmos NOS
Agenesis of eye
Aplasia of eye

Q11.2 Microphthalmos
Cryptophthalmos NOS
Dysplasia of eye
Hypoplasia of eye
Rudimentary eye

Excludes1: cryptophthalmos syndrome (Q87.0)

Q11.3 Macrophthalmos

Excludes1: 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
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

Use additional code for associated glaucoma (H42)

Q13.2 Other congenital malformations of iris
Anisocoria, congenital
Atresia of pupil
Congenital malformation of iris NOS
Corectopia

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

Q13.81 Rieger's anomaly

Use additional code for associated glaucoma (H42)

Q13.89 Other congenital malformations of anterior segment of eye

Q13.9 Congenital malformation of anterior segment of eye, unspecified

Q14 Congenital malformations of posterior segment of eye

Excludes2: optic nerve hypoplasia (H47.03-)

Q14.0 Congenital malformation of vitreous humor
Congenital vitreous opacity

Q14.1 Congenital malformation of retina
Congenital retinal aneurysm

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

Excludes1: congenital nystagmus (H55.01)
ocular albinism (E70.31-)
optic nerve hypoplasia (H47.03-)
retinitis pigmentosa (H35.52)

Q15.0 Congenital glaucoma
Axenfeld's anomaly
Buphthalmos
Glaucoma of childhood
Glaucoma of newborn
Hydrophthalmos
Keratoglobus, congenital, with glaucoma
Macrocornea with glaucoma
Macrophthalmos in congenital glaucoma

Megalocornea with glaucoma

Q15.8 Other specified congenital malformations of eye

Q15.9 Congenital malformation of eye, unspecified
Congenital anomaly of eye
Congenital deformity of eye

Q16 Congenital malformations of ear causing impairment of hearing

Excludes1: congenital deafness (H90.-)

Q16.0 Congenital absence of (ear) auricle

Q16.1 Congenital absence, atresia and stricture of auditory canal (external)
Congenital atresia or stricture of osseous meatus

Q16.2 Absence of eustachian tube

Q16.3 Congenital malformation of ear ossicles
Congenital fusion of ear ossicles

Q16.4 Other congenital malformations of middle ear
Congenital malformation of middle ear NOS

Q16.5 Congenital malformation of inner ear
Congenital anomaly of membranous labyrinth
Congenital anomaly of 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

Excludes1: congenital malformations of ear with impairment of hearing (Q16.0-Q16.9)
preauricular sinus (Q18.1)

Q17.0 Accessory auricle
Accessory tragus
Polyotia
Preauricular appendage or tag
Supernumerary ear
Supernumerary lobule

Q17.1 Macrotia

Q17.2 Microtia

Q17.3 Other misshapen ear
Pointed ear

Q17.4 Misplaced ear
Low-set ears

Excludes1: cervical auricle (Q18.2)

Q17.5 Prominent ear
Bat ear

Q17.8 Other specified congenital malformations of ear
Congenital absence of lobe of ear

Q17.9 Congenital malformation of ear, unspecified

Congenital anomaly of ear NOS

Q18 Other congenital malformations of face and neck

Excludes1: cleft lip and cleft palate (Q35-Q37)
conditions classified to Q67.0-Q67.4
congenital malformations of skull and face bones (Q75.-)
cyclopia (Q87.0)
dentofacial anomalies [including malocclusion] (M26.-)
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 fistula

Q18.2 Other branchial cleft malformations
Branchial cleft malformation 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
Medial fistula of face and neck
Medial sinus of face and neck

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

Congenital malformations of the circulatory system (Q20-Q28)

Q20 Congenital malformations of cardiac chambers and connections

Excludes1: dextrocardia with situs inversus (Q89.3)
mirror-image atrial arrangement with situs inversus (Q89.3)

Q20.0 Common arterial trunk
Persistent truncus arteriosus

Excludes1: aortic septal defect (Q21.4)

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

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

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

Q21 Congenital malformations of cardiac septa

Excludes1: acquired cardiac septal defect (I51.0)

Q21.0 Ventricular septal defect
Roger's disease

Q21.1 Atrial septal defect
Coronary sinus defect
Patent or persistent foramen ovale
Patent or persistent ostium secundum defect (type II)
Patent or persistent sinus venosus defect

Q21.2 Atrioventricular septal defect
Common atrioventricular canal
Endocardial cushion defect
Ostium primum atrial septal defect (type I)

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
Eisenmenger's defect
Pentalogy of Fallot

Code also if applicable:
Eisenmenger's complex (I27.83)
Eisenmenger's syndrome (I27.83)

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
Supernumerary cusps of pulmonary valve

Q22.4 Congenital tricuspid stenosis
Congenital 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
Congenital aortic stenosis NOS

Excludes1: congenital stenosis of aortic valve in hypoplastic left heart syndrome (Q23.4)
congenital subaortic stenosis (Q24.4)
supravalvular aortic stenosis (congenital) (Q25.3)

Q23.1 Congenital insufficiency of aortic valve
Bicuspid aortic valve
Congenital aortic insufficiency

Q23.2 Congenital mitral stenosis
Congenital mitral atresia

Q23.3 Congenital mitral insufficiency

Q23.4 Hypoplastic left heart syndrome

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

Excludes1: endocardial fibroelastosis (I42.4)

Q24.0 Dextrocardia

Excludes1: 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 Levocardia

Q24.2 Cor triatriatum

Q24.3 Pulmonary infundibular stenosis
Subvalvular pulmonic 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 heart
Congenital diverticulum of left ventricle
Congenital malformation of myocardium
Congenital malformation of pericardium
Malposition of heart
Uhl's disease

Q24.9 Congenital malformation of heart, unspecified
Congenital anomaly of heart
Congenital disease of heart

Q25 Congenital malformations of great arteries

Q25.0 Patent ductus arteriosus
Patent ductus Botallo
Persistent ductus arteriosus

Q25.1 Coarctation of aorta
Coarctation of aorta (preductal) (postductal)
Stenosis of aorta

Q25.2 Atresia of aorta

Q25.21 Interruption of aortic arch
Atresia of aortic arch

Q25.29 Other atresia of aorta
Atresia of aorta

Q25.3 Supravalvular aortic stenosis

Excludes1: congenital aortic stenosis NOS (Q23.0)
congenital stenosis of aortic valve (Q23.0)

Q25.4 Other congenital malformations of aorta

Excludes1: hypoplasia of aorta in hypoplastic left heart syndrome (Q23.4)

Q25.40 Congenital malformation of aorta unspecified

Q25.41 Absence and aplasia of aorta

Q25.42 Hypoplasia of aorta

Q25.43 Congenital aneurysm of aorta
Congenital aneurysm of aortic root
Congenital aneurysm of aortic sinus

Q25.44 Congenital dilation of aorta

Q25.45 Double aortic arch
Vascular ring of aorta

Q25.46 Tortuous aortic arch
Persistent convolutions of aortic arch

Q25.47 Right aortic arch
Persistent right aortic arch

Q25.48 Anomalous origin of subclavian artery

Q25.49 Other congenital malformations of aorta
Aortic arch
Bovine arch

Q25.5 Atresia of pulmonary artery

Q25.6 Stenosis of pulmonary artery
Supravalvular pulmonary stenosis

Q25.7 Other congenital malformations of pulmonary artery

Q25.71 Coarctation of pulmonary artery

Q25.72 Congenital pulmonary arteriovenous malformation
Congenital pulmonary arteriovenous aneurysm

Q25.79 Other congenital malformations of pulmonary artery
Aberrant pulmonary artery
Agenesis of pulmonary artery
Congenital aneurysm of pulmonary artery
Congenital anomaly of pulmonary artery
Hypoplasia of pulmonary artery

Q25.8 Other congenital malformations of other great arteries

Q25.9 Congenital malformation of great arteries, unspecified

Q26 Congenital malformations of great veins

Q26.0 Congenital stenosis of vena cava
Congenital stenosis of vena cava (inferior)(superior)

Q26.1 Persistent left superior vena cava

Q26.2 Total anomalous pulmonary venous connection
Total anomalous pulmonary venous return [TAPVR], subdiaphragmatic
Total anomalous pulmonary venous return [TAPVR], supradiaphragmatic

Q26.3 Partial anomalous pulmonary venous connection
Partial anomalous pulmonary venous return

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
Congenital anomaly of vena cava (inferior) (superior) NOS

Q27 Other congenital malformations of peripheral vascular system

Excludes2: anomalies of cerebral and precerebral vessels (Q28.0-Q28.3)
anomalies of coronary vessels (Q24.5)
anomalies of pulmonary artery (Q25.5-Q25.7)
congenital retinal aneurysm (Q14.1)
hemangioma and lymphangioma (D18.-)

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 Arteriovenous malformation (peripheral)
Arteriovenous aneurysm

Excludes1: acquired arteriovenous aneurysm (I77.0)

Excludes2: arteriovenous malformation of cerebral vessels (Q28.2)
arteriovenous malformation of precerebral vessels (Q28.0)

Q27.30 Arteriovenous malformation, site unspecified

Q27.31 Arteriovenous malformation of vessel of upper limb

Q27.32 Arteriovenous malformation of vessel of lower limb

Q27.33 Arteriovenous malformation of digestive system vessel

Q27.34 Arteriovenous malformation of renal vessel

Q27.39 Arteriovenous malformation, other site

Q27.4 Congenital phlebectasia

Q27.8 Other specified congenital malformations of peripheral vascular system
Absence of peripheral vascular system
Atresia of peripheral vascular system
Congenital aneurysm (peripheral)
Congenital stricture, artery
Congenital varix

Excludes1: arteriovenous malformation (Q27.3-)

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

Q28 Other congenital malformations of circulatory system

Excludes1: congenital aneurysm NOS (Q27.8)
congenital coronary aneurysm (Q24.5)
ruptured cerebral arteriovenous malformation (I60.8)
ruptured malformation of precerebral vessels (I72.0)

Excludes2: congenital peripheral aneurysm (Q27.8)
congenital pulmonary aneurysm (Q25.79)
congenital retinal aneurysm (Q14.1)

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
Congenital precerebral aneurysm (nonruptured)

Q28.2 Arteriovenous malformation of cerebral vessels
Arteriovenous malformation of brain NOS

Congenital arteriovenous cerebral aneurysm (nonruptured)

Q28.3 Other malformations of cerebral vessels
Congenital cerebral aneurysm (nonruptured)
Congenital malformation of cerebral vessels NOS
Developmental venous anomaly

Q28.8 Other specified congenital malformations of circulatory system
Congenital aneurysm, specified site NEC
Spinal vessel anomaly

Q28.9 Congenital malformation of circulatory system, unspecified

Congenital malformations of the respiratory system (Q30-Q34)

Q30 Congenital malformations of nose

Excludes1: congenital deviation of nasal septum (Q67.4)

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

Q30.1 Agenesis and underdevelopment of nose
Congenital absent 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

Excludes1: congenital laryngeal stridor NOS (P28.89)

Q31.0 Web of larynx
Glottic web of larynx
Subglottic web of larynx
Web of larynx NOS

Q31.1 Congenital subglottic stenosis

Q31.2 Laryngeal hypoplasia

Q31.3 Laryngocele

Q31.5 Congenital laryngomalacia

Q31.8 Other congenital malformations of larynx
Absence of larynx
Agenesis of larynx
Atresia of larynx
Congenital cleft thyroid cartilage
Congenital fissure of epiglottis
Congenital stenosis of larynx NEC
Posterior cleft of cricoid cartilage

Q31.9 Congenital malformation of larynx, unspecified

Q32 Congenital malformations of trachea and bronchus

Excludes1: congenital bronchiectasis (Q33.4)

Q32.0 Congenital tracheomalacia

Q32.1 Other congenital malformations of trachea
Atresia of trachea
Congenital anomaly of tracheal cartilage
Congenital dilatation of trachea
Congenital malformation of trachea
Congenital stenosis of trachea
Congenital tracheocele

Q32.2 Congenital bronchomalacia

Q32.3 Congenital stenosis of bronchus

Q32.4 Other congenital malformations of bronchus
Absence of bronchus
Agenesis of bronchus
Atresia of bronchus
Congenital diverticulum of bronchus
Congenital malformation of bronchus NOS

Q33 Congenital malformations of lung

Q33.0 Congenital cystic lung
Congenital cystic lung disease
Congenital honeycomb lung
Congenital polycystic lung disease

Excludes1: cystic fibrosis (E84.0)
cystic lung disease, acquired or unspecified (J98.4)

Q33.1 Accessory lobe of lung
Azygos lobe (fissured), lung

Q33.2 Sequestration of lung

Q33.3 Agenesis of lung
Congenital absence of lung (lobe)

Q33.4 Congenital bronchiectasis

Q33.5 Ectopic tissue in lung

Q33.6 Congenital hypoplasia and dysplasia of lung

Excludes1: pulmonary hypoplasia associated with short gestation (P28.0)

Q33.8 Other congenital malformations of lung

Q33.9 Congenital malformation of lung, unspecified

Q34 Other congenital malformations of respiratory system

Excludes2: congenital central alveolar hypoventilation syndrome (G47.35)

Q34.0 Anomaly of pleura

Q34.1 Congenital cyst of mediastinum

Q34.8 Other specified congenital malformations of respiratory system
Atresia of nasopharynx

Q34.9 Congenital malformation of respiratory system, unspecified
Congenital absence of respiratory system
Congenital anomaly of respiratory system NOS

Cleft lip and cleft palate (Q35-Q37)

Use additional code to identify associated malformation of the nose (Q30.2)

Excludes1: Robin's syndrome (Q87.0)

Q35 Cleft palate

Includes: fissure of palate
palatoschisis

Excludes1: cleft palate with cleft lip (Q37.-)

Q35.1 Cleft hard palate

Q35.3 Cleft soft palate

Q35.5 Cleft hard palate with cleft soft palate

Q35.7 Cleft uvula

Q35.9 Cleft palate, unspecified
Cleft palate NOS

Q36 Cleft lip

Includes: cheiloschisis
congenital fissure of lip
harelip
labium leporinum

Excludes1: cleft lip with cleft palate (Q37.-)

Q36.0 Cleft lip, bilateral

Q36.1 Cleft lip, median

Q36.9 Cleft lip, unilateral
Cleft lip NOS

Q37 Cleft palate with cleft lip

Includes: cheilopalatoschisis

Q37.0 Cleft hard palate with bilateral cleft lip

Q37.1 Cleft hard palate with unilateral cleft lip
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
Cleft soft palate with cleft lip NOS

Q37.4 Cleft hard and soft palate with bilateral cleft lip

Q37.5 Cleft hard and soft palate with unilateral cleft lip

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
Cleft palate with cleft lip NOS

Other congenital malformations of the digestive system (Q38-Q45)

Q38 Other congenital malformations of tongue, mouth and pharynx

Excludes1: dentofacial anomalies (M26.-)
macrostomia (Q18.4)
microstomia (Q18.5)

Q38.0 Congenital malformations of lips, not elsewhere classified
Congenital fistula of lip
Congenital malformation of lip NOS
Van der Woude's syndrome

Excludes1: cleft lip (Q36.-)
cleft lip with cleft palate (Q37.-)
macrocheilia (Q18.6)
microcheilia (Q18.7)

Q38.1 Ankyloglossia
Tongue tie

Q38.2 Macroglossia
Congenital hypertrophy of tongue

Q38.3 Other congenital malformations of tongue
Aglossia
Bifid tongue
Congenital adhesion of tongue
Congenital fissure of tongue
Congenital malformation of tongue NOS
Double tongue
Hypoglossia
Hypoplasia of tongue
Microglossia

Q38.4 Congenital malformations of salivary glands and ducts
Atresia of salivary glands and ducts
Congenital absence of salivary glands and ducts
Congenital accessory salivary glands and ducts
Congenital fistula of salivary gland

Q38.5 Congenital malformations of palate, not elsewhere classified
Congenital absence of uvula
Congenital malformation of palate NOS
Congenital high arched palate

Excludes1: cleft palate (Q35.-)
cleft palate with cleft lip (Q37.-)

Q38.6 Other congenital malformations of mouth
Congenital malformation of mouth NOS

Q38.7 Congenital pharyngeal pouch
Congenital diverticulum of pharynx

Excludes1: pharyngeal pouch syndrome (D82.1)

Q38.8 Other congenital malformations of pharynx
Congenital malformation of pharynx NOS
Imperforate pharynx

Q39 Congenital malformations of esophagus

Q39.0 Atresia of esophagus without fistula
Atresia of esophagus NOS

Q39.1 Atresia of esophagus with tracheo-esophageal fistula
Atresia of esophagus with broncho-esophageal fistula

Q39.2 Congenital tracheo-esophageal fistula without atresia
Congenital tracheo-esophageal fistula NOS

Q39.3 Congenital stenosis and stricture of esophagus

Q39.4 Esophageal web

Q39.5 Congenital dilatation of esophagus
Congenital cardiospasm

Q39.6 Congenital diverticulum of esophagus
Congenital esophageal pouch

Q39.8 Other congenital malformations of esophagus
Congenital absence of esophagus
Congenital displacement of esophagus
Congenital duplication of esophagus

Q39.9 Congenital malformation of esophagus, unspecified

Q40 Other congenital malformations of upper alimentary tract

Q40.0 Congenital hypertrophic pyloric stenosis
Congenital or infantile constriction
Congenital or infantile hypertrophy
Congenital or infantile spasm
Congenital or infantile stenosis
Congenital or infantile stricture

Q40.1 Congenital hiatus hernia
Congenital displacement of cardia through esophageal hiatus

Excludes1: congenital diaphragmatic hernia (Q79.0)

Q40.2 Other specified congenital malformations of stomach
Congenital displacement of stomach
Congenital diverticulum of stomach
Congenital hourglass stomach
Congenital duplication of stomach
Megalogastria
Microgastria

Q40.3 Congenital malformation of stomach, unspecified

Q40.8 Other specified congenital malformations of upper alimentary tract

Q40.9 Congenital malformation of upper alimentary tract, unspecified
Congenital anomaly of upper alimentary tract
Congenital deformity of upper alimentary tract

Q41 Congenital absence, atresia and stenosis of small intestine

Includes: congenital obstruction, occlusion or stricture of small intestine or intestine NOS

Excludes1: cystic fibrosis with intestinal manifestation (E84.11)
meconium ileus NOS (without cystic fibrosis) (P76.0)

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

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

Q42.1 Congenital absence, atresia and stenosis of rectum without fistula
Imperforate rectum

Q42.2 Congenital absence, atresia and stenosis of anus with fistula

Q42.3 Congenital absence, atresia and stenosis of anus without fistula
Imperforate anus

Q42.8 Congenital absence, atresia and stenosis of other parts of large intestine

Q42.9 Congenital absence, atresia and stenosis of large intestine, part unspecified

Q43 Other congenital malformations of intestine

Q43.0 Meckel's diverticulum (displaced) (hypertrophic)
Persistent omphalomesenteric duct
Persistent vitelline duct

Q43.1 Hirschsprung's disease
Aganglionosis
Congenital (aganglionic) megacolon

Q43.2 Other congenital functional disorders of colon
Congenital dilatation of colon

Q43.3 Congenital malformations of intestinal fixation
Congenital omental, anomalous adhesions [bands]
Congenital peritoneal adhesions [bands]
Incomplete rotation of cecum and colon
Insufficient rotation of cecum and colon
Jackson's membrane
Malrotation of colon
Rotation failure of cecum and colon
Universal mesentery

Q43.4 Duplication of intestine

Q43.5 Ectopic anus

Q43.6 Congenital fistula of rectum and anus

Excludes1: congenital fistula of anus with absence, atresia and stenosis (Q42.2)
congenital fistula of rectum with absence, atresia and stenosis (Q42.0)
congenital rectovaginal fistula (Q52.2)
congenital urethrorectal fistula (Q64.73)
pilonidal fistula or sinus (L05.-)

Q43.7 Persistent cloaca
Cloaca NOS

Q43.8 Other specified congenital malformations of intestine
Congenital blind loop syndrome
Congenital diverticulitis, colon
Congenital diverticulum, intestine
Dolichocolon
Megaloappendix
Megaloduodenum
Microcolon
Transposition of appendix
Transposition of colon
Transposition of intestine

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

Q44.2 Atresia of bile ducts

Q44.3 Congenital stenosis and stricture of bile ducts

Q44.4 Choledochal cyst

Q44.5 Other congenital malformations of bile ducts
Accessory hepatic duct
Biliary duct duplication
Congenital malformation of bile duct NOS
Cystic duct duplication

Q44.6 Cystic disease of liver
Fibrocystic disease of liver

Q44.7 Other congenital malformations of liver
Accessory liver
Alagille's syndrome
Congenital absence of liver
Congenital hepatomegaly
Congenital malformation of liver NOS

Q45 Other congenital malformations of digestive system

Excludes2: congenital diaphragmatic hernia (Q79.0)
congenital 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

Excludes1: congenital diabetes mellitus (E10.-)
cystic fibrosis (E84.0-E84.9)
fibrocystic disease of pancreas (E84.-)
neonatal diabetes mellitus (P70.2)

Q45.8 Other specified congenital malformations of digestive system
Absence (complete) (partial) of alimentary tract NOS
Duplication of digestive system
Malposition, congenital of digestive system

Q45.9 Congenital malformation of digestive system, unspecified
Congenital anomaly of digestive system
Congenital deformity of digestive system

Congenital malformations of genital organs (Q50-Q56)

Excludes1: androgen insensitivity 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

Excludes1: Turner's syndrome (Q96.-)

Q50.01 Congenital absence of ovary, unilateral

Q50.02 Congenital absence of ovary, bilateral

Q50.1 Developmental ovarian cyst

Q50.2 Congenital torsion of ovary

Q50.3 Other congenital malformations of ovary

Q50.31 Accessory ovary

Q50.32 Ovarian streak
46, XX with streak gonads

Q50.39 Other congenital malformation of ovary
Congenital malformation of ovary NOS

Q50.4 Embryonic cyst of fallopian tube
Fimbrial cyst

Q50.5 Embryonic cyst of broad ligament
Epoophoron cyst
Parovarian cyst

Q50.6 Other congenital malformations of fallopian tube and broad ligament
Absence of fallopian tube and broad ligament

Accessory fallopian tube and broad ligament
Atresia of fallopian tube and broad ligament
Congenital malformation of fallopian tube or broad ligament NOS

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

Q51.10 Doubling of uterus with doubling of cervix and vagina without obstruction
Doubling of uterus with doubling of cervix and vagina NOS

Q51.11 Doubling of uterus with doubling of cervix and vagina with obstruction

Q51.2 Other doubling of uterus
Doubling of uterus NOS
Septate uterus, complete or partial

Q51.3 Bicornate uterus
Bicornate uterus, complete or partial

Q51.4 Unicornate uterus
Unicornate uterus with or without a separate uterine horn
Uterus with only one functioning horn

Q51.5 Agenesis and aplasia of cervix
Congenital absence of cervix

Q51.6 Embryonic cyst of cervix

Q51.7 Congenital fistulae between uterus and digestive and urinary tracts

Q51.8 Other congenital malformations of uterus and cervix

Q51.81 Other congenital malformations of uterus

Q51.810 Arcuate uterus
Arcuatus uterus

Q51.811 Hypoplasia of uterus

Q51.818 Other congenital malformations of uterus
Müllerian anomaly of uterus NEC

Q51.82 Other congenital malformations of cervix

Q51.820 Cervical duplication

Q51.821 Hypoplasia of cervix

Q51.828 Other congenital malformations of cervix

Q51.9 Congenital malformation of uterus and cervix, unspecified

Q52 Other congenital malformations of female genitalia

Q52.0 Congenital absence of vagina
Vaginal agenesis, total or partial

Q52.1 Doubling of vagina

Excludes1: doubling of vagina with doubling of uterus and cervix (Q51.1-)

Q52.10 Doubling of vagina, unspecified
Septate vagina NOS

Q52.11 Transverse vaginal septum

Q52.12 Longitudinal vaginal septum

Q52.120 Longitudinal vaginal septum, nonobstructing

Q52.121 Longitudinal vaginal septum, obstructing, right side

Q52.122 Longitudinal vaginal septum, obstructing, left side

Q52.123 Longitudinal vaginal septum, microperforate, right side

Q52.124 Longitudinal vaginal septum, microperforate, left side

Q52.129 Other and unspecified longitudinal vaginal septum

Q52.2 Congenital rectovaginal fistula

Excludes1: cloaca (Q43.7)

Q52.3 Imperforate hymen

Q52.4 Other congenital malformations of vagina
Canal of Nuck cyst, congenital
Congenital malformation of vagina NOS
Embryonic vaginal cyst
Gartner's duct cyst

Q52.5 Fusion of labia

Q52.6 Congenital malformation of clitoris

Q52.7 Other and unspecified congenital malformations of vulva

Q52.70 Unspecified congenital malformations of vulva
Congenital malformation of vulva NOS

Q52.71 Congenital absence of vulva

Q52.79 Other congenital malformations of vulva
Congenital cyst of vulva

Q52.8 Other specified congenital malformations of female genitalia

Q52.9 Congenital malformation of female genitalia, unspecified

Q53 Undescended and ectopic testicle

Q53.0 Ectopic testis

Q53.00 Ectopic testis, unspecified

Q53.01 Ectopic testis, unilateral

Q53.02 Ectopic testes, bilateral

Q53.1 Undescended testicle, unilateral

Q53.10 Unspecified undescended testicle, unilateral

Q53.11 Abdominal testis, unilateral

Q53.111 Unilateral intraabdominal testis

Q53.112 Unilateral inguinal testis

Q53.12 Ectopic perineal testis, unilateral

Q53.13 Unilateral high scrotal testis

Q53.2 Undescended testicle, bilateral

Q53.20 Undescended testicle, unspecified, bilateral

Q53.21 Abdominal testis, bilateral

Q53.211 Bilateral intraabdominal testes

Q53.212 Bilateral inguinal testes

Q53.22 Ectopic perineal testis, bilateral

Q53.23 Bilateral high scrotal testes

Q53.9 Undescended testicle, unspecified
Cryptorchism NOS

Q54 Hypospadias

Excludes1: epispadias (Q64.0)

Q54.0 Hypospadias, balanic
Hypospadias, coronal
Hypospadias, glandular

Q54.1 Hypospadias, penile

Q54.2 Hypospadias, penoscrotal

Q54.3 Hypospadias, perineal

Q54.4 Congenital chordee
Chordee without hypospadias

Q54.8 Other hypospadias
Hypospadias with intersex state

Q54.9 Hypospadias, unspecified

Q55 Other congenital malformations of male genital organs

Excludes1: congenital hydrocele (P83.5)
hypospadias (Q54.-)

Q55.0 Absence and aplasia of testis
Monorchism

Q55.1 Hypoplasia of testis and scrotum
Fusion of testes

Q55.2 Other and unspecified congenital malformations of testis and scrotum

Q55.20 Unspecified congenital malformations of testis and scrotum
Congenital malformation of testis or scrotum NOS

Q55.21 Polyorchism

Q55.22 Retractile testis

Q55.23 Scrotal transposition

Q55.29 Other congenital malformations of testis and scrotum

Q55.3 Atresia of vas deferens

Code first any associated cystic fibrosis (E84.-)

Q55.4 Other congenital malformations of vas deferens, epididymis, seminal vesicles and prostate
Absence or aplasia of prostate
Absence or aplasia of spermatic cord
Congenital malformation of vas deferens, epididymis, seminal vesicles or prostate NOS

Q55.5 Congenital absence and aplasia of penis

Q55.6 Other congenital malformations of penis

Q55.61 Curvature of penis (lateral)

Q55.62 Hypoplasia of penis
Micropenis

Q55.63 Congenital torsion of penis

Excludes1: acquired torsion of penis (N48.82)

Q55.64 Hidden penis
Buried penis
Concealed penis

Excludes1: acquired buried penis (N48.83)

Q55.69 Other congenital malformation of penis
Congenital malformation of penis NOS

Q55.7 Congenital vasocutaneous fistula

Q55.8 Other specified congenital malformations of male genital organs

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

Q56 Indeterminate sex and pseudohermaphroditism

Excludes1: 46,XX true hermaphrodite (Q99.1)
androgen insensitivity syndrome (E34.5-)
chimera 46,XX/46,XY true hermaphrodite (Q99.0)
female pseudohermaphroditism with adrenocortical disorder (E25.-)
pseudohermaphroditism with specified chromosomal anomaly (Q96-Q99)
pure gonadal dysgenesis (Q99.1)

Q56.0 Hermaphroditism, not elsewhere classified
Ovotestis

Q56.1 Male pseudohermaphroditism, not elsewhere classified
46, XY with streak gonads
Male pseudohermaphroditism NOS

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

Q56.3 Pseudohermaphroditism, unspecified

Q56.4 Indeterminate sex, unspecified
Ambiguous genitalia

Congenital malformations of the urinary system (Q60-Q64)

Q60 Renal agenesis and other reduction defects of kidney

Includes: congenital absence of kidney
congenital atrophy of kidney
infantile atrophy 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

Q61 Cystic kidney disease

Excludes1: acquired cyst of kidney (N28.1)
Potter's syndrome (Q60.6)

Q61.0 Congenital renal cyst

Q61.00 Congenital renal cyst, unspecified
Cyst of kidney NOS (congenital)

Q61.01 Congenital single renal cyst

Q61.02 Congenital multiple renal cysts

Q61.1 Polycystic kidney, infantile type
Polycystic kidney, autosomal recessive

Q61.11 Cystic dilatation of collecting ducts

Q61.19 Other polycystic kidney, infantile type

Q61.2 Polycystic kidney, adult type
Polycystic kidney, autosomal dominant

Q61.3 Polycystic kidney, unspecified

Q61.4 Renal dysplasia
Multicystic dysplastic kidney
Multicystic kidney (development)
Multicystic kidney disease
Multicystic renal dysplasia

Excludes1: polycystic kidney disease (Q61.11-Q61.3)

Q61.5 Medullary cystic kidney
Nephronopthisis

Sponge kidney NOS

Q61.8 Other cystic kidney diseases
Fibrocystic kidney
Fibrocystic renal degeneration or disease

Q61.9 Cystic kidney disease, unspecified
Meckel-Gruber syndrome

Q62 Congenital obstructive defects of renal pelvis and congenital malformations of ureter

Q62.0 Congenital hydronephrosis

Q62.1 Congenital occlusion of ureter
Atresia and stenosis of ureter

Q62.10 Congenital occlusion of ureter, unspecified

Q62.11 Congenital occlusion of ureteropelvic junction

Q62.12 Congenital occlusion of ureterovesical orifice

Q62.2 Congenital megaureter
Congenital dilatation of ureter

Q62.3 Other obstructive defects of renal pelvis and ureter

Q62.31 Congenital ureterocele, orthotopic

Q62.32 Cecoureterocele
Ectopic ureterocele

Q62.39 Other obstructive defects of renal pelvis and ureter
Ureteropelvic junction obstruction NOS

Q62.4 Agenesis of ureter
Congenital absence ureter

Q62.5 Duplication of ureter
Accessory ureter
Double ureter

Q62.6 Malposition of ureter

Q62.60 Malposition of ureter, unspecified

Q62.61 Deviation of ureter

Q62.62 Displacement of ureter

Q62.63 Anomalous implantation of ureter
Ectopia of ureter
Ectopic ureter

Q62.69 Other malposition of ureter

Q62.7 Congenital vesico-uretero-renal reflux

Q62.8 Other congenital malformations of ureter
Anomaly of ureter NOS

Q63 Other congenital malformations of kidney

Excludes1: congenital nephrotic syndrome (N04.-)

Q63.0 Accessory kidney

Q63.1 Lobulated, fused and horseshoe kidney

Q63.2 Ectopic kidney
Congenital displaced kidney
Malrotation of kidney

Q63.3 Hyperplastic and giant kidney
Compensatory hypertrophy of kidney

Q63.8 Other specified congenital malformations of kidney
Congenital renal calculi

Q63.9 Congenital malformation of kidney, unspecified

Q64 Other congenital malformations of urinary system

Q64.0 Epispadias

Excludes1: hypospadias (Q54.-)

Q64.1 Exstrophy of urinary bladder

Q64.10 Exstrophy of urinary bladder, unspecified
Ectopia vesicae

Q64.11 Supravesical fissure of urinary bladder

Q64.12 Cloacal exstrophy of urinary bladder

Q64.19 Other exstrophy of urinary bladder
Extroversion of bladder

Q64.2 Congenital posterior urethral valves

Q64.3 Other atresia and stenosis of urethra and bladder neck

Q64.31 Congenital bladder neck obstruction
Congenital obstruction of vesicourethral orifice

Q64.32 Congenital stricture of urethra

Q64.33 Congenital stricture of urinary meatus

Q64.39 Other atresia and stenosis of urethra and bladder neck
Atresia and stenosis of urethra and bladder neck NOS

Q64.4 Malformation of urachus
Cyst of urachus
Patent urachus
Prolapse of urachus

Q64.5 Congenital absence of bladder and urethra

Q64.6 Congenital diverticulum of bladder

Q64.7 Other and unspecified congenital malformations of bladder and urethra

Excludes1: congenital prolapse of bladder (mucosa) (Q79.4)

Q64.70 Unspecified congenital malformation of bladder and urethra
Malformation of bladder or urethra NOS

Q64.71 Congenital prolapse of urethra

Q64.72 Congenital prolapse of urinary meatus

Q64.73 Congenital urethrorectal fistula

Q64.74 Double urethra

Q64.75 Double urinary meatus

Q64.79 Other congenital malformations of bladder and urethra

Q64.8 Other specified congenital malformations of urinary system

Q64.9 Congenital malformation of urinary system, unspecified
Congenital anomaly NOS of urinary system
Congenital deformity NOS of urinary system

Congenital malformations and deformations of the musculoskeletal system (Q65-Q79)

Q65 Congenital deformities of hip

Excludes1: clicking hip (R29.4)

Q65.0 Congenital dislocation of hip, unilateral

Q65.00 Congenital dislocation of unspecified hip, unilateral

Q65.01 Congenital dislocation of right hip, unilateral

Q65.02 Congenital dislocation of left hip, unilateral

Q65.1 Congenital dislocation of hip, bilateral

Q65.2 Congenital dislocation of hip, unspecified

Q65.3 Congenital partial dislocation of hip, unilateral

Q65.30 Congenital partial dislocation of unspecified hip, unilateral

Q65.31 Congenital partial dislocation of right hip, unilateral

Q65.32 Congenital partial dislocation of left hip, unilateral

Q65.4 Congenital partial dislocation of hip, bilateral

Q65.5 Congenital partial dislocation of hip, unspecified

Q65.6 Congenital unstable hip
Congenital dislocatable hip

Q65.8 Other congenital deformities of hip

Q65.81 Congenital coxa valga

Q65.82 Congenital coxa vara

Q65.89 Other specified congenital deformities of hip
Anteversion of femoral neck
Congenital acetabular dysplasia

Q65.9 Congenital deformity of hip, unspecified

Q66 Congenital deformities of feet

Excludes1: reduction defects of feet (Q72.-)
valgus deformities (acquired) (M21.0-)
varus deformities (acquired) (M21.1-)

Q66.0 Congenital talipes equinovarus

Q66.1 Congenital talipes calcaneovarus

Q66.2 Congenital metatarsus (primus) varus

Q66.21 Congenital metatarsus primus varus

Q66.22 Congenital metatarsus adductus
Congenital metatarsus varus

Q66.3 Other congenital varus deformities of feet
Hallux varus, congenital

Q66.4 Congenital talipes calcaneovalgus

Q66.5 Congenital pes planus
Congenital flat foot
Congenital rigid flat foot
Congenital spastic (everted) flat foot

Excludes1: pes planus, acquired (M21.4)

Q66.50 Congenital pes planus, unspecified foot

Q66.51 Congenital pes planus, right foot

Q66.52 Congenital pes planus, left foot

Q66.6 Other congenital valgus deformities of feet
Congenital metatarsus valgus

Q66.7 Congenital pes cavus

Q66.8 Other congenital deformities of feet

Q66.80 Congenital vertical talus deformity, unspecified foot

Q66.81 Congenital vertical talus deformity, right foot

Q66.82 Congenital vertical talus deformity, left foot

Q66.89 Other specified congenital deformities of feet
Congenital asymmetric talipes
Congenital clubfoot NOS
Congenital talipes NOS
Congenital tarsal coalition
Hammer toe, congenital

Q66.9 Congenital deformity of feet, unspecified

Q67 Congenital musculoskeletal deformities of head, face, spine and chest

Excludes1: congenital malformation syndromes classified to Q87.-
Potter's syndrome (Q60.6)

Q67.0 Congenital facial asymmetry

Q67.1 Congenital compression facies

Q67.2 Dolichocephaly

Q67.3 Plagiocephaly

Q67.4 Other congenital deformities of skull, face and jaw
Congenital depressions in skull
Congenital hemifacial atrophy or hypertrophy
Deviation of nasal septum, congenital
Squashed or bent nose, congenital

Excludes1: dentofacial anomalies [including malocclusion] (M26.-)
syphilitic saddle nose (A50.5)

Q67.5 Congenital deformity of spine
Congenital postural scoliosis
Congenital scoliosis NOS

Excludes1: infantile idiopathic scoliosis (M41.0)
scoliosis due to congenital bony malformation (Q76.3)

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

Excludes1: reduction defects of limb(s) (Q71-Q73)

Excludes2: congenital myotonic chondrodystrophy (G71.13)

Q68.0 Congenital deformity of sternocleidomastoid muscle
Congenital contracture of sternocleidomastoid (muscle)
Congenital (sternomastoid) torticollis
Sternomastoid tumor (congenital)

Q68.1 Congenital deformity of finger(s) and hand
Congenital clubfinger
Spade-like hand (congenital)

Q68.2 Congenital deformity of knee
Congenital dislocation of knee
Congenital genu recurvatum

Q68.3 Congenital bowing of femur

Excludes1: anteversion of femur (neck) (Q65.89)

Q68.4 Congenital bowing of tibia and fibula

Q68.5 Congenital bowing of long bones of leg, unspecified

Q68.6 Discoid meniscus

Q68.8 Other specified congenital musculoskeletal deformities
Congenital deformity of clavicle
Congenital deformity of elbow
Congenital deformity of forearm
Congenital deformity of scapula

Congenital deformity of wrist
Congenital dislocation of elbow
Congenital dislocation of shoulder
Congenital dislocation of wrist

Q69 Polydactyly

Q69.0 Accessory finger(s)

Q69.1 Accessory thumb(s)

Q69.2 Accessory toe(s)
Accessory hallux

Q69.9 Polydactyly, unspecified
Supernumerary digit(s) NOS

Q70 Syndactyly

Q70.0 Fused fingers
Complex syndactyly of fingers with synostosis

Q70.00 Fused fingers, unspecified hand

Q70.01 Fused fingers, right hand

Q70.02 Fused fingers, left hand

Q70.03 Fused fingers, bilateral

Q70.1 Webbed fingers
Simple syndactyly of fingers without synostosis

Q70.10 Webbed fingers, unspecified hand

Q70.11 Webbed fingers, right hand

Q70.12 Webbed fingers, left hand

Q70.13 Webbed fingers, bilateral

Q70.2 Fused toes
Complex syndactyly of toes with synostosis

Q70.20 Fused toes, unspecified foot

Q70.21 Fused toes, right foot

Q70.22 Fused toes, left foot

Q70.23 Fused toes, bilateral

Q70.3 Webbed toes
Simple syndactyly of toes without synostosis

Q70.30 Webbed toes, unspecified foot

Q70.31 Webbed toes, right foot

Q70.32 Webbed toes, left foot

Q70.33 Webbed toes, bilateral

Q70.4 Polysyndactyly, unspecified

Excludes1: specified syndactyly of hand and feet - code to specified conditions (Q70.0- -Q70.3-)

Q70.9 Syndactyly, unspecified
Symphalangy NOS

Q71 Reduction defects of upper limb

Q71.0 Congenital complete absence of upper limb

Q71.00 Congenital complete absence of unspecified upper limb

Q71.01 Congenital complete absence of right upper limb

Q71.02 Congenital complete absence of left upper limb

Q71.03 Congenital complete absence of upper limb, bilateral

Q71.1 Congenital absence of upper arm and forearm with hand present

Q71.10 Congenital absence of unspecified upper arm and forearm with hand present

Q71.11 Congenital absence of right upper arm and forearm with hand present

Q71.12 Congenital absence of left upper arm and forearm with hand present

Q71.13 Congenital absence of upper arm and forearm with hand present, bilateral

Q71.2 Congenital absence of both forearm and hand

Q71.20 Congenital absence of both forearm and hand, unspecified upper limb

Q71.21 Congenital absence of both forearm and hand, right upper limb

Q71.22 Congenital absence of both forearm and hand, left upper limb

Q71.23 Congenital absence of both forearm and hand, bilateral

Q71.3 Congenital absence of hand and finger

Q71.30 Congenital absence of unspecified hand and finger

Q71.31 Congenital absence of right hand and finger

Q71.32 Congenital absence of left hand and finger

Q71.33 Congenital absence of hand and finger, bilateral

Q71.4 Longitudinal reduction defect of radius
Clubhand (congenital)
Radial clubhand

Q71.40 Longitudinal reduction defect of unspecified radius

Q71.41 Longitudinal reduction defect of right radius

Q71.42 Longitudinal reduction defect of left radius

Q71.43 Longitudinal reduction defect of radius, bilateral

Q71.5 Longitudinal reduction defect of ulna

Q71.50 Longitudinal reduction defect of unspecified ulna

Q71.51 Longitudinal reduction defect of right ulna

Q71.52 Longitudinal reduction defect of left ulna

Q71.53 Longitudinal reduction defect of ulna, bilateral

Q71.6 Lobster-claw hand

Q71.60 Lobster-claw hand, unspecified hand

Q71.61 Lobster-claw right hand

Q71.62 Lobster-claw left hand

Q71.63 Lobster-claw hand, bilateral

Q71.8 Other reduction defects of upper limb

Q71.81 Congenital shortening of upper limb

Q71.811 Congenital shortening of right upper limb

Q71.812 Congenital shortening of left upper limb

Q71.813 Congenital shortening of upper limb, bilateral

Q71.819 Congenital shortening of unspecified upper limb

Q71.89 Other reduction defects of upper limb

Q71.891 Other reduction defects of right upper limb

Q71.892 Other reduction defects of left upper limb

Q71.893 Other reduction defects of upper limb, bilateral

Q71.899 Other reduction defects of unspecified upper limb

Q71.9 Unspecified reduction defect of upper limb

Q71.90 Unspecified reduction defect of unspecified upper limb

Q71.91 Unspecified reduction defect of right upper limb

Q71.92 Unspecified reduction defect of left upper limb

Q71.93 Unspecified reduction defect of upper limb, bilateral

Q72 Reduction defects of lower limb

Q72.0 Congenital complete absence of lower limb

Q72.00 Congenital complete absence of unspecified lower limb

Q72.01 Congenital complete absence of right lower limb

Q72.02 Congenital complete absence of left lower limb

Q72.03 Congenital complete absence of lower limb, bilateral

Q72.1 Congenital absence of thigh and lower leg with foot present

Q72.10 Congenital absence of unspecified thigh and lower leg with foot present

Q72.11 Congenital absence of right thigh and lower leg with foot present

Q72.12 Congenital absence of left thigh and lower leg with foot present

Q72.13 Congenital absence of thigh and lower leg with foot present, bilateral

Q72.2 Congenital absence of both lower leg and foot

Q72.20 Congenital absence of both lower leg and foot, unspecified lower limb

Q72.21 Congenital absence of both lower leg and foot, right lower limb

Q72.22 Congenital absence of both lower leg and foot, left lower limb

Q72.23 Congenital absence of both lower leg and foot, bilateral

Q72.3 Congenital absence of foot and toe(s)

Q72.30 Congenital absence of unspecified foot and toe(s)

Q72.31 Congenital absence of right foot and toe(s)

Q72.32 Congenital absence of left foot and toe(s)

Q72.33 Congenital absence of foot and toe(s), bilateral

Q72.4 Longitudinal reduction defect of femur
Proximal femoral focal deficiency

Q72.40 Longitudinal reduction defect of unspecified femur

Q72.41 Longitudinal reduction defect of right femur

Q72.42 Longitudinal reduction defect of left femur

Q72.43 Longitudinal reduction defect of femur, bilateral

Q72.5 Longitudinal reduction defect of tibia

Q72.50 Longitudinal reduction defect of unspecified tibia

Q72.51 Longitudinal reduction defect of right tibia

Q72.52 Longitudinal reduction defect of left tibia

Q72.53 Longitudinal reduction defect of tibia, bilateral

Q72.6 Longitudinal reduction defect of fibula

Q72.60 Longitudinal reduction defect of unspecified fibula

Q72.61 Longitudinal reduction defect of right fibula

Q72.62 Longitudinal reduction defect of left fibula

Q72.63 Longitudinal reduction defect of fibula, bilateral

Q72.7 Split foot

Q72.70 Split foot, unspecified lower limb

Q72.71 Split foot, right lower limb

Q72.72 Split foot, left lower limb

Q72.73 Split foot, bilateral

Q72.8 Other reduction defects of lower limb

Q72.81 Congenital shortening of lower limb

Q72.811 Congenital shortening of right lower limb

Q72.812 Congenital shortening of left lower limb

Q72.813 Congenital shortening of lower limb, bilateral

Q72.819 Congenital shortening of unspecified lower limb

Q72.89 Other reduction defects of lower limb

Q72.891 Other reduction defects of right lower limb

Q72.892 Other reduction defects of left lower limb

Q72.893 Other reduction defects of lower limb, bilateral

Q72.899 Other reduction defects of unspecified lower limb

Q72.9 Unspecified reduction defect of lower limb

Q72.90 Unspecified reduction defect of unspecified lower limb

Q72.91 Unspecified reduction defect of right lower limb

Q72.92 Unspecified reduction defect of left lower limb

Q72.93 Unspecified reduction defect of lower limb, bilateral

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 of limb NOS
Hemimelia of limb NOS
Reduction defect of limb NOS

Q74 Other congenital malformations of limb(s)

Excludes1: polydactyly (Q69.-)
reduction defect of limb (Q71-Q73)
syndactyly (Q70.-)

Q74.0 Other congenital malformations of upper limb(s), including shoulder girdle
Accessory carpal bones
Cleidocranial dysostosis
Congenital pseudarthrosis of clavicle
Macrodactylia (fingers)
Madelung's deformity
Radioulnar synostosis
Sprengel's deformity
Triphalangeal thumb

Q74.1 Congenital malformation of knee
Congenital absence of patella
Congenital dislocation of patella

Congenital genu valgum
Congenital genu varum
Rudimentary patella

Excludes1: congenital dislocation of knee (Q68.2)
congenital genu recurvatum (Q68.2)
nail patella syndrome (Q87.2)

Q74.2 Other congenital malformations of lower limb(s), including pelvic girdle
Congenital fusion of sacroiliac joint
Congenital malformation of ankle joint
Congenital malformation of sacroiliac joint

Excludes1: anteversion of femur (neck) (Q65.89)

Q74.3 Arthrogryposis multiplex congenita

Q74.8 Other specified congenital malformations of limb(s)

Q74.9 Unspecified congenital malformation of limb(s)
Congenital anomaly of limb(s) NOS

Q75 Other congenital malformations of skull and face bones

Excludes1: congenital malformation of face NOS (Q18.-)
congenital malformation syndromes classified to Q87.-
dentofacial anomalies [including malocclusion] (M26.-)
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
Acrocephaly
Imperfect fusion of skull
Oxycephaly
Trigonocephaly

Q75.1 Craniofacial dysostosis
Crouzon's disease

Q75.2 Hypertelorism

Q75.3 Macrocephaly

Q75.4 Mandibulofacial dysostosis
Franceschetti syndrome
Treacher Collins syndrome

Q75.5 Oculomandibular dysostosis

Q75.8 Other specified congenital malformations of skull and face bones
Absence of skull bone, congenital
Congenital deformity of forehead
Platybasia

Q75.9 Congenital malformation of skull and face bones, unspecified
Congenital anomaly of face bones NOS
Congenital anomaly of skull NOS

Q76 Congenital malformations of spine and bony thorax

Excludes1: congenital musculoskeletal deformities of spine and chest (Q67.5-Q67.8)

Q76.0 Spina bifida occulta

Excludes1: meningocele (spinal) (Q05.-)
spina bifida (aperta) (cystica) (Q05.-)

Q76.1 Klippel-Feil syndrome
Cervical fusion syndrome

Q76.2 Congenital spondylolisthesis
Congenital spondylolysis

Excludes1: spondylolisthesis (acquired) (M43.1-)
spondylolysis (acquired) (M43.0-)

Q76.3 Congenital scoliosis due to congenital bony malformation
Hemivertebra fusion or failure of segmentation with scoliosis

Q76.4 Other congenital malformations of spine, not associated with scoliosis

Q76.41 Congenital kyphosis

Q76.411 Congenital kyphosis, occipito-atlanto-axial region

Q76.412 Congenital kyphosis, cervical region

Q76.413 Congenital kyphosis, cervicothoracic region

Q76.414 Congenital kyphosis, thoracic region

Q76.415 Congenital kyphosis, thoracolumbar region

Q76.419 Congenital kyphosis, unspecified region

Q76.42 Congenital lordosis

Q76.425 Congenital lordosis, thoracolumbar region

Q76.426 Congenital lordosis, lumbar region

Q76.427 Congenital lordosis, lumbosacral region

Q76.428 Congenital lordosis, sacral and sacrococcygeal region

Q76.429 Congenital lordosis, unspecified region

Q76.49 Other congenital malformations of spine, not associated with scoliosis
Congenital absence of vertebra NOS
Congenital fusion of spine NOS
Congenital malformation of lumbosacral (joint) (region) NOS
Congenital malformation of spine NOS
Hemivertebra NOS
Malformation of spine NOS
Platyspondylisis NOS
Supernumerary vertebra NOS

Q76.5 Cervical rib
Supernumerary rib in cervical region

Q76.6 Other congenital malformations of ribs
Accessory rib

Congenital absence of rib
Congenital fusion of ribs
Congenital malformation of ribs NOS

Excludes1: short rib syndrome (Q77.2)

Q76.7 Congenital malformation of sternum
Congenital absence of sternum
Sternum bifidum

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

Excludes1: mucopolysaccharidosis (E76.0-E76.3)

Excludes2: congenital myotonic chondrodystrophy (G71.13)

Q77.0 Achondrogenesis
Hypochondrogenesis

Q77.1 Thanatophoric short stature

Q77.2 Short rib syndrome
Asphyxiating thoracic dysplasia [Jeune]

Q77.3 Chondrodysplasia punctata

Excludes1: Rhizomelic chondrodysplasia punctata (E71.43)

Q77.4 Achondroplasia
Hypochondroplasia
Osteosclerosis congenita

Q77.5 Diastrophic dysplasia

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

Q77.9 Osteochondrodysplasia with defects of growth of tubular bones and spine, unspecified

Q78 Other osteochondrodysplasias

Excludes2: congenital myotonic chondrodystrophy (G71.13)

Q78.0 Osteogenesis imperfecta
Fragilitas ossium
Osteopsathyrosis

Q78.1 Polyostotic fibrous dysplasia
Albright(-McCune)(-Sternberg) syndrome

Q78.2 Osteopetrosis
Albers-Schönberg syndrome
Osteosclerosis NOS

Q78.3 Progressive diaphyseal dysplasia
Camurati-Engelmann syndrome

Q78.4 Enchondromatosis
Maffucci's syndrome
Ollier's disease

Q78.5 Metaphyseal dysplasia
Pyle's syndrome

Q78.6 Multiple congenital exostoses
Diaphyseal aclasis

Q78.8 Other specified osteochondrodysplasias
Osteopoikilosis

Q78.9 Osteochondrodysplasia, unspecified
Chondrodystrophy NOS
Osteodystrophy NOS

Q79 Congenital malformations of musculoskeletal system, not elsewhere classified

Excludes2: congenital (sternomastoid) torticollis (Q68.0)

Q79.0 Congenital diaphragmatic hernia

Excludes1: congenital hiatus hernia (Q40.1)

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

Q79.2 Exomphalos
Omphalocele

Excludes1: umbilical hernia (K42.-)

Q79.3 Gastroschisis

Q79.4 Prune belly syndrome
Congenital prolapse of bladder mucosa
Eagle-Barrett syndrome

Q79.5 Other congenital malformations of abdominal wall

Excludes1: umbilical hernia (K42.-)

Q79.51 Congenital hernia of bladder

Q79.59 Other congenital malformations of abdominal wall

Q79.6 Ehlers-Danlos syndrome

Q79.8 Other congenital malformations of musculoskeletal system
Absence of muscle
Absence of tendon
Accessory muscle
Amyotrophia congenita
Congenital constricting bands
Congenital shortening of tendon
Poland syndrome

Q79.9 Congenital malformation of musculoskeletal system, unspecified
Congenital anomaly of musculoskeletal system NOS
Congenital deformity of musculoskeletal system NOS

Other congenital malformations (Q80-Q89)

Q80 Congenital ichthyosis

Excludes1: Refsum's disease (G60.1)

Q80.0 Ichthyosis vulgaris

Q80.1 X-linked ichthyosis

Q80.2 Lamellar ichthyosis
Collodion baby

Q80.3 Congenital bullous ichthyosiform erythroderma

Q80.4 Harlequin fetus

Q80.8 Other congenital ichthyosis

Q80.9 Congenital ichthyosis, unspecified

Q81 Epidermolysis bullosa

Q81.0 Epidermolysis bullosa simplex

Excludes1: 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

Excludes1: acrodermatitis enteropathica (E83.2)
congenital erythropoietic porphyria (E80.0)
pilonidal cyst or sinus (L05.-)
Sturge-Weber (-Dimitri) syndrome (Q85.8)

Q82.0 Hereditary lymphedema

Q82.1 Xeroderma pigmentosum

Q82.2 Congenital cutaneous mastocytosis
Congenital diffuse cutaneous mastocytosis
Congenital maculopapular cutaneous mastocytosis
Congenital urticaria pigmentosa

Excludes1: cutaneous mastocytosis NOS (D47.01)
diffuse cutaneous mastocytosis (with onset after newborn period) (D47.01)
malignant mastocytosis (C96.2-)
systemic mastocytosis (D47.02)
urticaria pigmentosa (non-congenital) (with onset after newborn period) (D47.01)

Q82.3 Incontinentia pigmenti

Q82.4 Ectodermal dysplasia (anhidrotic)

Excludes1: Ellis-van Creveld syndrome (Q77.6)

Q82.5 Congenital non-neoplastic nevus
Birthmark NOS
Flammeus Nevus
Portwine Nevus
Sanguineous Nevus
Strawberry Nevus
Vascular Nevus NOS
Verrucous Nevus

Excludes2: Café au lait spots (L81.3)
lentigo (L81.4)
nevus NOS (D22.-)
araneus nevus (I78.1)
melanocytic nevus (D22.-)
pigmented nevus (D22.-)
spider nevus (I78.1)
stellar nevus (I78.1)

Q82.6 Congenital sacral dimple
Parasacral dimple

Excludes2: pilonidal cyst with abscess (L05.01)
pilonidal cyst without abscess (L05.91)

Q82.8 Other specified congenital malformations of skin
Abnormal palmar creases
Accessory skin tags
Benign familial pemphigus [Hailey-Hailey]
Congenital poikiloderma
Cutis laxa (hyperelastica)
Dermatoglyphic anomalies
Inherited keratosis palmaris et plantaris
Keratosis follicularis [Darier-White]

Excludes1: Ehlers-Danlos syndrome (Q79.6)

Q82.9 Congenital malformation of skin, unspecified

Q83 Congenital malformations of breast

Excludes2: absence of pectoral muscle (Q79.8)
hypoplasia of breast (N64.82)
micromastia (N64.82)

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

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

Excludes1: Menkes' kinky hair syndrome (E83.0)

Q84.2 Other congenital malformations of hair
Congenital hypertrichosis
Congenital malformation of hair NOS
Persistent lanugo

Q84.3 Anonychia

Excludes1: 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
Congenital koilonychia
Congenital malformation of nail NOS

Q84.8 Other specified congenital malformations of integument
Aplasia cutis congenita

Q84.9 Congenital malformation of integument, unspecified
Congenital anomaly of integument NOS
Congenital deformity of integument NOS

Q85 Phakomatoses, not elsewhere classified

Excludes1: ataxia telangiectasia [Louis-Bar] (G11.3)
familial dysautonomia [Riley-Day] (G90.1)

Q85.0 Neurofibromatosis (nonmalignant)

Q85.00 Neurofibromatosis, unspecified

Q85.01 Neurofibromatosis, type 1
Von Recklinghausen disease

Q85.02 Neurofibromatosis, type 2
Acoustic neurofibromatosis

Q85.03 Schwannomatosis

Q85.09 Other neurofibromatosis

Q85.1 Tuberous sclerosis
Bourneville's disease
Epiloia

Q85.8 Other phakomatoses, not elsewhere classified
Peutz-Jeghers Syndrome
Sturge-Weber(-Dimitri) syndrome
von Hippel-Lindau syndrome

Excludes1: Meckel-Gruber syndrome (Q61.9)

Q85.9 Phakomatosis, unspecified
Hamartosis NOS

Q86 Congenital malformation syndromes due to known exogenous causes, not elsewhere classified

Excludes2: 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
Meadow's syndrome

Q86.2 Dysmorphism due to warfarin

Q86.8 Other congenital malformation syndromes due to known exogenous causes

Q87 Other specified congenital malformation syndromes affecting multiple systems

Use additional code(s) to identify all associated manifestations

Q87.0 Congenital malformation syndromes predominantly affecting facial appearance
Acrocephalopolysyndactyly
Acrocephalosyndactyly [Apert]
Cryptophthalmos syndrome
Cyclopia
Goldenhar syndrome
Moebius syndrome
Oro-facial-digital syndrome
Robin syndrome
Whistling face

Q87.1 Congenital malformation syndromes predominantly associated with short stature
Aarskog syndrome
Cockayne syndrome
De Lange syndrome
Dubowitz syndrome
Noonan syndrome
Prader-Willi syndrome
Robinow-Silverman-Smith syndrome
Russell-Silver syndrome
Seckel syndrome

Excludes1: Ellis-van Creveld syndrome (Q77.6)
Smith-Lemli-Opitz syndrome (E78.72)

Q87.2 Congenital malformation syndromes predominantly involving limbs
Holt-Oram syndrome
Klippel-Trenaunay-Weber syndrome
Nail patella syndrome
Rubinstein-Taybi syndrome
Sirenomelia syndrome
Thrombocytopenia with absent radius [TAR] syndrome
VATER syndrome

Q87.3 Congenital malformation syndromes involving early overgrowth
Beckwith-Wiedemann syndrome
Sotos syndrome
Weaver syndrome

Q87.4 Marfan's syndrome

Q87.40 Marfan's syndrome, unspecified

Q87.41 Marfan's syndrome with cardiovascular manifestations

Q87.410 Marfan's syndrome with aortic dilation

Q87.418 Marfan's syndrome with other cardiovascular manifestations

Q87.42 Marfan's syndrome with ocular manifestations

Q87.43 Marfan's syndrome with skeletal manifestation

Q87.5 Other congenital malformation syndromes with other skeletal changes

Q87.8 Other specified congenital malformation syndromes, not elsewhere classified

Excludes1: Zellweger syndrome (E71.510)

Q87.81 Alport syndrome

Use additional code to identify stage of chronic kidney disease (N18.1-N18.6)

Q87.82 Arterial tortuosity syndrome

Q87.89 Other specified congenital malformation syndromes, not elsewhere classified
Laurence-Moon (-Bardet)-Biedl syndrome

Q89 Other congenital malformations, not elsewhere classified

Q89.0 Congenital absence and malformations of spleen

Excludes1: isomerism of atrial appendages (with asplenia or polysplenia) (Q20.6)

Q89.01 Asplenia (congenital)

Q89.09 Congenital malformations of spleen
Congenital splenomegaly

Q89.1 Congenital malformations of adrenal gland

Excludes1: adrenogenital disorders (E25.-)
congenital adrenal hyperplasia (E25.0)

Q89.2 Congenital malformations of other endocrine glands
Congenital malformation of parathyroid or thyroid gland
Persistent thyroglossal duct
Thyroglossal cyst

Excludes1: congenital goiter (E03.0)
congenital hypothyroidism (E03.1)

Q89.3 Situs inversus
Dextrocardia with situs inversus
Mirror-image atrial arrangement with situs inversus
Situs inversus or transversus abdominalis
Situs inversus or transversus thoracis
Transposition of abdominal viscera
Transposition of thoracic viscera

Excludes1: dextrocardia NOS (Q24.0)

Q89.4 Conjoined twins
Craniopagus
Dicephaly

Pygopagus
Thoracopagus

Q89.7 Multiple congenital malformations, not elsewhere classified
Multiple congenital anomalies NOS
Multiple congenital deformities NOS

Excludes1: congenital malformation syndromes affecting multiple systems (Q87.-)

Q89.8 Other specified congenital malformations

Use additional code(s) to identify all associated manifestations

Q89.9 Congenital malformation, unspecified
Congenital anomaly NOS
Congenital deformity NOS

Chromosomal abnormalities, not elsewhere classified (Q90-Q99)

Excludes2: mitochondrial metabolic disorders (E88.4-)

Q90 Down syndrome

Use additional code(s) to identify any associated physical conditions and degree of intellectual disabilities (F70-F79)

Q90.0 Trisomy 21, nonmosaicism (meiotic nondisjunction)

Q90.1 Trisomy 21, mosaicism (mitotic nondisjunction)

Q90.2 Trisomy 21, translocation

Q90.9 Down syndrome, unspecified
Trisomy 21 NOS

Q91 Trisomy 18 and Trisomy 13

Q91.0 Trisomy 18, nonmosaicism (meiotic nondisjunction)

Q91.1 Trisomy 18, mosaicism (mitotic nondisjunction)

Q91.2 Trisomy 18, translocation

Q91.3 Trisomy 18, unspecified

Q91.4 Trisomy 13, nonmosaicism (meiotic nondisjunction)

Q91.5 Trisomy 13, mosaicism (mitotic nondisjunction)

Q91.6 Trisomy 13, translocation

Q91.7 Trisomy 13, unspecified

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

Includes: unbalanced translocations and insertions

Excludes1: trisomies of chromosomes 13, 18, 21 (Q90-Q91)

Q92.0 Whole chromosome trisomy, nonmosaicism (meiotic nondisjunction)

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

Q92.2 Partial trisomy
Less than whole arm duplicated

Whole arm or more duplicated

Excludes1: partial trisomy due to unbalanced translocation (Q92.5)

Q92.5 Duplications with other complex rearrangements
Partial trisomy due to unbalanced translocations

Code also any associated deletions due to unbalanced translocations, inversions and insertions (Q93.7)

Q92.6 Marker chromosomes
Trisomies due to dicentrics
Trisomies due to extra rings
Trisomies due to isochromosomes
Individual with marker heterochromatin

Q92.61 Marker chromosomes in normal individual

Q92.62 Marker chromosomes in abnormal individual

Q92.7 Triploidy and polyploidy

Q92.8 Other specified trisomies and partial trisomies of autosomes
Duplications identified by fluorescence in situ hybridization (FISH)
Duplications identified by in situ hybridization (ISH)
Duplications seen only at prometaphase

Q92.9 Trisomy and partial trisomy of autosomes, unspecified

Q93 Monosomies and deletions from the autosomes, not elsewhere classified

Q93.0 Whole chromosome monosomy, nonmosaicism (meiotic nondisjunction)

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

Q93.2 Chromosome replaced with ring, dicentric or isochromosome

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
Angelman syndrome

Q93.7 Deletions with other complex rearrangements
Deletions due to unbalanced translocations, inversions and insertions

Code also any associated duplications due to unbalanced translocations, inversions and insertions (Q92.5)

Q93.8 Other deletions from the autosomes

Q93.81 Velo-cardio-facial syndrome
Deletion 22q11.2

Q93.88 Other microdeletions
Miller-Dieker syndrome
Smith-Magenis syndrome

Q93.89 Other deletions from the autosomes
Deletions identified by fluorescence in situ hybridization (FISH)
Deletions identified by in situ hybridization (ISH)
Deletions seen only at prometaphase

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

Q95.5 Individual with autosomal fragile site

Q95.8 Other balanced rearrangements and structural markers

Q95.9 Balanced rearrangement and structural marker, unspecified

Q96 Turner's syndrome

Excludes1: Noonan syndrome (Q87.1)

Q96.0 Karyotype 45, X

Q96.1 Karyotype 46, X iso (Xq)
Karyotype 46, isochromosome Xq

Q96.2 Karyotype 46, X with abnormal sex chromosome, except iso (Xq)
Karyotype 46, X with abnormal sex chromosome, except isochromosome 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

Excludes1: Turner's syndrome (Q96.-)

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

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 syndrome karyotype 47, XXY

Q98.1 Klinefelter syndrome, male with more than two X chromosomes

Q98.3 Other male with 46, XX karyotype

Q98.4 Klinefelter 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