New York State Department of Health
Birth Defects Registry

ICD-10 Coding Manual

List of all Reportable Congenital Malformations

Last updated 11/16/2023

Table of Contents

Reporting Requirements and Instructions

Children to Report:

Hospitals and healthcare providers are required to report children born or living in New York State (NYS) who have been diagnosed, before two years of age (unless otherwise specified), with birth defects, chromosomal anomalies, or were stillborn or other non-liveborn. Certain birth defects are required to be reported for children up to age 10 (specified in red). For purposes of this registry, a birth defect (also referred to as anomaly or malformation) is defined as any structural, functional, and/or specific biochemical abnormalities which were determined genetically or induced during gestation and not due to birthing events. This includes newborns and pediatric patients with a birth defect, even if they are transferred to or from another hospital, and any child with a birth defect who dies in the hospital (inpatient or emergency room).

What to Report:

This coding manual contains a complete list of NYS Birth Defects Registry (BDR) reportable anomalies. Rows with grey backgrounds represent non-billable codes identified by the ICD-10 nomenclature. When reporting syndromes which have several defects, report each individual defect as well as the associated syndrome. Tables contain the following three columns:

1st Column called 'ICD-10' – ICD-10 codes that correspond to the child's birth defect.

2nd Column called 'Description' – Lists the standard narrative birth defect description(s) and may provide specific directions (e.g., must specify, female only, male only, birth weight criteria, etc.)

- All major defects and stillbirths must be reported
  - Minor defects, descriptions with an *(m)* (e.g. *(m) ankyloglossia*), are only required to be reported if the child also has one or more major defects or was stillborn or other non-liveborn.
- Some birth defects are only reportable if the child is a certain sex or was born above a certain birth weight. Please only report them if they meet the given criteria or the criteria is unknown. Examples:
  - Only report patent ductus arteriosus (PDA) if the child's birth weight is 1,500 grams or greater. PDA is normal and necessary as a fetus develops and should close at birth.
  - Only report K40-, inguinal hernia, for female children
- **Please include as much specific detail from the medical record as possible for each defect**
  - Specific narrative descriptions of birth defects are **required** for reporting of all birth defects
  - Indicate the laterality if applicable (e.g., talipes equinovarus, left)
  - Avoid reporting vague descriptions starting with "Other" or those with codes ending in -8 or -9
  - Vague Descriptions state '(must specify)' as a reminder to provide the actual birth defect diagnosis. For example, "Other specified congenital malformations of heart (must specify)" needs to be updated to the actual diagnosis in the medical record.

3rd Column called 'ICD-9': ICD-9 code that corresponds with the respective ICD-10 code.

Syndrome and chromosomal defects: please report the cytogenetic diagnoses or test results. These results are also required for defects under ICD-10 code Q56 (indeterminate sex or pseudohermaphroditism).

Common Acronyms:

(m) = Minor defect
NOS = Not Otherwise Specified
NEC = Not Elsewhere Classified

Color Coding:

Black font: From ICD-10 Codebook
Blue font: Additional synonyms or descriptions of other conditions also included under this code.
Red font: Key instructions to follow

Common Notation:

Includes: Further defines, or gives examples of, the content of the code or category
Excludes 1 (Type 1 Excludes): Indicates the code identified under the 'excludes' statement and the respective ICD10 code cannot be reported together because the two conditions cannot appear together.

Excludes 2 (Type 2 Excludes): Condition excluded is not part of the condition it is excluded from, but a patient may have both conditions at the same time. Report both codes if the patient has both conditions.

Please see Revision History section for changes in ICD-10 codes over time.

Congenital Malformations of the Nervous System (Q00-Q07)

ICD-10	Description	ICD-9
Q00	**Anencephaly and similar malformations**	
Q00.0	Anencephaly/Anencephalus Acephaly Acrania Amyelencephaly/Amyelencephalus Congenital brain aplasia Hemianencephaly Hemicephaly	740.0
Q00.1	Craniorachischisis (fissure skull/vertebral column)	740.1
Q00.2	Iniencephaly	740.2
Q01	**Encephalocele** **Includes:** Arnold-Chiari syndrome, type III, encephalocystocele, encephalomyelocele, hydroencephalocele, hydromeningocele (cranial), meningocele (cerebral), meningoencephalocele **Excludes 1:** Meckel-Gruber syndrome (Q61.9)	
Q01.0	Frontal encephalocele Encephalocystocele, frontal Encephalomyelocele, frontal Hydroencephalocele, frontal Hydromeningocele, frontal Meningocele, frontal Meningoencephalocele, frontal	742.0
Q01.1	Nasofrontal encephalocele Encephalocystocele, nasofrontal Encephalomyelocele, nasofrontal Hydroencephalocele, nasofrontal Hydromeningocele, nasofrontal Meningocele, nasofrontal Meningoencephalocele, nasofrontal	742.0
Q01.2	Occipital encephalocele Encephalocystocele, occipital Encephalomyelocele, occipital Hydroencephalocele, occipital Hydromeningocele, occipital Meningocele, occipital Menigoencephalocele, occipital	742.0
Q01.8	Encephalocele of other sites: cranial, cerebral, nasal, parietal, other specified site (must specify) Encephalocystocele (cranial, cerebral, nasal, parietal or other specified site) Encephalomyelocele (cranial, cerebral, nasal, parietal or other specified site) Hydroencephalocele (cranial, cerebral, nasal, parietal or other specified site) Hydromeningocele (cranial, cerebral, nasal, parietal or other specified site) Meningocele (cranial, cerebral, nasal, parietal or other specified site) Meningoencephalocele (cranial, cerebral, nasal, parietal or other specified site)	742.0
Q01.9	Encephalocele, unspecified (try not to use this code) (must specify) Arnold-Chiari syndrome, type 3 Congenital cerebral meningocele Encephalocystocele, unspecified	742.0

	Encephalomyelocele, unspecified Hydroencephalocele, unspecified Hydromeningocele, cranial, unspecified Meningocele, cerebral, unspecified Meningoencephalocele, unspecified	
Q02	**Microcephaly** Code first, if applicable, congenital Zika virus disease **Includes:** hydromicrocephaly, micrencephalon **Excludes 1:** Meckel-Gruber syndrome (Q61.9)	
Q02	Microcephaly Congenital brain hypoplasia Galloway syndrome Hydromicrocephaly Micrencephalon Micrencephaly Microcephalus Primary microcephaly **Use Additional Code**, if applicable, to identify congenital Zika virus disease	742.1
Q03	**Congenital hydrocephalus** **Excludes 1:** 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 (must specify) Obstruction of aqueduct of Sylvius, congenital Stenosis of aqueduct of Sylvius	742.3
Q03.1	Atresia of foramina of Magendie and Luschka Dandy-Walker syndrome	742.3
Q03.8	Other congenital hydrocephalus (must specify) Communicating hydrocephaly/enlarged Cisterna Magna Other specified hydrocephaly (do not code if Intraventricular hemorrhage/bleed) Ventriculomegaly of the Head (verify not ventriculomegaly of the heart, do not report/code if Mild dilatation)	742.3
Q03.9	Congenital hydrocephalus, unspecified	742.3
Q04	**Other congenital malformations of the brain** **Excludes 1:** cyclopia (Q87.0), macrocephaly (Q75.3)	
Q04.0	Congenital malformations of corpus callosum (must specify) Agenesis of corpus callosum Aplasia corpus callosum Hypoplasia, hypoplastic corpus callosum Other anomalies of the corpus callosum (must specify) Partial agenesis of the corpus callosum Absent corpus callosum Hypogenesis of corpus callosum	742.2
Q04.1	Arhinencephaly Arhinia	742.2
Q04.2	Holoprosencephaly Holoprosencephaly sequence	742.2
Q04.3	Other reduction deformities of the brain (must specify) Absence of part of brain Agenesis of part brain Agyria Aplasia of part of brain Hydranencephaly Hypoplasia of part of brain Lissencephaly Microgyria	742.2

	Pachygyria Polymicrogyria **Excludes 1**: congenital malformations of corpus callosum (Q04.0)	
Q04.4	Septo-optic dysplasia of brain Septo-optic dysplasia sequence De Morsier syndrome	742.4
Q04.5	Megalencephaly Enlarged brain and/or head	742.4
Q04.6	Congenital cerebral cyst(s) Colloid cyst of third ventricle Congenital pseudoporencephaly Porencephaly Schizencephaly *(m) Congenital choroid plexus cyst* Porencephalic cysts **Excludes 1**: acquired porencephalic cyst (G93.0)	742.4
Q04.8	Other specified congenital malformation of brain (must specify) Arnold-Chiari syndrome, type IV Cerebral ventriculomegaly Macrogyria Anomalies of cerebellum, includes atrophy Anomalies of hypothalamus Colpocephaly Cortical atrophy Cranial nerve defects Cystic encephalomalacia Encephalomalacia Other specified anomalies of brain Small brain Ulegyria Ventricular cysts	742.4
Q04.9	Congenital malformation of brain, unspecified (try not to use this code) (must specify) Brain anomaly Congenital disease or lesion NOS of brain Multiple anomalies NOS of brain, congenital	742.9
Q05	**Spina bifida (SB)** **Includes:** hydromeningocele (spinal), meningocele (spinal), meningomyelocele, myelocele, myelomeningocele, rachischisis, spina bifida (aperta)(cystica), syringomyelocele **Excludes 1:** Arnold-Chiari syndrome, type II (Q07.0-), spina bifida occulta (Q76.0)	
Q05.0	Cervical spina bifida (SB) with hydrocephalus Cervical SB with hydrocephalus and hydromeningocele Cervical SB with hydrocephalus and meningocele Cervical SB with hydrocephalus and meningomyelocele Cervical SB with hydrocephalus and myelocele Cervical SB with hydrocephalus and myelomeningocele Cervical SB with hydrocephalus and rachischisis Cervical SB with hydrocephalus and syringohydromyelia Cervical SB with hydrocephalus and lipomeningomyelocele Cervical SB with hydrocephalus and lipomyelomeningocele Cervical SB with hydrocephalus and lipomeningocele Cervical SB with hydrocephalus and spina bifida (aperta)(cystica)	741.01
Q05.1	Thoracic spina bifida (SB) with hydrocephalus Dorsal spina bifida Thoracolumbar spina bifida (SB) with hydrocephalus Thoracic/dorsal/thoracolumbar SB with hydrocephalus and hydromeningocele Thoracic/dorsal/thoracolumbar SB with hydrocephalus and meningocele	741.02

	Thoracic/dorsal/thoracolumbar SB with hydrocephalus and meningomyelocele Thoracic/dorsal/thoracolumbar SB with hydrocephalus and myelocele Thoracic/dorsal/thoracolumbar SB with hydrocephalus and myelomeningocele Thoracic/dorsal/thoracolumbar SB with hydrocephalus and rachischisis Thoracic/dorsal/thoracolumbar SB with hydrocephalus and syringohydromyelia Thoracic/dorsal/thoracolumbar SB with hydrocephalus and lipomeningomyelocele Thoracic/dorsal/thoracolumbar SB with hydrocephalus and lipomyelomeningocele Thoracic/dorsal/thoracolumbar SB with hydrocephalus and lipomeningocele Thoracic/dorsal/thoracolumbar SB with hydrocephalus and SB (aperta)(cystica)	
Q05.2	Lumbar spina bifida (SB) with hydrocephalus Lumbosacral spina bifida (SB) with hydrocephalus Lumbar/lumbosacral SB with hydrocephalus and hydromeningocele Lumbar/lumbosacral SB with hydrocephalus and meningocele Lumbar/lumbosacral SB with hydrocephalus and meningomyelocele Lumbar/lumbosacral SB with hydrocephalus and myelocele Lumbar/lumbosacral SB with hydrocephalus and myelomeningocele Lumbar/lumbosacral SB with hydrocephalus and rachischisis Lumbar/lumbosacral SB with hydrocephalus and syringohydromyelia Lumbar/lumbosacral SB with hydrocephalus and lipomeningomyelocele Lumbar/lumbosacral SB with hydrocephalus and lipomyelomeningocele Lumbar/lumbosacral SB with hydrocephalus and lipomeningocele Lumbar/lumbosacral SB with hydrocephalus and SB (aperta)(cystica)	741.03
Q05.3	Sacral spina bifida (SB) with hydrocephalus Sacral SB with hydrocephalus and hydromeningocele Sacral SB with hydrocephalus and meningocele Sacral SB with hydrocephalus and meningomyelocele Sacral SB with hydrocephalus and myelocele Sacral SB with hydrocephalus and myelomeningocele Sacral SB with hydrocephalus and rachischisis Sacral SB with hydrocephalus and syringohydromyelia Sacral SB with hydrocephalus and lipomeningomyelocele Sacral SB with hydrocephalus and lipomyelomeningocele Sacral SB with hydrocephalus and lipomeningocele Sacral SB with hydrocephalus and SB (aperta)(cystica)	741.03
Q05.4	Unspecified spina bifida (SB) with hydrocephalus Unspecified SB with hydrocephalus and hydromeningocele Unspecified SB with hydrocephalus and meningocele Unspecified SB with hydrocephalus and meningomyelocele Unspecified SB with hydrocephalus and myelocele Unspecified SB with hydrocephalus and myelomeningocele Unspecified SB with hydrocephalus and rachischisis Unspecified SB with hydrocephalus and syringohydromyelia Unspecified SB with hydrocephalus and lipomeningomyelocele Unspecified SB with hydrocephalus and lipomyelomeningocele Unspecified SB with hydrocephalus and lipomeningocele Unspecified SB with hydrocephalus and SB (aperta)(cystica)	741.00
Q05.5	Cervical spina bifida (SB) without hydrocephalus Cervical SB without hydrocephalus, with hydromeningocele Cervical SB without hydrocephalus, with lipomeningomyelocele Cervical SB without hydrocephalus, with lipomyelomeningocele Cervical SB without hydrocephalus, with lipomeningocele Cervical SB without hydrocephalus, with meningocele Cervical SB without hydrocephalus, with meningomyelocele Cervical SB without hydrocephalus, with myelocele Cervical SB without hydrocephalus, with myelomeningocele Cervical SB without hydrocephalus, with rachischisis Cervical SB without hydrocephalus, with SB (aperta)(cystica)	741.91

	Cervical SB without hydrocephalus, with syringohydromyelia	
Q05.6	Thoracic spina bifida (SB) without hydrocephalus Dorsal spina bifida (SB), NOS Thoracolumbar spina bifida(SB), NOS Thoracic/dorsal/thoracolumbar SB without hydrocephaly with hydromeningocele Thoracic/dorsal/thoracolumbar SB without hydrocephaly with lipomeningomyelocele Thoracic/dorsal/thoracolumbar SB without hydrocephaly with lipomyelomeningocele Thoracic/dorsal/thoracolumbar SB without hydrocephaly with lipomeningocele Thoracic/dorsal/thoracolumbar SB without hydrocephaly with meningocele Thoracic/dorsal/thoracolumbar SB without hydrocephaly with meningomyelocele Thoracic/dorsal/thoracolumbar SB without hydrocephaly with myelocele Thoracic/dorsal/thoracolumbar SB without hydrocephaly with myelomeningocele Thoracic/dorsal/thoracolumbar SB without hydrocephaly with rachischisis Thoracic/dorsal/thoracolumbar SB without hydrocephaly with SB (aperta)(cystica) Thoracic/dorsal/thoracolumbar SB without hydrocephaly with syringohydromyelia	741.92
Q05.7	Lumbar spina bifida (SB) without hydrocephalus Lumbosacral SB, NOS Lumbar/lumbosacral SB without hydrocephalus, with hydromeningocele Lumbar/lumbosacral SB without hydrocephalus, with lipomeningomyelocele Lumbar/lumbosacral SB without hydrocephalus, with lipomyelomeningocele Lumbar/lumbosacral SB without hydrocephalus, with lipomeningocele Lumbar/lumbosacral SB without hydrocephalus, with meningocele Lumbar/lumbosacral SB without hydrocephalus, with meningomyelocele Lumbar/lumbosacral SB without hydrocephalus, with myelocele Lumbar/lumbosacral SB without hydrocephalus, with myelomeningocele Lumbar/lumbosacral SB without hydrocephalus, with rachischisis Lumbar/lumbosacral SB without hydrocephalus, with SB (aperta)(cystica) Lumbar/lumbosacral SB without hydrocephalus, with syringohydromyelia	741.93
Q05.8	Sacral spina bifida (SB) without hydrocephalus Sacral SB without hydrocephalus, with hydromeningocele Sacral SB without hydrocephalus, with lipomeningomyelocele Sacral SB without hydrocephalus, with lipomyelomeningocele Sacral SB without hydrocephalus, with lipomeningocele Sacral SB without hydrocephalus, with meningocele Sacral SB without hydrocephalus, with meningomyelocele Sacral SB without hydrocephalus, with myelocele Sacral SB without hydrocephalus, with myelomeningocele Sacral SB without hydrocephalus, with rachischisis Sacral SB without hydrocephalus, with spina bifida (aperta)(cystica) Sacral SB without hydrocephalus, with syringohydromyelia	741.90
Q05.9	Spina bifida (SB) unspecified (without hydrocephalus) (must specify) Congenital meningocele Decubitus ulcer due to spina bifida Hydromeningocele Hydromeningomyelocele Hydromyelocele Lipoma of spina bifida Lipomyelomeningocele Meningomyelocele Myelocele Myelomeningocele Myelomeningocele without hydrocephalus Neurogenic bladder due to spina bifida Pressure ulcer due to spina bifida Rachischisis SB without hydrocephalus Unspecified SB without hydrocephalus, with lipomeningomyelocele	741.90

	Unspecified SB without hydrocephalus, with lipomeningocele Unspecified SB without hydrocephalus, with meningocele Unspecified SB without hydrocephalus, with SB (aperta)(cystica) Unspecified SB without hydrocephalus, with syringohydromyelia	
Q06	**Other congenital malformations of spinal cord**	
Q06.0	Amyelia (congenital absence of spinal cord)	742.59
Q06.1	Hypoplasia or dysplasia of spinal cord Atelomyelia (spinal cord defect) Myelatelia Myelodysplasia of spinal cord	742.59
Q06.2	Diastematomyelia	742.51
Q06.3	Other congenital cauda equina malformations (must specify)	742.59
Q06.4	Hydromyelia Hydrorachis	742.53
Q06.8	Other specified congenital malformations of spinal cord (must specify) Lumbosacral dermal sinus tract Occult spinal dysraphism sequence Tethered cord syndrome, congenital Tethered cord, filum Low lying spinal chord Tethered filum terminale *(m) Fibrolipoma of filum terminale*	742.59
Q06.9	Congenital malformation of spinal cord, unspecified (must specify) Cervical spine dysraphism Congenital anomaly NOS of spinal cord Congenital deformity NOS of spinal cord Congenital disease or lesion NOS of spinal cord Spinal cord anomaly Split spinal cord	742.9
Q07	**Other congenital malformations of nervous system** **Excludes 2:** congenital central alveolar hypoventilation syndrome (G47.35), familial dysautonomia [Riley-Day] (G90.1), neurofibromatosis (nonmalignant) (Q85.0-)	
Q07.0	**Arnold-Chiari syndrome (type II), without hydrocephalus** **Excludes 1**: Arnold-Chiari syndrome, type III (Q01.-), Arnold-Chiari syndrome, type IV (Q04.8)	
Q07.00	Arnold-Chiari syndrome (type II) without spina bifida or hydrocephalus Arnold-Chiari type 2 without hydrocephalus	741.90
Q07.01	Arnold-Chiari syndrome (type II) with spina bifida (SB) Arnold-Chiari syndrome (type II) with SB and hydromeningocele Arnold-Chiari syndrome (type II) with SB and meningocele Arnold-Chiari syndrome (type II) with SB and meningomyelocele Arnold-Chiari syndrome (type II) with SB and myelocele Arnold-Chiari syndrome (type II) with SB and myelomeningocele Arnold-Chiari syndrome (type II) with SB and rachischisis Arnold-Chiari syndrome (type II) with SB and syringohydromyelia Arnold-Chiari syndrome (type II) with SB and lipomeningomyelocele Arnold-Chiari syndrome (type II) with SB and lipomyelomeningocele Arnold-Chiari syndrome (type II) with SB and lipomeningocele Arnold-Chiari syndrome (type II) with SB and SB (aperta)(cystica) Specify (cervical, thoracic, dorsal, thoracolumbar, lumbar, lumbosacral, sacral, unspecified)	741.90 741.91 741.92 741.93
Q07.02	Arnold-Chiari syndrome (type II) with hydrocephalus Arnold-Chiari Type 2 with hydrocephalus	742.0
Q07.03	Arnold-Chiari syndrome (type II) with spina bifida (SB) and hydrocephalus Arnold-Chiari syndrome (type II) with SB, hydrocephalus and hydromeningocele Arnold-Chiari syndrome (type II) with SB, hydrocephalus and meningocele Arnold-Chiari syndrome (type II) with SB, hydrocephalus and meningomyelocele	741.00 741.01 741.02 741.03

	Arnold-Chiari syndrome (type II) with SB, hydrocephalus and myelocele Arnold-Chiari syndrome (type II) with SB, hydrocephalus and myelomeningocele Arnold-Chiari syndrome (type II) with SB, hydrocephalus and rachischisis Arnold-Chiari syndrome (type II) with SB, hydrocephalus and syringohydromyelia Arnold-Chiari syndrome (type II) with SB, hydrocephalus and lipomeningomyelocele Arnold-Chiari syndrome (type II) with SB, hydrocephalus and lipomyelomeningocele Arnold-Chiari syndrome (type II) with SB, hydrocephalus and lipomeningocele Arnold-Chiari syndrome (type II) with SB, hydrocephalus and SB (aperta)(cystica) Specify (cervical, thoracic, dorsal, thoracolumbar, lumbar, lumbosacral, sacral, unspecified)	
Q07.8	Other specified congenital malformations of nervous system (must specify) Agenesis of nerve Congenital optic nerve pit Displacement of brachial plexus Incomplete development of nerve Jaw-winking syndrome Marcus Gunn's syndrome Marcus Gunn jaw winking syndrome Synkinesis, eyelid	742.8
Q07.9	Congenital malformation of nervous system, unspecified (must specify) Central nervous system (CNS) malformation, fetal Congenital disease or lesion NOS of nervous system Nervous system anomaly Neutral tube defect	742.9

Congenital Malformations of Eye, Ear, Face and Neck (Q10-Q18)

ICD-10	Description	ICD-9
Q10	**Congenital malformation of eyelid, lacrimal apparatus and orbit** **Excludes 1:** cryptophthalmos NOS (Q11.2), cryptophthalmos syndrome (Q87.0)	
Q10.0	Congenital ptosis Bilateral congenital ptosis Left congenital ptosis Right congenital ptosis Blepharoptosis	743.61
Q10.1	Congenital ectropion	743.62
Q10.2	Congenital entropion	743.62
Q10.3	Other congenital malformations of eyelid (must specify) Ablepharon Blepharophimosis, congenital Coloboma of eyelid Congenital absence or agenesis of eyelid Congenital accessory eyelid Congenital accessory eye muscle Telecanthus *(m) Congenital absence or agenesis of cilia* *(m) Congenital malformation of eyelid NOS* *(m) Absence of eyelashes* *(m) Epicanthal folds* *(m) Fused eyelids (if birth weight greater than 1000 grams)* *(m) Slanted eyes* *(m) Small or narrow palpebral fissures* *(m) Upward eyeslant* *(m) Weakness of eyelids* Do not report pseudostrabismus	743.62 743.63
Q10.4	*(m) Absence and agenesis of lacrimal apparatus* *(m) Congenital absence of punctum lacrimal* *(m) Nasolacrimal duct obstruction*	743.65
Q10.5	*(m) Congenital stenosis and stricture of lacrimal duct* *(m) Congenital blocked tear duct* *(m) Congenital dacryostenosis (left/ right/ bilateral)* *(m) Obstruction of lacrimal duct(s)*	743.65
Q10.6	*(m) Other congenital malformations of lacrimal apparatus* (must specify) *(m) Congenital abnormality of lacrimal drainage system* *(m) Congenital absence of lacrimal drainage structure/lacrimal apparatus* *(m) Congenital anomaly of lacrimal gland* *(m) Accessory lacrimal canal*	743.65
Q10.7	Congenital malformation/anomaly of orbit/ orbit proper (must specify) Congenital superior sulcus anomaly of orbit	743.66
Q11	**Anophthalmos, microphthalmos and macrophthalmos**	
Q11.0	Cystic eyeball Congenital cystic globe	743.03
Q11.1	Other anophthalmos (must specify) Agenesis of eye Anophthalmos, NOS Aplasia of eye Congenital absence of the eye	743.00
Q11.2	Microphthalmos (eye condition) Cryptophthalmos Dysplasia of eye Hypoplasia of eye	743.06 743.10 743.11 743.12

	Rudimentary eye Microphthalmos associated with other anomalies of eye and adnexa **Excludes 1**: cryptophthalmos syndrome (Q87.0)	
Q11.3	Macrophthalmos **Excludes 1**: macrophthalmos in congenital glaucoma (Q15.0)	743.8
Q12	**Congenital lens malformations**	
Q12.0	Congenital cataract (Please specify right, left or bilateral) Anterior and posterior axial embryonal cataract Capsular and subcapsular cataract Cataract, anterior polar Cataract, NOS Cataract, other specified Combined form cataract Cortical and zonular cataract Nuclear cataract Total and subtotal cataract, congenital	743.30 743.31 743.32 743.33 743.34
Q12.1	Congenital displaced lens Congenital ectopic lens Ectopia lentis	743.37
Q12.2	Coloboma of lens	743.36
Q12.3	Congenital aphakia Absence of lens	743.35
Q12.4	Spherophakia	743.36
Q12.8	Other congenital lens malformations (must specify) Microphakia Anomalies of lens shape Mittendorf Dot is not reportable	743.36
Q12.9	Congenital lens malformation, unspecified (must specify) Congenital anomaly of lens shape Unspecified lens anomaly	743.39
Q13	**Congenital malformations of anterior segment of eye**	
Q13.0	Coloboma of iris Coloboma, NOS	743.46
Q13.1	Absence of iris Aniridia	743.45
Q13.2	Other congenital malformations of iris (must specify) *(m) Anisocoria, congenital* *(m) Anomaly of iris and/or ciliary body* *(m) Atresia of pupil* *(m) Congenital heterochromia iridis* *(m) Congenital iris heterochromia* *(m) Congenital pupil anomaly* *(m) Corectopia* *(m) Deformed pupil* *(m) Dyscoria* *(m) Glaucoma assoc with or due to iris anomaly* *(m) Brushfield spots* *(m) Ectopic pupil* *(m) Iridial remnants* *(m) Polycoria*	743.46
Q13.3	Congenital corneal opacity Congenital arcus juvenilis Corneal opacities, interfering with vision, congenital	743.42
Q13.4	Other congenital corneal malformations (size and shape) (must specify) Congenital malformations of cornea, NOS Irido-corneo-trabecular dysgenesis	743.22 743.41

	Microcornea Peter's anomaly *(m) Megalocornea (without glaucoma)* *(m) Congenital megalocornea (without glaucoma)* *(m) Enlarged cornea (without glaucoma)*	
Q13.5	*(m) Blue sclera* *(m) Scleral anomaly* *(m) Specified anomalies of sclera*	743.47
Q13.8	**Other congenital malformations of anterior segment of eye**	
Q13.81	Rieger's anomaly Irido-trabecular dysgenesis Rieger eye malformation sequence	743.44
Q13.89	Other congenital malformations of anterior segment of eye (must specify) Multiple and combined anomaly of anterior segment Other specified colobomas and anomalies of anterior segments *(m) Congenital scleral show*	743.47 743.48 743.49
Q13.9	Congenital malformations of anterior segment of eye, unspecified (must specify) Anterior segment anomaly, eye Congenital anomaly of anterior chamber or segment of eye Glaucoma associated with or due to chamber angle anomaly Multiple anterior segment anomalies Unspecified anomalies of anterior segments or anterior chamber	743.49
Q14	**Congenital malformations of posterior segment of eye** **Excludes 2:** optic nerve hypoplasia (H47.03-)	
Q14.0	Congenital malformation of vitreous humor Congenital vitreous opacity Persistent hyperplastic primary vitreous	743.51
Q14.1	Congenital malformations of retina (must specify) Congenital retinal aneurysm Congenital macular changes Other retinal changes, congenital Specified anomaly of retina	743.55 743.56 743.58
Q14.2	Congenital malformations of optic disc (must specify) Coloboma of optic disc Congenital optic disc coloboma Optic nerve pit, congenital Pit of optic disc Specified anomaly of optic disc	743.57
Q14.3	Congenital malformation of choroid (must specify) Chorioretinal degeneration, congenital Coloboma of choroid folds Congenital choroidal folds	743.53
Q14.8	Other congenital malformations of posterior segment of eye (must specify) Chroioretinal coloboma Coloboma of the fundus Congenital hypoplasia of fovea centralis Fovea hypoplasia Congenital folds and cysts of posterior segment Vascular anomalies	743.52 743.58
Q14.9	Congenital malformation of posterior segment of eye, unspecified (must specify) Unspecified anomaly of posterior segment of eye	743.59
Q15	**Other congenital malformations of eye** **Excludes 1:** 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	743.20 743.21 743.22

	Enlarged eye, NOS Glaucoma of childhood Glaucoma of newborn Hydrophthamos, simple, unspecified Juvenile glaucoma Keratoglobus, congenital, with glaucoma Macrocornea with glaucoma Macrophthalmos in congenital glaucoma Megalocornea with glaucoma Primary congenital glaucoma Simple buphthalmos Buphthalmos associated with other ocular anomalies Congenital glaucoma, simple	743.44
Q15.8	Other specified congenital malformations of eye (must specify) Epibulbar dermoid cyst	743.8
Q15.9	Congenital malformation of eye, unspecified (try not to use) (must specify) Congenital anomaly of eye Congenital deformity of eye	743.9
Q16	**Congenital malformations of ear causing impairment of hearing** **Excludes 1:** congenital deafness (H90.-)	
Q16.0	Congenital absence of (ear) auricle (Anotia) Absence of external ear	744.01
Q16.1	Congenital absence, atresia or stricture of auditory canal (external) (must specify) Congenital atresia or stricture of osseous meatus Congenital atresia of external auditory canal Congenital atresia of external ear (left/right/bilateral) Aural atresia Absence of auditory canal Absence of external auditory meatus *(m) Narrow external auditory meatus*	744.02
Q16.2	Absence of Eustachian tube	744.24
Q16.3	Congenital malformation of ear ossicles (must specify) Congenital fusion of ear ossicles	744.04
Q16.4	Other congenital malformations of middle ear (must specify) Congenital malformations of middle ear, NOS	744.03
Q16.5	Congenital malformation of inner ear (must specify) Congenital anomaly of membranous labyrinth Congenital anomaly of organ of Corti	744.05
Q16.9	Congenital malformation of ear causing impairment of hearing, unspecified (must specify) Congenital absence of ear, NOS Congenital external ear anomaly with hearing impairment	744.00
Q17	**Other congenital malformations of ear Excludes 1:** congenital malformations of ear with impairment of hearing (Q16.0-Q16.9), preauricular sinus (Q18.1)	
Q17.0	*(m) Accessory auricle of ear* *(m) Accessory tragus* *(m) Polyotia* *(m) Preauricular appendage or skin tag* *(m) Supernumerary ear* *(m) Supernumerary lobule* *(m) Ear, double lobule* *(m) Other appendage, tag, or lobule* *(m) Papillomas, ear tags* *(m) Skin tag of pinna*	744.1
Q17.1	*(m) Macrotia (enlarged pinna)*	744.22
Q17.2	Microtia (hypoplastic pinna)	744.23

Q17.3	*(m) Other misshapen ear* *(m) Lop ear* *(m) Pointed ear* *(m) Absent or decreased cartilage* *(m) Cleft ear* *(m) Crumpled ears* *(m) Cupped ears* *(m) Darwin's tubercle* *(m) Ears, absent tragus* *(m) Ears, bridged concha* *(m) Ears, lack of helical fold* *(m) Ears, thickened or over folded helix* *(m) Extra cartilage* *(m) Malformed ear* *(m) Notch in ear* *(m) Posteriorly rotated ears* *(m) Protruding ears*	744.29
Q17.4	*(m) Misplaced ear* *(m) Low-set ears* **Excludes 1**: cervical auricle (Q18.2)	744.29
Q17.5	*(m) Prominent ear* *(m) Bat ear*	744.29
Q17.8	*(m) Other specified congenital malformations of ear* (must specify) *(m) Congenital absence of lobe of ear* *(m) Congenital split ear lobes (left/right/bilateral)* *(m) Cryptotia*	744.21 744.24 743.29
Q17.9	*(m) Congenital malformation of ear, unspecified* (must specify) *(m) Congenital external ear (pinna) anomaly*	744.3
Q18	**Other congenital malformations of face and neck Excludes 1:** 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	*(m) Sinus, fistula and cyst of branchial cleft* *(m) Branchial vestage* *(m) Branchial cleft fistula* *(m) Branchial cleft sinus* *(m) Branchial cleft cyst* *(m) Branchial cleft pit*	744.41
Q18.1	*(m) Preauricular sinus and cyst* *(m) Cervicoaural fistula* *(m) Fistula of auricle, congenital* *(m) Ear pit* *(m) Preauricalar fistula* *(m) Preauricular pits* *(m) Preauricular sinus*	744.46 744.47
Q18.2	*(m) Other branchial cleft malformations* (must specify) *(m) Branchial cleft malformations, NOS* *(m) Cervical auricle* *(m) Otocephaly* *(m) Dermal sinus of head* *(m) Other branchial cleft anomaly*	744.43 744.49
Q18.3	*(m) Webbing of neck* *(m) Pterygium colli* *(m) Redundant neck skin folds*	744.5
Q18.4	Macrostomia	744.83

Q18.5	Microstomia	744.84
Q18.6	*(m) Macrocheilia (large lips)* *(m) Hypertrophy of lip, congenital*	744.81
Q18.7	*(m) Microcheilia*	744.82
Q18.8	Other specified congenital malformations of face and neck (must specify) Medial cyst of face and neck Medial fistula of face and neck Medial sinus of face and neck Cleft face (excluding cleft lip and palate) *(m) Short neck*	744.89
Q18.9	Congenital malformations of face and/or neck, unspecified (try not to use) (must specify) Congenital anomaly NOS of face and/or neck *(m) Abnormal facies* *(m) Craniofacial dysmorphosis* *(m) Dysmorphic facies/face*	744.89 744.9

Congenital Malformations of the Circulatory System (Q20-Q28)

ICD-10	Descriptions	ICD-9
Q20	**Congenital malformations of cardiac chambers and connections** **Excludes 1:** dextrocardia with situs inversus (Q89.3), mirror-image atrial arrangement with situs inversus (Q89.3)	
Q20.0	Common arterial trunk Persistent truncus arteriosus Absent septum between aorta and pulmonary artery Common aortopulmonary trunk **Excludes 1:** aortic septal defect (Q21.4)	745.0
Q20.1	Double outlet right ventricle Taussig-Bing syndrome	745.11
Q20.2	Double outlet left ventricle	745.19
Q20.3	Discordant ventriculoarterial connection Dextraposition of aorta Dextro-transposition of aorta Dextroposition of aorta Dextrotransposition of the great arteries Incomplete great vessel transposition Incomplete transposition of the great vessels L-transposition of the great vessels Levotransposition of the great arteries Transposition of great vessels (complete) D-transposition	745.10
Q20.4	Double inlet ventricle (right or left) Common ventricle Cor triloculare biatriatum Single ventricle	745.3
Q20.5	Discordant atrioventricular connection Corrected transposition Levo-transposition Ventricular inversion L-transposition, ventricular inversion Other specific transposition of great vessels Unspecified transposition of great vessels	745.12
Q20.6	Isomerism of atrial appendages Isomerism of atrial appendages with asplenia or polysplenia	745.8 759.0
Q20.8	Other congenital malformations of cardiac chambers and connections (must specify) Cor binoculare Absence of atrial and ventricular septa Two chambered heart	745.19 745.7 745.8
Q20.9	Congenital malformation of cardiac chambers and connections, unspecified (must specify)	746.9
Q21	**Congenital malformations of cardiac septa** **Excludes 1:** acquired cardiac septal defect (I51.0)	
Q21.0	Ventricular septal defect (VSD) (must specify) Roger's disease VSD, Malalignment (type I, subarterial, subaortic, subpulmonic; outlet, supracristal) VSD, Perimembranous (type II, membranous, infracristal, subaortic, Gerbode) VSD, Endocardial cushion defect (type III; inlet, inflow type, subtricuspid, canal-type) VSD, Muscular (type IV; trabecular, swiss cheese, central, apical, marginal, Roger's disease) Interventricular septal defect	745.4
Q21.1	**Atrial septal defect** (ASD) Excludes 2: ostium primum atrial septal defect (type I) (Q21.20)	

Q21.10	Atrial septal defect, unspecified (must specify)	745.5
Q21.11	Secundum atrial septal defect Fenestrated atrial septum Patent or persistent ostium secundum defect (type II)	745.5
Q21.12	*(m) Patent foramen ovale* *(m) Persistent foramen ovale*	745.5
Q21.13	Coronary sinus atrial septal defect Coronary sinus defect Unroofed coronary sinus	745.5
Q21.14	Superior sinus venosus atrial septal defect Superior vena cava type atrial septal defect	745.5
Q21.15	Inferior sinus venosus atrial septal defect Inferior vena cava type atrial septal defect	745.5
Q21.16	Sinus venosus atrial septal defect, unspecified Sinus venosus defect, NOS	745.5
Q21.19	Other specified atrial septal defect (must specify) Common atrium	745.5
Q21.2	**Atrioventricular septal defect** Atrioventricular canal defect	
Q21.20	Atrioventricular septal defect (AVSD), unspecified as to partial or complete Atrioventricular canal, NOS Endocardial cushion defect, NOS Ostium primum atrial septal defect (type I) NOS	745.60
Q21.21	Partial atrioventricular septal defect Incomplete atrioventricular canal Incomplete atrioventricular septal defect Incomplete endocardial cushion defect Ostium primum atrial septal defect (type I) with separate atrioventricular valves Partial atrioventricular canal Partial endocardial cushion defect	
Q21.22	Transitional atrioventricular septal defect Intermediate atrioventricular canal Intermediate atrioventricular septal defect Intermediate endocardial cushion defect Ostium primum atrial septal defect (type I) with separate atrioventricular valves and a small or restrictive inlet VSD Transitional atrioventricular canal Transitional endocardial cushion defect	
Q21.23	Complete atrioventricular septal defect Common atrioventricular canal Common atrioventricular septal defect Common endocardial cushion defect Ostium primum atrial septal defect (type I) with common atrioventricular valve and a moderate or larger inlet VSD	
Q21.3	Tetralogy of Fallot (ventricular septal defect with pulmonary stenosis or atresia, dextroposition of aorta and hypertrophy of right ventricle) (also specify all individual defects) Dextraposition of aorta in Fallot's tetralogy Tetralogy of Fallot with absent pulmonary valve Tetralogy of Fallot with pulmonary atresia or stenosis Tetralogy of Fallot with ventricular septal defect	745.2
Q21.4	Aortopulmonary septal defect Abnormal communication between aorta and pulmonary artery Aortic septal defect Aortopulmonary window	745.0
Q21.8	Other congenital malformations of cardiac septa (must specify)	745.8

	Eisenmenger's defect Pentalogy of Fallot Fallot's tetralogy plus atrial septal defect	745.2
Q21.9	Congenital malformations of cardiac septum, unspecified (must specify) Septal (heart) defect, NOS	745.9
Q22	**Congenital malformations of pulmonary and tricuspid valves**	
Q22.0	Pulmonary valve atresia Congenital absence of pulmonary valve	746.01
Q22.1	Congenital pulmonary valve stenosis (narrowing)	746.02
Q22.2	Congenital pulmonary valve insufficiency (do not code if mild, trivial or physiologic) Congenital pulmonary valve regurgitation (do not code if mild, trivial or physiologic) Trilogy of Fallot	746.09
Q22.3	Other congenital malformations of pulmonary valve (must specify) Congenital malformation of pulmonary valve, NOS Supernumerary cusps of pulmonary valve Bicuspid pulmonary valve Other specified anomaly of pulmonary valve Pulmonary valve hypoplasia Quadricuspid pulmonary valve	746.00 746.09
Q22.4	Congenital tricuspid valve stenosis (narrowing) Congenital tricuspid atresia Absence of tricuspid valve Tricuspid hypoplasia	746.1
Q22.5	Ebstein's anomaly Ebstein's anomaly of tricuspid valve	746.2
Q22.6	Hypoplastic right heart syndrome	756.89
Q22.8	Other congenital malformations of tricuspid valve (must specify) Insufficiency of tricuspid valve Congenital tricuspid valve regurgitation	746.89
Q22.9	Congenital malformations of tricuspid valve, unspecified (must specify)	746.89
Q23	**Congenital malformations of aortic and mitral valves**	
Q23.0	Congenital stenosis of aortic valve Congenital aortic atresia Congenital aortic stenosis, NOS **Excludes 1:** Congenital stenosis of aortic valve in hypoplastic left heart syndrome (Q23.4), congenital subaortic stenosis (Q24.4), supravalvular aortic stenosis (congenital) (Q25.3v)	746.3
Q23.1	Congenital insufficiency of aortic valve (do not code if mild, trivial or physiologic) Bicuspid aortic valve Congenital aortic insufficiency Congenital aortic valve regurgitation	746.4
Q23.2	Congenital mitral stenosis (narrowing) Congenital mitral atresia Congenital supravalvular mitral stenosis Absence of mitral valve Atresia of mitral valve Hypoplasia of mitral valve/cleft mitral valve	746.5
Q23.3	Congenital mitral insufficiency (do not code if mild, trivial or physiologic) Congenital mitral valve regurgitation Mitral valve prolapse	746.6
Q23.4	Hypoplastic left heart syndrome	746.7
Q23.8	Other congenital malformations of aortic and mitral valves (must specify) Cleft leaflet of mitral valve Aortic valve atresia/absence Fused commissure of mitral valve Other specified anomaly of aortic valves	746.89

	Parachute deformity of mitral valve Supernumerary cusps	
Q23.9	Congenital malformation of aortic and mitral valves, unspecified (must specify) Unspecified anomaly of the aortic valves	746.89
Q24	**Other congenital malformations of heart Excludes 1:** endocardial fibroelastosis (I42.4)	
Q24.0	Dextrocardia Dextrocardia with no mention of situs inversus Dextrocardia without situs inversus (situs solitus) **Excludes 1:** 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)	746.87
Q24.1	Levocardia Situs inversus with levocardia	746.87
Q24.2	Cor triatriatum Cor triatriatum, dexter/left sided/right sided/sinister Divided left atrium Divided right atrium	746.82
Q24.3	Pulmonary infundibular stenosis (narrowing) Infundibular pulmonic stenosis Stenosis of infundibulum of right ventricle Subvalvular pulmonic stenosis	746.83
Q24.4	Congenital subaortic stenosis Congenital subvalvular aortic stenosis	746.81
Q24.5	Malformation of coronary vessels (must specify) Congenital coronary (artery) aneurysm Congenital coronary artery fistula Absence of coronary artery Anomalies of coronary artery or sinus Anomalous origin or communication of coronary artery Coronary artery arising from aorta or pulmonary trunk Single coronary artery	746.85
Q24.6	Congenital heart block Complete or incomplete atrioventricular (AV) block	746.86
Q24.8	Other specified congenital malformations of heart (must specify) Congenital diverticulum of left ventricle Congenital malformation of myocardium Congenital malformation of pericardium Malposition of heart Uhl's disease (hypoplastic right ventricle) Abdominal heart Absence of papillary muscle of luthisis Anomalies of myocardium Anomalies of pericardium Anomaly of papillary muscle Atresia of cardiac vein Ectopia (ectopic) cordis (mesocordia) Hypoplasia of cardiac vein Hypoplastic left ventricle Hypoplastic ventricle, NOS Shones syndrome (Complex) = 1) Parachute Mitral Valve; 2) supravalvular ring of L atrium; 3) subaortic stenosis; 4) coarctation of aorta [Code the stenosis and coarctation separately] Ventricular hypertrophy (right or left) *(m) Congenital cardiomegaly* *Cardiomyopathy, hypertrophic is not reportable*	746.84 746.87 746.89
Q24.9	Congenital malformations of heart, unspecified (try not to use) (must specify)	746.9

	Complex congenital heart defect (code component defects) Congenital anomaly of heart Congenital disease of heart	
Q25	**Congenital malformations of great arteries**	
Q25.0	Patent ductus arteriosus (PDA) (do not report if birth weight <1500 grams) Patent ductus Botallo Persistent ductus arteriosus Patent ductus arteriosis closed with medication	747.0
Q25.1	Coarctation of aorta Coarctation of aorta (preductal) (postductal) Unspecified coarctation of aorta	747.10
Q25.2	Atresia of aorta	
Q25.21	Interruption of aortic arch Atresia of aortic arch Interrupted aortic arch Interrupted aortic arch. Type A Interrupted aortic arch. Type B	747.11
Q25.29	Other atresia of aorta Atresia of aorta	747.22
Q25.3	Supravalvular aortic stenosis (narrowing) Small aorta, narrowing of aorta Stenosis of aorta Stricture of aorta **Excludes 1:** congenital aortic stenosis NOS (Q23.0); congenital stenosis of aortic valve (Q23.0)	747.22
Q25.4	Other congenital malformations of aorta (must specify) Kommerells diverticulum Overriding aorta Persistent right aortic arch Pseudocoarctation of aorta Aberrant subclavian artery Hypoplastic aortic isthmus Vascular ring compression of trachea **Excludes 1:** hypoplasia of aorta in hypoplastic left heart syndrome (Q23.4)	
Q25.40	*(m) Congenital malformation of aorta unspecified* (must specify)	747.20
Q25.41	Absence and aplasia of aorta	747.29
Q25.42	Hypoplasia of aorta	747.29
Q25.43	Congenital aneurysm of aorta Congenital aneurysm of aortic root Congenital aneurysm of aortic sinus	747.29
Q25.44	*(m) Congenital dilation of aorta*	747.29
Q25.45	Double aortic arch Vascular ring of aorta	747.21
Q25.46	*(m) Tortuous aortic arch* *(m) Persistent convolutions of aortic arch*	747.21
Q25.47	*(m) Right aortic arch* *(m) Persistent right aortic arch*	747.21
Q25.48	*(m) Anomalous origin of subclavian artery*	747.29
Q25.49	*(m) Other congenital malformations of aorta* (must specify) *(m) Aortic arch* *(m) Bovine arch*	747.29
Q25.5	Atresia of pulmonary artery Pulmonary artery absence Pulmonary artery agenesis Pulmonary artery atresia	747.31

Q25.6	Stenosis of pulmonary artery Supravalvular pulmonary stenosis (narrowing) *Peripheral pulmonary artery stenosis (do not report if birth weight <2500 g)* *Peripheral pulmonic stenosis (PPS) (do not report if birth weight <2500 g)*	747.31 747.39
Q25.7	**Other congenital malformations of pulmonary artery**	
Q25.71	Coarctation of pulmonary artery	747.31
Q25.72	Congenital pulmonary arteriovenous malformation Congenital pulmonary arteriovenous aneurysm Aneurysm of pulmonary artery	747.32
Q25.79	Other congenital malformations of pulmonary artery (must specify) Aberrant pulmonary artery Agenesis of pulmonary artery Congenital aneurysm of pulmonary artery Congenital anomaly of pulmonary artery Hypoplasia of pulmonary artery Dilatation of pulmonary artery Unspecified anomaly of pulmonary artery	747.39
Q25.8	Other congenital malformations of other great arteries (must specify) Anomalous origin of right subclavian artery	747.29
Q25.9	Congenital malformations of great arteries, unspecified (must specify)	747.29
Q26	**Congenital malformations of great veins**	
Q26.0	Congenital stenosis of vena cava Congenital stenosis of vena cava (inferior or superior)	747.49
Q26.1	Persistent left superior vena cava	747.49
Q26.2	Total anomalous pulmonary venous connection Total anomalous pulmonary venous return [TAPVR], subdiaphragmatic Total anomalous pulmonary venous return [TAPVR], supradiaphragmatic Total anomalous lung veins to coronary sinus/liver veins/right heart/vena cavae	747.41
Q26.3	Partial anomalous pulmonary venous connection Partial anomalous pulmonary venous return [PAPVR]	747.42
Q26.4	Anomalous pulmonary venous connection, unspecified (must specify)	747.42
Q26.5	Anomalous portal venous connection	747.61
Q26.6	Portal vein-hepatic artery fistula	747.61
Q26.8	Other congenital malformations of great veins (must specify) Absence of vena cava (inferior or superior) Atresia of vena cava (inferior or superior) Azygos continuation of inferior vena cava Persistent left posterior cardinal vein Pulmonary vein stenosis Scimitar syndrome Anomalous portal vein termination Stenosis of vena cava (inferior or superior) Transposition of pulmonary veins, NOS	747.49
Q26.9	Congenital malformation of great vein, unspecified (must specify) Congenital anomaly of vena cava (inferior) (superior), NOS Anomaly NOS of pulmonary veins	747.40
Q27	**Other congenital malformations of peripheral vascular system** **Excludes 2:** 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	*(m) Congenital absence and hypoplasia of umbilical artery* *(m)* Single umbilical artery *(m) Two vessels in umbilical cord*	747.5
Q27.1	Congenital renal artery stenosis	747.62
Q27.2	Other congenital malformations of renal artery (must specify) Congenital malformation of renal artery, NOS	747.62

	Multiple renal arteries Renal vessel anomaly	
Q27.3	**Arteriovenous malformation (AVM) (peripheral)** Arteriovenous aneurysm	
Q27.30	Arteriovenous malformation, site unspecified (must specify) Peripheral arteriovenous malformation	747.69
Q27.31	Arteriovenous malformation of vessel of upper limb	747.63
Q27.32	Arteriovenous malformation of vessel of lower limb	747.64
Q27.33	Arteriovenous malformation of digestive system vessel Arteriovenous malformation of duodenum and jejunum Arteriovenous malformation of intestinal vessels Congenital arteriovenous malformation of the gastrointestinal tract Congenital duodenal and jejunal arteriovenous malformation	747.61
Q27.34	Arteriovenous malformation of renal vessel	747.62
Q27.39	Arteriovenous malformation, other site Arteriovenous malformation of spine	747.69
Q27.4	Congenital phlebectasia	747.89
Q27.8	Other specified congenital malformations of peripheral vascular system (must specify) Absence of peripheral vascular system Atresia of peripheral vascular system Congenital aneurysm (peripheral) Congenital stricture, artery Congenital varix Absence of artery, NEC Anomaly of spinal vessel Atresia of artery Gastrointestinal vessel anomaly Other anomaly of peripheral arteries Other anomaly of peripheral veins **Excludes 1:** arteriovenous malformation (Q27.3-)	747.69
Q27.9	Congenital malformation of peripheral vascular system, unspecified (must specify) Anomaly of artery or vein, NOS Arteriovenous malformation Congenital anomaly of peripheral blood vessel Congenital vascular malformation of orbit Vascular or venous malformation	747.60
Q28	**Other congenital malformations of circulatory system** **Excludes 1:** congenital aneurysm NOS (Q27.8), congenital coronary aneurysm (Q24.5), ruptured cerebral arteriovenous malformation (I60.8), ruptured malformation of precerebral vessels (I72.0) **Excludes 2:** 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 (non-ruptured)	747.89
Q28.1	Congenital malformation of precerebral vessels, NOS (must specify) Congenital precerebral aneurysm (nonruptured)	747.89
Q28.2	Arteriovenous malformation of cerebral vessels (must specify) Arteriovenous malformation of brain, NOS Congenital arteriovenous cerebral aneurysm (nonruptured) Cerebral arteriovenous malformation Anomalies of cerebrovascular system Anomalies of vein of Galen Other anomaly of cerebral vessels	747.81
Q28.3	Other malformations of cerebral vessels (must specify) Congenital cerebral aneurysm (nonruptured)	747.81

	Congenital malformation of cerebral vessels, NOS Developmental venous anomaly	
Q28.8	Other specified congenital malformations of circulatory system (must specify) Congenital aneurysm, specified site NEC Spinal vessel anomaly	747.89 747.82
Q28.9	Congenital malformation of circulatory system, unspecified (must specify)	747.9

Congenital Malformations of the Respiratory System (Q30-Q34)

ICD-10	Description	ICD-9
Q30	**Congenital malformations of nose Excludes 1:** congenital deviation of nasal septum (Q67.4)	
Q30.0	Choanal atresia Atresia of nares (anterior) (posterior) Congenital stenosis of nares (anterior) (posterior) Choanal stenosis	748.0
Q30.1	Agenesis and underdevelopment of nose Agenesis of nose Congenital absent of nose	748.1
Q30.2	Fissured, notched and cleft nose Cleft nose anomaly Congenital cleft nose Congenital cleft nose anomaly Midline fissured, notched and cleft nose	748.1
Q30.3	*(m) Congenital perforated nasal septum*	748.1
Q30.8	*(m) Other congenital malformations of nose (must specify)* *(m) Accessory nose* *(m) Congenital anomaly of nasal sinus wall* *(m) Congenital malformation of nasal septum (nasal septal defect)* *(m) Absent nasal septum* *(m) Anteverted nares* *(m) Flat or wide nasal bridge or other minor nose malformation* *(m) Notched or hypoplastic alae nasi* *(m) Other anomalies of nose* *(m) Small nares* *(m) Small nose and nostril* *(m) Tubular nose, single nostril, proboscis* *(m) Upturned nose* *(m) Wide nasal bridge*	748.1
Q30.9	*(m) Congenital malformation of nose, unspecified*	748.1
Q31	**Congenital malformation of larynx Excludes 1:** congenital laryngeal stridor, NOS (P28.89)	
Q31.0	Web of larynx Glottic web of larynx Laryngeal web Subglottic web Subglottic web of larynx	748.2
Q31.1	Congenital subglottic stenosis	748.3
Q31.2	Laryngeal hypoplasia	748.3
Q31.3	*(m) Laryngocele*	748.3
Q31.5	*(m) Congenital laryngomalacia*	748.3
Q31.8	Other congenital malformations of larynx (must specify) Absence of larynx Agenesis of larynx Atresia of larynx Congenital cleft larynx Congenital cleft thyroid cartilage Congenital fissure of epiglottis Congenital stenosis of larynx, NEC Laryngeal cleft Posterior cleft of cricoid cartilage Anomalies of larynx and supporting cartilage Anomaly of epiglottis Anomaly of thyroid cartilage Atresia of epiglottis	748.3

	Atresia glottis Cleft larynx, laryngotracheoesophageal cleft Cleft thyroid, cartilage, congenital	
Q31.9	Congenital malformation of larynx, unspecified (must specify) Congenital laryngeal anomaly	748.3
Q32	**Congenital malformations of trachea and bronchus** **Excludes 1:** congenital bronchiectasis (Q33.4)	
Q32.0	*(m) Congenital tracheomalacia*	748.3
Q32.1	Other congenital malformations of trachea (must specify) Atresia of trachea Congenital anomaly of tracheal cartilage Congenital dilation of trachea Congenital malformation of trachea Congenital stenosis of trachea Congenital tracheocele Congenital laryngotracheal anomaly Diverticulum of trachea Rudimentary tracheal bronchus	748.3
Q32.2	Congenital bronchomalacia	748.3
Q32.3	Congenital stenosis of bronchus	748.3
Q32.4	Other congenital malformations of bronchus (must specify) Absence of bronchus Agenesis of bronchus Atresia of bronchus Congenital diverticulum of bronchus Congenital malformations of bronchus, NOS	748.3
Q33	**Congenital malformations of lung**	
Q33.0	Congenital cystic lung Bronchogenic cyst Congenital cystic adenomatoid malformation of lung Congenital cystic lung disease Congenital honeycomb lung Congenital polycystic lung disease Single lung cyst CCAM (congenital cystic adenomatoid malformation) Cystic adenomatoid dysplasia of lung Cystic adenomatoid lung Lobar adenomatosis Multiple cysts, lung Polycystic lung (do not use this code for broncopulmonary dysplasia) **Excludes 1:** Cystic Fibrosis (E84.0), cystic lung disease, acquired or unspecified (J98.4)	748.4
Q33.1	Accessory lobe of lung Accessory lung Azygos lobe (fissured), lung	748.69
Q33.2	Sequestration of lung Congenital sequestration of lung Pulmonary sequestration	748.5
Q33.3	Agenesis of lung Congenital absence of lung (lobe) Congenital absence of lung	748.5
Q33.4	Congenital bronchiectasis	748.61
Q33.5	Ectopic tissue in lung	748.69
Q33.6	Congenital hypoplasia or dysplasia of lung Congenital pulmonary dysplasia Hypoplasia of lung	748.5

	Pulmonary hypoplasia Hypoplasia of lobe of lung **Excludes 1:** pulmonary hypoplasia associated with short gestation (P28.0)	
Q33.8	Other congenital malformations of lung (must specify) Aplasia of lung Bilobar right lung Congenital lobar emphysema Trilobal left lung	748.69
Q33.9	Congenital malformations of lung, unspecified Congenital anomaly of lung	748.60
Q34	**Other congenital malformations of respiratory system** **Excludes 2:** congenital central alveolar hypoventilation syndrome (G47.35)	
Q34.0	Anomaly of pleura Congenital anomaly of pleural folds Abnormal communication between pericardial/pleural sacs	748.8
Q34.1	Congenital cyst of mediastinum	748.8
Q34.8	Other specified congenital malformation of respiratory system (must specify) Atresia of nasopharynx Immotile cilia syndrome Primary ciliary dyskinesia	748.8
Q34.9	Congenital malformation of respiratory system, unspecified (try not to use this code) (must specify) Congenital absence of respiratory system Congenital anomaly of respiratory system, NOS	748.9

Congenital Malformations of the Cleft Lip and Cleft Palate (Q35-Q37)

Excludes 2: Robin's Syndrome (Q87.0)

ICD-10	Description	ICD-9
Q35	**Cleft palate Excludes 1:** cleft palate with cleft lip (Q37.-)	
Q35.1	Cleft hard palate Cleft hard palate, central Cleft hard palate, unilateral Cleft hard palate, unilateral, left Cleft hard palate, unilateral, right Cleft hard palate, bilateral	749.00
Q35.3	Cleft soft palate Cleft palate, submucous Submucous cleft palate Cleft soft palate alone, central Cleft soft palate alone, unilateral Cleft soft palate alone, unilateral, left Cleft soft palate alone, unilateral, right Cleft soft palate alone, bilateral	749.00
Q35.5	Cleft hard palate with cleft soft palate Complete cleft palate, central Complete cleft palate, unilateral Complete cleft palate, unilateral, left Complete cleft palate, unilateral, right Complete cleft palate, bilateral Uranostaphyloschisis	749.01 749.03
Q35.7	*(m) Cleft uvula* *(m) Bifid uvula*	749.02
Q35.9	Cleft palate, unspecified (try not to use this code) Cleft palate, NOS Cleft secondary palate Palatoschisis	749.00 749.02 749.04
Q36	**Cleft lip** **Includes:** cheiloschisis, congenital fissure of lip, harelip, labium leporinum **Excludes 1:** cleft lip with cleft palate (Q37.-)	
Q36.0	Cleft lip, bilateral Complete cleft lip, bilateral Incomplete cleft lip, bilateral	749.13 749.14
Q36.1	Cleft lip, median Central cleft lip	749.10
Q36.9	Cleft lip, unilateral Cleft lip, NOS Cleft gingiva Cleft gum Cleft lip, unilateral, left Cleft lip, unilateral, right Complete cleft lip, unilateral Complete cleft lip, unilateral, left Complete cleft lip, unilateral, right Incomplete cleft lip, unilateral Incomplete cleft lip, unilateral, left Incomplete cleft lip, unilateral, right Incomplete form of cleft lip (microform)	749.10 749.11 749.12
Q37	**Cleft palate with cleft lip** **Includes:** Cheilopalatoschisis	
Q37.0	Cleft hard palate with bilateral cleft lip	749.24

		749.25
Q37.1	Cleft hard palate with unilateral cleft lip Cleft hard palate with cleft lip, NOS	749.22 749.25
Q37.2	Cleft soft palate with bilateral cleft lip	749.24 749.25
Q37.3	Cleft soft palate with unilateral cleft lip Cleft soft palate with cleft lip, NOS	749.22 749.25
Q37.4	Cleft hard and soft palate with bilateral cleft lip Complete cleft palate and bilateral cleft lip	749.23 749.24
Q37.5	Cleft hard and soft palate with unilateral cleft lip Cleft hard and soft palate with cleft lip, NOS Complete cleft palate and cleft lip, unilateral Complete cleft palate and cleft lip, unilateral, left Complete cleft palate and cleft lip, unilateral, right Complete cleft palate and cleft lip, NOS	 749.21 749.22
Q37.8	Unspecified cleft palate with bilateral cleft lip Incomplete cleft palate and bilateral cleft lip Bilateral cleft lip, with any cleft palate	 749.24
Q37.9	Unspecified cleft palate with unilateral cleft lip Cleft palate with cleft lip, NOS Other combinations cleft palate and lip	 749.20

Congenital Malformations of the Digestive System (Q38-Q45)

ICD-10	Descriptions	ICD-9
Q38	**Other congenital malformations of tongue, mouth and pharynx** **Excludes 1:** dentofacial anomalies (M26.-), macrostomia (Q18.4), microstomia (Q18.5)	
Q38.0	Congenital malformations of lips, NEC (must specify) Van der Woude syndrome *(m) Congenital fistula of lip or pits* *(m) Congenital palsy of the lower lip* *(m) Long philtrum (only code if with Q86.-)* *(m) Lip tie* *(m) Notched lip (only code if with Q86.-)* *(m) Prominent philtrum (only code if with Q86.-)* **Excludes 1:** cleft lip (Q36.-), cleft lip with cleft palate (Q37.-), macrocheilia (Q18.6), microcheilia (Q18.7)	750.25
Q38.1	*(m) Ankyloglossia* *(m) Tongue tie* *(m) Short frenulum of tongue* *(m) Aberrant frenula*	750.0
Q38.2	*(m) Macroglossia (large tongue)* *(m) Congenital hypertrophy of tongue*	750.15
Q38.3	Other congenital malformations of tongue (must specify) Aglossia (absence of tongue) *(m) Bifid tongue* *(m) Congenital adhesion of tongue* *(m) Congenital fissure of tongue* *(m) Double tongue* *(m) Hypoglossia (small tongue)* *(m) Hypoplasia of tongue* *(m) Microglossia* *(m) Cleft tongue or split tongue* *(m) Glossoptosis*	750.10 750.11 750.12 750.13 750.16 750.19
Q38.4	Congenital malformations of salivary glands and ducts (must specify) Atresia of salivary glands and ducts Congenital absence of salivary glands and ducts Congenital accessory salivary glands and ducts Congenital fistula of salivary gland Congenital ranula	750.21 750.22 750.23 750.24
Q38.5	Congenital malformations of palate, NEC (must specify) *(m) Congenital absence of uvula* *(m) Congenital high arched palate* **Excludes 1:** cleft palate (Q35.-), cleft palate with cleft lip (Q37.-)	750.26
Q38.6	*(m) Other congenital malformations of mouth (must specify)* *(m) Congenital malformations of mouth, NOS* *(m) Fordyce spots of mouth* *(m) Fordyce's disease* *(m) Low lying maxillary frenulum*	750.26
Q38.7	Congenital pharyngeal pouch Congenital diverticulum of pharynx **Excludes 1:** pharyngeal pouch syndrome (D82.1)	750.27
Q38.8	Other congenital malformations of pharynx (must specify) Congenital malformation of pharynx, NOS Congenital anomaly of pharynx Congenital velopharyngeal dysfunction Congenital velopharyngeal impairment Congenital velopharyngeal incompetence	750.29

	Imperforate pharynx Do not report tonsil asymmetry	
Q39	**Congenital malformations of esophagus**	
Q39.0	Atresia of esophagus without fistula Atresia of esophagus, NOS Esophageal atresia Imperforate esophagus	750.3
Q39.1	Atresia of esophagus with tracheoesophageal fistula Atresia of esophagus with broncho-esophageal fistula Esophageal atresia with tracheoesophageal fistula	750.3
Q39.2	Congenital trachea-esophageal fistula without atresia Congenital trachea-esophageal fistula, NOS Congenital trachea esophageal fistula without atresia of esophagus Esophagotracheal fistula, congenital T E fistula without esophageal atresia Tracheoesophageal fistula “h” type	750.3
Q39.3	Congenital stenosis and stricture of esophagus Congenital esophageal ring Congenital Schatzkis ring Congenital stenosis of esophagus	750.3
Q39.4	Esophageal web	750.3
Q39.5	Congenital dilatation of esophagus Congenital cardiospasm Congenital achalasia Giant esophagus	750.4
Q39.6	Congenital diverticulum of esophagus Congenital esophageal pouch	750.4
Q39.8	Other congenital malformations of esophagus (must specify) Congenital absence of esophagus Congenital displacement of esophagus Congenital duplication of esophagus Esophageal duplication Other tracheoesophageal anomaly	750.4
Q39.9	Congenital malformation of esophagus, unspecified (must specify)	750.4
Q40	**Other congenital malformations of upper alimentary tract**	
Q40.0	Congenital hypertrophic pyloric stenosis Congenital or infantile constriction of pylorus Congenital or infantile hypertrophy of pylorus Congenital of infantile stenosis of pylorus Congenital or infantile stricture of pylorus Other congenital pyloric obstruction Do not report epileptic/infantile spasms	750.5
Q40.1	Congenital hiatus hernia Congenital displacement of cardia through esophageal hiatus Congenital hiatal hernia **Excludes 1**: congenital diaphragmatic hernia (Q79.0)	750.6
Q40.2	Other specified congenital malformations of stomach (must specify) Congenital displacement of stomach Congenital diverticulum of stomach Congenital duplication of stomach Congenital hourglass stomach Megalogastria Microgastria Displacement or transposition of stomach	750.7
Q40.3	Congenital malformation of stomach, unspecified (must specify) Congenital gastric anomaly	750.7

Q40.8	Other specified congenital malformations of upper alimentary tract (must specify)	750.8
Q40.9	Congenital malformation of upper alimentary tract, unspecified (must specify) Congenital anomaly of upper alimentary tractCongenital upper GI tract anomaly	750.9
Q41	**Congenital absence, atresia or stenosis of small intestine** **Includes:** congenital obstruction, occlusion or stricture of small intestine or intestine, NOS **Excludes 1:** cystic fibrosis with intestinal manifestation (E84.11), meconium ileus NOS (without cystic fibrosis) (P76.0)	
Q41.0	Congenital absence, atresia or stenosis of duodenum (must specify) Absence of duodenum/ duodenal absence Atresia of duodenum/ duodenal atresia Stenosis of duodenum/ duodenal stenosis	751.1
Q41.1	Congenital absence, atresia or stenosis of jejunum (must specify) Apple peel syndrome Imperforated jejunum Absence of jejunum Atresia of jejunum Stenosis of jejunum	751.1
Q41.2	Congenital absence, atresia or stenosis of ileum (must specify) Absence of ileum/ ileal absence Atresia of ileum/ ileal atresia Stenosis of ileum/ ileal stenosis	751.1
Q41.8	Congenital absence, atresia or stenosis of other specified parts of small intestine (must specify) Absence of other specified parts of small intestine (specify) Atresia of other specified parts of small intestine (specify) Stenosis of other specified parts of small intestine (specify)	751.1
Q41.9	Congenital absence, atresia and stenosis of small intestine, part unspecified Absence of small intestine Atresia of small intestine Stenosis of small intestine	751.1
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 (must specify) Absence of rectum with fistula Atresia of rectum with fistula Stenosis of rectum with fistula	751.2
Q42.1	Congenital absence, atresia and stenosis of rectum without fistula (must specify) Imperforated rectum Absence of rectum without fistula/rectal absence Atresia of rectum without fistula/ rectal atresia Stenosis of rectum without fistula/ rectal stenosis	751.2
Q42.2	Congenital absence, atresia and stenosis of anus with fistula (must specify) Absence of anus with fistula Atresia of anus with fistula Stenosis of anus with fistula Congenital or infantile occlusion of anus with fistula Congenital infantile stricture of anus with fistula Imperforated anus with fistula	751.2
Q42.3	Congenital absence, atresia and stenosis of anus without fistula (must specify) Imperforated anus Absence of anus Atresia of anus Congenital or infantile occlusion of anus without fistula Congenital infantile stricture of anus without fistula Stenosis of anus	751.2

Q42.8	Congenital absence, atresia and stenosis of other parts of large intestine (must specify) Absence of appendix Atresia of appendix Stenosis of appendix	751.2
Q42.9	Congenital absence, atresia, or stenosis of large intestine, part unspecified (try not to code) Absence of large intestine Atresia of colon Stenosis of colon	751.2
Q43	**Other congenital malformations of intestine**	
Q43.0	*(m) Meckel's diverticulum (displaced) (hypertrophic)* *(m) Persistent vitelline duct* *(m) Persistent omphalomesenteric duct*	751.0
Q43.1	Hirschsprung's disease Aganglionosis Congenital (aganglionic) megacolon Megacolon, congenital Long segment Hirschsprung's disease Short segment Hirschsprung's-Recto-Sigmoid	751.3
Q43.2	Other congenital functional disorders of colon (must specify) Congenital dilation of colon Congenital megacolon, not aganglionic	751.3
Q43.3	Congenital malformations of intestinal fixation (must specify) Congenital omentum, anomalous adhesions [bands] Congenital peritoneal adhesions [bands] Incomplete rotation of cecum and colon Insufficient rotation of cecum and colon Intestinal malrotation Jackson's membrane Malrotation of colon Malrotation of intestine Rotation failure of cecum and colon Universal mesentery Anomalies of mesentery Ladd's bands Malrotation of cecum Malrotation of small intestine alone Other specified malrotation Unspecified malrotation	751.4
Q43.4	Duplication of intestine Duplication of anus Duplication of appendix Duplication of cecum Duplication of cyst ileum Duplication of pylorus	751.5
Q43.5	Ectopic anus Anterior anus	751.5
Q43.6	Congenital fistula of rectum and anus **Excludes 1:** 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.-)	751.5
Q43.7	Persistent cloaca Cloaca, NOS	751.5

Q43.8	Other specified congenital malformations of intestine (must specify) Congenital anal fissure Congenital blind loop syndrome Congenital diverticulitis, colon Congenital diverticulum, intestine Congenital rectal fissure Dolichocolon Megaloappendix Megaloduodenum Microcolon Transposition of appendix Transposition of colon Transposition of intestine Enterogenous cyst Duodenal web Volvus	751.5
Q43.9	Congenital malformation of intestine, unspecified (must specify)	751.5
Q44	**Congenital malformations of gallbladder, bile ducts and liver**	
Q44.0	Agenesis, aplasia or hypoplasia of gallbladder Congenital absence of gallbladder Gallbladder hypoplasia Gallbladder aplasia Gallbladder agenesis	751.69
Q44.1	Other congenital malformations of gallbladder (must specify) Intrahepatic gallbladder Duplication gallbladder Floating gallbladder	751.69 751.60
Q44.2	Atresia of bile ducts Biliary atresia Carlois disease with obstruction Congenital dilatation of lobar intrahepatic bile duct with obstruction	751.61
Q44.3	Congenital stenosis or stricture of bile ducts Congenital biliary obstruction Congenital hypoplasia of bile duct (common) or passage	751.61
Q44.4	Choledochal cyst	751.69
Q44.5	Other congenital malformations of bile ducts (must specify) Accessory hepatic duct Biliary duct duplication Carlois disease without obstruction Congenital dilated bile duct Congenital dilatation of lobar intrahepatic bile duct Congenital malformation of bile duct, NOS Cystic duct duplication Agenesis or atresia of hepatic ducts Anomalies of biliary tract, NEC Other anomaly of hepatic or bile ducts	751.69 751.60
Q44.6	Cystic disease of liver Fibrocystic disease of liver	751.62
Q44.7	Other congenital malformation of liver (must specify)	
Q44.70	Other congenital malformations of liver, unspecified (must specify) Congenital malformation of liver, NOS	751.60
Q44.71	Alagille syndrome Alagille-Watson syndrome	751.69
Q44.79	Other congenital malformations of liver (must specify) Accessory liver Congenital absence of liver	751.69

	(m) Congenital hepatomegaly	
Q45	**Other congenital malformations of digestive system** **Excludes 2:** congenital diaphragmatic hernia (Q79.0), congenital hiatus hernia (Q40.1)	
Q45.0	Agenesis, aplasia or hypoplasia of pancreas Congenital absence of pancreas Pancreatic agenesis Pancreatic aplasia Pancreatic hypoplasia	751.7
Q45.1	Annular pancreas	751.7
Q45.2	Congenital pancreatic cyst	751.7
Q45.3	Other congenital malformations of pancreas or pancreatic duct (must specify) Accessory pancreas Congenital malformation of pancreas or pancreatic duct, NOS Ectopic pancreas Ectopic pancreatic tissue Pancreatic heterotopia **Excludes 1:** congenital diabetes mellitus (E10.-), cystic fibrosis (E84.0-E84.9), fibrocystic disease of pancreas (E84.-), neonatal diabetes mellitus (P70.2)	751.7
Q45.8	Other specified congenital malformations of digestive system (must specify) Absence (complete) (partial) of alimentary tract, NOS Duplication of digestive system Malposition, congenital of digestive system Congenital malposition of digestive organs, NOS Cyst of mesenteric remnant Duplication of alimentary tract Ectopic digestive organs, NOS	751.8
Q45.9	Congenital malformation of digestive system, unspecified (must specify)	751.9

Congenital Malformations of Genital Organs (Q50-Q56)

ICD-10 codes 56.0 to 56.4 require chromosome results.

ICD- 10	Description	ICD-9
Q50	**Congenital malformations of ovaries, fallopian tubes and broad ligaments**	
Q50.0	**Congenital absence of ovary [Only applicable to female patients.]** **Excludes 1:** Turner's Syndrome (Q96.-)	
Q50.01	Congenital absence or agenesis of ovary, unilateral	752.0
Q50.02	Congenital absence or agenesis of ovary, bilateral	752.0
Q50.1	Developmental ovarian cyst Cystic ovary Multiple ovarian cysts	752.0
Q50.2	Congenital torsion of ovary	752.0
Q50.3	**Other congenital malformations of ovary [Only applicable to female patients.]**	
Q50.31	Accessory ovary	752.0
Q50.32	Ovarian streak 46.XX with streak gonads	752.0
Q50.39	Other congenital malformation of ovary Congenital malformation of ovary, NOS Ectopic ovary	752.0
Q50.4	Embryonic cyst of fallopian tube Fimbrial cyst	752.11
Q50.5	Embryonic cyst of broad ligament Cyst of hydatid of Morgagni Epoophoron cyst Parovarian cyst	752.11
Q50.6	Other congenital malformations of fallopian tube and broad ligament (must specify) 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 board ligament, NOS	752.10 752.19
Q51	**Congenital malformations of uterus and cervix [Only applicable to female patients.]**	
Q51.0	Agenesis and aplasia of uterus Congenital absence of uterus	752.31
Q51.1	**Doubling of uterus with doubling of cervix and vagina [Only applicable to female patients.]**	
Q51.10	Doubling of uterus with doubling of cervix and vagina without obstruction Doubling of uterus with doubling of cervix and vagina, NOS	752.2
Q51.11	Doubling of uterus with doubling of cervix and vagina with obstruction	752.2
Q51.2	Other doubling of uterus **[Only applicable to female patients.]** Septate uterus	
Q51.21	Complete doubling of uterus Complete septate uterus	
Q51.22	Partial doubling of uterus Partial septate uterus	
Q51.28	Other and unspecified doubling of uterus Septate uterus, NOS	752.35
Q51.3	Bicornate uterus Bicornate uterus, complete or partial	752.34
Q51.4	Unicornate uterus Unicornate uterus with or without a separate uterine horn Uterus with only one functioning horn	752.33
Q51.5	Agenesis and aplasia of cervix Congenital absence of cervix	752.43
Q51.6	*(m) Embryonic cyst of cervix*	752.41
Q51.7	Congenital fistula between uterus and digestive and urinary tracts	752.49

	Congenital fistula between uterus and digestive tracts Congenital fistula between uterus and urinary tract Congenital uterointestinal fistula	752.39
Q51.8	**Other congenital malformations of uterus and cervix [Only applicable to female patients.]**	
Q51.81	**Other congenital malformations of uterus [Only applicable to female patients.]**	
Q51.810	Arcuate uterus Arcuatus uterus	752.36
Q51.811	Hypoplasia of uterus Infantile uterus	752.32
Q51.818	Other congenital malformations of uterus (must specify) Müllerian anomaly of uterus, NEC	752.39
Q51.82	**Other congenital malformations of cervix [Only applicable to female patients.]**	
Q51.820	Cervical duplication Congenital duplication of cervix	752.44
Q51.821	Hypoplasia of cervix	752.49
Q51.828	Other congenital malformations of cervix (must specify) Congenital imperforate cervix Congenital stenosis of cervical canal Congenital stenosis of uterine cervix Congenital stenosis or stricture of cervical canal	752.49
Q51.9	Congenital malformation of uterus and cervix, unspecified (must specify) Congenital uterine anomaly Inadequate development of endometrium	752.39 752.49
Q52	**Other congenital malformations of female genitalia [Only applicable to female patients.]**	
Q52.0	Congenital absence of vagina Vaginal agenesis, total or partial	752.45
Q52.1	**Doubling of vagina [Only applicable to female patients.]** **Excludes 1:** doubling of vagina with doubling of uterus and cervix (Q51.1-)	
Q52.10	Doubling of vagina, unspecified Congenital duplication of vagina Congenital vaginal septum Septate vagina, NOS	752.47
Q52.11	Transverse vaginal septuma Congenital transverse septated vagina	752.46
Q52.12	Longitudinal vaginal septum Congenital longitudinal vaginal septum Longitudinal septate vagina	
Q52.120	Longitudinal vaginal septum, non-obstructing (female only)	752.47
Q52.121	Longitudinal vaginal septum, obstructing, right side (female only)	752.47
Q52.122	Longitudinal vaginal septum, obstructing, left side (female only)	752.47
Q52.123	Longitudinal vaginal septum, microperforate, right side (female only)	752.47
Q52.124	Longitudinal vaginal septum, microperforate, left side (female only)	752.47
Q52.129	Other and unspecified longitudinal vaginal septum (female only)	752.47
Q52.2	Congenital rectovaginal fistula **Excludes 1:** cloaca (Q43.7)	752.49
Q52.3	*(m) Imperforate hymen*	752.42
Q52.4	Other congenital malformation of vagina (must specify) Congenital malformation of vagina, NOS (must specify) *(m) Canal of Nuck cyst, congenital* *(m) Congenital atresia of vagina* *(m) Embryonic vaginal cyst* *(m) Gartner's duct cyst* *(m) Vaginal adenosis* *(m) Cyst of vagina, vulva* *(m) Congenital stenosis or stricture of vagina*	752.41 752.49

Q52.5	Fusion of labia *(m) Labial adhesions, congenital*	752.49
Q52.6	Congenital malformation of clitoris (must specify) Absence of clitoris Agenesis of clitoris *(m) Clitoromegaly* *(m) Enlarged clitoris* *(m) Prominent clitoris*	752.49
Q52.7	**Other and unspecified congenital malformations of vulva [Only applicable to female patients.]**	
Q52.70	Unspecified congenital malformations of vulva (must specify) Congenital anomaly of external female genitalia Congenital malformation of vulva, NOS Fusion of vulva	752.49
Q52.71	Congenital absence of vulva	752.49
Q52.79	Other congenital malformations of vulva (must specify) *(m) Congenital cyst of vulva*	752.49
Q52.8	Other specified congenital malformation of female genitalia (must specify) *(m) Vaginal or hymenal tags* *(m) Hypoplastic majora*	752.49
Q52.9	Congenital malformation of female genitalia, unspecified (must specify)	752.9
Q53	**Undescended and ectopic testicle [Only applicable to male patients.]**	
Q53.0	**Ectopic testis [Only applicable to male patients.]**	
Q53.00	Ectopic testis, unspecified (birth weight greater than 2,500 grams)	752.51
Q53.01	Ectopic testis, unilateral (birth weight greater than 2,500 grams) Ectopic testis, left only (birth weight greater than 2,500 grams) Ectopic testis, right only (birth weight greater than 2,500 grams)	752.51
Q53.02	Ectopic testes, bilateral (birth weight greater than 2,500 grams)	752.51
Q53.1	**Undescended testicle, unilateral [Only applicable to male patients.]**	
Q53.10	Unspecified undescended testicle, unilateral (birth weight greater than 2,500 grams) Cryptorchidism, left only (birth weight greater than 2,500 grams) Cryptorchidism, right only (birth weight greater than 2,500 grams) Cryptorchidism, unilateral (birth weight greater than 2,500 grams) Left undescended testicle (birth weight greater than 2,500 grams) Nonpalpable testis, unilateral (birth weight greater than 2,500 grams) Right undescended testicle (birth weight greater than 2,500 grams)	752.51
Q53.11	**Abdominal testis, unilateral (birth weight greater than 2,500 grams)**	752.51
Q53.111	Intraabdominal testis, unilateral (birth weight greater than 2,500 grams)	752.51
Q53.112	Inguinal testis, unilateral (birth weight greater than 2,500 grams)	752.51
Q53.12	Ectopic perineal testis, unilateral (birth weight greater than 2,500 grams)	752.51
Q53.13	*(m) High scrotal testis, unilateral* (birth weight greater than 2,500 grams)	752.51
Q53.2	**Undescended testicle, bilateral [Only applicable to male patients.]**	
Q53.20	Undescended testicle, unspecified, bilateral (birth weight greater than 2,500 grams) Cryptorchidism, bilateral (birth weight greater than 2,500 grams) Nonpalpable testes (birth weight greater than 2,500 grams) Undescended testicle, bilateral (birth weight greater than 2,500 grams)	752.51
Q53.21	**Abdominal testis, bilateral (birth weight greater than 2,500 grams)**	752.51
Q53.211	Intraabdominal testes, bilateral (birth weight greater than 2,500 grams)	752.51
Q53.212	Inguinal testes, bilateral (birth weight greater than 2,500 grams)	752.51
Q53.22	Ectopic perineal testis, bilateral (birth weight greater than 2,500 grams)	752.51
Q53.23	*(m) Bilateral high scrotal testes* (birth weight greater than 2,500 grams)	
Q53.9	Undescended testicle, unspecified (birth weight greater than 2,500 grams) Cryptorchidism, NOS (birth weight greater than 2,500 grams) Nonpalpable testis (birth weight greater than 2,500 grams)	752.51
Q54	**Hypospadias PLEASE SPECIFY DEGREE OR SITE OF HYPOSPADIAS [Only applicable to male patients.]**	

	Excludes 1: epispadias (Q64.0)	
Q54.0	Hypospadias, balanic Hypospadias, coronal Hypospadias, glandular Hypospadias 1 degree, glandular, coronal	752.61
Q54.1	Hypospadias, penile Hypospadias 2 degree, penile, subcoronal	752.61
Q54.2	Hypospadias, penoscrotal Hypospadias 3 degree, scrotal, penile scrotal	752.61
Q54.3	Hypospadias, perineal Hypospadias 3 degree, perineal, scrotal, penile scrotal	752.61
Q54.4	Congenital chordee (report only with hypospadias (ventral, dorsal, NOS)) *(m) Chordee without hypospadias*	752.63
Q54.8	Other hypospadias (must specify) Hypospadias with intersex state	752.61
Q54.9	Hypospadias, unspecified (must specify) Paraspadias	752.61
Q55	**Other congenital malformations of male genital organs [Only applicable to male patients.]** **Excludes 1:** congenital hydrocele (P83.5), hypospadias (Q54.-)	
Q55.0	Absence and aplasia of testis Congenital absence of testicle Monorchism	752.89
Q55.1	Hypoplasia of testis and scrotum Fusion of testes	752.89
Q55.2	**Other and unspecified congenital malformations of testis and scrotum [Only applicable to male patients.]**	
Q55.20	Unspecified congenital malformations of testis and scrotum (must specify) Congenital malformation of testis or scrotum, NOS	752.9
Q55.21	Polyorchidism	752.89
Q55.22	*(m) Retractile testis*	752.52
Q55.23	Scrotal transposition Penoscrotal transposition	752.81
Q55.29	Other congenital malformations of testis and scrotum (must specify)	752.89
Q55.3	Atresia of vas deferens	752.89
Q55.4	Other congenital malformations of vas deferens, epididymis, seminal vesicles and prostate (must specify) Absence or aplasia of prostate Absence or aplasia of spermatic cord Appendix testis cyst Wolffian duct cyst	752.89
Q55.5	Congenital absence and aplasia of penis	752.69
Q55.6	**Other congenital malformations of penis**	
Q55.61	Curvature of penis (lateral)	752.69
Q55.62	Hypoplasia of penis Micropenis	752.64
Q55.63	*(m) Congenital torsion of penis* *Phimosis is not reportable* **Excludes 1:** acquired torsion of penis (N48.82)	752.69
Q55.64	*(m) Buried penis* *(m)* Concealed penis *(m) Hidden penis* *(m) Webbed penis* **Excludes 1:** acquired buried penis (N48.83)	752.65

Q55.69	Other congenital malformation of penis (must specify) Congenital malformation of penis, NOS Absent or hooded foreskin Incomplete foreskin	752.69
Q55.7	Congenital vasocutaneous fistula	752.89
Q55.8	Other specified congenital malformations of male genital organs (must specify)	752.89
Q55.9	Congenital malformations of male genital organ, unspecified (must specify) Congenital anomaly of male genital organ Congenital deformity of male genital organ	752.9
Q56	**Indeterminate sex and pseudohermaphroditism** **PLEASE REPORT CHROMOSOME TEST RESULTS** **Excludes 1:** 46,XX true hermaphrodite (Q99.1), androgen insensitivity syndrome (E34.5-), chimera 46,XXX/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, NEC Ovotestis	752.7
Q56.1	Male pseudohermaphroditism, NEC	752.7
Q56.2	Female pseudohermaphroditism, NEC	752.7
Q56.3	Pseudohermaphroditism, unspecified	752.7
Q56.4	Indeterminate sex, unspecified Ambiguous genitalia Disorder of sexual differentiation Sexual differentiation disorder	752.7

Congenital Malformations of the Urinary System (Q60-Q64)

ICD-10	Description	ICD-9
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 Single kidney Solitary kidney	753.0
Q60.1	Renal agenesis, bilateral	753.0
Q60.2	Renal agenesis, unspecified	753.0
Q60.3	Renal hypoplasia, unilateral	753.0
Q60.4	Renal hypoplasia, bilateral	753.0
Q60.5	Renal hypoplasia, unspecified Oligomeganephronic hypoplasia of kidney	753.0
Q60.6	Potter's syndrome Oligohydramnios sequence	753.0
Q61	**Cystic kidney disease** **Excludes 1:** 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)	753.10
Q61.01	Congenital single renal cyst	753.11
Q61.02	Congenital multiple renal cysts	753.19
Q61.1	**Polycystic kidney, infantile type** Polycystic kidney, autosomal recessive	
Q61.11	Cystic dilation of collecting ducts	753.14
Q61.19	Other polycystic kidney, infantile type Autosomal recessive infantile polycystic kidney disease Polycystic kidney, autosomal recessive Congenital PKD	753.14
Q61.2	Polycystic kidney, adult type Polycystic kidney, autosomal dominant	753.13
Q61.3	Polycystic kidney, unspecified	753.12
Q61.4	Renal dysplasia Congenital renal dysplasia Multicystic dysplastic kidney Multicystic kidney (development) Multicystic kidney disease (MCDK) Multicystic renal dysplasia Dysplastic kidney, bilateral Renal dysplasia, bilateral Renal dysplasia, NOS; dysplastic kidney, NOS Unilateral dysplasia of kidney **Excludes 1:** polycystic kidney disease (Q61.11-Q61.3)	753.15 753.19
Q61.5	Medullary cystic kidney Medullary cystic kidney, juvenile type Medullary cystic kidney, adult type Medullary sponge kidney Nephronophthisis Sponge kidney, NOS	753.16 753.17
Q61.8	Other cystic kidney diseases (must specify) Fibrocystic kidney Fibrocystic renal degeneration of disease	753.19
Q61.9	Cystic kidney disease, unspecified (must specify) Cystic kidney disease Meckel-Gruber syndrome	753.10

Q62	**Congenital obstructive defects of renal pelvis and congenital malformations of ureter**	
Q62.0	Congenital hydronephrosis (swelling of kidney) Congenital hydronephrosis, bilateral Congenital hydronephrosis, left only Congenital hydronephrosis, NOS Congenital hydronephrosis, right only Congenital hydronephrosis, unilateral *(m) Pelvicaliectasis/Pelviectasis* *(m) Pyelectasis*	753.29
Q62.1	**Congenital occlusion of ureter** Atresia and stenosis of ureter	
Q62.10	Congenital occlusion of ureter, unspecified Stenosis of ureter Atresia, stricture, or stenosis of ureter, bilateral Atresia, stricture, or stenosis of ureter, left Atresia, stricture, or stenosis of ureter, NOS Atresia, stricture, or stenosis of ureter, right Atresia, stricture, or stenosis of ureter, unilateral Congenital occlusion of ureter, bilateral Congenital occlusion of ureter, left Congenital occlusion of ureter, right Congenital occlusion of ureter, unilateral	753.29
Q62.11	Congenital occlusion of ureteropelvic junction Stricture of pelviureteric junction Ureteropelvic junction (UPJ) obstruction/stenosis, left Ureteropelvic junction obstruction/stenosis, right Ureteropelvic junction obstruction/stenosis, unilateral Ureteropelvic junction obstruction/stenosis, bilateral	753.21
Q62.12	Congenital occlusion of ureterovesical orifice Ureterovesical junction obstruction/stenosis, NOS Ureterovesical junction obstruction/stenosis, left Ureterovesical junction obstruction/stenosis, right Ureterovesical junction obstruction/stenosis, unilateral Ureterovesical junction obstruction/stenosis, bilateral	753.22
Q62.2	Congenital megaureter Congenital dilation of ureter Congenital hydroureter, NOS Congenital hydroureter, right Congenital hydroureter, left Congenital hydroureter, unilateral Congenital hydroureter, bilateral Megaureter, left Megaureter, right Megaureter, unilateral Megaureter, bilateral	753.22
Q62.3	**Other obstructive defects of renal pelvis and ureter**	
Q62.31	Congenital ureterocele, orthotopic Simple ureterocele Ureterocele is reportable; Hydrocele is NOT reportable	753.23
Q62.32	Cecoureterocele Ectopic ureterocele	753.23
Q62.39	Other obstructive defects of renal pelvis ureter Congenital hydroureter Congenital occlusion of ureter Congenital ureteral obstruction Congenital ureterovesical obstruction	753.20

	Duplicated collecting system with obstruction Ureteropelvic junction obstruction, NOS Urinary tract obstruction due to duplicated collecting system	
Q62.4	Agenesis of ureter Congenital absence ureter	753.4
Q62.5	Duplication of ureter Accessory ureter Double ureter, duplex collecting system Duplication of collecting system	753.4
Q62.6	**Malposition of ureter**	
Q62.60	Malposition of ureter, unspecified	753.4
Q62.61	Deviation of ureter	753.4
Q62.62	Displacement of ureter	753.4
Q62.63	Anomalous implantation of ureter Ectopia of ureter Ectopic ureter Displaced ureteric orifice	753.4
Q62.69	Other malposition of ureter	753.4
Q62.7	Congenital vesico-uretero-renal reflux Ureteral reflux Variations of vesicoureteral reflux	753.4
Q62.8	Other congenital malformations of ureter (must specify) Anomaly of ureter, NOS Hypoplastic ureter, NOS Hypoplastic ureter, left Hypoplastic ureter, right Hypoplastic ureter, bilateral Unspecified obstructive defects of renal pelvis and ureter	753.4
Q63	**Other congenital malformations of kidney Excludes 1:** congenital nephrotic syndrome (N04.-)	
Q63.0	Accessory kidney Double kidney and pelvis, pyelon duplex Trifid kidney (pelvis) Triple kidney and pelvis, pyelon triplex	753.3
Q63.1	Lobulated, fused or horseshoe kidney Fused kidney Horseshoe kidney Lobulated kidney	753.3
Q63.2	Ectopic kidney/Pelvic Kidney Congenital displaced kidney Malrotation of kidney	753.3
Q63.3	Hyperplastic or giant kidney Compensatory hypertrophy of kidney	753.3
Q63.8	Other specified congenital malformations of kidney (must specify) Congenital renal caluli/ calculus Congenital renal pelvis and ureteral defect Discoid kidney Duplicated collecting system without obstruction Inequal in size of kidney Megacalycosis, congenital	753.3
Q63.9	Congenital malformation of kidney, unspecified	753.3
Q64	**Other congenital malformations of urinary system**	
Q64.0	Epispadias Anaspadias Ventral meatus **Excludes 1:** hypospadias (Q54.-) [Q64.0 is only applicable to male patients.]	752.62

Q64.10	Exstrophy of urinary bladder, unspecified Ectopia vesicae Exstrophy of bladder sequence	753.5
Q64.11	Supravesical fissure of urinary bladder	753.8
Q64.12	Cloacal exstrophy of urinary bladder Exstrophy of cloaca sequence	753.5
Q64.19	Other exstrophy of urinary bladder Estroversion of bladder	753.5
Q64.2	Congenital posterior urethral valves	753.6
Q64.3	**Other atresia and stenosis of urethra and bladder neck**	
Q64.31	Congenital bladder neck obstruction Congenital bladder neck stricture Congenital obstruction of vesicourethral orifice	753.6
Q64.32	Congenital stricture of urethra	753.6
Q64.33	Congenital stricture of urinary meatus	753.6
Q64.39	Other atresia and stenosis of urethra and bladder neck Atresia and stenosis of urethra and bladder neck, NOS	753.6
Q64.4	Malformation of urachus (must specify) Cyst of urachus Fistula of urachus Patent urachus Prolapse of urachus	753.7
Q64.5	Congenital absence of bladder and urethra	753.8
Q64.6	Congenital diverticulum of bladder	753.8
Q64.7	**Other and unspecified congenital malformations of bladder and urethra** **Excludes 1:** congenital prolapse of bladder (mucosa) (Q79.4)	
Q64.70	Unspecified congenital malformations of bladder and urethra (must specify) Malformation of bladder or urethra, NOS	753.9
Q64.71	Congenital prolapse of urethra	753.8
Q64.72	Congenital prolapse of urinary meatus	753.8
Q64.73	Congenital urethrorectal fistula	753.8
Q64.74	Double urethra	753.8
Q64.75	Double urinary meatus	753.8
Q64.79	Other congenital malformations of bladder and urethra (must specify)	753.8
Q64.8	Other specified congenital malformations of urinary system (must specify)	753.8
Q64.9	Congenital malformation of urinary system, unspecified (must specify)	753.9

Congenital Malformations of the Musculoskeletal System (Q65-Q79)

ICD-10	Description	ICD 9
Q65	**Congenital deformities of hip Excludes 1:** clicking hip (R29.4)	
Q65.0	**Congenital dislocation of hip, unilateral**	
Q65.00	Congenital dislocation of unspecified hip, unilateral	754.30
Q65.01	Congenital dislocation of right hip, unilateral	754.30
Q65.02	Congenital dislocation of left hip, unilateral	754.30
Q65.1	Congenital dislocation of hip, bilateral	754.31
Q65.2	Congenital dislocation of hip, unspecified	754.30
Q65.3	**Congenital partial dislocation of hip, unilateral**	
Q65.30	Congenital partial dislocation of unspecified hip, unilateral Congenital subluxation of unspecified hip, unilateral	754.32
Q65.31	Congenital partial dislocation of right hip, unilateral Congenital subluxation of right hip	754.32
Q65.32	Congenital partial dislocation of left hip, unilateral Congenital subluxation of left hip	754.32
Q65.4	Congenital partial dislocation of hip, bilateral Congenital subluxation of hip, bilateral	754.33
Q65.5	Congenital partial dislocation of hip, unspecified Congenital subluxation of hip	754.32
Q65.6	Congenital unstable hip Congenital dislocatable hip Predislocation status of hip at birth, unilateral, left Predislocation status of hip at birth, unilateral, right Predislocation status of hip at birth, unilateral Unstable hip, unilateral Unstable hip, unilateral, left Unstable hip, unilateral, right	754.32
Q65.8	**Other congenital deformities of hip**	
Q65.81	Congenital coxa valga	755.61
Q65.82	Congenital coxa vara	755.62
Q65.89	Other specified congenital deformities of hip (must specify) Anteversion of femoral neck/ femoral anteversion (do not report if normal x-ray) Congenital acetabular dysplasia Hip dysplasia, NOS Bilateral hip dysplasia Congenital flexion deformity, hip or thigh, left Congenital flexion deformity, hip or thigh, right Developmental dysplasia of hip (DDH) Unilateral hip dysplasia	755.63
Q65.9	Congenital deformity of hip, unspecified (try not to use; must specify)	755.63
Q66	**Congenital deformities of feet** **Excludes 1:** reduction defects of feet (Q72.-), valgus deformities (acquired) (M21.0-), varus deformities (acquired) (M21.1-)	
Q66.0	Congenital talipes equinovarus	
Q66.00	Congenital talipes equinovarus, unspecified foot Talipes equinovarus, NOS Talipes equinovarus, unilateral Talipes equinovarus, bilateral	754.51
Q66.01	Talipes equinovarus, right	754.51
Q66.02	Talipes equinovarus, left	754.51
Q66.1	**Congenital talipes calcaneovarus**	
Q66.10	Congenital talipes calcaneovarus, unspecified foot Talipes calcaneovarus, unilateral Talipes calcaneovarus, bilateral	754.59

Q66.11	Congenital talipes calcaneovarus, right	754.59
Q66.12	Congenital talipes calcaneovarus, left	754.59
Q66.2	**Congenital metatarsus adductus** Congenital metatarsus varus Congenital metatarsus primus varus	
Q66.21	**Congenital metatarsus (primus) varus**	
Q66.211	Congenital metatarsus primus varus, right foot	754.52
Q66.212	Congenital metatarsus primus varus, left foot	754.52
Q66.219	Congenital metatarsus primus varus, unspecified foot	754.52
Q66.22	**Congenital metatarsus adductus** Congenital metatarsus varus	
Q66.221	Congenital metatarsus adductus, right foot Congenital metatarsus varus, right foot	754.53
Q66.222	Congenital metatarsus adductus, left foot Congenital metatarsus varus, left foot	754.53
Q66.229	Congenital metatarsus adductus, unspecified foot Congenital metatarsus varus, unspecified foot	754.53
Q66.3	**Other congenital varus deformities of feet** Hallux varus, congenital Talipes varus Cubitus varus deformity of feet	
Q66.30	Other congenital varus deformities of feet, unspecified foot (must specify) Talipes varus	754.59
Q66.31	Other congenital varus deformities of feet, right foot (must specify) Talipes varus, right	754.59
Q66.32	Other congenital varus deformities of feet, left foot (must specify) Talipes varus, left	754.59
Q66.4	**Congenital talipes calcaneovalgus**	
Q66.40	Congenital talipes calcaneovalgus, unspecified foot Congenital talipes calcaneovalgus, unilateral Congenital talipes calcaneovalgus, bilateral	754.62
Q66.41	Congenital talipes calcaneovalgus, right foot	754.62
Q66.42	Congenital talipes calcaneovalgus, left foot	754.62
Q66.5	**Congenital pes planus** Congenital flat foot Congenital rigid flat foot Congenital spastic (everted) flat foot *(m) Congenital rocker bottom foot* **Excludes 1:** pes planus, acquired (M21.4)	
Q66.50	Congenital pes planus, unspecified foot	754.61
Q66.51	Congenital pes planus, right foot	754.61
Q66.52	Congenital pes planus, left foot	754.61
Q66.6	Other congenital valgus deformities of feet (must specify) Congenital metatarsus valgus Hallus valgus Pes valgoplanus congenital Talipes valgus Talipes equinovalgus	754.69
Q66.7	**Congenital pes cavus** Talipes cavus	
Q66.70	Congenital pes cavus, unspecified foot	754.71
Q66.71	Congenital pes cavus, right foot	754.71
Q66.72	Congenital pes cavus, left foot	754.71
Q66.8	**Other congenital deformities of feet**	
Q66.80	Congenital vertical talus deformity, unspecified foot	754.61

Q66.81	Congenital vertical talus deformity, right foot	754.61
Q66.82	Congenital vertical talus deformity, left foot	754.61
Q66.89	Other specified congenital deformities of feet (must specify) Brachydactyly of toes Brachymetatarsia Congenital asymmetric talipes Congenital contracture of toe Congenital talipes, NOS Congenital tarsal coalition Congenital clubfoot, NOS Fusion of calcaneus (heel bone) Tarsal coalitions (specify laterality) Astragalo-scaphoid synostosis Calcaneonavicular bar Clubfoot, NOS Congenital hallux valgus Congenital hallux varus Dorsiflexion of foot (fixed only) Other specified deformities of toes (must specify) Plantar furrow Prominent heel Talipes calcaneus Talipes equinus Talonavicular synostosis *(m) Hammer toe, congenital* *(m) Curly toe* *(m) Gap between toes (1-2)* *(m) Long toes* *(m) Overlapping toes* *(m) Recessed toes (4,5)* *(m) Short or broad great toe* *(m) Wide spaced first and second toes*	 754.79 755.67
Q66.9	**Congenital deformity of feet, unspecified (must specify)** Congenital deformity of toe(s) (must specify)	
Q66.90	Congenital deformity of feet, unspecified, unspecified foot (must specify)	754.79
Q66.91	Congenital deformity of feet, unspecified, right foot (must specify)	754.79
Q66.92	Congenital deformity of feet, unspecified, left foot (must specify)	754.79
Q67	**Congenital musculoskeletal deformities of head, face, spine and chest** **Excludes 1:** congenital malformation syndromes classified to Q87.-, Potter's syndrome (Q60.6)	
Q67.0	*(m) Congenital facial asymmetry* *(m) Asymmetry of face* *(m) Facial asymmetry* *(m) Head asymmetry*	 754.0
Q67.1	*(m) Congenital compression facies*	754.0
Q67.2	*(m) Dolichocephaly* *(m) Long narrow head* **Excludes 1:** sagittal craniosynostosis (Q75.01)	754.0
Q67.3	*(m) Plagiocephaly* *(m) Flat occiput* **Excludes 1:** coronal craniosynostosis (Q75.02-), lambdoid craniosynostosis (Q75.04-)	 754.0
Q67.4	*(m) Other congenital deformities of skull, face and jaw (must specify)* *(m) Congenital hemifacial atrophy or hypertrophy* *(m) Congenital depressions in skull* *(m) Deviation of nasal septum, congenital* *(m) Squashed or bent nose, congenital* *(m) Oxycephaly*	 748.1 754.0

	(m) Trigonocephaly *(m) Large or small fontanelles* *(m) Metopic suture open to bregma* *(m) Scaphocephaly (if due to craniosynostosis)* *(m) Trigonocephaly, other head deformations without synostosis* **Excludes 1:** dentofacial anomalies [including malocclusion] (M26.-), syphilitic saddle nose (A50.5)	
Q67.5	Congenital deformity of spine Congenital postural scoliosis Congenital scoliosis, NOS **Excludes 1:** infantile idiopathic scoliosis (M41.0), scoliosis due to congenital bony malformation (Q76.3)	754.2
Q67.6	Pectus excavatum Congenital funnel chest	754.81
Q67.7	Pectus carinatum Congenital pigeon chest	754.82
Q67.8	Other congenital deformities of chest (must specify) Congenital deformity of chest wall, NOS Congenital anomaly of thoracic cage *(m) Bell shaped chest*	754.89
Q68	**Other congenital musculoskeletal deformities** **Excludes 1:** reduction defects of limb(s) (Q71-Q73) **Excludes 2:** congenital myotonic chondrodystrophy (G71.13)	
Q68.0	Congenital deformity of sternocleidomastoid muscle (must specify) Congenital anomaly of sternocleidomastoid muscle Congenital contracture of sternocleidomastoid (muscle) Congenital (sternomastoid) torticollis Sternomastoid tumor (congenital) Absent or hypoplastic sternocleidomastoid muscle Congenital wryneck Sternomastoid tumor	754.1
Q68.1	Congenital deformity of finger(s) and hand (must specify) Congenital club finger Congenital crooked finger Spade-like hand (congenital) Anthrogryposis Flexion contractures of individual joints of the hand	754.89
Q68.2	Congenital deformity of knee Congenital dislocation of knee Congenital genu recurvatum	755.64 754.41 754.40
Q68.3	Congenital bowing of femur **Excludes 1:** anteversion of femur (neck) (Q65.89)	754.42
Q68.4	Congenital bowing of tibia and fibula	754.43
Q68.5	Congenital bowing of long bones of leg, unspecified	754.44
Q68.6	Discoid meniscus Discoid meniscus of knee	717.5
Q68.8	Other specified congenital musculoskeletal deformities (must specify) 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 Congenital dislocation of radial head Congenital elevation of scapula	756.9 755.51 755.52

	Generalized flexion contractures of lower limb joints	
Q69	**Polydactyly**	
Q69.0	Accessory finger(s) Radial polydactyly Accessory digits hand, NOS Polydactyly of fingers	755.01
Q69.1	Accessory thumb(s)	755.01
Q69.2	Accessory toe(s) (do not report if skin tag) Accessory hallux Polydactyly of toes Accessory big toe (preaxial) Accessory toes (postaxial) Accessory digits foot, NOS	755.02
Q69.9	Polydactyly, unspecified (must specify) Supernumerary digit(s), NOS Postaxial polydactyly, NOS Accessory digits, NOS (hand/foot not specified)	755.00
Q70	**Syndactyly**	
Q70.0	**Fused fingers** **Complex syndactyly of fingers and/or thumb with synostosis**	
Q70.00	Fused fingers and/or thumb, unspecified hand Syndactyly of fingers with fusion of bones	755.12
Q70.01	Fused fingers and/or thumb, right hand Syndactyly of fingers of right hand with fusion of bones	755.12
Q70.02	Fused fingers and/or thumb, left hand Syndactyly of fingers of left hand with fusion of bones	755.12
Q70.03	Fused fingers and/or thumb, bilateral Syndactyly of fingers, bilateral, with fusion of bones	755.12
Q70.1	***(m) Webbed fingers*** ***(m) Simple syndactyly of fingers without synostosis***	
Q70.10	*(m) Webbed fingers, unspecified hand*	755.11
Q70.11	*(m) Webbed fingers, right hand*	755.11
Q70.12	*(m) Webbed fingers, left hand*	755.11
Q70.13	*(m) Webbed fingers, bilateral*	755.11
Q70.2	**Fused toes** **Complex syndactyly of toes with synostosis**	
Q70.20	Fused toes, unspecified foot Syndactyly of toes with fusion of bones	755.14
Q70.21	Fused toes, right foot Syndactyly of toes of right foot, with fusion of bones	755.14
Q70.22	Fused toes, left foot Syndactyly of toes of left foot, with fusion of bones	755.14
Q70.23	Fused toes, bilateral Syndactyly of toes of both feet, with fusion of bones	755.14
Q70.3	***(m) Webbed toes*** ***(m) Simple syndactyly of toes without synostosis***	
Q70.30	*(m) Webbed toes, unspecified foot* *(m) Simple syndactyly of toes*	755.13
Q70.31	*(m) Webbed toes, right foot*	755.13
Q70.32	*(m) Webbed toes, left foot*	755.13
Q70.33	*(m) Webbed toes, bilateral*	755.13
Q70.4	Polysyndactyly, unspecified (must specify) **Excludes 1:** specified syndactyly of hand and feet - code to specified conditions (Q70.0--Q70.3-)	755.00 755.10
Q70.9	Syndactyly, unspecified (must specify) Symphalangy, NOS	755.10

	Syndactyly of multiple and unspecified sites	
Q71	**Reduction defects of upper limb**	
Q71.0	**Congenital complete absence of upper limb** **Transverse deficiency of upper limb**	
Q71.00	Congenital complete absence of unspecified upper limb Congenital complete absence of arm Transverse deficiency of arm Congenital amputation of upper limb, NOS Transverse hemimelia of upper limb	755.21
Q71.01	Congenital complete absence of right upper limb	755.21
Q71.02	Congenital complete absence of left upper limb	755.21
Q71.03	Congenital complete absence of upper limb, bilateral	755.21
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 Complete phocomelia of upper limb Distal phocomelia of upper limb (forearm missing, upper arm/hand present) Proximal phocomelia of upper limb Total absence of forearm only (radius and ulna)	755.23
Q71.11	Congenital absence of right upper arm and forearm with hand present	755.23
Q71.12	Congenital absence of left upper arm and forearm with hand present	755.23
Q71.13	Congenital absence of upper arm and forearm with hand present, bilateral Complete phocomelia of bilateral upper limbs	755.23
Q71.2	**Congenital absence of both forearm and hand** **Transverse deficiency, radioulnar, complete or partial**	
Q71.20	Congenital absence of both forearm and hand, unspecified upper limb	755.25
Q71.21	Congenital absence of both forearm and hand, right upper limb	755.25
Q71.22	Congenital absence of both forearm and hand, left upper limb	755.25
Q71.23	Congenital absence of both forearm and hand, bilateral	755.25
Q71.3	**Congenital absence of hand and finger**	
Q71.30	Congenital absence of unspecified hand and finger Congenital absence of hand Congenital deficiency of hand bones Absence of thumb	755.29
Q71.31	Congenital absence of right hand and finger Hypoplasia of right thumb	755.29
Q71.32	Congenital absence of left hand and finger Hypoplasia of left thumb	755.29
Q71.33	Congenital absence of hand and finger, bilateral Hypoplasia of thumbs, bilateral	755.29
Q71.4	**Longitudinal reduction defect of radius (congenital)** **Radial clubhand**	
Q71.40	Longitudinal reduction defect of unspecified radius Radial hemimelia Absence of radius Congenital absence of radius with or without absence of some distal elements Longitudinal deficiency, radial, complete or partial Preaxial (longitudinal) reduction deficiency of upper limb, radial ray defect	755.26
Q71.41	Longitudinal reduction defect of right radius	755.26
Q71.42	Longitudinal reduction defect of left radius	755.26
Q71.43	Longitudinal reduction of radius, bilateral	755.26
Q71.5	**Longitudinal reduction defect of ulna**	
Q71.50	Longitudinal reduction defect of unspecified ulna Longitudinal deficiency of ulna Absence of ulna Congenital absence ulna, with or without absence of some distal elements Longitudinal deficiency ulna, complete or partial	755.27

	Postaxial (longitudinal) reduction deficiency of upper limb, ulna ray defect	
Q71.51	Longitudinal reduction defect of right ulna	755.27
Q71.52	Longitudinal reduction defect of left ulna	755.27
Q71.53	Longitudinal reduction defect of ulna, bilateral	755.27
Q71.6	**Lobster-claw hand**	
Q71.60	Lobster-claw hand, unspecified hand Cleft hand	755.58
Q71.61	Lobster-claw right hand Cleft hand, right hand	755.58
Q71.62	Lobster-claw left hand Cleft hand, left hand	755.58
Q71.63	Lobster-claw hand, bilateral Cleft hand, bilateral	755.58
Q71.8	**Other reduction defect of upper limb**	
Q71.81	**Congenital shortening of upper limb**	
Q71.811	Congenital shortening of right upper limb	755.20
Q71.812	Congenital shortening of left upper limb	755.20
Q71.813	Congenital shortening of upper limb, bilateral	755.20
Q71.819	Congenital shortening of unspecified upper limb	755.20
Q71.89	**Other reduction defects of upper limb**	
Q71.891	Other reduction defects of right upper limb Brachydactyly of right fingers	755.20
Q71.892	Other reduction defects of left upper limb Brachydactyly of left fingers	755.20
Q71.893	Other reduction defects of upper limb, bilateral	755.20
Q71.899	Other reduction defects of unspecified upper limb (must specify) Ectromelia NOS of upper limb Hemimelia of upper limb, NOS Longitudinal reduction deficiency of upper limb, unspecified ray defect	755.20
Q71.9	**Unspecified reduction defect of upper limb**	
Q71.90	Unspecified reduction defect of unspecified upper limb	755.20
Q71.91	Unspecified reduction defect of right upper limb	755.20
Q71.92	Unspecified reduction defect of left upper limb	755.20
Q71.93	Unspecified reduction defect of upper limb, bilateral	755.20
Q72	**Reduction defects of lower limb**	
Q72.0	**Congenital complete absence of lower limb**	
Q72.00	Congenital complete absence of unspecified lower limb Congenital complete absence of unspecified leg Transverse deficiency unspecified lower limb Transverse hemimelia of unspecified lower leg	755.31
Q72.01	Congenital complete absence of right lower limb Congenital complete absence of right leg Transverse deficiency right lower limb Transverse hemimelia of right leg	755.31
Q72.02	Congenital complete absence of left lower limb Congenital complete absence of left leg Transverse deficiency left lower limb Transverse hemimelia of left leg	755.31
Q72.03	Congenital complete absence of lower limbs, bilateral Congenital complete absence of legs, bilateral Transverse deficiency lower limbs, bilateral Transverse hemimelia of lower legs, bilateral	755.31
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 Complete phocomelia of lower limb (upper and lower leg missing, foot present)	755.33

	Absence of lower leg only (femur and foot present) Distal phocomelia of lower limb (lower leg absent, femur/foot present) Proximal phocomelia of lower limb (femur absent, lower leg, foot present)	
Q72.11	Congenital absence of right thigh and lower leg with foot present	755.33
Q72.12	Congenital absence of left thigh and lower leg with foot present	755.33
Q72.13	Congenital absence of thigh and lower leg with foot present, bilateral	755.33
Q72.2	**Congenital absence of both lower leg and foot**	
Q72.20	Congenital absence of both lower leg and foot, unspecified lower limb Transverse deficiency lower limb - knee level	755.35
Q72.21	Congenital absence of both lower leg and foot, right lower limb	755.35
Q72.22	Congenital absence of both lower leg and foot, left lower limb	755.35
Q72.23	Congenital absence of both lower leg and foot, bilateral	755.35
Q72.3	**Congenital absence of foot and toe(s)**	
Q72.30	Congenital absence of unspecified foot and toe(s) Longitudinal deficiency of phalanges of foot Longitudinal reduction defect of toes	755.38
Q72.31	Congenital absence of right foot and toe(s) Longitudinal deficiency of right toes Longitudinal reduction defect of right toes	755.38
Q72.32	Congenital absence of left foot and toe(s) Longitudinal deficiency of left toes Longitudinal reduction defect of left toes	755.38 755.39
Q72.33	Congenital absence of foot and toe(s), bilateral Longitudinal deficiency of bilateral toes Longitudinal reduction defect of bilateral toes	755.38 755.39
Q72.4	**Longitudinal reduction defect of femur**	
Q72.40	Longitudinal reduction defect of unspecified femur Longitudinal deficiency of unspecified femur Proximal focal deficiency of unspecified femur Absent unspecified femur, congenital	755.34
Q72.41	Longitudinal reduction defect of right femur Longitudinal deficiency of right femur Absent right femur, congenital	755.34
Q72.42	Longitudinal reduction defect of left femur Longitudinal deficiency of left femur Congenital absence of left femur	755.34
Q72.43	Longitudinal reduction defect of femur, bilateral Longitudinal deficiency of femur, bilateral Absent bilateral femurs, congenital	755.34
Q72.5	**Longitudinal reduction defect of tibia**	
Q72.50	Longitudinal reduction defect of unspecified tibia Congenital absence of tibia Longitudinal deficiency of tibia Tibial hemimelia	755.36
Q72.51	Longitudinal reduction defect of right tibia Longitudinal deficiency of right tibia Congenital absence of right tibia Tibial hemimelia, right	755.36
Q72.52	Longitudinal reduction defect of left tibia Longitudinal deficiency of left tibia Congenital absence of left tibia Tibial hemimelia, left	755.36
Q72.53	Longitudinal reduction defect of tibia, bilateral Congenital absence of tibia, bilateral Agenesis of tibia, bilateral Tibial hemimelia, bilateral	755.36

Q72.6	**Longitudinal reduction defect of fibula** Congenital absence of fibula	
Q72.60	Longitudinal reduction defect of unspecified fibula Congenital absence of fibula Fibular hemimelia Longitudinal deficiency of fibula Agenesis of unspecified fibula	755.37
Q72.61	Longitudinal reduction defect of right fibula Longitudinal deficiency of right fibula Congenital absence of right fibula Agenesis of right fibula	755.37
Q72.62	Longitudinal reduction defect of left fibula Longitudinal deficiency of left fibula Congenital absence of left fibula Agenesis of left fibula	755.37
Q72.63	Longitudinal reduction defect of fibula, bilateral Bilateral longitudinal deficiency of fibulas Congenital absence of fibula, bilateral Bilateral agenesis of fibula	755.37
Q72.7	**Split foot** Cleft foot	
Q72.70	Split foot, unspecified lower limb	755.67
Q72.71	Split foot, right lower limb Right split foot	755.67
Q72.72	Split foot, left lower limb Left split foot	755.67
Q72.73	Split foot, bilateral	755.67
Q72.8	**Other reduction defects of lower limb**	
Q72.81	**Congenital shortening of lower limb**	
Q72.811	Congenital shortening of right lower limb/leg	755.30
Q72.812	Congenital shortening of left lower limb/leg	755.30
Q72.813	Congenital shortening of lower limb/leg, bilateral	755.30
Q72.819	Congenital shortening of unspecified lower limb/leg	755.30
Q72.89	**Other reduction defects of lower limb**	
Q72.891	Other reductions defects of right lower limb Brachydactyly of right toes Longitudinal deficiency of right leg Longitudinal reduction defect of right leg	755.30
Q72.892	Other reductions defects of left lower limb Brachydactyly of left toes Longitudinal deficiency of left leg Longitudinal reduction defect of left leg	755.30
Q72.893	Other reduction defects of lower limb, bilateral	755.30
Q72.899	Other reduction defects of unspecified lower limb Longitudinal deficiency of lower limb Longitudinal reduction defect of leg	755.30
Q72.9	**Unspecified reduction defect of lower limb**	
Q72.90	Unspecified reduction defect of unspecified lower limb/leg	755.32
Q72.91	Unspecified reduction defect of right lower limb/leg	755.32
Q72.92	Unspecified reduction defect of left lower limb/leg	755.32
Q72.93	Unspecified reduction defect of lower limb/leg, bilateral	755.32
Q73	**Reduction defects of unspecified limb (complete or partial)**	
Q73.0	Congenial absence of unspecified limb(s)	755.4
Q73.1	Phocomelia, unspecified limb(s)	755.4

	Phocomelia, NOS	
Q73.8	Other reduction defects of unspecified limb(s) (must specify) Ectromelia of limb, NOS Hemimelia of limb, NOS Longitudinal reduction deformity of unspecified limb(s) Reduction defect of limb, NOS Absent digits, NOS Amputation of unspecified limb Unspecified reduction defect of unspecified limb	755.4
Q74	**Other congenital malformations of limb(s)** **Excludes 1:** polydactyly (Q69.-), reduction defect of limb (Q71-Q73), syndactyly (Q70.-)	
Q74.0	Other congenital malformations of upper limb(s), including shoulder girdle (must specify) Accessory carpal bones Cleidocranial dysostosis Congenital abnormal fusion of carpal bone Congenital carpal coalition Congenital glenoid dysplasia Congenital hypoplasia of ulna Congenital pseudarthrosis of clavicle Congenital trigger finger and trigger thumb Madelung's deformity Radioulnar synostosis Sprengel's deformity Triphalangeal thumb Anomalies of hand (must specify) Anomalies of elbow and upper arm (must specify) Anomalies of shoulder (must specify) Anomalies of wrist (must specify) Bifid thumb Cubitus valgus, congenital Cubitus varus, congenital Hyperextensibilty of upper limb Hypoplasia of arms Hypoplasia of upper limb Other specified deformity of hands Radioulnar dysostosis *(m) Camptodactyly* *(m) Clinodactyly* *(m) Macrodactyly of finger(s)* *(m) Anomalies of finger* *(m) Incurving fingers* *(m) Long fingers* *(m) Long fingers and toes* *(m) Overlapping fingers* *(m) Tapered fingers*	755.53 755.54 755.56 755.57 755.59
Q74.1	Congenital malformation of knee (and joint) (must specify) Bifid patella Bipartite patella (kneecap) Congenital absence of patella Congenital dislocation of patella (knee-cap) Congenital genu valgum (knock-knee) Congenital genu varum (bowleg) Rudimentary patella **Excludes 1**: congenital dislocation of knee (Q68.2), congenital genu recurvatum (Q68.2), nail patella syndrome (Q87.2)	755.64

Q74.2	Other congenital malformations of lower limb(s), including pelvic girdle (must specify) Accessory tarsal bones Accessory tarsal navicular bone Congenital fusion of sacroiliac joint Congenital malformation of ankle joint Congenital malformation of sacroiliac joint Anomalies of ankle (must specify) Anomalies of pelvis (must specify) Anomalies of upper leg (must specify) Congenital deformity of sacroiliac (joint) Deformity of leg, NOS Dorsiflexion of foot Femoral hypoplasia syndrome Hypoplasia of feet Other specified anomaly of lower limb (must specify) *(m) Congenital internal tibial torsion* *(m) Macrodactylia of toe(s)* *(m) Anomalies of lower leg* *(m) Congenital angulation of tibia* **Excludes 1:** anteversion of femur (neck) (Q65.89)	755.69
Q74.3	Arthrogryposis multiplex congenita	754.89
Q74.8	Other specified congenital malformations of limb(s) (must specify) Larsen's syndrome	755.8
Q74.9	Unspecified congenital malformations of limb(s) (try not to use; must specify) Congenital anomaly of limb(s), NOS Dysplasia of limb Anomalies of forearm, NOS Other anomaly of whole arm Unspecified anomalies of upper limb	755.9
Q75	**Other congenital malformations of skull and face bones** **Excludes 1:** 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.-), and hydrocephalus (Q03.-), microcephaly (Q02)	
Q75.0	Craniosynostosis	756.0
Q75.00	Craniosynostosis unspecified (must specify) Closed skull sutures, NOS Premature closure of cranial sutures	756.0
Q75.001	Craniosynostosis unspecified, unilateral	756.0
Q75.002	Craniosynostosis unspecified, bilateral	756.0
Q75.009	Craniosynostosis unspecified Imperfect fusion of skull	756.0
Q75.01	Sagittal Craniosynostosis Non-deformational dolichocephaly Non-deformational scaphocephaly **Excludes1:** plagiocephaly (Q67.3)	756.0
Q75.02	Coronal Craniosynostosis Non-deformational anterior plagiocephaly **Excludes1:** dolichocephaly (Q67.2)	756.0
Q75.021	Coronal Craniosynostosis, unilateral Non-deformational anterior plagiocephaly	756.0
Q75.022	Coronal Craniosynostosis, bilateral Non-deformational brachycephaly	756.0
Q75.029	Coronal Craniosynostosis, unspecified	756.0
Q75.03	Metopic Craniosynostosis	756.0

	Trigonocephaly	
Q75.04	Lambdoid Craniosynostosis Non-deformational posterior plagiocephaly **Excludes1:** dolichocephaly (Q67.2)	756.0
Q75.041	Lambdoid Craniosynostosis, unilateral	756.0
Q75.042	Lambdoid Craniosynostosis, bilateral	756.0
Q75.049	Lambdoid Craniosynostosis, unspecified	756.0
Q75.05	Multi-suture Craniosynostosis (must specify)	756.0
Q75.051	Cloverleaf skull Kleeblattschaedel skull	756.0
Q75.052	Pansynostosis	756.0
Q75.058	Other multi-suture craniosynostosis (must specify) **Excludes 1:** coronal craniosynostosis, bilateral (Q75.022), lambdoid craniosynostosis, bilateral (Q75.042)	756.0
Q75.08	Other single-suture Craniosynostosis (must specify)	756.0
Q75.1	Craniofacial dysostosis Crouzon's disease Crouzon syndrome	756.0
Q75.2	Hypertelorism Orbital hypertelorism Wide set eyes	756.0
Q75.3	Macrocephaly Macroencephaly	756.0
Q75.4	Mandibulofacial dysostosis Franceschetti syndrome Treacher Collins syndrome	756.0
Q75.5	Oculomandibular Dysostosis	756.0
Q75.8	Other specified congenital malformations of skull and face bones (must specify) Asymmetry of head (skull bones) Absence of skull bone, congenital Absence, cranial bone Basilar skull invagination Congenital deformity of forehead Delayed closure of anterior fontanel Forehead deformity, congenital Invagination of basilar skull Late closure of anterior fontanel Oculoauricular vertebral dysplasia Platybasia Cleft mandible malar hypoplasia Goldenhar syndrome Hallermann-Streiff syndrome Hemifacial microsomia Oculoauriculovertebral (OAV) dysplasia Oculomandibulofacial syndrome Orbital dystopia Other craniofacial syndromes Prominent occiput *(m) Bony occipital spur* *(m) Frontal bossing* *(m) Hypotelorism* *(m) Localized skull defects* *(m) Midfacial hypoplasia*	756.0
Q75.9	Congenital malformation of skull and face bones, unspecified (try not to use; must specify) Congenital anomaly of face bones, NOS	756.0

	Congenital anomaly of skull, NOS	
Q76	**Congenital malformations of spine and bony thorax** **Excludes 1:** congenital musculoskeletal deformities of spine and chest (Q67.5 - Q67.8)	
Q76.0	Spina bifida occulta **Excludes 1:** meningocele (spinal) (Q05.-), spina bifida (aperta) (cystica) (Q05.-)	756.17
Q76.1	Klippel-Feil syndrome Cervical fusion syndrome Klippel feil sequence Thoracolumbar kissing spine Wildervanck syndrome Cervicooculaoaccoustic syndrome	756.16
Q76.2	Congenital spondylolisthesis Congenital spondylolysis Congenital cervical spondylolisthesis Congenital lumbar spondylolisthesis Congenital lumbosacral spondylolysis Prespondylolisthesis, lumbosacral Spondyloysis, lumbosacral **Excludes 1:** spondylolisthesis (acquired) (M43.1-), spondylolysis (acquired) (M43.0-)	756.12
Q76.3	Congenital scoliosis due to congenital bony malformation Hemivertebra fusion or failure of segmentation with scoliosis	754.2
Q76.4	**Other congenital malformations of spine, not associated with scoliosis**	
Q76.41	**Congenital kyphosis**	
Q76.411	Congenital kyphosis, occipito-atlanto-axial region	756.19
Q76.412	Congenital kyphosis, cervical region	756.19
Q76.413	Congenital kyphosis, cervicothoracic region	756.19
Q76.414	Congenital kyphosis, thoracic region	756.19
Q76.415	Congenital kyphosis, thoracolumbar region	756.19
Q76.419	Congenital kyphosis, unspecified region	756.19
Q76.42	**Congenital lordosis**	
Q76.425	Congenital lordosis, thoracolumbar region	754.2
Q76.426	Congenital lordosis, lumbar region	754.2
Q76.427	Congenital lordosis, lumbosacral region	754.2
Q76.428	Congenital lordosis, sacral and sacrococcygeal region	754.2
Q76.429	Congenital lordosis, unspecified region	754.2
Q76.49	Other congenital malformations of spine, not associated with scoliosis (must specify) Agenesis of sacrum Aplasia odontoid Atlanto-occipital fusion Caudal agenesis (incomplete development of lower spine) Congenital absence of vertebra, NOS Congenital fusion of spine, NOS Congenital malformation of lumbosacral (joint) (region), NOS Congenital malformation of spine, NOS Dysgenesis, odontoid Dysraphysm, cervical (neck) spine Fusion of atlantooccipital spine Hemivertebra, NOS Malformation of spine, NOS Occipitoatlantoaxial spine ankylosis Platyspondylisis, NOS Sacral agenesis Spinal dysraphism Supernumerary vertebra, NOS Agenesis of cervical vertebrae Agenesis of lumbar vertebrae	756.19 756.13 756.15 756.14

	Agenesis of thoracic vertebrae Hemivertebrae, cervical vertebrae Hemivertebrae, thoracic vertebrae Hemivertebrae, lumbar vertebrae Sacral mass, NOS Sacrococcygeal anomaly	
Q76.5	*(m) Cervical rib* *(m) Supernumerary rib in cervical region*	756.2
Q76.6	Other congenital malformations of ribs (must specify) Accessory rib Congenital absence of rib Congenital fusion of ribs Congenital malformation of ribs, NOS **Excludes 1:** short rib syndrome (Q77.2)	756.3
Q76.7	Congenital malformations of sternum (must specify) Congenital absence of sternum Sternum bifidum Anomalies of thoracic cage, unspecified Congenital fissure of sternum Double ossification center in the manubrium Short sternum	756.3
Q76.8	Other congenital malformations of bony thorax	756.3
Q76.9	Congenital malformation of bony thorax, unspecified	756.3
Q77	**Osteochondrodysplasia with defects of growth of tubular bones and spine** **Excludes 1:** mucopolysaccharidosis (E76.0-E76.3) **Excludes 2:** congenital myotonic chondrodystrophy (G71.13)	
Q77.0	Achondrogenesis Hypochondrogenesis	756.4
Q77.1	Thanatophoric short stature Thanatophoric dysplasia	756.4
Q77.2	Short rib syndrome Asphyxiating thoracic dysplasia (Jeune) Short rib dysplasia	756.3
Q77.3	Chondrodysplasia punctate Chondrodysplasia punctata (stippled epiphyses) group Conradi syndrome **Excludes 1:** Rhizomelic chondrodysplasia punctata (E71.43)	756.59
Q77.4	Achondroplasia Hypochondroplasia Osteosclerosis congenital	756.4
Q77.5	Diastrophic dysplasia	756.4
Q77.6	Chondroectodermal dysplasia Ellis-van Creveld syndrome	756.55
Q77.7	Spondyloepiphyseal dysplasia Spondyloepimetaphyseal disorder	756.4
Q77.8	Other osteochondrodysplasia with defects of growth of tubular bones and spine	756.4
Q77.9	Osteochondrodysplasia with defects of growth of tubular bones and spine, unspecified	756.4
Q78	**Other osteochondrodysplasias** **Excludes 2:** congenital myotonic chondrodystrophy (G71.13)	
Q78.0	Osteogenesis imperfecta Brittle bones disease Fragilitas ossium Osteopsathyrosis	756.51
Q78.1	Polyostotic fibrous dysplasia Albright (McCune) (Sternberg) syndrome Osteitis fibrosa disseminata	756.54

Q78.2	Osteopetrosis Albers-Schönberg syndrome Osteosclerosis, NOS Marble bones	756.52
Q78.3	Progressive diaphyseal dysplasia (multiple epiphyseal dysplasia) Camurati-Engelmann syndrome Engelmann syndrome	756.56
Q78.4	Enchondromatosis Maffucci's syndrome Multiple enchondromatosis Ollier's disease	756.4
Q78.5	Metaphyseal dysplasia Pyle metaphyseal dysplasia Pyle's syndrome	756.59
Q78.6	Multiple congenital exostoses Diaphyseal aclasis	756.6
Q78.8	Other specified osteochondrodysplasia (must specify) Dysplasia of epiphysis Epiphyseal dysplasia Homozygous Leri Weill dyschondrosteosis syndrome Langer mesomelic dysplasia syndrome Osteopoikilosis Melnick-needles syndrome	756.59
Q78.9	Osteochondrodysplasia, unspecified Chondrodysplasia Chondrodystrophy, NOS Osteodystrophy, NOS	756.50
Q79	**Congenital malformations of musculoskeletal system, NEC** **Excludes 2:** congenital (sternomastoid) torticollis (Q68.0)	
Q79.0	Congenital diaphragmatic hernia **Excludes 1:** congenital hiatus hernia (Q40.1)	756.6
Q79.1	Other congenital malformations of diaphragm (must specify) Absence of diaphragm Congenital malformation of diaphragm, NOS Eventration of diaphragm	756.6
Q79.2	Exomphalos Omphalocele **Excludes 1:** umbilical hernia (K42.-)	756.72
Q79.3	Gastroschisis	756.73
Q79.4	Prune belly syndrome Congenital prolapse of bladder mucosa Eagle-Barrett syndrome Early urethral obstruction sequence	756.71
Q79.5	**Other congenital malformations of abdominal wall Excludes 1:** umbilical hernia (K42.-)	
Q79.51	Congenital hernia of bladder	756.71
Q79.59	Other congenital malformations of abdominal wall (must specify) *(m) Diastasis recti* *(m) Unspecified anomaly of abdominal wall*	756.79
Q79.6	**Ehlers-Danlos syndromes** Cutis laxa hyperelastica	
Q79.60	Ehlers-Danlos syndrome, unspecified	756.83
Q79.61	Classical Ehlers-Danlos syndrome Classical EDS (cEDS)	756.83
Q79.62	Hypermobile Ehlers-Danlos syndrome Hypermobile EDS (hEDS)	756.83
Q79.63	Vascular Ehlers-Danlos syndrome	756.83

	Vascular EDS (vEDS)	
Q79.69	Other Ehlers-Danlos syndromes (must specify)	756.83
Q79.8	Other congenital malformations of musculoskeletal system (must specify) Absence of muscle Absence of pectoral muscle, congenital Absence of tendon Accessory muscle Amyoplasia, disruptive sequence Amyotrophia congenital Congenital constricting bands Congenital shortening of tendon Contracture of gastrocnemius, congenital Hereditary osteoonychodysplasia Nail-patella syndrome Poland anomalad Poland syndrome Popliteal pterygium syndrome Schwartz Jampel syndrome Waardenburg syndrome Hypoplastic Depressor Angularis Muscle (face) Other absent or hypoplastic muscle Other specified anomaly muscle, tendon, fascia, connective tissue	756.9 756.81 756.82
Q79.9	Congenital malformations of musculoskeletal system, unspecified (try not to use; must specify) Unspecified anomaly of bone Unspecified anomaly of cartilage Unspecified anomaly of connective tissue Unspecified anomaly of muscle Unspecified anomaly of musculoskeletal system Unspecified anomaly of tendon	756.9

Other Congenital Malformations (Q80-Q89)

ICD-10	Description	ICD-9
Q80	**Congenital ichthyosis Excludes 1:** Refsum's disease (G60.1)	
Q80.0	Ichthyosis vulgaris	757.1
Q80.1	X-linked ichthyosis Placental sulfatase deficiency (X-linked steryl-sulfatase deficiency) in a female	757.1
Q80.2	Lamellar ichthyosis Collodion baby	757.1
Q80.3	Congenital bullous ichthyosiform erythroderma Ichthyosiform erythroderma	757.1
Q80.4	Harlequin fetus	757.1
Q80.8	Other congenital ichthyosis	757.1
Q80.9	Congenital ichthyosis, unspecified Keratitis ichthyosis and deafness (KID) syndrome Netherton syndrome	757.1
Q81	**Epidermolysis bullosa**	
Q81.0	Epidermolysis bullosa simplex **Excludes 1:** Cockayne's syndrome (Q87.19)	757.39
Q81.1	Epidermolysis bullosa letalis Herlitz' syndrome	757.39
Q81.2	Epidermolysis bullosa dystrophica	757.39
Q81.8	Other epidermolysis bullosa	757.39
Q81.9	Epidermolysis bullosa, unspecified	757.39
Q82	**Other congenital malformations of skin** **Excludes 1:** acrodermatitis enteropathica (E83.2), congenital erythropoietic porphyria E80.0), pilonidal cyst or sinus (L05.-), Sturge-Weber (-Dimitri) syndrome (Q85.89)	
Q82.0	Hereditary lymphedema Hereditary edema of leg(s) Meige syndrome Primary congenital lymphedema Hereditary trophedema Milroy's disease	757.0
Q82.1	Xeroderma pigmentosum	757.33
Q82.2	Congenital cutaneous mastocytosis Congenital diffuse cutaneous mastocytosis Congenital maculopapular cutaneous mastocytosis Congenital urticaria pigmentosa Urticaria pigmentosa **Excludes 1:** 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)	757.33
Q82.3	Incontinentia pigmenti	757.33
Q82.4	Ectodermal dysplasia (anhidrotic) Hay Wells syndrome of ectodermal dysplasia X-linked type ectodermal dysplasia Other specified ectodermal dysplasias **Excludes 1:** Ellis-van Creveld syndrome (Q77.6)	757.31
Q82.5	*(m) Congenital non-neoplastic nevus* *(m) Birthmark, NOS* *(m) Flammeus nevus* *(m) Nevus anemicus* *(m) Portwine nevus* *(m) Sanguineous nevus* *(m) Strawberry nevus*	757.32

	(m) Vascular nevus, NOS *(m) Verrucous nevus* *(m) Angel bite or kiss* *(m) Salmon patch* *(m) Stork bites* *(m) Vascular hamartomas* **Excludes 2:** 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	*(m) Congenital sacral dimple* *(m) Parasacral dimple* **Excludes 2:** pilonidal cyst with abscess (L05.01), pilonidal cyst without abscess (L05.91)	757.39
Q82.8	Other specified congenital malformations of skin (must specify) Acrokeratosis verruciformis Benign familial pemphigus [Hailey-Hailey disease] Bloom syndrome Congenital poikiloderma Cutis laxa (hyperelastica) Goltz syndrome Goltz-Gorlin (dermal hypoplasia) syndrome Howel Evans syndrome Inherited keratosis palmaris et plantaris Keratosis follicularis [Darier-White] Absence of skin Cutis aplasia *(m) Abnormal palmar creases* *(m) Darier disease* *(m) Mongolian spot* *(m) Porokeratosis* *(m) Congenital Dermal Melanocytosis* *(m) Cutis marmorata* *(m) Cutis marmorata telangiectatica congenital (CMTC)* *(m) Depigmented or hyperpigmented spot* *(m) Extra or absent creases* *(m) Hyperpigmented areas* *(m) Sidney line* *(m) Simian creases, transverse palmar creases* *(m) Single transverse palmar crease* *Congenital scars and non-ear skin tags are not reportable* **Excludes 1:** Ehlers-Danlos syndromes (Q79.6-)	757.39
Q82.9	Congenital malformation of skin, unspecified (must specify)	757.39
Q83	**Congenital malformations of breast** **Excludes 2:** absence of pectoral muscle (Q79.8), hypoplasia of breast (N64.82), micromastia (N64.82)	
Q83.0	Congenital absence of breast with absent nipple	757.6
Q83.1	*(m) Accessory breast* *(m) Supernumerary breast*	757.6
Q83.2	Absent nipple	757.6
Q83.3	*(m) Accessory nipple (congenital)* *(m) Supernumerary nipple*	757.6
Q83.8	Other congenital malformations of breast (must specify) Congenital absence of breast Hypoplastic breast with hypoplastic nipple *(m) Asymmetry of breasts, congenital* *(m) Widely spaced nipples* *(m) Small nipple (hypoplastic)*	757.6

Q83.9	Congenital malformation of breast, unspecified (try not to use)	757.6
Q84	**Other congenital malformations of integument**	
Q84.0	*(m) Congenital alopecia* *(m) Congenital atrichosis*	757.4
Q84.1	*(m) Congenital morphological disturbances of hair, NEC* (must specify) *(m) Beaded hair* *(m) Monilethrix* *(m) Pili annulati* **Excludes 1*:*** Menkes' kinky hair syndrome (E83.09)	757.4
Q84.2	*(m) Other congenital malformations of hair (must specify)* *(m) Congenital hypertrichosis* *(m) Congenital malformation of hair, NOS* *(m) Persistent lanugo* *(m) Hirsutism* *(m) Aberrant scalp hair patterning* *(m) Low hair line* *(m) Other specified anomaly of hair*	757.4
Q84.3	*(m) Anonychia* *(m) Absent nails* **Excludes 1:** nail patella syndrome (Q87.2)	757.5
Q84.4	*(m) Congenital leukonychia*	757.5
Q84.5	*(m) Enlarged and hypertrophic nails* *(m) Congenital onychauxis* *(m) Pachyonychia* *(m) Hyperconvex or thickened toenails*	757.5
Q84.6	Hypoplastic, hyperconvex, or duplicated fingernails *(m) Other congenital malformations of nails (must specify)* *(m) Congenital clubnail* *(m) Congenital koilonychias* *(m) Congenital malformations of nail, NOS* *(m) Hypoplastic (small) fingernails and/or toenails*	757.5
Q84.8	Other specified congenital malformations of integument (must specify) Aplasia cutis congenital *(m) Scalp defects*	757.8
Q84.9	*(m) Congenital malformation of integument, unspecified* *(m) Congenital anomaly of integument, NOS* *(m) Congenital deformity of integument, NOS* *(m) Unspecified anomaly of hair* *(m) Unspecified anomaly of nail*	757.9
Q85	**Phakomatoses, NEC** **Excludes 1:** ataxia telangiectasia [Louis-Bar] (G11.3), familial dysautonomia [Riley-Day] (G90.1)	
Q85.0	**Neurofibromatosis (nonmalignant)**	
Q85.00	Neurofibromatosis, unspecified	237.70
Q85.01	Neurofibromatosis, Type 1 Von Recklinghausen disease	237.71
Q85.02	Neurofibromatosis, Type 2 Acoustic neurofibromatosis	237.72
Q85.03	Schwannomatosis	237.73
Q85.09	Other neurofibromatosis	237.79
Q85.1	Tuberous sclerosis Bourneville's disease Epiloia	759.5
Q85.8	**Other phakomatoses, not elsewhere classified**	
Q85.81	PTEN hamartoma tumor syndrome PHTS PTEN related Cowden syndrome	

Q85.82	Other Cowden syndrome	759.6
Q85.83	Von Hippel-Lindau syndrome	759.6
Q85.89	Other phakomatoses, NEC (must specify) Peutz-Jeghers Syndrome Sturdge-Weber (-Dimitri) syndrome Neuro-cutaneous syndrome	759.6
Q85.9	Phakomatosis, unspecified Hamartosis, NOS Hamartoma of retina Vascular hamartoma of skin	759.6
Q86	**Congenital malformations syndromes due to known exogenous causes, NEC** **Excludes 2:** iodine-deficiency-related hypothyroidism (E00-E02), nonteratogenic effects of substances transmitted via placenta or breast milk (P04.-)	
Q86.0	Fetal alcohol syndrome (dysmorphic) (reportable to age 10)	760.71
Q86.1	Fetal hydantoin syndrome Meadow's syndrome	760.77
Q86.2	Dysmorphism due to warfarin	760.79
Q86.8	Other congenital malformation syndromes due to known exogenous causes (must specify)	760.76
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 (must specify) Acrocephalopolysyndactyly Acrocephalosyndactyly [Apert syndrome] Branchio oto renal syndrome [Melnick-Fraser syndrome] Carpenter's syndrome Cryptophthalmos syndrome Cyclopia Freeman Sheldon syndrome Goldenhar syndrome (oculo-auriculovertebral) Gorlin-Chaudhry-Moss syndrome Hallerman-Streiff syndrome Moebius syndrome Oro-facial-digital syndrome Oto palato digital (OPD) syndrome Pfeiffer syndrome Pierre Robin syndrome Saethre Chotzen syndrome Whistling face Antley-Bixler Branchio-oculo-facial syndrome	755.55 756.0 759.89
Q87.1	Congenital malformation syndromes predominantly associated w/ short stature (must specify)	
Q87.11	Prader-Willi syndrome	759.81
Q87.19	Other congenital malformation syndromes predominantly associated w/ short stature (must specify) Aarskog syndrome Bonnevie-Ullrich syndrome Cornelia de Lange syndrome Cockayne syndrome De Lange syndrome Dubowitz syndrome Noonan syndrome Robinow-Silverman-Smith syndrome Russell-Silver syndrome Seckel syndrome	759.89

	Sjogren-Larsson syndrome	
Q87.2	Congenital malformation syndromes predominantly involving limbs (must specify) Holt-Oram syndrome Klippel-Trenaunay-Weber syndrome Nail patella syndrome Rubinstein-Taybi syndrome Sirenomelia syndrome Thrombocytopenia with absent radius [TAR] syndrome VATER syndrome	759.89
Q87.3	Congenital malformation syndromes involving early overgrowth (must specify) Beckwith-Wiedemann syndrome Proteus syndrome Sotos syndrome Weaver syndrome	759.89
Q87.4	**Marfan syndrome**	
Q87.40	Marfan syndrome, unspecified	759.82
Q87.41	**Marfan syndrome with cardiovascular manifestations**	
Q87.410	Marfan syndrome with aortic dilation	759.82
Q87.418	Marfan syndrome with other cardiovascular manifestations	759.82
Q87.42	Marfan syndrome with ocular manifestations	759.82
Q87.43	Marfan syndrome with skeletal manifestation	759.82
Q87.5	Other congenital malformation syndromes with other skeletal changes (must specify)	759.89
Q87.8	**Other specified congenital malformation syndromes, NEC** **Excludes 1:** Zellweger syndrome (E71.510)	
Q87.81	Alport syndrome Hereditary nephritis	759.89
Q87.82	Arterial tortuosity syndrome	759.89
Q87.83	Bardet-Biedl syndrome	
Q87.84	Laurence-Moon syndrome	
Q87.85	MED 13L syndrome Asadollahi-Rauch syndrome Mediator complex subunit 13L syndrome	
Q87.89	Other specified congenital malformation syndromes, NEC (must specify) Basal cell nevus syndrome Gorlin syndrome	759.89
Q89	**Other congenital malformations, NEC**	
Q89.0	**Congenital absence and malformations of spleen** **Excludes 1:** isomerism of atrial appendages (with asplenia or polysplenia) (Q20.6)	
Q89.01	Asplenia (congenital)	759.0
Q89.09	Congenital malformations of spleen (must specify) Congenital splenomegaly	759.0
Q89.1	Congenital malformations of adrenal gland **Excludes 1:** adrenogenital disorders (E25.-), congenital adrenal hyperplasia (E25.0)	759.1
Q89.2	Congenital malformations of other endocrine glands Congenital malformation of parathyroid or thyroid gland Persistent thyroglossal duct Thyroglossal cyst **Excludes 1:** congenital goiter (E03.0), congenital hypothyroidism (E03.1)	759.2
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	759.3

	Excludes 1: dextrocardia NOS (Q24.0)	
Q89.4	Conjoined twins Craniopagus Cicephaly Pygopagus Thoracopagus	759.4
Q89.7	Multiple congenital malformations, NEC (try not to code) Multiple congenital anomalies, NOS Multiple congenital deformities, NOS **Excludes 1:** congenital malformation syndromes affecting multiple systems (Q87.-)	759.7
Q89.8	Other specified congenital malformations (must specify) Charge syndrome Kabuki syndrome Stickler syndrome Williams syndrome Use additional code(s) to identify all associated manifestations	759.89
Q89.9	Congenital malformation, unspecified (try not to code; must specify)	759.9

Chromosomal Abnormalities, Not Elsewhere Classified (NEC) (Q90-Q99)

**** Please report karyotype (chromosome test results) in addition to the ICD code!**

ICD-10	Description	ICD-9
Q90	**Trisomy 21, Down Syndrome** Code also associated physical condition(s), such as atrioventricular spetal defect (Q21.2)	
Q90.0	Trisomy 21, nonmosaicism (meiotic nondisjunction) Karyotype trisomy 21 (47,XX or XY,+21)	758.0
Q90.1	Trisomy 21, mosaicism (mitotic nondisjunction) (46,XX/47,XX+21 or 46,XY/47,XY+21)	758.0
Q90.2	Trisomy 21, translocation t(13:21), t(14:21), t(15:21), t(21:22) Translocation trisomy duplication of a 21[t(21:21)]	758.0
Q90.9	Down syndrome, unspecified (must specify) Trisomy 21, NOS	758.0
Q91	**Trisomy 18 and Trisomy 13**	
Q91.0	Trisomy 18, nonmosaicism (meiotic nondisjunction) Edwards syndrome, karyotype trisomy 18(47,XX or XY+18)	758.2
Q91.1	Trisomy 18, mosaicism (mitotic nondisjunction) (46,XX/47,XX+18 or 46,XY/47,XY+18)	758.2
Q91.2	Trisomy 18, translocation	758.2
Q91.3	Trisomy 18, unspecified Trisomy 18, complete Edwards syndrome, NOS	758.2
Q91.4	Trisomy 13, nonmosaicism (meiotic nondisjunction) Patau syndrome, karyotype trisomy 13(47,XX or XY,+13)	758.1
Q91.5	Trisomy 13, mosaicism nondisjunction) (46,XX/47, XX+13 or 46,XY/47,XY+13)	758.1
Q91.6	Trisomy 13, (translocation) t(13:14), t(13:21), t(13:15), t(13:22), t(13:13)	758.1
Q91.7	Trisomy 13, unspecified Patau syndrome, NOS	758.1
Q92	**Other trisomies and partial trisomies of the autosome, NEC** **Includes:** unbalanced transolocations and insertions **Excludes 1:** trisomies of chromosomes 13, 18, 21 (Q90-Q91)	
Q92.0	Whole chromosomes trisomy, nonmosaicism (meiotic nondisjunction) • Trisomy 1 • Trisomy 2 • Trisomy 3 • Trisomy 4 • Trisomy 5 • Trisomy 6 • Trisomy 7 • Trisomy 8 • Trisomy 9 • Trisomy 10 • Trisomy 11 • Trisomy 12 • Trisomy 14 • Trisomy 15 • Trisomy 16 • Trisomy 17 • Trisomy 19 • Trisomy 20 • Trisomy 22	758.5
Q92.1	Whole chromosome trisomy, mosaicism (mitotic nondisjunction) Mosaic trisomy of any chromosome except 21, 13 or 18	758.5
Q92.2	Partial trisomy Less than whole arm duplicated Whole arm or more duplicated **Excludes 1:** partial trisomy due to unbalanced translocation (Q92.5)	758.2
Q92.5	Duplications with other complex rearrangements Partial trisomy due to unbalanced translocations Partial trisomy syndromes (unbalanced, not defined) **Also code any associated deletions due to unbalanced translocations, inversions and insertions	758.5
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	758.5
Q92.62	Marker chromosomes in abnormal individual	758.5
Q92.7	Triploidy and polyploidy Triploidy, 69 chromosomes Other Polyploidy (96 chromosomes or other multiples of 23, except 46,69)	758.5
Q92.8	Other specified trisomies and partial trisomies of autosomes (chromosomes 1-22) (must specify) Duplication identified by fluorescence in situ hybridization (FISH) Duplications identified by in situ hybridization (ISH) Duplications seen only at prometaphase	758.5
Q92.9	Trisomy and partial trisomy of autosomes, unspecified Trisomy, NOS	758.5
Q93	**Monosomies and deletions from the autosomes, NEC**	
Q93.0	Whole chromosomes monosomy, nonmosaicism (meiotic nondisjunction) E.g. (45, XX, -21) or (45,XY,-21) for any chromosome #1-22	758.5
Q93.1	Whole chromosome monosomy, mosaicism (mitotic nondisjunction) Monosomy 21 mosaicism: e.g.: (46,XX/45,XX,-21) or (46,XY/45,XY,-21) Monosomy of other whole chromosomes #1-22 in mosaic form	758.5
Q93.2	Chromosome replaced with ring, dicentric or isochromosomes	758.5
Q93.3	Deletion of short arm of chromosome 4 4p deletion syndrome Del4p15 Deletion 4p15-16 Wolff-Hirschorn syndrome	758.39
Q93.4	Deletion of short arm of chromosome 5 Cri-du-chat syndrome (5p-)	758.31
Q93.5	**Other deletions of part of a chromosome**	
Q93.51	Angelman syndrome	
Q93.52	Phelan-Dermid syndrome 22q13.3 deletion syndrome	
Q93.59	Other deletions of part of a chromosome Antimongolism syndrome: partial or total deletion of 21 [del (21)] Deletion of long arm 13: (del(13q)) or 13q- Deletion of long arm 17: (del(17q)) or 17q- Deletion of long arm 18: (del(18q)) or 18q- Deletion of short arm 17: (del(17p)) or 17p- Deletion of short arm 18: (del(18p)) or 18p-	
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	758.39
Q93.8	**Other deletions from the autosomes**	
Q93.81	Velo-cardio-facial syndrome Deletion 22q11.2	758.32
Q93.82	Williams syndrome	
Q93.88	Other microdeletions (must specify) Miller-Dieker syndrome Smith-Magenis syndrome	758.33
Q93.89	Other deletions from the autosomes (chromosomes 1-22) (must specify) Deletions identified by fluorescence in situ hybridization (FISH) Deletions identified by in situ hybridization (IS) Deletions seen only at prometaphase	758.39

Q93.9	Deletion from autosomes, unspecified (try not to use) Autosomal deletion syndrome Chromosomal deletion	758.39
Q95	**Balanced rearrangements and structural markers, NEC** **Includes:** Robertsonian and balanced reciprocal translocations and insertions	
Q95.0	Balanced translocation and insertion in normal individual	758.4
Q95.1	Chromosome inversion in normal individual	758.4
Q95.2	Balanced autosomal rearrangement in abnormal individual	758.5
Q95.3	Balanced sex/autosomal rearrangement in abnormal individual	758.5
Q95.5	Individual with autosomal fragile site Autosomal fragile site [fra (_)]	758.4
Q95.8	Other balanced rearrangements and structural markers (must specify)	758.4
Q95.9	Balanced rearrangement and structural marker, unspecified	758.4
Q96	**Turner's syndrome** **XO syndrome** **Bonnevie-Ullrich syndrome** **[Only applicable to female patients.]** **Excludes 1:** Noonan syndrome (Q87.19), pure gonadal dysgenesis (Q99.1)	
Q96.0	Karyotype 45,X Turner's phenotype, karyotype 45,X	758.6
Q96.1	Karyotype 46, X iso (Xq) Karyotype 46, isochromosome Xq	758.6
Q96.2	Karyotype 46, X with abnormal sex chromosome, except iso (Xq) Karyotype 46, X with abnormal sex chromosome, except isochromosome Xq Turner syndrome variant, [46,X,r(X)] Turner syndrome variant, [46,Xdel(Xq)] Turner syndrome variant, [46,X,del(Xp)] **[Only applicable to female patients.]**	758.6
Q96.3	Mosaicism, 45, X/46, XX or XY **[Only applicable to female patients.]**	758.6
Q96.4	Mosaicism, 45, X/other cell line(s) with abnormal sex chromosome **[Only applicable to female patients.]**	756.6
Q96.8	Other variants of Turner's syndrome	758.6
Q96.9	Turner's syndrome, unspecified Turner Syndrome, karyotype unspecified	758.6
Q97	**Other sex chromosome abnormalities, female phenotype, NEC** **[Only applicable to female patients.]** **Excludes 1:** Turner's syndrome (Q96.-)	
Q97.0	Karyotype 47, XXX	758.81
Q97.1	Female with more than three X chromosomes	758.81
Q97.2	Mosaicism, lines with various numbers of X chromosomes	758.81
Q97.3	Female with 46, XY karyotype	758.81
Q97.8	Other specified sex chromosome abnormalities, female phenotype (must specify)	758.81
Q97.9	Sex chromosome abnormalities, female phenotype unspecified	758.81
Q98	**Other sex chromosome abnormalities, male phenotype, NEC** **[Only applicable to male patients.]**	
Q98.0	Klinefelter syndrome karyotype 47, XXY	758.7
Q98.1	Klinefelter syndrome, male with more than two X chromosome 48,XXXY 48,XXYY 49,XXXXY	758.7
Q98.3	Other male with 46, XX karyotype	758.7
Q98.4	Klinefelter syndrome, unspecified	758.7
Q98.5	Karyotype 47, XYY	758.81
Q98.6	Male with structurally abnormal sex chromosome	758.81
Q98.7	Male with sex chromosome mosaicism	758.81

	Klinefelter syndrome mosaic (46,XY/47,XXY) Mosaic X/XY, 45X/46XY Mosaic XY/XXY, 46XY/47XXY Mosaic including XXXXY,49XXXXY Mosaic XYY male[46,XY/47XYY] Sex chromosomes mosaicism	
Q98.8	Other specified sex chromosome abnormalities, male phenotype (must specify)	758.81
Q98.9	Sex chromosome abnormality, male phenotype unspecified	758.81
Q99	**Other chromosome abnormalities, NEC**	
Q99.0	Chimera 46, XX/46, XY Chimera 46, XX/46, XY true hermaphrodite	758.81
Q99.1	46, XX true hermaphrodite 46, XX with streak gonads 46, XY with streak gonads Pure gonadal dysgenesis	758.81
Q99.2	Fragile X chromosome Fragile X syndrome	759.83
Q99.8	Other specified chromosome abnormalities (must specify) Accessory autosomes, NOS Additional chromosome(s), NOS Deletion of chromosome(s), NOS Derivative chromosomes, NOS [not X or Y;der (_)] Double minute chromosomes (+dmidn) Duplication of chromosome(s), NOS Insertion, autosome [inv (_)] Inversion, autosome [inv (_)] Isochromosome, autosome [I (_)] Mosaicism, NOS Other specified anomaly of autosomes, includes satellites, stalks, excess Unspecified sex chromosome anomaly Variable region, autosome [var (_)]	758.89
Q99.9	Chromosomal abnormality, unspecified (must specify) (try not to use)	758.9

Other Selected Reportable Conditions

ICD-10	Notes & Description	ICD-9
A50	**Congenital syphilis**	
A50.0	Early congenital syphilis, symptomatic	
A50.01	Early congenital syphilitic oculopathy	
A50.02	Early congenital syphilitic osteochondropathy	
A50.03	Early congenital syphilitic pharyngitis	
A50.04	Early congenital syphilitic pneumonia	
A50.05	Early congenital syphilitic rhinitis	
A50.06	Early cutaneous congenital syphilis	
A50.07	Early mucocutaneous congenital syphilis	
A50.08	Early visceral congenital syphilis	
A50.09	Other early congenital syphilis, symptomatic	
A50.1	Early congenital syphilis, latent	
A50.2	Early congenital syphilis, unspecified	
A50.3	Late congenital syphilitic oculopathy	
A50.30	Late congenital syphilitic oculopathy, unspecified	
A50.31	Late congenital syphilitic interstitial keratitis	
A50.32	Late congenital syphilitic chorioretinitis	
A50.39	Other late congenital syphilitic oculopathy	
A50.4	Late congenital neurosyphilis [juvenile neurosyphilis]	
A50.40	Late congenital neurosyphilis, unspecified	
A50.41	Late congenital syphilitic meningitis	
A50.42	Late congenital syphilitic encephalitis	
A50.43	Late congenital syphilitic polyneuropathy	
A50.44	Late congenital syphilitic optic nerve atrophy	
A50.45	Juvenile general paresis Dementia paralytica juvenilis	
A50.49	Other late congenital neurosyphilis Juvenile tabes dorsalis	
A50.5	Other late congenital syphilis, symptomatic	
A50.51	Clutton's joints	
A50.52	Hutchinson's teeth	
A50.53	Hutchinson's triad	
A50.54	Late congenital cardiovascular syphilis	
A50.55	Late congenital syphilitic arthropathy	
A50.56	Late congenital syphilitic osteochondropathy	
A50.57	Syphilitic saddle nose	
A50.59	Other late congenital syphilis, symptomatic	
A50.6	Late congenital syphilis, latent	
A50.7	Late congenital syphilis, unspecified	
A50.9	Congenital syphilis, unspecified	
D82	**Immunodeficiency associated with other major defects** **Excludes 1:** ataxia telangiectasia [Louis-Bar] (G11.3)	
D82.1	Di George's syndrome Also code 22q deletion separately if reported Pharyngeal pouch syndrome Thymic alymphoplasia Thymic aplasia or hypoplasia with immunodeficiency	279.11
E25	**Adrenogenital disorders** Please report cytogenetic diagnosis **Includes:** adrenogenital syndromes, virilizing or feminizing, whether acquired or due to adrenal hyperplasia consequent on inborn enzyme defects in hormone synthesis, Female adrenal pseudohermaphroditism, Female heterosexual precocious pseudopuberty, Male isosexual	

	precocious pseudopuberty, Male macrogenitosomia praecox, Male sexual precocity with adrenal hyperplasia, Male virilization (female) **Excludes 1:** indeterminate sex and pseudohermaphroditism (Q56), chromosomal abnormalities (Q90-Q99)	
E25.0	Congenital adrenogenital disorders associated with enzyme deficiency Congenital adrenal hyperplasia (CAH) 21-Hydroxylase deficiency Salt-losing congenital adrenal hyperplasia	255.2
E25.8	Other adrenogenital disorders idiopathic adrenogenital disorder	255.2
E25.9	Adrenogenital disorder, unspecified Adrenogenital syndrome NOS	255.2
E34	**Other endocrine disorders ** only report if congenital or genetic** **Excludes 1:** pseudohypoparathyroidism (E20.1)	
E34.3	**Short stature due to endocrine disorder**	
E34.30	Short stature due to endocrine disorder, unspecified (must specify)	259.4
E34.31	Constitutional short stature Constitutional delay of growth, puberty, or maturation	259.4
E34.32	**Genetic causes of short stature**	
E34.321	Primary insulin-like growth factor-1 (IGF-1) deficiency Acid-labile subunit gene (IGFALS) defect Growth hormone gene 1 (GH1) defect with growth hormone neutralizing antibodies Growth hormone insensitivity syndrome (GHIS) Insulin-like growth factor 1 gene (IGF1) defect Laron type short stature Severe primary insulin-like growth factor-1 deficiency (SPIGFD) Signal transducer and activator of transcription 5B gene (STAT5b) defect	259.4
E34.322	Insulin-like growth factor-1 (IGF-1) resistance Genetic syndrome with resistance to insulin-like growth factor-1 Insulin-like growth factor-1 receptor (IGF-1R) defect Post-insulin-like growth factor-1 receptor signaling defect	259.4
E34.328	Other genetic causes of short stature Short stature due to ACAN gene variant Short stature due to aggrecan deficiency Short stature due to NPR-2 gene variant	259.4
E34.329	Unspecified genetic causes of short stature	259.4
E34.39	Other short stature due to endocrine disorder	259.4
E34.5	**Androgen insensitivity syndrome**	
E34.50	Androgen insensitivity syndrome, unspecified	259.5
E34.51	Complete androgen insensitivity syndrome Complete androgen insensitivity de Quervain syndrome Goldberg-Maxwell syndrome Testicular feminization	259.5
E34.52	Partial androgen insensitivity syndrome Partial androgen insensitivity Reifenstein syndrome	259.5
E78	Disorders of lipoprotein metabolism and other lipidemias **Excludes 1:** sphingolipidosis (E75.0-E75.3)	
E78.72	Smith-Lemli-Opitz syndrome	759.89
G12	**Spinal muscular atrophy and related syndromes**	
G12.0	Infantile spinal muscular atrophy, type I Werdnig-Hoffman disease	335.0
G12.1	Other inherited spinal muscular atrophy (must specify) Adult form spinal muscular atrophy Childhood form, type II spinal muscular atrophy	335.11

	Distal spinal muscular atrophy Juvenile form, type III spinal muscular atrophy [Kugelberg-Welander] Progressive bulbar palsy of childhood [Fazio-Londe] Scapuloperoneal form spinal muscular atrophy	
G12.9	Spinal muscular atrophy, unspecified	335.10
G60	**Hereditary and idiopathic neuropathy**	
G60.0	Hereditary motor and sensory neuropathy Charcot-Marie-Tooth disease Déjérine-Sottas disease Hereditary motor and sensory neuropathy, types I-IV Hypertrophic neuropathy of infancy Peroneal muscular atrophy (axonal type) (hypertrophic type) Roussy-Levy syndrome	356.0
G60.1	Refsum disease Infantile Refsum disease	356.3
G60.2	Neuropathy in association with hereditary ataxia	356.0
G71	**Primary disorders of muscles** **Excludes 2:** arthrogryposis multiplex congenita (Q74.3), metabolic disorders (E70-E88), myositis (M60.-)	
G71.0	**Muscular dystrophy**	
G71.00	Muscular dystrophy, unspecified (must specify) (reportable to age 10)	359.1
G71.01	Duchenne or Becker muscular dystrophy (reportable to age 10) Autosomal recessive, childhood type, muscular dystrophy resembling Duchenne or Becker muscular dystrophy Benign [Becker] muscular dystrophy Severe [Duchenne] muscular dystrophy	359.1
G71.02	Facioscapulohumeral muscular dystrophy (reportable to age 10) Scapulohumeral muscular dystrophy	359.1
G71.03	**Limb girdle muscular dystrophies** (reportable to age 10)	
G71.031	Autosomal dominant limb girdle muscular dystrophy (reportable to age 10) LGMD D4 calpain-3-related LGMD D5 collagen 6-related Limb girdle muscular dystrophy type 1	
G71.032	Autosomal recessive limb girdle muscular dystrophy due to calpain-3 dysfunction (reportable to age 10) Limb girdle muscular dystrophy type 2A LGMD R1 calpain-3-related Primary calpainopathy	
G71.033	Limb girdle muscular dystrophy due to dysferlin dysfunction (reportable to age 10) Dysferlinopathy LGMD R2 dysferlin-related Limb girdle muscular dystrophy type 2B Miyoshi Myopathy type 1	
G71.034	**Limb girdle muscular dystrophy due to sarcoglycan dysfunction** (reportable to age 10)	
G71.0340	Limb girdle muscular dystrophy due to sarcoglycan dysfunction, unspecified (reportable to age 10) Sarcoglycanopathy, NOS	
G71.0341	Limb girdle muscular dystrophy due to alpha sarcoglycan dysfunction (reportable to age 10) Alpha sarcoglycanopathy Limb-girdle muscular dystrophy due to alpha-sarcoglycan deficiency Limb girdle muscular dystrophy type 2D	
G71.0342	Limb girdle muscular dystrophy due to beta sarcoglycan dysfunction (reportable to age 10) Beta sarcoglycanopathy	

	Limb girdle muscular dystrophy due to beta-sarcoglycan deficiency Limb girdle muscular dystrophy type 2E	
G71.0349	Limb girdle muscular dystrophy due to other sarcoglycan dysfunction (reportable to age 10) Delta sarcoglycanopathy Delta-sarcoglycan-related LGMD R6 Gamma sarcoglycanopathy Gamma-sarcoglycan-related LGMD R5 Limb girdle muscular dystrophy type 2C Limb girdle muscular dystrophy type 2F	
G71.035	Limb girdle muscular dystrophy due to anoctamin-5 dysfunction (reportable to age 10) Anoctamin-5-related LGMD R12 Anoctaminopathy Autosomal recessive limb girdle muscular dystrophy type 2L Miyoshi myopathy type 3	
G71.038	Other limb girdle muscular dystrophy (reportable to age 10) LGMD R9 FKRP-related LGMD R22 collagen 6-related Limb girdle muscular dystrophy due to fukutin related protein dysfunction Limb girdle muscular dystrophy type 2I Other autosomal recessive limb girdle muscular dystrophy	
G71.039	Limb girdle muscular dystrophy, unspecified (reportable to age 10)	
G71.09	Other specified muscular dystrophies (must specify) (reportable to age 10) Benign scapuloperoneal muscular dystrophy with early contractures [Emery-Dreifuss] Congenital muscular dystrophy NOS Congenital muscular dystrophy with specific morphological abnormalities of the muscle fiber Distal muscular dystrophy Ocular muscular dystrophy Oculopharyngeal muscular dystrophy Scapuloperoneal muscular dystrophy	359.1
G71.1	**Myotonic disorders**	
G71.11	Myotonic muscular dystrophy (reportable to age 10) Dystrophia myotonica [Steinert] Myotonia atrophica Myotonic dystrophy Proximal myotonic myopathy (PROMM) Steinert disease	359.21
G71.12	Myotonia congenita Acetazolamide responsive myotonia congenita Dominant myotonia congenita [Thomsen disease] Myotonia levior Recessive myotonia congenita [Becker disease]	359.22
G71.13	Myotonic chondrodystrophy Chondrodystrophic myotonia Congenital myotonic chondrodystrophy Schwartz-Jampel disease	359.23
G71.19	Other specified myotonic disorders (must specify) Isaac-Mertens syndrome Myotonia fluctuans Myotonia permanens Neuromyotonia [Isaacs] Paramyotonia congenita (of von Eulenburg) Pseudomyotonia Symptomatic myotonia	359.29
G71.2	**Congenital myopathies Excludes 1**: arthrogryposis multiplex congenita (Q74.3)	359.0

G71.20	Congenital myopathy, unspecified Fiber-type disproportion	
G71.21	Nemaline myopathy	
G71.22	**Centronuclear myopathy**	
G71.220	X-linked myotubular myopathy Myotubular (centronuclear) myopathy	
G71.228	Other centronuclear myopathy Autosomal centronuclear myopathy Autosomal dominant centronuclear myopathy Autosomal recessive centronuclear myopathy Centronuclear myopathy, NOS	
G71.29	Other congenital myopathy (must specify) Central core disease Minicore disease Multicore disease Multiminicore disease	
G71.3	Mitochondrial myopathy, not elsewhere classified **Excludes 1**: Kearns-Sayre syndrome (H49.81), Leber's disease (H47.21), Leigh's encephalopathy (G31.82), mitochondrial metabolism disorders (E88.4.-), Reye's syndrome (G93.7)	359.89
G71.8	Other primary disorders of muscles Congenital hemihypertrophy	359.89
G71.9	Primary disorder of muscle, unspecified Hereditary myopathy, NOS	359.9
G80	**Cerebral Palsy is Not Reportable**	
K40	**Inguinal Hernia (report for female only)** **Includes:** bubonocele, direct inguinal hernia, double inguinal hernia, indirect inguinal hernia, inguinal hernia NOS, oblique inguinal hernia, scrotal hernia	
K40.00	Bilateral inguinal hernia, with obstruction, without gangrene, not specified as recurrent Bilateral inguinal hernia, with obstruction, without gangrene, NOS	550.12
K40.1	**Bilateral inguinal hernia, with gangrene (report for female only)**	
K40.10	Bilateral inguinal hernia, with gangrene, not specified as recurrent Bilateral inguinal hernia, with gangrene, NOS	550.02
K40.2	**Bilateral inguinal hernia, without obstruction or gangrene (female only)**	
K40.20	Bilateral inguinal hernia, without obstruction or gangrene, not specified as recurrent Bilateral inguinal hernia, NOS	550.92
K40.3	**Unilateral inguinal hernia, with obstruction, without gangrene (female only)** • Inguinal hernia (unilateral) causing obstruction without gangrene • Incarcerated inguinal hernia (unilateral) without gangrene • Irreducible inguinal hernia (unilateral) without gangrene • Strangulated inguinal hernia (unilateral) without gangrene Indicate right, left, or unspecified	
K40.30	Unilateral inguinal hernia, with obstruction, without gangrene, not specified as recurrent Inguinal hernia, with obstruction, NOS Unilateral inguinal hernia, with obstruction, without gangrene, NOS	550.1
K40.40	Unilateral inguinal hernia, with gangrene, not specified as recurrent Inguinal hernia with gangrene, NOS Unilateral inguinal hernia with gangrene, NOS	550
K40.9	**Unilateral inguinal hernia, without obstruction or gangrene (female only)** Indicate right, left, or unspecified	
K40.90	Unilateral inguinal hernia, without obstruction or gangrene, not specified as recurrent Direct inguinal hernia Indirect inguinal hernia Inguinal hernia, NOS Left direct inguinal hernia Left indirect inguinal hernia	550.9

	Left inguinal hernia Right direct inguinal hernia Right indirect inguinal hernia Right inguinal hernia	
K41	**Femoral hernia (female only)**	
K41.0	**Bilateral femoral hernia, with obstruction, without gangrene (female only)** • Femoral hernia (bilateral) causing obstruction, without gangrene • Incarcerated femoral hernia (bilateral), without gangrene • Irreducible femoral hernia (bilateral), without gangrene • Strangulated femoral hernia (bilateral), without gangrene	
K41.00	Bilateral femoral hernia, with obstruction, without gangrene, not specified as recurrent Bilateral femoral hernia, with obstruction, without gangrene, NOS	552.02
K41.1	**Bilateral femoral hernia, with gangrene (female only)**	
K41.10	Bilateral femoral hernia, with gangrene, not specified as recurrent Bilateral femoral hernia, with gangrene, NOS	551.02
K41.2	**Bilateral femoral hernia, without obstruction or gangrene (female only)**	
K41.20	Bilateral femoral hernia, without obstruction or gangrene, not specified as recurrent Bilateral femoral hernia, NOS	553.02
K41.3	**Unilateral femoral hernia, with obstruction, without gangrene (female only)** • Femoral hernia (unilateral) causing obstruction, without gangrene • Incarcerated femoral hernia (unilateral), without gangrene • Irreducible femoral hernia (unilateral), without gangrene • Strangulated femoral hernia (unilateral), without gangrene Indicate right, left, or unspecified	
K41.30	Unilateral femoral hernia, with obstruction, without gangrene, not specified as recurrent Femoral hernia, with obstruction NOS Unilateral femoral hernia, with obstruction NOS	552
K41.4	**Unilateral femoral hernia, with gangrene (female only)** Indicate right, left, or unspecified	
K41.40	Unilateral femoral hernia, with gangrene, not specified as recurrent Femoral hernia, with gangrene NOS Unilateral femoral hernia, with gangrene NOS	551
K41.9	**Unilateral femoral hernia, without obstruction or gangrene (female only)** Indicate right, left, or unspecified	
K41.90	Unilateral femoral hernia, without obstruction or gangrene, not specified as recurrent Femoral hernia, NOS Unilateral femoral hernia, NOS	553
L05	***(m) Pilonidal cyst and sinus***	
L05.9	***(m) Pilonidal cyst and sinus without abscess***	
L05.91	*(m) Pilonidal cyst without abscess* *(m) Pilonidal dimple* *(m) Pilonidal cyst NOS* *(m) Postanal dimple* **Excludes 2:** congenital sacral dimple (Q82.6), parasacral dimple (Q82.6)	685.1
L05.92	*(m) Pilonidal sinus without abscess* *(m) Coccygeal fistula* *(m) Coccygeal sinus without abscess* *(m) Pilonidal fistula*	685.1
L81	***(m) Other disorders of pigmentation*** **Excludes 1:** birthmark NOS (Q82.5), Peutz-Jeghers syndrome (Q85.89) **Excludes 2:** nevus - see Alphabetical Index	
L81.3	*(m) Café au lait spots*	709.09
L81.4	*(m) Other melanin hyperpigmentation*	709.09
L81.9	*(m) Disorder of pigmentation, unspecified*	709
M26	***(m) Dentofacial anomalies [including malocclusion]***	
M26.0	***(m) Major anomalies of jaw size***	

	Excludes 1: acromegaly (E22.0), Robin's syndrome (Q87.0)	
M26.01	*(m) Maxillary hyperplasia* *(m) Prominent maxilla*	524.01
M26.02	*(m) Maxillary hypoplasia*	524.03
M26.03	*(m) Mandibular hyperplasia*	524.02
M26.04	*(m) Mandibular hypoplasia*	524.04
M26.05	*(m) Macrogenia*	524.05
M26.06	*(m) Microgenia*	524.06
M26.09	Micrognathia Macrognathia *(m) Other specified anomalies of jaw (specify)*	524.09
M26.1	**Anomalies of jaw-cranial base relationship**	
M26.19	Other specified anomalies of jaw-cranial base relationship (must specify) Retrognathia *(m) Congenital prognathism* *(m) Mandibular prognathism* *(m) Maxillary retrognathia* *(m) Maxillary retrognathism*	524.19
P02	**Newborn (suspected to be) affected by complications of placenta, cord and membranes** **Includes:** conditions that have their origin in the fetal or perinatal period (before birth through the first 28 days after birth) even if morbidity occurs later Code first any current condition in newborn	
P02.8	Newborn (suspected to be) affected by other abnormalities of membranes Amniotic band syndrome Amniotic bands (constricting bands) Streeter's (Simonart's) bands P02.8 is only applicable to newborns of age 0 years.	762.8
P35	**Congenital viral diseases** **Includes:** infections acquired in utero or during birth Only applicable to newborns of age 0 years.	
P35.0	Congenital Rubella Syndrome Rubella, congenital Congenital rubella pneumonitis	771.0
P35.1	Congenital cytomegalovirus infection (CMV) Cytomegalovirus infection, congenital	771.1
P35.2	Congenital herpesviral [herpes simplex] infection Herpes simplex Encephalitis herpes Meningoencephalitis herpes	771.2
P35.3	Congenital viral hepatitis	771.2
P35.4	Congenital Zika virus disease **Use Additional Code** to identify manifestations of congenital Zika virus disease	
P35.8	Congenital infection, other specified Congenital varicella [chickenpox]	771.2
P35.9	Congenital viral disease, unspecified	771.2
P37	**Other congenital infectious and parasitic diseases** **Excludes 2**: congenital syphilis (A50.), infectious neonatal diarrhea (A00-A09), necrotizing enterocolitis in newborn (P77.-), noninfectious neonatal diarrhea (P78.3), ophthalmia neonatorum due to gonococcus (A54.31), tetanus neonatorum (A33)	
P37.0	Congenital tuberculosis P37.0 is only applicable to newborns of age 0 years.	771.2
P37.1	Congenital toxoplasmosis Hydrocephalus due to congenital toxoplasmosis P37.1 is only applicable to newborns of age 0 years.	771.2
P96	Other conditions originating in the perinatal period	
P96.1	Neonatal withdrawal symptoms from maternal use of drugs of addiction	

Stillbirth Codes (O36.4, Z37)

ICD-10	Notes & Description	ICD-9
O36.4	**Maternal care for intrauterine death** • **Maternal care for intrauterine fetal death NOS** • **Maternal care for intrauterine fetal death after completion of 20 weeks of gestation** • **Maternal care for late fetal death** • **Maternal care for missed delivery** **Only applicable to female maternity patients aged 12 - 55 years inclusive.** **Should only be used on the maternal record - not on the newborn record.** **Use a code under Z3A to document the exact week during the pregnancy.** **Excludes 1:** missed abortion (O02.1), stillbirth (P95)	
O36.4XX0	Maternal care for intrauterine death, not applicable or unspecified Dead fetus in utero Fetal death from asphyxia and/or anoxia, not clear if noted before or after onset of labor Fetal death from asphyxia or anoxia Pregnancy loss	656.40 656.41 656.43
O36.4XX1	Maternal care for intrauterine death, fetus 1	656.41 656.43
O36.4XX2	Maternal care for intrauterine death, fetus 2	656.41 656.43
O36.4XX3	Maternal care for intrauterine death, fetus 3	656.41 656.43
O36.4XX4	Maternal care for intrauterine death, fetus 4	656.41 656.43
O36.4XX5	Maternal care for intrauterine death, fetus 5	656.41 656.43
O36.4XX9	Maternal care for intrauterine death, other fetus	656.41 656.43
P95	Stillbirth Fetal death of unspecified cause **Excludes 1:** maternal care for intrauterine death (O36.4), missed abortion (O02.1), outcome of delivery, stillbirth (Z37.1, Z37.3, Z37.4, Z37.7)	779.6
Z37.1	Outcome of delivery, single stillborn	V27.1
Z37.3	Outcome of delivery, twins, one liveborn and one stillborn	V27.3
Z37.4	Outcome of delivery, twins, both stillborn	V27.4
Z37.6	Outcome of delivery, other multiple birth, some liveborn	V27.6
Z37.60	Multiple births, unspecified, some liveborn	V27.6
Z37.61	Triplets, some liveborn	V27.6
Z37.62	Quadruplets, some liveborn	V27.6
Z37.63	Quintuplets, some liveborn	V27.6
Z37.64	Sextuplets, some liveborn	V27.6
Z37.69	Other multiple births, some liveborn	V27.6
Z37.7	Outcome of delivery, other multiple birth, all stillborn	V27.7

Revision History

Since the December 2015 version, we added “(must specify)” next to some of the non-specific narrative descriptions. In addition, we added more synonyms to the list of narrative descriptions.

Added/modified Codes due to October 2023 ICD10 revision:

Q44.7	Q75.001	Q75.022	Q75.049	Q87.83
Q44.70	Q75.002	Q75.029	Q75.05	Q87.84
Q44.71	Q75.009	Q75.03	Q75.051	Q87.85
Q44.79	Q75.01	Q75.04	Q75.052	Q93.52
Q75.0	Q75.02	Q75.041	Q75.058	
Q75.00	Q75.021	Q75.042	Q75.08	

Added/modified Codes due to October 2022 ICD10 revision:

Q21.1	Q21.19	Q85.82	E34.328	G71.0342
Q21.10	Q21.2	Q85.83	E34.329	G71.0349
Q21.11	Q21.20	Q85.89	E34.39	G71.035
Q21.12	Q21.21	E34.3	G71.031	G71.038
Q21.13	Q21.22	E34.30	G71.032	G71.039
Q21.14	Q21.23	E34.31	G71.033	
Q21.15	Q85.8	E34.321	G71.0340	
Q21.16	Q85.81	E34.322	G71.0341	

Added/modified Codes due to October 2021 ICD10 revision:

Q61.5

Added/modified Codes due to October 2020 ICD10 revision:

A50	A50.2	A50.45	A50.7	G71.02
A50.0	A50.3	A50.49	A50.9	G71.09
A50.01	A50.30	A50.5	Q35-Q37	G71.11
A50.02	A50.31	A50.51	Q51.20	G71.2
A50.03	A50.32	A50.52	Q51.21	G71.20
A50.04	A50.39	A50.53	Q51.22	G71.21
A50.05	A50.4	A50.54	Q51.28	G71.22
A50.06	A50.40	A50.55	Q82.8	G71.220
A50.07	A50.41	A50.56	Q86.0	G71.228
A50.08	A50.42	A50.57	A50.9	G71.29
A50.09	A50.43	A50.59	G71.00	P96.1
A50.1	A50.44	A50.6	G71.01	

Added/modified Codes due to October 2019 ICD10 revision:

Q02	Q66.211	Q66.32	Q66.90	Q82.8
Q66.0	Q66.212	Q66.4	Q66.91	Q87.1
Q66.00	Q66.219	Q66.40	Q66.92	Q87.11
Q66.01	Q66.22	Q66.41	Q79.6	Q87.19
Q66.02	Q66.221	Q66.42	Q79.60	Q96
Q66.1	Q66.222	Q66.7	Q79.61	E34.3
Q66.10	Q66.229	Q66.70	Q79.62	
Q66.11	Q66.3	Q66.71	Q79.63	
Q66.12	Q66.30	Q66.72	Q79.69	
Q66.21	Q66.31	Q66.9	Q81.0	

Added/modified Codes due to October 2018 ICD10 revision:

Q02	Q51.2	Q51.20	Q51.21	Q51.28

Q93.5	Q93.59	G71.0	G71.01	G71.09
Q93.51	Q93.82	G71.00	G71.02	P35.4

Added/modified Codes due to October 2017 ICD10 revision:

Q25.49	Q53.111	Q53.13	Q53.211	Q53.23
Q53.11	Q53.112	Q53.21	Q53.212	Q82.2

Added/modified Codes due to October 2016 ICD10 revision:

Q25.2	Q25.42	Q25.48	Q52.123	Q82.6
Q25.21	Q25.43	Q25.49	Q52.124	Q87.82
Q25.29	Q25.44	Q52.12	Q52.129	
Q25.4	Q25.45	Q52.120	Q66.2	
Q25.40	Q25.46	Q52.121	Q66.21	
Q25.41	Q25.47	Q52.122	Q66.22	

Major/Minor/Reportable Classification Changes (effective October of the year listed)

Year Effective	Code	Defect	Previous classification	Current classification
2022	Q53.13	High scrotal testis, unilateral	Major	Minor
2022	Q53.23	Bilateral high scrotal testes	Major	Minor
2022	Q62.0	Pelvicaliectasis/Pelviectasis	Major	Minor
2022	Q62.0	Pyelectasis	Major	Minor
2019	Q55.64	Hidden penis	Major	Minor
2019	Q55.64	Buried penis	Major	Minor
2019	Q04.6	Congenital choroid plexus cyst	Major	Minor
2018	Q25.6	Peripheral pulmonic stenosis (PPS)	Minor	Major, only report if ≥2500g at birth
2018	Q55.63	Congenital torsion of penis	Major	Minor
2018	Q55.63	Phimosis	Major	Not Reportable

- P95 moved to stillbirth section from 'other selected reportable conditions' section, October 2018.

Removed codes from the Other Selected Reportable Conditions section, March 2017:

- All D codes besides D82.1 Di George's Syndrome
- E codes up until and including E23.2
- E34.8
- E70-E78.71
- E79-88.89
- G11
- G23-52.7
- H Codes
- P04

Reclassified codes (effective March 2017):

Description	Previous	Revised	Notes
Sacral dimple	L05.91	Q82.6	Description under "Excludes" for L05.91
Parasacral dimple	L05.91	Q82.6	
(m) Scaphocephaly (if due to craniosynostosis)	Q75.0	Q67.4	
(m) Trigonocephaly, other head deformations without synostosis	Q75.0	Q67.4	