New York State Department of Health
Congenital Malformations Registry

# ICD-10 Coding Manual

## List of all Reportable Congenital Malformations

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# 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, with congenital malformations or chromosomal anomalies, or were stillborn. For purposes of this registry, a congenital 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 congenital malformation who dies in the hospital (inpatient or emergency room).

## *What to Report:*

This coding manual contains a complete list of NYS Congenital Malformations Registry (CMR) 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 malformations and stillbirths must be reported
  - Minor anomalies, descriptions with an *(m)* (e.g. *(m) ankyloglossia*), are only required to be reported if the child also has one or more major malformations or was stillborn.
- 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**
  - 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
  - Add more specificity if you see '(must specify)' next to an ICD-10 Description

**NOTE:** Specific narrative descriptions of birth defects are required for reporting of all birth defects.

**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 and descriptions over time.

# Congenital Malformations of the Nervous System (Q00-Q07)

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

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

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

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

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

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| | Rachischisis<br>SB without hydrocephalus<br>Unspecified SB without hydrocephalus, with lipomeningomyelocele<br>Unspecified SB without hydrocephalus, with lipomeningocele<br>Unspecified SB without hydrocephalus, with meningocele<br>Unspecified SB without hydrocephalus, with SB (aperta)(cystica)<br>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<br>Atelomyelia (spinal cord defect)<br>Myelatelia<br>Myelodysplasia of spinal cord | 742.59 |
| **Q06.2** | Diastematomyelia | 742.51 |
| **Q06.3** | Other congenital cauda equina malformations | 742.59 |
| **Q06.4** | Hydromyelia<br>Hydrorachis | 742.53 |
| **Q06.8** | Other specified congenital malformations of spinal cord (must specify)<br>Lumbosacral dermal sinus tract<br>Occult spinal dysraphism sequence<br>Tethered cord syndrome, congenital<br>Tethered cord, filum<br>Low lying spinal chord<br>Tethered filum terminale<br>*(m) Fibrolipoma of filum terminale* | 742.59 |
| **Q06.9** | Congenital malformation of spinal cord, unspecified<br>Cervical spine dysraphism<br>Congenital anomaly NOS of spinal cord<br>Congenital deformity NOS of spinal cord<br>Congenital disease or lesion NOS of spinal cord<br>Spinal cord anomaly<br>Split spinal cord | 742.9 |
| **Q07** | **Other congenital malformations of nervous system**<br>**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**<br>**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<br>Arnold-Chiari type 2 without hydrocephalus | 741.90 |
| **Q07.01** | Arnold-Chiari syndrome (type II) with spina bifida (SB)<br>Arnold-Chiari syndrome (type II) with SB and hydromeningocele<br>Arnold-Chiari syndrome (type II) with SB and meningocele<br>Arnold-Chiari syndrome (type II) with SB and meningomyelocele<br>Arnold-Chiari syndrome (type II) with SB and myelocele<br>Arnold-Chiari syndrome (type II) with SB and myelomeningocele<br>Arnold-Chiari syndrome (type II) with SB and rachischisis<br>Arnold-Chiari syndrome (type II) with SB and syringohydromyelia<br>Arnold-Chiari syndrome (type II) with SB and lipomeningomyelocele<br>Arnold-Chiari syndrome (type II) with SB and lipomyelomeningocele<br>Arnold-Chiari syndrome (type II) with SB and lipomeningocele<br>Arnold-Chiari syndrome (type II) with SB and SB (aperta)(cystica)<br>Specify (cervical, thoracic, dorsal, thoracolumbar, lumbar, lumbosacral, sacral, unspecified) | 741.90<br>741.91<br>741.92<br>741.93 |
| **Q07.02** | Arnold-Chiari syndrome (type II) with hydrocephalus<br>Arnold-Chiari Type 2 with hydrocephalus | 742.0 |
| **Q07.03** | Arnold-Chiari syndrome (type II) with spina bifida (SB) and hydrocephalus<br>Arnold-Chiari syndrome (type II) with SB, hydrocephalus and hydromeningocele | 741.00<br>741.01 |

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| | Arnold-Chiari syndrome (type II) with SB, hydrocephalus and meningocele<br>Arnold-Chiari syndrome (type II) with SB, hydrocephalus and meningomyelocele<br>Arnold-Chiari syndrome (type II) with SB, hydrocephalus and myelocele<br>Arnold-Chiari syndrome (type II) with SB, hydrocephalus and myelomeningocele<br>Arnold-Chiari syndrome (type II) with SB, hydrocephalus and rachischisis<br>Arnold-Chiari syndrome (type II) with SB, hydrocephalus and syringohydromyelia<br>Arnold-Chiari syndrome (type II) with SB, hydrocephalus and lipomeningomyelocele<br>Arnold-Chiari syndrome (type II) with SB, hydrocephalus and lipomyelomeningocele<br>Arnold-Chiari syndrome (type II) with SB, hydrocephalus and lipomeningocele<br>Arnold-Chiari syndrome (type II) with SB, hydrocephalus and SB (aperta)(cystica)<br>Specify (cervical, thoracic, dorsal, thoracolumbar, lumbar, lumbosacral, sacral, unspecified) | 741.02<br>741.03 |
| **Q07.8** | Other specified congenital malformations of nervous system (must specify)<br>Agenesis of nerve<br>Congenital optic nerve pit<br>Displacement of brachial plexus<br>Incomplete development of nerve<br>Jaw-winking syndrome<br>Marcus Gunn's syndrome<br>Marcus Gunn jaw winking syndrome<br>Pit of optic disc<br>Synkinesis, eyelid | 742.8 |
| **Q07.9** | Congenital malformation of nervous system, unspecified<br>Central nervous system (CNS) malformation, fetal<br>Congenital anomaly NOS of nervous system<br>Congenital deformity NOS of nervous system<br>Congenital disease or lesion NOS of nervous system<br>Nervous system anomaly<br>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**<br>**Excludes 1:** cryptophthalmos NOS (Q11.2), cryptophthalmos syndrome (Q87.0) | |
| **Q10.0** | Congenital ptosis<br>Bilateral congenital ptosis<br>Left congenital ptosis<br>Right congenital ptosis<br>Blepharoptosis | 743.61 |
| **Q10.1** | Congenital ectropion | 743.62 |
| **Q10.2** | Congenital entropion | 743.62 |
| **Q10.3** | Other congenital malformations of eyelid (must specify)<br>Ablepharon<br>Blepharophimosis, congenital<br>Coloboma of eyelid<br>Congenital absence or agenesis of eyelid<br>Congenital accessory eyelid<br>Congenital accessory eye muscle<br>Telecanthus<br>*(m) Congenital absence or agenesis of cilia*<br>*(m) Congenital malformation of eyelid NOS*<br>*(m) Absence of eyelashes*<br>*(m) Epicanthal folds*<br>*(m) Fused eyelids (if birth weight greater than 1000 grams)*<br>*(m) Slanted eyes*<br>*(m) Small or narrow palpebral fissures*<br>*(m) Upward eyeslant*<br>*(m) Weakness of eyelids*<br>Do not report pseudostrabismus | 743.62<br>743.63 |
| **Q10.4** | *(m) Absence and agenesis of lacrimal apparatus*<br>*(m) Congenital absence of punctum lacrimal*<br>*(m) Nasolacrimal duct obstruction* | 743.65 |
| **Q10.5** | *(m) Congenital stenosis and stricture of lacrimal duct*<br>*(m) Congenital blocked tear duct*<br>*(m) Congenital dacryostenosis (left/ right/ bilateral)*<br>*(m) Obstruction of lacrimal duct(s)* | 743.65 |
| **Q10.6** | *(m) Other congenital malformations of lacrimal apparatus*<br>*(m) Congenital abnormality of lacrimal drainage system*<br>*(m) Congenital absence of lacrimal drainage structure/lacrimal apparatus*<br>*(m) Congenital anomaly of lacrimal gland*<br>*(m) Accessory lacrimal canal* | 743.65 |
| **Q10.7** | Congenital malformation/anomaly of orbit/ orbit proper<br>Congenital superior sulcus anomaly of orbit | 743.66 |
| **Q11** | **Anophthalmos, microphthalmos and macrophthalmos** | |
| **Q11.0** | Cystic eyeball<br>Congenital cystic globe | 743.03 |
| **Q11.1** | Other anophthalmos<br>Anophthalmos, NOS<br>Agenesis of eye<br>Aplasia of eye<br>Congenital absence of the eye | 743.00 |
| **Q11.2** | Microphthalmos (eye condition)<br>Cryptophthalmos<br>Dysplasia of eye<br>Hypoplasia of eye<br>Rudimentary eye | 743.06<br>743.10<br>743.11<br>743.12 |

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

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

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| | Enlarged eye, NOS<br>Glaucoma of childhood<br>Glaucoma of newborn<br>Hydrophthamos, simple, unspecified<br>Juvenile glaucoma<br>Keratoglobus, congenital, with glaucoma<br>Macrocornea with glaucoma<br>Macrophthalmos in congenital glaucoma<br>Megalocornea with glaucoma<br>Primary congenital glaucoma<br>Simple buphthalmos<br>Buphthalmos associated with other ocular anomalies<br>Congenital glaucoma, simple | 743.44 |
| Q15.8 | Other specified congenital malformations of eye (must specify)<br>Epibulbar dermoid cyst | 743.8 |
| Q15.9 | Congenital malformation of eye, unspecified (try not to use)<br>Congenital anomaly of eye<br>Congenital deformity of eye | 743.9 |
| Q16 | **Congenital malformations of ear causing impairment of hearing**<br>**Excludes 1:** congenital deafness (H90.-) | |
| Q16.0 | Congenital absence of (ear) auricle (Anotia)<br>Absence of external ear | 744.01 |
| Q16.1 | Congenital absence, atresia or stricture of auditory canal (external) (must specify)<br>Congenital atresia or stricture of osseous meatus<br>Congenital atresia of external auditory canal<br>Congenital atresia of external ear (left/right/bilateral)<br>Aural atresia<br>Absence of auditory canal<br>Absence of external auditory meatus<br>*(m) Narrow external auditory meatus* | 744.02 |
| Q16.2 | Absence of Eustachian tube | 744.24 |
| Q16.3 | Congenital malformation of ear ossicles<br>Congenital fusion of ear ossicles | 744.04 |
| Q16.4 | Other congenital malformations of middle ear<br>Congenital malformations of middle ear, NOS | 744.03 |
| Q16.5 | Congenital malformation of inner ear<br>Congenital anomaly of membranous labyrinth<br>Congenital anomaly of organ of Corti | 744.05 |
| Q16.9 | Congenital malformation of ear causing impairment of hearing, unspecified<br>Congenital absence of ear, NOS<br>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*<br>*(m) Accessory tragus*<br>*(m) Polyotia*<br>*(m) Preauricular appendage or skin tag*<br>*(m) Supernumerary ear*<br>*(m) Supernumerary lobule*<br>*(m) Ear, double lobule*<br>*(m) Other appendage, tag, or lobule*<br>*(m) Papillomas, ear tags*<br>*(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*<br>*(m) Lop ear* | 744.29 |

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| | *(m) Pointed ear*<br>*(m) Absent or decreased cartilage*<br>*(m) Cleft ear*<br>*(m) Crumpled ears*<br>*(m) Cupped ears*<br>*(m) Darwin's tubercle*<br>*(m) Ears, absent tragus*<br>*(m) Ears, bridged concha*<br>*(m) Ears, lack of helical fold*<br>*(m) Ears, thickened or over folded helix*<br>*(m) Extra cartilage*<br>*(m) Malformed ear*<br>*(m) Notch in ear*<br>*(m) Posteriorly rotated ears*<br>*(m) Protruding ears* | |
| **Q17.4** | *(m) Misplaced ear*<br>*(m) Low-set ears*<br>**Excludes 1**: cervical auricle (Q18.2) | 744.29 |
| **Q17.5** | *(m) Prominent ear*<br>*(m) Bat ear* | 744.29 |
| **Q17.8** | *(m) Other specified congenital malformations of ear* (must specify)<br>*(m) Congenital absence of lobe of ear*<br>*(m) Congenital split ear lobes (left/right/bilateral)*<br>*(m) Cryptotia* | 744.21<br>744.24<br>743.29 |
| ***Q17.9*** | *(m) Congenital malformation of ear, unspecified*<br>*(m) Congenital anomaly of ear, NOS*<br>*(m) Congenital external ear (pinna) anomaly*<br>*(m) Congenital anomaly of ear (left/right/bilateral)* | 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*<br>*(m) Branchial vestage*<br>*(m) Branchial cleft fistula*<br>*(m) Branchial cleft sinus*<br>*(m) Branchial cleft cyst*<br>*(m) Branchial cleft pit* | 744.41 |
| **Q18.1** | *(m) Preauricular sinus and cyst*<br>*(m) Cervicoaural fistula*<br>*(m) Fistula of auricle, congenital*<br>*(m) Ear pit*<br>*(m) Preauricalar fistula*<br>*(m) Preauricular pits*<br>*(m) Preauricular sinus* | 744.46<br>744.47 |
| **Q18.2** | *(m) Other branchial cleft malformations* (must specify)<br>*(m) Branchial cleft malformations, NOS*<br>*(m) Cervical auricle*<br>*(m) Otocephaly*<br>*(m) Dermal sinus of head*<br>*(m) Other branchial cleft anomaly* | 744.43<br>744.49 |
| **Q18.3** | *(m) Webbing of neck*<br>*(m) Pterygium colli*<br>*(m) Redundant neck skin folds* | 744.5 |
| **Q18.4** | Macrostomia | 744.83 |
| **Q18.5** | Microstomia | 744.84 |

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| **Q18.6** | *(m) Macrocheilia (large lips)*<br>*(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)<br>Medial cyst of face and neck<br>Medial fistula of face and neck<br>Medial sinus of face and neck<br>Cleft face (excluding cleft lip and palate)<br>*(m) Short neck* | 744.89 |
| **Q18.9** | Congenital malformations of face and/or neck, unspecified (try not to use)<br>Congenital anomaly NOS of face and/or neck<br>*(m) Abnormal facies*<br>*(m) Craniofacial dysmorphosis*<br>*(m) Dysmorphic facies/face* | 744.89<br>744.9 |

# Congenital Malformations of the Circulatory System (Q20-Q28)

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

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| Q21.1 | Atrial septal defect (ASD)<br>Coronary sinus defect<br>Patent or persistent ostium secundum defect (type II)<br>Atrial septal defect, NOS<br>Auricular septal defect, NOS<br>Secundum atrial septal defect<br>*(m) Patent foraman ovale (PFO)*<br>*(m) Patent or persistent sinus venosus defect*<br>*(m) Persistent foraman ovale (PFO)*<br>*(m) Fossa ovalis defect*<br>*(m) Foramen ovale*<br>*(m) Nonclosure of foramen ovale, NOS*<br>*(m) Partial foramen ovale* | 745.5 |
| Q21.2 | Atrioventricular septal defect (AVSD)<br>Atrial septal defect, primum<br>Common atrioventricular canal<br>Endocardial cushion defect<br>Ostium primum atrial septal defect (ASD) (type I)<br>Ostium primum defect<br>Absence of atrial septum<br>Artrioventricular canal type VSD<br>Common atrioventricular canal with VSD<br>Other endocardial cushion defects<br>Persistent ostium primum<br>Single common atrium<br>Type III VSD, Canal-type VSD | 745.60<br>745.61<br>745.69 |
| 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)<br>Dextraposition of aorta in Fallot's tetralogy<br>Tetralogy of Fallot with absent pulmonary valve<br>Tetralogy of Fallot with pulmonary atresia or stenosis<br>Tetralogy of Fallot with ventricular septal defect | 745.2 |
| Q21.4 | Aortopulmonary septal defect<br>Abnormal communication between aorta and pulmonary artery<br>Aortic septal defect<br>Aortopulmonary window | 745.0 |
| Q21.8 | Other congenital malformations of cardiac septa (must specify)<br>Eisenmenger's defect<br>Pentalogy of Fallot<br>Fallot's tetralogy plus atrial septal defect | 745.8<br>745.2 |
| Q21.9 | Congenital malformations of cardiac septum, unspecified<br>Septal (heart) defect, NOS | 745.9 |
| Q22 | **Congenital malformations of pulmonary and tricuspid valves** | |
| Q22.0 | Pulmonary valve atresia<br>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)<br>Congenital pulmonary valve regurgitation (do not code if mild, trivial or physiologic)<br>Trilogy of Fallot | 746.09 |
| Q22.3 | Other congenital malformations of pulmonary valve (must specify)<br>Congenital malformation of pulmonary valve, NOS<br>Supernumerary cusps of pulmonary valve<br>Bicuspid pulmonary valve<br>Other specified anomaly of pulmonary valve<br>Pulmonary valve hypoplasia<br>Quadricuspid pulmonary valve | 746.00<br>746.09 |

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| | Unicuspid pulmonary valve | |
| Q22.4 | Congenital tricuspid valve stenosis (narrowing)<br>Congenital tricuspid atresia<br>Absence of tricuspid valve<br>Tricuspid hypoplasia | 746.1 |
| Q22.5 | Ebstein's anomaly<br>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)<br>Insufficiency of tricuspid valve<br>Congenital tricuspid valve regurgitation | 746.89 |
| Q22.9 | Congenital malformations of tricuspid valve, unspecified | 746.89 |
| **Q23** | **Congenital malformations of aortic and mitral valves** | |
| Q23.0 | Congenital stenosis of aortic valve<br>Congenital aortic atresia<br>Congenital aortic stenosis, NOS<br>**Type 1 Excludes:** Congenital stenosis of aortic valve in hypoplastic left heart syndrome (Q23.4), congenital subaortic stenosis (Q24.4), supravalvular aortic stenosis (congenital) (Q25.3) | 746.3 |
| Q23.1 | Congenital insufficiency of aortic valve (do not code if mild, trivial or physiologic)<br>Bicuspid aortic valve<br>Congenital aortic insufficiency<br>Congenital aortic valve regurgitation | 746.4 |
| Q23.2 | Congenital mitral stenosis (narrowing)<br>Congenital mitral atresia<br>Congenital supravalvular mitral stenosis<br>Absence of mitral valve<br>Atresia of mitral valve<br>Hypoplasia of mitral valve/cleft mitral valve | 746.5 |
| Q23.3 | Congenital mitral insufficiency (do not code if mild, trivial or physiologic)<br>Congenital mitral valve regurgitation<br>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)<br>Cleft leaflet of mitral valve<br>Aortic valve atresia/absence<br>Fused commissure of mitral valve<br>Other specified anomaly of aortic valves<br>Parachute deformity of mitral valve<br>Supernumerary cusps | 746.89 |
| Q23.9 | Congenital malformation of aortic and mitral valves, unspecified<br>Unspecified anomaly of the aortic valves | 746.89 |
| **Q24** | **Other congenital malformations of heart Excludes 1:** endocardial fibroelastosis (I42.4) | |
| Q24.0 | Dextrocardia<br>Dextrocardia with no mention of situs inversus<br>Dextrocardia without situs inversus (situs solitus)<br>**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<br>Situs inversus with levocardia | 746.87 |
| Q24.2 | Cor triatriatum<br>Cor triatriatum, dexter/left sided/right sided/sinister<br>Divided left atrium<br>Divided right atrium | 746.82 |
| Q24.3 | Pulmonary infundibular stenosis (narrowing)<br>Infundibular pulmonic stenosis | 746.83 |

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| | Stenosis of infundibulum of right ventricle<br>Subvalvular pulmonic stenosis | |
| Q24.4 | Congenital subaortic stenosis<br>Congenital subvalvular aortic stenosis | 746.81 |
| Q24.5 | Malformation of coronary vessels<br>Congenital coronary (artery) aneurysm<br>Congenital coronary artery fistula<br>Absence of coronary artery<br>Anomalies of coronary artery or sinus<br>Anomalous origin or communication of coronary artery<br>Coronary artery arising from aorta or pulmonary trunk<br>Single coronary artery | 746.85 |
| Q24.6 | Congenital heart block<br>Complete or incomplete atrioventricular (AV) block | 746.86 |
| Q24.8 | Other specified congenital malformations of heart (must specify)<br>Congenital diverticulum of left ventricle<br>Congenital malformation of myocardium<br>Congenital malformation of pericardium<br>Malposition of heart<br>Uhl's disease (hypoplastic right ventricle)<br>Abdominal heart<br>Absence of papillary muscle of luthisis<br>Anomalies of myocardium<br>Anomalies of pericardium<br>Anomaly of papillary muscle<br>Atresia of cardiac vein<br>Congenital pericardial defect<br>Ectopia (ectopic) cordis (mesocordia)<br>Hypoplasia of cardiac vein<br>Hypoplastic left ventricle<br>Hypoplastic ventricle, NOS<br>Obstructive anomalies of heart, NEC<br>Conduction defects, NOS<br>Other defects of the atria<br>Other specified anomaly of heart<br>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]<br>Ventricular hypertrophy (right or left)<br>*(m) Congenital cardiomegaly*<br>*Cardiomyopathy, hypertrophic is not reportable* | 746.84<br>746.87<br>746.89 |
| Q24.9 | Congenital malformations of heart, unspecified (try not to use)<br>Complex congenital heart defect (code component defects)<br>Congenital anomaly of heart<br>Congenital disease of heart | 746.9 |
| Q25 | **Congenital malformations of great arteries** | |
| Q25.0 | Patent ductus arteriosus (PDA) (do not report if birth weight <1500 grams)<br>Patent ductus Botallo<br>Persistent ductus arteriosus<br>Patent ductus arteriosis closed with medication | 747.0 |
| Q25.1 | Coarctation of aorta<br>Coarctation of aorta (preductal) (postductal)<br>Unspecified coarctation of aorta | 747.10 |
| Q25.2 | Atresia of aorta | |
| Q25.21 | Interruption of aortic arch<br>Atresia of aortic arch<br>Interrupted aortic arch | 747.11 |

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| | Interrupted aortic arch. Type A<br>Interrupted aortic arch. Type B | |
| Q25.29 | Other atresia of aorta<br>Atresia of aorta | 747.22 |
| Q25.3 | Supravalvular aortic stenosis (narrowing)<br>Small aorta, narrowing of aorta<br>Stenosis of aorta<br>Stricture of aorta<br>**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)<br>Double aortic arch (Vascular ring of aorta)<br>Hypoplasia of aorta<br>Hypoplasia of aortic arch, congenital<br>Hypoplastic aorta syndrome<br>Kommerells diverticulum<br>Overriding aorta<br>Persistent right aortic arch<br>Pseudocoarctation of aorta<br>Right aortic arch<br>Aberrant subclavian artery<br>Anomalous origin, right subclavian artery<br>Dextroposition of aorta<br>Hypoplastic aortic isthmus<br>Pseudotruncus arteriosus<br>Tubular hypoplasia of aorta<br>Vascular ring compression of trachea<br>**Excludes 1:** hypoplasia of aorta in hypoplastic left heart syndrome (Q23.4) | |
| Q25.40 | *(m) Congenital malformation of aorta unspecified* | 747.20 |
| Q25.41 | Absence and aplasia of aorta | 747.29 |
| Q25.42 | Hypoplasia of aorta | 747.29 |
| Q25.43 | Congenital aneurysm of aorta<br>Congenital aneurysm of aortic root<br>Congenital aneurysm of aortic sinus | 747.29 |
| Q25.44 | *(m) Congenital dilation of aorta* | 747.29 |
| Q25.45 | Double aortic arch<br>Vascular ring of aorta | 747.21 |
| Q25.46 | *(m) Tortuous aortic arch*<br>*(m) Persistent convolutions of aortic arch* | 747.21 |
| Q25.47 | *(m) Right aortic arch*<br>*(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)<br>*(m) Aortic arch*<br>*(m) Bovine arch* | 747.29 |
| Q25.5 | Atresia of pulmonary artery<br>Pulmonary artery absence<br>Pulmonary artery agenesis<br>Pulmonary artery atresia | 747.31 |
| Q25.6 | Stenosis of pulmonary artery<br>Supravalvular pulmonary stenosis (narrowing)<br>*Peripheral pulmonary artery stenosis (do not report if birth weight <2500 g)*<br>*Peripheral pulmonic stenosis (PPS) (do not report if birth weight <2500 g)* | 747.31<br>747.39 |
| **Q25.7** | **Other congenital malformations of pulmonary artery** | |
| Q25.71 | Coarctation of pulmonary artery | 747.31 |
| Q25.72 | Congenital pulmonary arteriovenous malformation<br>Congenital pulmonary arteriovenous aneurysm | 747.32 |

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| | Aneurysm of pulmonary artery | |
| Q25.79 | Other congenital malformations of pulmonary artery (must specify)<br>Aberrant pulmonary artery<br>Agenesis of pulmonary artery<br>Congenital aneurysm of pulmonary artery<br>Congenital anomaly of pulmonary artery<br>Hypoplasia of pulmonary artery<br>Dilatation of pulmonary artery<br>Unspecified anomaly of pulmonary artery | 747.39 |
| Q25.8 | Other congenital malformations of other great arteries<br>Anomalous origin of right subclavian artery | 747.29 |
| Q25.9 | Congenital malformations of great arteries, unspecified | 747.29 |
| Q26 | **Congenital malformations of great veins** | |
| Q26.0 | Congenital stenosis of vena cava<br>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<br>Total anomalous pulmonary venous return [TAPVR], subdiaphragmatic<br>Total anomalous pulmonary venous return [TAPVR], supradiaphragmatic<br>Total anomalous lung veins to coronary sinus/liver veins/right heart/vena cavae | 747.41 |
| Q26.3 | Partial anomalous pulmonary venous connection<br>Partial anomalous pulmonary venous return [PAPVR] | 747.42 |
| Q26.4 | Anomalous pulmonary venous connection, unspecified | 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)<br>Absence of vena cava (inferior or superior)<br>Atresia of vena cava (inferior or superior)<br>Azygos continuation of inferior vena cava<br>Persistent left posterior cardinal vein<br>Pulmonary vein stenosis<br>Scimitar syndrome<br>Stenosis of vena cava (inferior or superior)<br>Anomalous portal vein termination<br>Transposition of pulmonary veins, NOS | 747.49 |
| Q26.9 | Congenital malformation of great vein, unspecified<br>Congenital anomaly of vena cava (inferior) (superior), NOS<br>Anomaly NOS of pulmonary veins | 747.40 |
| Q27 | **Other congenital malformations of peripheral vascular system**<br>**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*<br>*(m)* Single umbilical artery<br>*(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)<br>Congenital malformation of renal artery, NOS<br>Multiple renal arteries<br>Other anomaly of renal artery<br>Renal vessel anomaly | 747.62 |
| Q27.3 | **Arteriovenous malformation (AVM) (peripheral)**<br>Arteriovenous aneurysm | |
| Q27.30 | Arteriovenous malformation, site unspecified<br>Peripheral arteriovenous malformation | 747.69 |
| Q27.31 | Arteriovenous malformation of vessel of upper limb | 747.63 |

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| Q27.32 | Arteriovenous malformation of vessel of lower limb | 747.64 |
| Q27.33 | Arteriovenous malformation of digestive system vessel<br>Arteriovenous malformation of duodenum and jejunum<br>Arteriovenous malformation of intestinal vessels<br>Congenital arteriovenous malformation of the gastrointestinal tract<br>Congenital duodenal and jejunal arteriovenous malformation | 747.61 |
| Q27.34 | Arteriovenous malformation of renal vessel | 747.62 |
| Q27.39 | Arteriovenous malformation, other site<br>Arteriovenous malformation of spine | 747.69 |
| Q27.4 | Congenital phlebectasia | 747.89 |
| Q27.8 | Other specified congenital malformations of peripheral vascular system (must specify)<br>Absence of peripheral vascular system<br>Anomalous origin of right subclavian artery<br>Atresia of peripheral vascular system<br>Congenital aneurysm (peripheral)<br>Congenital stricture, artery<br>Congenital varix<br>Absence of artery, NEC<br>Anomaly of spinal vessel<br>Atresia of artery<br>Gastrointestinal vessel anomaly<br>Other anomaly of peripheral arteries<br>Other anomaly of peripheral veins<br>**Excludes 1:** arteriovenous malformation (Q27.3-) | 747.69 |
| Q27.9 | Congenital malformation of peripheral vascular system, unspecified<br>Anomaly of artery or vein, NOS<br>Arteriovenous malformation<br>Congenital anomaly of peripheral blood vessel<br>Congenital vascular malformation of orbit<br>Vascular or venous malformation | 747.60 |
| Q28 | **Other congenital malformations of circulatory system**<br>**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)<br>**Excludes 2:** congenital peripheral aneurysm (Q27.8), congenital pulmonary aneurysm (Q25.79), congenital retinal aneurysm (Q14.1) | |
| Q28.0 | Arteriovenous malformation of precerebral vessels<br>Congenital arteriovenous precerebral aneurysm (non-ruptured) | 747.89 |
| Q28.1 | Congenital malformation of precerebral vessels, NOS<br>Congenital precerebral aneurysm (nonruptured) | 747.89 |
| Q28.2 | Arteriovenous malformation of cerebral vessels<br>Anomaly of cerebral arteriovenous system<br>Arteriovenous malformation of brain, NOS<br>Congenital arteriovenous cerebral aneurysm (nonruptured)<br>Cerebral arteriovenous malformation<br>Anomalies of cerebrovascular system<br>Anomalies of vein of Galen<br>Other anomaly of cerebral vessels | 747.81 |
| Q28.3 | Other malformations of cerebral vessels (must specify)<br>Congenital cerebral aneurysm (nonruptured)<br>Congenital malformation of cerebral vessels, NOS<br>Developmental venous anomaly | 747.81 |
| Q28.8 | Other specified congenital malformations of circulatory system (must specify)<br>Congenital aneurysm, specified site NEC<br>Spinal vessel anomaly | 747.89<br>747.82 |
| Q28.9 | Congenital malformation of circulatory system, unspecified | 747.9 |

# Congenital Malformations of the Respiratory System (Q30-Q34)

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

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

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| **Q33.6** | Congenital hypoplasia or dysplasia of lung<br>Congenital pulmonary dysplasia<br>Hypoplasia of lung<br>Pulmonary hypoplasia<br>Hypoplasia of lobe of lung<br>Other specified dysplasia of lung<br>Unspecified dysplasia of lung<br>**Excludes 1:** pulmonary hypoplasia associated with short gestation (P28.0) | 748.5 |
| **Q33.8** | Other congenital malformations of lung (must specify)<br>Aplasia of lung<br>Bilobar right lung<br>Congenital lobar emphysema<br>Fusion of lobes of lung<br>Lymphangiectasia of lungs<br>Right lung with left lung bronchial pattern<br>Trilobal left lung | 748.69 |
| **Q33.9** | Congenital malformations of lung, unspecified<br>Congenital anomaly of lung | 748.60 |
| **Q34** | **Other congenital malformations of respiratory system**<br>**Excludes 2:** congenital central alveolar hypoventilation syndrome (G47.35) | |
| **Q34.0** | Anomaly of pleura<br>Congenital anomaly of pleural folds<br>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)<br>Atresia of nasopharynx<br>Immotile cilia syndrome<br>Primary ciliary dyskinesia | 748.8 |
| **Q34.9** | Congenital malformation of respiratory system, unspecified (try not to use this code)<br>Congenital absence of respiratory system<br>Congenital anomaly of respiratory system, NOS | 748.9 |

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

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

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| **Q37.0** | Cleft hard palate with bilateral cleft lip | 749.24<br>749.25 |
| **Q37.1** | Cleft hard palate with unilateral cleft lip<br>Cleft hard palate with cleft lip, NOS | 749.22<br>749.25 |
| **Q37.2** | Cleft soft palate with bilateral cleft lip | 749.24<br>749.25 |
| **Q37.3** | Cleft soft palate with unilateral cleft lip<br>Cleft soft palate with cleft lip, NOS | 749.22<br>749.25 |
| **Q37.4** | Cleft hard and soft palate with bilateral cleft lip<br>Complete cleft palate and bilateral cleft lip | 749.23<br>749.24 |
| **Q37.5** | Cleft hard and soft palate with unilateral cleft lip<br>Cleft hard and soft palate with cleft lip, NOS<br>Complete cleft palate and cleft lip, unilateral, left<br>Complete cleft palate and cleft lip, unilateral, right<br>Complete cleft palate and cleft lip, unilateral<br>Complete cleft palate and cleft lip, NOS<br>Uranostaphyloschisis with cleft lip | 749.21<br>749.22 |
| **Q37.8** | Unspecified cleft palate with bilateral cleft lip<br>Incomplete cleft palate and bilateral cleft lip<br>Bilateral cleft lip, with any cleft palate | 749.24 |
| **Q37.9** | Unspecified cleft palate with unilateral cleft lip<br>Cleft palate with cleft lip, NOS<br>Cheilopalatoschisis<br>Cleft palate with cleft lip<br>Cleft palate with cleft lip, unspecified<br>Other combinations cleft palate and lip | 749.20 |

# Congenital Malformations of the Digestive System (Q38-Q45)

| ICD-10 | Descriptions | ICD-9 |
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| **Q38** | **Other congenital malformations of tongue, mouth and pharynx**<br>**Excludes 1:** dentofacial anomalies (M26.-), macrostomia (Q18.4), microstomia (Q18.5) | |
| **Q38.0** | Congenital malformations of lips, NEC<br>Van der Woude syndrome<br>*(m) Congenital fistula of lip or pits*<br>*(m) Congenital palsy of the lower lip*<br>*(m) Long philtrum (only code if with Q86.02)*<br>*(m) Lip tie*<br>*(m) Notched lip (only code if with Q86.02)*<br>*(m) Other lip anomaly (only code if with Q86.02)*<br>*(m) Prominent philtrum (only code if with Q86.02)*<br>**Excludes 1:** cleft lip (Q36.-), cleft lip with cleft palate (Q37.-), macrocheilia (Q18.6), microcheilia (Q18.7) | 750.25 |
| **Q38.1** | *(m) Ankyloglossia*<br>*(m) Tongue tie*<br>*(m) Short frenulum of tongue*<br>*(m) Aberrant frenula* | 750.0 |
| **Q38.2** | *(m) Macroglossia (large tongue)*<br>*(m) Congenital hypertrophy of tongue* | 750.15 |
| **Q38.3** | Other congenital malformations of tongue (must specify)<br>Aglossia (absence of tongue)<br>*(m) Bifid tongue*<br>*(m) Congenital abnormal tongue position anomaly*<br>*(m) Congenital adhesion of tongue*<br>*(m) Congenital anomaly of tongue*<br>*(m) Congenital fissure of tongue*<br>*(m) Double tongue*<br>*(m) Hypoglossia (small tongue)*<br>*(m) Hypoplasia of tongue*<br>*(m) Microglossia*<br>*(m) Cleft tongue or split tongue*<br>*(m) Dislocation or displacement of tongue*<br>*(m) Glossoptosis* | 750.10<br>750.11<br>750.12<br>750.13<br>750.16<br>750.19 |
| **Q38.4** | Congenital malformations of salivary glands and ducts (must specify)<br>Atresia of salivary glands and ducts<br>Congenital absence of salivary glands and ducts<br>Congenital accessory salivary glands and ducts<br>Congenital fistula of salivary gland<br>Congenital ranula<br>Congenital salivary gland anomaly<br>Imperforate salivary duct<br>Other anomaly of salivary glands or ducts | 750.21<br>750.22<br>750.23<br>750.24 |
| **Q38.5** | Congenital malformations of palate, NEC<br>*(m) Congenital absence of uvula*<br>*(m) Congenital malformation of palate, NOS*<br>*(m) Congenital high arched palate*<br>**Excludes 1:** cleft palate (Q35.-), cleft palate with cleft lip (Q37.-) | 750.26 |
| **Q38.6** | *(m) Other congenital malformations of mouth (must specify)*<br>*(m) Congenital malformations of mouth, NOS*<br>*(m) Fordyce spots of mouth*<br>*(m) Fordyce's disease*<br>*(m) Low lying maxillary frenulum* | 750.26 |
| **Q38.7** | Congenital pharyngeal pouch<br>Congenital diverticulum of pharynx | 750.27 |

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| | **Excludes 1:** pharyngeal pouch syndrome (D82.1) | |
| **Q38.8** | Other congenital malformations of pharynx (must specify)<br>Congenital malformation of pharynx, NOS<br>Congenital anomaly of pharynx<br>Congenital velopharyngeal dysfunction<br>Congenital velopharyngeal impairment<br>Congenital velopharyngeal incompetence<br>Imperforate pharynx<br>Do not report tonsil asymmetry | 750.29 |
| **Q39** | **Congenital malformations of esophagus** | |
| **Q39.0** | Atresia of esophagus without fistula<br>Atresia of esophagus, NOS<br>Esophageal atresia<br>Imperforate esophagus | 750.3 |
| **Q39.1** | Atresia of esophagus with tracheoesophageal fistula<br>Atresia of esophagus with broncho-esophageal fistula<br>Esophageal atresia with tracheoesophageal fistula | 750.3 |
| **Q39.2** | Congenital trachea-esophageal fistula without atresia<br>Congenital trachea-esophageal fistula, NOS<br>Congenital trachea esophageal fistula without atresia of esophagus<br>Esophagotracheal fistula, congenital<br>T E fistula without esophageal atresia<br>Tracheoesophageal fistula “h” type | 750.3 |
| **Q39.3** | Congenital stenosis and stricture of esophagus<br>Congenital esophageal ring<br>Congenital Schatzkis ring<br>Congenital stenosis of esophagus | 750.3 |
| **Q39.4** | Esophageal web | 750.3 |
| **Q39.5** | Congenital dilatation of esophagus<br>Congenital cardiospasm<br>Congenital achalasia<br>Giant esophagus | 750.4 |
| **Q39.6** | Congenital diverticulum of esophagus<br>Congenital esophageal pouch | 750.4 |
| **Q39.8** | Other congenital malformations of esophagus (must specify)<br>Congenital absence of esophagus<br>Congenital displacement of esophagus<br>Congenital duplication of esophagus<br>Esophageal duplication<br>Other tracheoesophageal anomaly | 750.4 |
| **Q39.9** | Congenital malformation of esophagus, unspecified | 750.4 |
| **Q40** | **Other congenital malformations of upper alimentary tract** | |
| **Q40.0** | Congenital hypertrophic pyloric stenosis<br>Congenital or infantile constriction<br>Congenital or infantile hypertrophy<br>Congenital or infantile spasm of pylorus<br>Congenital of infantile stenosis<br>Congenital or infantile stricture<br>Other congenital pyloric obstruction<br>Do not report epileptic/infantile spasms | 750.5 |
| **Q40.1** | Congenital hiatus hernia<br>Congenital hiatal hernia<br>Congenital displacement of cardia through esophageal hiatus<br>**Excludes 1**: congenital diaphragmatic hernia (Q79.0) | 750.6 |
| **Q40.2** | Other specified congenital malformations of stomach (must specify)<br>Congenital displacement of stomach<br>Congenital diverticulum of stomach | 750.7 |

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| | Congenital hourglass stomach<br>Congenital duplication of stomach<br>Megalogastria<br>Microgastria<br>Displacement or transposition of stomach | |
| Q40.3 | Congenital malformation of stomach, unspecified<br>Congenital gastric anomaly | 750.7 |
| Q40.8 | Other specified congenital malformations of upper alimentary tract | 750.8 |
| Q40.9 | Congenital malformation of upper alimentary tract, unspecified<br>Congenital anomaly of upper alimentary tract<br>Congenital deformity of upper alimentary tract<br>Congenital upper GI tract anomaly | 750.9 |
| Q41 | **Congenital absence, atresia or stenosis of small intestine**<br>**Includes:** congenital obstruction, occlusion or stricture of small intestine or intestine, NOS<br>**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)<br>Absence of duodenum/ duodenal absence<br>Atresia of duodenum/ duodenal atresia<br>Stenosis of duodenum/ duodenal stenosis | 751.1 |
| Q41.1 | Congenital absence, atresia or stenosis of jejunum (must specify)<br>Apple peel syndrome<br>Imperforated jejunum<br>Absence of jejunum<br>Atresia of jejunum<br>Stenosis of jejunum | 751.1 |
| Q41.2 | Congenital absence, atresia or stenosis of ileum (must specify)<br>Absence of ileum/ ileal absence<br>Atresia of ileum/ ileal atresia<br>Stenosis of ileum/ ileal stenosis | 751.1 |
| Q41.8 | Congenital absence, atresia or stenosis of other specified parts of small intestine (must specify)<br>Absence of other specified parts of small intestine (specify)<br>Atresia of other specified parts of small intestine (specify)<br>Stenosis of other specified parts of small intestine (specify) | 751.1 |
| Q41.9 | Congenital absence, atresia and stenosis of small intestine, part unspecified<br>Absence of small intestine<br>Atresia of small intestine<br>Stenosis of small intestine | 751.1 |
| Q42 | **Congenital absence, atresia and stenosis of large intestine**<br>**Includes:** congenital obstruction, occlusion and stricture of large intestine | |
| Q42.0 | Congenital absence, atresia and stenosis of rectum with fistula (must specify)<br>Absence of rectum with fistula<br>Atresia of rectum with fistula<br>Stenosis of rectum with fistula | 751.2 |
| Q42.1 | Congenital absence, atresia and stenosis of rectum without fistula (must specify)<br>Imperforated rectum<br>Absence of rectum without fistula/rectal absence<br>Atresia of rectum without fistula/ rectal atresia<br>Stenosis of rectum without fistula/ rectal stenosis | 751.2 |
| Q42.2 | Congenital absence, atresia and stenosis of anus with fistula (must specify)<br>Absence of anus with fistula<br>Atresia of anus with fistula<br>Stenosis of anus with fistula<br>Imperforated anus with fistula<br>Congenital or infantile occlusion of anus with fistula<br>Congenital infantile stricture of anus with fistula | 751.2 |

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| Q42.3 | Congenital absence, atresia and stenosis of anus without fistula (must specify)<br>Imperforated anus<br>Absence of anus<br>Atresia of anus<br>Congenital or infantile occlusion of anus without fistula<br>Congenital infantile stricture of anus without fistula<br>Stenosis of anus | 751.2 |
| Q42.8 | Congenital absence, atresia and stenosis of other parts of large intestine (must specify)<br>Absence of appendix<br>Atresia of appendix<br>Stenosis of appendix | 751.2 |
| Q42.9 | Congenital absence, atresia, or stenosis of large intestine, part unspecified (try not to code)<br>Absence of large intestine<br>Atresia of colon<br>Stenosis of colon | 751.2 |
| **Q43** | **Other congenital malformations of intestine** | |
| Q43.0 | *(m) Meckel's diverticulum (displaced) (hypertrophic)*<br>*(m) Persistent vitelline duct*<br>*(m) Persistent omphalomesenteric duct* | 751.0 |
| Q43.1 | Hirschsprung's disease<br>Aganglionosis<br>Congenital (aganglionic) megacolon<br>Megacolon, congenital<br>Long segment Hirschsprung's disease<br>Short segment Hirschsprung's-Recto-Sigmoid | 751.3 |
| Q43.2 | Other congenital functional disorders of colon (must specify)<br>Congenital dilation of colon<br>Congenital megacolon, not aganglionic | 751.3 |
| Q43.3 | Congenital malformations of intestinal fixation (must specify)<br>Congenital omentum, anomalous adhesions [bands]<br>Congenital peritoneal adhesions [bands]<br>Incomplete rotation of cecum and colon<br>Insufficient rotation of cecum and colon<br>Intestinal malrotation<br>Jackson's membrane<br>Malrotation of colon<br>Malrotation of intestine<br>Rotation failure of cecum and colon<br>Universal mesentery<br>Anomalies of mesentery<br>Ladd's bands<br>Malrotation of cecum<br>Malrotation of small intestine alone<br>Other specified malrotation<br>Unspecified malrotation | 751.4 |
| Q43.4 | Duplication of intestine<br>Duplication of cyst ileum<br>Duplication of anus<br>Duplication of appendix<br>Duplication of pylorus<br>Duplication of cecum | 751.5 |
| Q43.5 | Ectopic anus<br>Anterior anus | 751.5 |
| Q43.6 | Congenital fistula of rectum and anus | 751.5 |

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

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| **Q44.7** | Other congenital malformation of liver (must specify)<br>Accessory liver<br>Alagille's syndrome<br>Arteriohepatic dysplasia<br>Congenital absence of liver<br>Congenital focal nodular hyperplasia of liver<br>Congenital malformation of liver, NOS<br>Absence or agenesis of liver, partial<br>Absence or agenesis of liver, total<br>Floating liver<br>Other anomaly of liver<br>*(m) Congenital hepatomegaly* | 751.60<br>751.69 |
| **Q45** | **Other congenital malformations of digestive system**<br>**Excludes 2:** congenital diaphragmatic hernia (Q79.0), congenital hiatus hernia (Q40.1) | |
| **Q45.0** | Agenesis, aplasia or hypoplasia of pancreas<br>Congenital absence of pancreas<br>Pancreatic agenesis<br>Pancreatic aplasia<br>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)<br>Accessory pancreas<br>Congenital malformation of pancreas or pancreatic duct, NOS<br>Ectopic pancreas<br>Ectopic pancreatic tissue<br>Pancreatic heterotopia<br>**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)<br>Absence(complete) (partial) of alimentary tract, NOS<br>Duplication of digestive system<br>Malposition, congenital of digestive system<br>Congenital malposition of digestive organs, NOS<br>Cyst of mesenteric remnant<br>Duplication of alimentary tract<br>Ectopic digestive organs, NOS | 751.8 |
| **Q45.9** | Congenital malformation of digestive system, unspecified<br>Congenital anomaly of digestive system<br>Congenital deformity of digestive system | 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.]**<br>**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<br>Cystic ovary<br>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<br>46.XX with streak gonads | 752.0 |
| Q50.39 | Other congenital malformation of ovary<br>Congenital malformation of ovary, NOS<br>Ectopic ovary | 752.0 |
| Q50.4 | Embryonic cyst of fallopian tube<br>Fimbrial cyst | 752.11 |
| Q50.5 | Embryonic cyst of broad ligament<br>Cyst of hydatid of Morgagni<br>Epoophoron cyst<br>Parovarian cyst | 752.11 |
| Q50.6 | Other congenital malformations of fallopian tube and broad ligament (must specify)<br>Absence of fallopian tube and broad ligament<br>Accessory fallopian tube and broad ligament<br>Atresia of fallopian tube and broad ligament<br>Congenital malformation of fallopian tube or board ligament, NOS | 752.10<br>752.19 |
| Q51 | **Congenital malformations of uterus and cervix [Only applicable to female patients.]** | |
| Q51.0 | Agenesis and aplasia of uterus<br>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<br>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.]**<br>Septate uterus | |
| Q51.20 | Other doubling of uterus, unspecified<br>Septate uterus, unspecified | 752.35 |
| Q51.21 | Other complete doubling of uterus<br>Complete septate uterus | |
| Q51.22 | Other partial doubling of uterus<br>Partial septate uterus | |
| Q51.28 | Other doubling of uterus, other specified<br>Septate uterus, other specified | |
| Q51.3 | Bicornate uterus<br>Bicornate uterus, complete or partial | 752.34 |
| Q51.4 | Unicornate uterus<br>Unicornate uterus with or without a separate uterine horn<br>Uterus with only one functioning horn<br>Uterus unicornis | 752.33 |
| Q51.5 | Agenesis and aplasia of cervix<br>Congenital absence of cervix | 752.43 |

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| Q51.6 | *(m) Embryonic cyst of cervix* | 752.41 |
| Q51.7 | Congenital fistula between uterus and digestive and urinary tracts<br>Congenital fistula between uterus and digestive tracts<br>Congenital fistula between uterus and urinary tract<br>Congenital uterointestinal fistula | <br>752.49<br>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<br>Arcuatus uterus | 752.36 |
| Q51.811 | Hypoplasia of uterus<br>Infantile uterus | 752.32 |
| Q51.818 | Other congenital malformations of uterus (must specify)<br>Müllerian anomaly of uterus, NEC | 752.39 |
| Q51.82 | **Other congenital malformations of cervix [Only applicable to female patients.]** | |
| Q51.820 | Cervical duplication<br>Congenital duplication of cervix | 752.44 |
| Q51.821 | Hypoplasia of cervix | 752.49 |
| Q51.828 | Other congenital malformations of cervix (must specify)<br>Congenital imperforate cervix<br>Congenital stenosis of cervical canal<br>Congenital stenosis of uterine cervix<br>Congenital stenosis or stricture of cervical canal | <br><br>752.49 |
| Q51.9 | Congenital malformation of uterus and cervix, unspecified<br>Congenital uterine anomaly<br>Inadequate development of endometrium | 752.39<br>752.49 |
| Q52 | **Other congenital malformations of female genitalia [Only applicable to female patients.]** | |
| Q52.0 | Congenital absence of vagina<br>Vaginal agenesis, total or partial | 752.45 |
| Q52.1 | **Doubling of vagina [Only applicable to female patients.]**<br>**Excludes 1:** doubling of vagina with doubling of uterus and cervix (Q51.1-) | |
| Q52.10 | Doubling of vagina, unspecified<br>Congenital duplication of vagina<br>Congenital vaginal septum<br>Septate vagina, NOS | 752.47 |
| Q52.11 | Transverse vaginal septuma<br>Congenital transverse septated vagina | 752.46 |
| Q52.12 | Longitudinal vaginal septum<br>Congenital longitudinal vaginal septum<br>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)<br>Congenital malformation of vagina, NOS<br>*(m) Canal of Nuck cyst, congenital*<br>*(m) Congenital atresia of vagina*<br>*(m) Embryonic vaginal cyst*<br>*(m) Gartner's duct cyst*<br>*(m) Vaginal adenosis*<br>*(m) Cyst of vagina, vulva* | <br><br><br>752.41<br>752.49 |

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| | *(m) Congenital stenosis or stricture of vagina* | |
| Q52.5 | Fusion of labia<br>*(m) Labial adhesions, congenital* | 752.49 |
| Q52.6 | Congenital malformation of clitoris (must specify)<br>Absence of clitoris<br>Agenesis of clitoris<br>*(m) Clitoromegaly*<br>*(m) Enlarged clitoris*<br>*(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<br>Congenital anomaly of external female genitalia<br>Congenital malformation of vulva, NOS<br>Fusion of vulva | 752.49 |
| Q52.71 | Congenital absence of vulva | 752.49 |
| Q52.79 | Other congenital malformations of vulva<br>*(m) Congenital cyst of vulva* | 752.49 |
| Q52.8 | Other specified congenital malformation of female genitalia<br>*(m) Vaginal or hymenal tags*<br>*(m) Hypoplastic majora* | 752.49 |
| Q52.9 | Congenital malformation of female genitalia, unspecified | 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 | 752.51 |
| Q53.01 | Ectopic testis, unilateral<br>Ectopic testis, left only<br>Ectopic testis, right only | 752.51 |
| Q53.02 | Ectopic testes, bilateral | 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)<br>Cryptorchidism, left only (birth weight greater than 2,500 grams)<br>Cryptorchidism, right only (birth weight greater than 2,500 grams)<br>Cryptorchidism, unilateral (birth weight greater than 2,500 grams)<br>Left undescended testicle (birth weight greater than 2,500 grams)<br>Nonpalpable testis (specify laterality) (birth weight greater than 2,500 grams)<br>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 | 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)<br>Cryptorchidism, bilateral (birth weight greater than 2,500 grams)<br>Nonpalpable testes (birth weight greater than 2,500 grams)<br>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 | Bilateral high scrotal testes (birth weight greater than 2,500 grams) | |
| Q53.9 | Undescended testicle, unspecified (birth weight greater than 2,500 grams)<br>Cryptorchidism, NOS (birth weight greater than 2,500 grams)<br>Nonpalpable testis (birth weight greater than 2,500 grams) | 752.51 |

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| **Q54** | **Hypospadias PLEASE SPECIFY DEGREE OR SITE OF HYPOSPADIAS [Only applicable to male patients.]**<br>**Excludes 1:** epispadias (Q64.0) | |
| **Q54.0** | Hypospadias, balanic<br>Hypospadias, coronal<br>Hypospadias, glandular<br>Hypospadias 1 degree, glandular, coronal | 752.61 |
| **Q54.1** | Hypospadias, penile<br>Hypospadias 2 degree, penile, subcoronal | 752.61 |
| **Q54.2** | Hypospadias, penoscrotal<br>Hypospadias 3 degree, scrotal, penile scrotal | 752.61 |
| **Q54.3** | Hypospadias, perineal<br>Hypospadias 3 degree, perineal, scrotal, penile scrotal | 752.61 |
| **Q54.4** | Congenital chordee (report only with hypospadias (ventral, dorsal, NOS))<br>*(m) Chordee without hypospadias* | 752.63 |
| **Q54.8** | Other hypospadias (must specify)<br>Hypospadias with intersex state | 752.61 |
| **Q54.9** | Hypospadias, unspecified (must specify)<br>Paraspadias | 752.61 |
| **Q55** | **Other congenital malformations of male genital organs [Only applicable to male patients.]**<br>**Excludes 1:** congenital hydrocele (P83.5), hypospadias (Q54.-) | |
| **Q55.0** | Absence and aplasia of testis<br>Congenital absence of testicle<br>Monorchism | 752.89 |
| **Q55.1** | Hypoplasia of testis and scrotum<br>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<br>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<br>Penoscrotal transposition | 752.81 |
| **Q55.29** | Other congenital malformations of testis and scrotum | 752.89 |
| **Q55.3** | Atresia of vas deferens<br>Code first any associated cystic fibrosis (E84) | 752.89 |
| **Q55.4** | Other congenital malformations of vas deferens, epididymis, seminal vesicles and prostate (must specify)<br>Absence or aplasia of prostate<br>Absence or aplasia of spermatic cord<br>Atresia of ejaculatory duct<br>Appendix testis cyst<br>Cysts of embryonic remnants<br>Hydatid of morgagni cyst<br>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<br>Micropenis | 752.64 |
| **Q55.63** | *(m) Congenital torsion of penis*<br>*Phimosis is not reportable*<br>**Excludes 1:** acquired torsion of penis (N48.82) | 752.69 |

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| **Q55.64** | *(m) Buried penis*<br>*(m)* Concealed penis<br>*(m) Hidden penis*<br>*(m) Webbed penis*<br>**Excludes 1:** acquired buried penis (N48.83) | 752.65 |
| **Q55.69** | Other congenital malformation of penis<br>Congenital malformation of penis, NOS<br>Absent or hooded foreskin<br>Incomplete foreskin | 752.69 |
| **Q55.7** | Congenital vasocutaneous fistula | 752.89 |
| **Q55.8** | Other specified congenital malformations of male genital organs | 752.89 |
| **Q55.9** | Congenital malformations of male genital organ, unspecified<br>Congenital anomaly of male genital organ<br>Congenital deformity of male genital organ | 752.9 |
| **Q56** | **Indeterminate sex and pseudohermaphroditism**<br>**PLEASE REPORT CHROMOSOME TEST RESULTS**<br>**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<br>Ovotestis<br>Gynandrism | 752.7 |
| **Q56.1** | Male pseudohermaphroditism, NEC<br>46, XY with streak gonads<br>Male pseudohermaphroditism, NOS | 752.7 |
| **Q56.2** | Female pseudohermaphroditism, NEC<br>Female pseudohermaphroditism, NOS | 752.7 |
| **Q56.3** | Pseudohermaphroditism, unspecified | 752.7 |
| **Q56.4** | Indeterminate sex, unspecified<br>Ambiguous genitalia<br>Disorder of sexual differentiation<br>Sexual differentiation disorder | 752.7 |

# Congenital Malformations of the Urinary System (Q60-Q64)

| ICD-10 | Description | ICD-9 |
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| Q60 | **Renal agenesis and other reduction defects of kidney**<br>**Includes:** congenital absence of kidney, congenital atrophy of kidney, infantile atrophy of kidney | |
| Q60.0 | Renal agenesis, unilateral<br>Single kidney<br>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<br>Oligomeganephronic hypoplasia of kidney | 753.0 |
| Q60.6 | Potter's syndrome<br>Oligohydramnios sequence | 753.0 |
| Q61 | **Cystic kidney disease**<br>**Excludes 1:** acquired cyst of kidney (N28.1), Potter's syndrome (Q60.6) | |
| Q61.0 | **Congenital renal cyst** | |
| Q61.00 | Congenital renal cyst, unspecified<br>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**<br>Polycystic kidney, autosomal recessive | |
| Q61.11 | Cystic dilation of collecting ducts | 753.14 |
| Q61.19 | Other polycystic kidney, infantile type<br>Autosomal recessive infantile polycystic kidney disease<br>Polycystic kidney, autosomal recessive<br>Congenital PKD | 753.14 |
| Q61.2 | Polycystic kidney, adult type<br>Polycystic kidney, autosomal dominant | 753.13 |
| Q61.3 | Polycystic kidney, unspecified | 753.12 |
| Q61.4 | Renal dysplasia<br>Congenital renal dysplasia<br>Multicystic dysplastic kidney<br>Multicystic kidney (development)<br>Multicystic kidney disease (MCDK)<br>Multicystic renal dysplasia<br>Dysplastic kidney, bilateral<br>Renal dysplasia, bilateral<br>Renal dysplasia, NOS; dysplastic kidney, NOS<br>Unilateral dysplasia of kidney<br>**Excludes 1:** polycystic kidney disease (Q61.11-Q61.3) | 753.15<br>753.19 |
| Q61.5 | Medullary cystic kidney<br>Medullary cystic kidney, juvenile type<br>Medullary cystic kidney, adult type<br>Medullary sponge kidney<br>Nephronopthisis<br>Sponge kidney, NOS | 753.16<br>753.17 |
| Q61.8 | Other cystic kidney diseases (must specify)<br>Fibrocystic kidney<br>Fibrocystic renal degeneration of disease | 753.19 |
| Q61.9 | Cystic kidney disease, unspecified (must specify)<br>Cystic kidney disease<br>Meckel-Gruber syndrome | 753.10 |
| Q62 | **Congenital obstructive defects of renal pelvis and congenital malformations of ureter** | |

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| Q62.0 | Congenital hydronephrosis (swelling of kidney)<br>Congenital hydronephrosis, bilateral<br>Congenital hydronephrosis, left only<br>Congenital hydronephrosis, NOS<br>Congenital hydronephrosis, right only<br>Congenital hydronephrosis, unilateral<br>Pelvicaliectasis/Pelviectasis<br>Pyelectasis | 753.29 |
| Q62.1 | **Congenital occlusion of ureter**<br>Atresia and stenosis of ureter | |
| Q62.10 | Congenital occlusion of ureter, unspecified<br>Stenosis of ureter<br>Atresia, stricture, or stenosis of ureter, bilateral<br>Atresia, stricture, or stenosis of ureter, left<br>Atresia, stricture, or stenosis of ureter, NOS<br>Atresia, stricture, or stenosis of ureter, right<br>Atresia, stricture, or stenosis of ureter, unilateral<br>Congenital occlusion of ureter, bilateral<br>Congenital occlusion of ureter, left<br>Congenital occlusion of ureter, right<br>Congenital occlusion of ureter, unilateral<br>Impervious ureter, bilateral<br>Impervious ureter, left<br>Impervious ureter, NOS<br>Impervious ureter, right<br>Impervious ureter, unilateral<br>Ureteral valves, bilateral<br>Ureteral valves, left<br>Ureteral valves, NOS<br>Ureteral valves, right<br>Ureteral valves, unilateral | 753.29 |
| Q62.11 | Congenital occlusion of ureteropelvic junction<br>Stricture of pelviureteric junction<br>Ureteropelvic junction (UPJ) obstruction/stenosis, left<br>Ureteropelvic junction obstruction/stenosis, right<br>Ureteropelvic junction obstruction/stenosis, unilateral<br>Ureteropelvic junction obstruction/stenosis, bilateral | 753.21 |
| Q62.12 | Congenital occlusion of ureterovesical orifice<br>Ureterovesical junction obstruction/stenosis, NOS<br>Ureterovesical junction obstruction/stenosis, left<br>Ureterovesical junction obstruction/stenosis, right<br>Ureterovesical junction obstruction/stenosis, unilateral<br>Ureterovesical junction obstruction/stenosis, bilateral | 753.22 |
| Q62.2 | Congenital megaureter<br>Congenital dilation of ureter<br>Congenital hydroureter, bilateral<br>Congenital hydroureter, left<br>Congenital hydroureter, NOS<br>Congenital hydroureter, right<br>Congenital hydroureter, unilateral<br>Megaureter, NOS<br>Megaureter, left<br>Megaureter, right<br>Megaureter, unilateral<br>Megaureter, bilateral | 753.22 |
| Q62.3 | **Other obstructive defects of renal pelvis and ureter** | |
| Q62.31 | Congenital ureterocele, orthotopic | 753.23 |

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| | Simple ureterocele<br>Ureterocele is reportable; Hydrocele is NOT reportable | |
| Q62.32 | Cecoureterocele<br>Ectopic ureterocele | 753.23 |
| Q62.39 | Other obstructive defects of renal pelvis ureter<br>Congenital hydroureter<br>Congenital occlusion of ureter<br>Congenital ureteral obstruction<br>Congenital ureterovesical obstruction<br>Duplicated collecting system with obstruction<br>Ureteropelvic junction obstruction, NOS<br>Urinary tract obstruction due to duplicated collecting system | 753.20 |
| Q62.4 | Agenesis of ureter<br>Congenital absence ureter | 753.4 |
| Q62.5 | Duplication of ureter<br>Accessory ureter<br>Double ureter, duplex collecting system<br>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<br>Ectopia of ureter<br>Ectopic ureter<br>Displaced ureteric orifice | 753.4 |
| Q62.69 | Other malposition of ureter | 753.4 |
| Q62.7 | Congenital vesico-uretero-renal reflux<br>Ureteral reflux<br>Variations of vesicoureteral reflux | 753.4 |
| Q62.8 | Other congenital malformations of ureter (must specify)<br>Anomaly of ureter, NOS<br>Hypoplastic ureter, bilateral<br>Hypoplastic ureter, left<br>Hypoplastic ureter, NOS<br>Hypoplastic ureter, right<br>Other specified anomalies of ureter<br>Unspecified anomaly of ureter<br>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<br>Double kidney and pelvis, pyelon duplex<br>Trifid kidney (pelvis)<br>Triple kidney and pelvis, pyelon triplex | 753.3 |
| Q63.1 | Lobulated, fused or horseshoe kidney<br>Fused kidney<br>Horseshoe kidney<br>Lobulated kidney | 753.3 |
| Q63.2 | Ectopic kidney/Pelvic Kidney<br>Congenital displaced kidney<br>Malrotation of kidney | 753.3 |
| Q63.3 | Hyperplastic or giant kidney<br>Compensatory hypertrophy of kidney | 753.3 |
| Q63.8 | Other specified congenital malformations of kidney<br>Congenital renal caluli/ calculus<br>Congenital renal pelvis and ureteral defect | 753.3 |

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| | Discoid kidney<br>Duplicated collecting system without obstruction<br>Inequal in size of kidney<br>Megacalycosis, congenital | |
| Q63.9 | Congenital malformation of kidney, unspecified | 753.3 |
| Q64 | **Other congenital malformations of urinary system** | |
| Q64.0 | Epispadias<br>Anaspadias<br>Ventral meatus<br>**Excludes 1:** hypospadias (Q54.-)<br>[Q64.0 is only applicable to male patients.] | 752.62 |
| Q64.10 | Exstrophy of urinary bladder, unspecified<br>Ectopia vesicae<br>Exstrophy of bladder sequence | 753.5 |
| Q64.11 | Supravesical fissure of urinary bladder | 753.8 |
| Q64.12 | Cloacal exstrophy of urinary bladder<br>Exstrophy of cloaca sequence | 753.5 |
| Q64.19 | Other exstrophy of urinary bladder<br>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<br>Congenital bladder neck stricture<br>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<br>Atresia and stenosis of urethra and bladder neck, NOS<br>Congenital posterior urethral valve | 753.6 |
| Q64.4 | Malformation of urachus<br>Cyst of urachus<br>Fistula of urachus<br>Patent urachus<br>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**<br>**Excludes 1:** congenital prolapse of bladder (mucosa) (Q79.4) | |
| Q64.70 | Unspecified congenital malformations of bladder and urethra<br>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 | 753.8 |
| Q64.8 | Other specified congenital malformations of urinary system | 753.8 |
| Q64.9 | Congenital malformation of urinary system, unspecified | 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 |

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| 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<br>Congenital subluxation of unspecified hip, unilateral | 754.32 |
| Q65.31 | Congenital partial dislocation of right hip, unilateral<br>Congenital subluxation of right hip | 754.32 |
| Q65.32 | Congenital partial dislocation of left hip, unilateral<br>Congenital subluxation of left hip | 754.32 |
| Q65.4 | Congenital partial dislocation of hip, bilateral<br>Congenital subluxation of hip, bilateral | 754.33 |
| Q65.5 | Congenital partial dislocation of hip, unspecified<br>Congenital subluxation of hip | 754.32 |
| Q65.6 | Congenital unstable hip<br>Congenital dislocatable hip<br>Predislocation status of hip at birth, unilateral, left<br>Predislocation status of hip at birth, unilateral, right<br>Predislocation status of hip at birth, unilateral<br>Preluxation of hip, congenital, left<br>Preluxation of hip, congenital, right<br>Preluxation of hip, congenital, unilateral<br>Unstable hip, unilateral<br>Unstable hip, unilateral, left<br>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)<br>Anteversion of femoral neck/ femoral anteversion (do not report if normal x-ray)<br>Congenital acetabular dysplasia<br>Hip dysplasia, NOS<br>Bilateral hip dysplasia<br>Congenital flexion deformity, hip or thigh, left<br>Congenital flexion deformity, hip or thigh, right<br>Developmental dysplasia of hip (DDH)<br>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**<br>**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<br>Talipes equinovarus, NOS<br>Talipes equinovarus, unilateral<br>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<br>Talipes calcaneovarus, unilateral<br>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<br>Congenital metatarsus varus | |

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|---|---|---|
| | 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<br>Congenital metatarsus varus | |
| Q66.221 | Congenital metatarsus adductus, right foot<br>Congenital metatarsus varus, right foot | 754.53 |
| Q66.222 | Congenital metatarsus adductus, left foot<br>Congenital metatarsus varus, left foot | 754.53 |
| Q66.229 | Congenital metatarsus adductus, unspecified foot<br>Congenital metatarsus varus, unspecified foot | 754.53 |
| Q66.3 | Other congenital varus deformities of feet<br>Hallux varus, congenital<br>Talipes varus<br>Cubitus varus deformity of feet | |
| Q66.30 | Other congenital varus deformities of feet, unspecified foot<br>Talipes varus | 754.59 |
| Q66.31 | Other congenital varus deformities of feet, right foot<br>Talipes varus, right | 754.59 |
| Q66.32 | Other congenital varus deformities of feet, left foot<br>Talipes varus, left | 754.59 |
| Q66.4 | Congenital talipes calcaneovalgus | |
| Q66.40 | Congenital talipes calcaneovalgus, unspecified foot<br>Congenital talipes calcaneovalgus, unilateral<br>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**<br>Congenital flat foot<br>Congenital rigid flat foot<br>Congenital spastic (everted) flat foot<br>*(m) Congenital rocker bottom foot*<br>**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)<br>Congenital metatarsus valgus<br>Hallus valgus<br>Pes valgoplanus congenital<br>Talipes valgus<br>Claw foot<br>Talipes equinovalgus | 754.69 |
| Q66.7 | Congenital pes cavus<br>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 |

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| **Q66.89** | Other specified congenital deformities of feet (must specify)<br>Brachydactyly of toes<br>Brachymetatarsia<br>Congenital asymmetric talipes<br>Congenital contracture of toe<br>Congenital talipes, NOS<br>Congenital tarsal coalition<br>Congenital clubfoot, NOS<br>Fusion of calcaneus (heel bone)<br>Tarsal coalitions (specify laterality)<br>Anomalies of foot<br>Astragalo-scaphoid synostosis<br>Calcaneonavicular bar<br>Clubfoot, NOS<br>Congenital hallux valgus<br>Congenital hallux varus<br>Coalition of calcaneus<br>Dorsiflexion of foot (fixed only)<br>Other specified deformities of toes<br>Plantar furrow<br>Prominent heel<br>Talipes calcaneus<br>Talipes equinus<br>Talonavicular synostosis<br>*(m) Hammer toe, congenital*<br>*(m) Curly toe*<br>*(m) Gap between toes (1-2)*<br>*(m) Long toes*<br>*(m) Overlapping toes*<br>*(m) Recessed toes (4,5)*<br>*(m) Short or broad great toe*<br>*(m) Wide spaced first and second toes* | 754.79<br>755.67 |
| **Q66.9** | Congenital deformity of feet, unspecified (must specify)<br>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**<br>**Excludes 1:** congenital malformation syndromes classified to Q87.-, Potter's syndrome (Q60.6) | |
| **Q67.0** | *(m) Congenital facial asymmetry*<br>*(m) Asymmetry of face*<br>*(m) Facial asymmetry*<br>*(m) Head asymmetry* | 754.0 |
| **Q67.1** | *(m) Congenital compression facies* | 754.0 |
| **Q67.2** | *(m) Dolichocephaly*<br>*(m) Long narrow head* | 754.0 |
| **Q67.3** | *(m) Plagiocephaly*<br>*(m) Flat occiput* | 754.0 |
| **Q67.4** | *(m) Other congenital deformities of skull, face and jaw (must specify)*<br>*(m) Congenital hemifacial atrophy or hypertrophy*<br>*(m) Congenital depressions in skull*<br>*(m) Deviation of nasal septum, congenital*<br>*(m) Squashed or bent nose, congenital*<br>*(m) Oxycephaly*<br>*(m) Trigonocephaly*<br>*(m) Metopic suture open to bregma*<br>*(m) Large or small fontanelles* | 748.1<br>754.0 |

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| | *(m) Scaphocephaly (if due to craniosynostosis)*<br>*(m) Third fontanel*<br>*(m) Trigonocephaly, other head deformations without synostosis*<br>**Excludes 1:** dentofacial anomalies [including malocclusion] (M26.-), syphilitic saddle nose (A50.5) | |
| **Q67.5** | Congenital deformity of spine<br>Congenital postural scoliosis<br>Congenital scoliosis, NOS<br>**Excludes 1:** infantile idiopathic scoliosis (M41.0 ), scoliosis due to congenital bony malformation (Q76.3) | 754.2 |
| **Q67.6** | Pectus excavatum<br>Congenital funnel chest | 754.81 |
| **Q67.7** | Pectus carinatum<br>Congenital pigeon chest | 754.82 |
| **Q67.8** | Other congenital deformities of chest (must specify)<br>Congenital deformity of chest wall, NOS<br>Congenital anomaly of thoracic cage<br>Shield chest<br>*(m) Bell shaped chest* | 754.89 |
| **Q68** | **Other congenital musculoskeletal deformities**<br>**Excludes 1:** reduction defects of limb(s) (Q71-Q73)<br>**Excludes 2:** congenital myotonic chondrodystrophy (G71.13) | |
| **Q68.0** | Congenital deformity of sternocleidomastoid muscle<br>Congenital anomaly of sternocleidomastoid muscle<br>Congenital contracture of sternocleidomastoid (muscle)<br>Congenital (sternomastoid) torticollis<br>Sternomastoid tumor (congenital)<br>Absent or hypoplastic sternocleidomastoid muscle<br>Congenital wryneck<br>Sternomastoid tumor | 754.1 |
| **Q68.1** | Congenital deformity of finger(s) and hand (must specify)<br>Congenital club finger<br>Congenital crooked finger<br>Spade-like hand (congenital)<br>Anthrogryposis<br>Flexion contractures of individual joints of the hand | 754.89 |
| **Q68.2** | Congenital deformity of knee<br>Congenital dislocation of knee<br>Congenital genu recurvatum | 755.64<br>754.41<br>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<br>Discoid meniscus of knee | 717.5 |
| **Q68.8** | Other specified congenital musculoskeletal deformities (must specify)<br>Congenital deformity of clavicle<br>Congenital deformity of elbow<br>Congenital deformity of forearm<br>Congenital deformity of scapula<br>Congenital deformity of wrist<br>Congenital dislocation of elbow<br>Congenital dislocation of shoulder<br>Congenital dislocation of wrist<br>Congenital dislocation of radial head<br>Congenital elevation of scapula<br>Generalized flexion contractures of lower limb joints | 756.9<br>755.51<br>755.52 |
| **Q69** | **Polydactyly** | |

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| Q69.0 | Accessory finger(s)<br>Radial polydactyly<br>Accessory digits hand, NOS<br>Polydactyly of fingers | 755.01 |
| Q69.1 | Accessory thumb(s) | 755.01 |
| Q69.2 | Accessory toe(s) (do not report if skin tag)<br>Accessory hallux<br>Polydactyly of toes<br>Accessory big toe (preaxial)<br>Accessory toes (postaxial)<br>Accessory digits foot, NOS | 755.02 |
| Q69.9 | Polydactyly, unspecified<br>Supernumerary digit(s), NOS<br>Postaxial polydactyly, NOS<br>Accessory digits, NOS (hand/foot not specified) | 755.00 |
| Q70 | **Syndactyly** | |
| Q70.0 | **Fused fingers**<br>**Complex syndactyly of fingers and/or thumb with synostosis** | |
| Q70.00 | Fused fingers and/or thumb, unspecified hand<br>Syndactyly of fingers with fusion of bones | 755.12 |
| Q70.01 | Fused fingers and/or thumb, right hand<br>Syndactyly of fingers of right hand with fusion of bones | 755.12 |
| Q70.02 | Fused fingers and/or thumb, left hand<br>Syndactyly of fingers of left hand with fusion of bones | 755.12 |
| Q70.03 | Fused fingers and/or thumb, bilateral<br>Syndactyly of fingers, bilateral, with fusion of bones | 755.12 |
| Q70.1 | ***(m) Webbed fingers***<br>***(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**<br>**Complex syndactyly of toes with synostosis** | |
| Q70.20 | Fused toes, unspecified foot<br>Syndactyly of toes with fusion of bones | 755.14 |
| Q70.21 | Fused toes, right foot<br>Syndactyly of toes of right foot, with fusion of bones | 755.14 |
| Q70.22 | Fused toes, left foot<br>Syndactyly of toes of left foot, with fusion of bones | 755.14 |
| Q70.23 | Fused toes, bilateral<br>Syndactyly of toes of both feet, with fusion of bones | 755.14 |
| Q70.3 | ***(m) Webbed toes***<br>***(m) Simple syndactyly of toes without synostosis*** | |
| Q70.30 | *(m) Webbed toes, unspecified foot*<br>*(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<br>**Excludes 1:** specified syndactyly of hand and feet - code to specified conditions (Q70.0--Q70.3-) | 755.00<br>755.10 |
| Q70.9 | Syndactyly, unspecified<br>Symphalangy, NOS<br>Syndactyly of multiple and unspecified sites | 755.10 |
| Q71 | **Reduction defects of upper limb** | |

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| **Q71.0** | **Congenital complete absence of upper limb**<br>**Transverse deficiency of upper limb** | |
| **Q71.00** | Congenital complete absence of unspecified upper limb<br>Congenital complete absence of arm<br>Transverse deficiency of arm<br>Congenital amputation of upper limb, NOS<br>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<br>Complete phocomelia of upper limb<br>Proximal phocomelia of upper limb<br>Distal phocomelia of upper limb (forearm missing. upper arm/hand present)<br>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<br>Complete phocomelia of bilateral upper limbs | 755.23 |
| **Q71.2** | **Congenital absence of both forearm and hand**<br>**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<br>Congenital absence of hand<br>Congenital deficiency of hand bones<br>Congenital longitudinal deficiency of fingers<br>Longitudinal deficiency of phalanges of hand<br>Absence of finger(s), congenital (includes individual missing fingers/phalanges)<br>Absence of preaxial fingers<br>Absence of thumb | 755.29 |
| **Q71.31** | Congenital absence of right hand and finger<br>Congenital longitudinal deficiency of right fingers<br>Hypoplasia of right finger(s)<br>Hypoplasia of right thumb | 755.29 |
| **Q71.32** | Congenital absence of left hand and finger<br>Congenital longitudinal deficiency of left fingers<br>Hypoplasia of left finger(s)<br>Hypoplasia of left thumb | 755.29 |
| **Q71.33** | Congenital absence of hand and finger, bilateral<br>Hypoplasia of fingers, bilateral<br>Hypoplasia of thumbs, bilateral | 755.29 |
| **Q71.4** | **Longitudinal reduction defect of radius (congenital)**<br>**Radial clubhand** | |
| **Q71.40** | Longitudinal reduction defect of unspecified radius<br>Radial hemimelia<br>Absence of radius<br>Congenital absence of radius with or without absence of some distal elements<br>Longitudinal deficiency, radial, complete or partial<br>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 |

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| Q71.43 | Longitudinal reduction of radius, bilateral | 755.26 |
| Q71.5 | **Longitudinal reduction defect of ulna** | |
| Q71.50 | Longitudinal reduction defect of unspecified ulna<br>Longitudinal deficiency of ulna<br>Absence of ulna<br>Congenital absence ulna, with or without absence of some distal elements<br>Longitudinal deficiency ulna, complete or partial<br>Postaxial (longitudinal) reduction deficiency of upper limb, ulna ray defect | 755.27 |
| 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<br>Cleft hand | 755.58 |
| Q71.61 | Lobster-claw right hand<br>Cleft hand, right hand | 755.58 |
| Q71.62 | Lobster-claw left hand<br>Cleft hand, left hand | 755.58 |
| Q71.63 | Lobster-claw hand, bilateral<br>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<br>Brachydactyly of right fingers | 755.20 |
| Q71.892 | Other reduction defects of left upper limb<br>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)<br>Ectromelia NOS of upper limb<br>Hemimelia of upper limb, NOS<br>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<br>Longitudinal deficiency of right upper arm | 755.20 |
| Q71.92 | Unspecified reduction defect of left upper limb<br>Longitudinal deficiency of left upper arm | 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<br>Congenital complete absence of unspecified leg<br>Transverse deficiency unspecified lower limb<br>Transverse hemimelia of unspecified lower leg | 755.31 |
| Q72.01 | Congenital complete absence of right lower limb<br>Congenital complete absence of right leg<br>Transverse deficiency right lower limb<br>Transverse hemimelia of right leg | 755.31 |
| Q72.02 | Congenital complete absence of left lower limb<br>Congenital complete absence of left leg<br>Transverse deficiency left lower limb | 755.31 |

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| | Transverse hemimelia of left leg | |
| Q72.03 | Congenital complete absence of lower limbs, bilateral<br>Congenital complete absence of legs, bilateral<br>Transverse deficiency lower limbs, bilateral<br>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<br>Complete phocomelia of lower limb (upper and lower leg missing, foot present)<br>Absence of lower leg only (femur and foot present)<br>Distal phocomelia of lower limb (lower leg absent, femur/foot present)<br>Proximal phocomelia of lower limb (femur absent, lower leg, foot present) | 755.33 |
| 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<br>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)<br>Longitudinal deficiency of phalanges of foot<br>Longitudinal reduction defect of toes | 755.38 |
| Q72.31 | Congenital absence of right foot and toe(s)<br>Longitudinal deficiency of right toes<br>Longitudinal reduction defect of right toes | 755.38 |
| Q72.32 | Congenital absence of left foot and toe(s)<br>Longitudinal deficiency of left toes<br>Longitudinal reduction defect of left toes | 755.38<br>755.39 |
| Q72.33 | Congenital absence of foot and toe(s), bilateral<br>Longitudinal deficiency of bilateral toes<br>Longitudinal reduction defect of bilateral toes | 755.38<br>755.39 |
| Q72.4 | **Longitudinal reduction defect of femur** | |
| Q72.40 | Longitudinal reduction defect of unspecified femur<br>Longitudinal deficiency of unspecified femur<br>Proximal focal deficiency of unspecified femur<br>Absent unspecified femur, congenital | 755.34 |
| Q72.41 | Longitudinal reduction defect of right femur<br>Longitudinal deficiency of right femur<br>Absent right femur, congenital | 755.34 |
| Q72.42 | Longitudinal reduction defect of left femur<br>Longitudinal deficiency of left femur<br>Congenital absence of left femur | 755.34 |
| Q72.43 | Longitudinal reduction defect of femur, bilateral<br>Longitudinal deficiency of femur, bilateral<br>Absent bilateral femurs, congenital | 755.34 |
| Q72.5 | **Longitudinal reduction defect of tibia** | |
| Q72.50 | Longitudinal reduction defect of unspecified tibia<br>Congenital absence of tibia<br>Longitudinal deficiency of tibia<br>Tibial hemimelia | 755.36 |
| Q72.51 | Longitudinal reduction defect of right tibia<br>Longitudinal deficiency of right tibia<br>Congenital absence of right tibia<br>Tibial hemimelia, right | 755.36 |

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| Q72.52 | Longitudinal reduction defect of left tibia<br>Longitudinal deficiency of left tibia<br>Congenital absence of left tibia<br>Tibial hemimelia, left | 755.36 |
| Q72.53 | Longitudinal reduction defect of tibia, bilateral<br>Congenital absence of tibia, bilateral<br>Agenesis of tibia, bilateral<br>Tibial hemimelia, bilateral | 755.36 |
| **Q72.6** | **Longitudinal reduction defect of fibula**<br>**Congenital absence of fibula** | |
| Q72.60 | Longitudinal reduction defect of unspecified fibula<br>Congenital absence of fibula<br>Fibular hemimelia<br>Longitudinal deficiency of fibula<br>Agenesis of unspecified fibula | 755.37 |
| Q72.61 | Longitudinal reduction defect of right fibula<br>Longitudinal deficiency of right fibula<br>Congenital absence of right fibula<br>Agenesis of right fibula | 755.37 |
| Q72.62 | Longitudinal reduction defect of left fibula<br>Longitudinal deficiency of left fibula<br>Congenital absence of left fibula<br>Agenesis of left fibula | 755.37 |
| Q72.63 | Longitudinal reduction defect of fibula, bilateral<br>Bilateral longitudinal deficiency of fibulas<br>Congenital absence of fibula, bilateral<br>Bilateral agenesis of fibula | 755.37 |
| **Q72.7** | **Split foot**<br>**Cleft foot**<br>**Clawfoot or lobster claw foot** | |
| Q72.70 | Split foot, unspecified lower limb | 755.67 |
| Q72.71 | Split foot, right lower limb<br>Right split foot | 755.67 |
| Q72.72 | Split foot, left lower limb<br>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<br>Brachydactyly of right toes<br>Longitudinal deficiency of right leg<br>Longitudinal reduction defect of right leg | 755.30 |
| Q72.892 | Other reductions defects of left lower limb<br>Brachydactyly of left toes<br>Longitudinal deficiency of left leg<br>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<br>Longitudinal deficiency of lower limb<br>Longitudinal reduction defect of leg | 755.30 |
| **Q72.9** | **Unspecified reduction defect of lower limb** | |

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| 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)<br>Phocomelia, NOS | 755.4 |
| Q73.8 | Other reduction defects of unspecified limb(s) (must specify)<br>Ectromelia of limb, NOS<br>Hemimelia of limb, NOS<br>Longitudinal reduction deformity of unspecified limb(s)<br>Reduction defect of limb, NOS<br>Absent digits, NOS<br>Amputation of unspecified limb<br>Unspecified reduction defect of unspecified limb | 755.4 |
| **Q74** | **Other congenital malformations of limb(s)**<br>**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)<br>Accessory carpal bones<br>Cleidocranial dysostosis<br>Congenital abnormal fusion of carpal bone<br>Congenital carpal coalition<br>Congenital elevation of scapula<br>Congenital glenoid dysplasia<br>Congenital hypoplasia of ulna<br>Congenital pseudarthrosis of clavicle<br>Congenital trigger finger and trigger thumb<br>Madelung's deformity<br>Radioulnar synostosis<br>Sprengel's deformity<br>Triphalangeal thumb<br>Anomalies of hand (must specify)<br>Anomalies of elbow and upper arm (must specify)<br>Anomalies of shoulder (must specify)<br>Anomalies of wrist (must specify)<br>Bifid thumb<br>Cubitus valgus, congenital<br>Cubitus varus, congenital<br>Hyperextensibilty of upper limb<br>Hypoplasia of arms<br>Hypoplasia of upper limb<br>Other specified deformity of hands<br>Radioulnar dysostosis<br>*(m) Camptodactyly*<br>*(m) Clinodactyly*<br>*(m) Macrodactyly of finger(s)*<br>*(m) Anomalies of finger*<br>*(m) Brachydactyly*<br>*(m) Incurving fingers*<br>*(m) Long fingers*<br>*(m) Long fingers and toes*<br>*(m) Overlapping fingers*<br>*(m) Tapered fingers* | 755.53<br>755.54<br>755.56<br>755.57<br>755.59 |
| Q74.1 | Congenital malformation of knee (and joint) (must specify)<br>Bifid patella<br>Bipartite patella (kneecap) | 755.64 |

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| | Congenital absence of patella<br>Congenital dislocation of patella (knee-cap)<br>Congenital genu valgum (knock-knee)<br>Congenital genu varum (bowleg)<br>Rudimentary patella<br>**Excludes 1**: congenital dislocation of knee (Q68.2), congenital genu recurvatum (Q68.2), nail patella syndrome (Q87.2) | |
| **Q74.2** | Other congenital malformations of lower limb(s), including pelvic girdle (must specify)<br>Accessory tarsal bones<br>Accessory tarsal navicular bone<br>Congenital fusion of sacroiliac joint<br>Congenital malformation of ankle joint<br>Congenital malformation of sacroiliac joint<br>Anomalies of ankle (must specify)<br>Anomalies of pelvis (must specify)<br>Anomalies of upper leg (must specify)<br>Congenital deformity of sacroiliac (joint)<br>Deformity of leg, NOS<br>Dorsiflexion of foot<br>Femoral hypoplasia syndrome<br>Hypoplasia of feet<br>Other specified anomaly of lower limb (must specify)<br>*(m) Congenital internal tibial torsion*<br>*(m) Macrodactylia of toe(s)*<br>*(m) Anomalies of lower leg*<br>*(m) Congenital angulation of tibia*<br>**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)<br>Larsen's syndrome | 755.8 |
| **Q74.9** | Unspecified congenital malformations of limb(s) (try not to use; must specify)<br>Congenital anomaly of limb(s), NOS<br>Dysplasia of limb<br>Anomalies of forearm, NOS<br>Other anomaly of whole arm<br>Unspecified anomalies of upper limb | 755.9 |
| **Q75** | **Other congenital malformations of skull and face bones**<br>**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<br>Acrocephaly<br>Complex craniosynostosis<br>Craniosynostosis, NOS<br>Imperfect fusion of skull<br>Simple craniosynostosis<br>Basilar craniosynostosis<br>Closed skull sutures, NOS<br>Cloverleaf skull (Keeblatt)<br>Coronal craniosynostosis<br>Kleeblattschädel<br>Lambdoidal<br>Metopic craniosynostosis<br>Premature closure of cranial sutures<br>Sagittal craniosynostosis | 756.0 |

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| **Q75.1** | Craniofacial dysostosis<br>Crouzon's disease<br>Crouzon syndrome | 756.0 |
| **Q75.2** | Hypertelorism<br>Orbital hypertelorism<br>Wide set eyes | 756.0 |
| **Q75.3** | Macrocephaly<br>Macroencephaly | 756.0 |
| **Q75.4** | Mandibulofacial dysostosis<br>Franceschetti syndrome<br>Treacher Collins syndrome | 756.0 |
| **Q75.5** | Oculomandibular Dysostosis | 756.0 |
| **Q75.8** | Other specified congenital malformations of skull and face bones (must specify)<br>Asymmetry of head (skull bones)<br>Absence of skull bone, congenital<br>Absence, cranial bone<br>Basilar skull invagination<br>Congenital deformity of forehead<br>Delayed closure of anterior fontanel<br>Forehead deformity, congenital<br>Invagination of basilar skull<br>Late closure of anterior fontanel<br>Oculoauricular vertebral dysplasia<br>Platybasia<br>Cleft mandible malar hypoplasia<br>Goldenhar syndrome<br>Hallermann-Streiff syndrome<br>Hemifacial microsomia<br>Oculoauriculovertebral (OAV) dysplasia<br>Oculomandibulofacial syndrome<br>Orbital dystopia<br>Other craniofacial syndromes<br>Prominent occiput<br>*(m) Bony occipital spur*<br>*(m) Frontal bossing*<br>*(m) Hypotelorism*<br>*(m) Localized skull defects*<br>*(m) Midfacial hypoplasia* | 756.0 |
| **Q75.9** | Congenital malformation of skull and face bones, unspecified (try not to use; must specify)<br>Congenital anomaly of face bones, NOS<br>Congenital anomaly of skull, NOS | 756.0 |
| **Q76** | **Congenital malformations of spine and bony thorax**<br>**Excludes 1:** congenital musculoskeletal deformities of spine and chest (Q67.5 - Q67.8) | |
| **Q76.0** | Spina bifida occulta<br>**Excludes 1:** meningocele (spinal) (Q05.-), spina bifida (aperta) (cystica) (Q05.-) | 756.17 |
| **Q76.1** | Klippel-Feil syndrome<br>Cervical fusion syndrome<br>Klippel feil sequence<br>Thoracolumbar kissing spine<br>Wildervanck syndrome<br>Cervicooculaoaccoustic syndrome | 756.16 |
| **Q76.2** | Congenital spondylolisthesis<br>Congenital spondylolysis<br>Congenital cervical spondylolisthesis<br>Congenital lumbar spondylolisthesis<br>Congenital lumbosacral spondylolysis | 756.12 |

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| | Prespondylolisthesis, lumbosacral<br>Spondyloysis, lumbosacral<br>**Excludes 1:** spondylolisthesis (acquired) (M43.1-), spondylolysis (acquired) (M43.0-) | |
| Q76.3 | Congenital scoliosis due to congenital bony malformation<br>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)<br>Agenesis of sacrum<br>Aplasia odontoid<br>Atlanto-occipital fusion<br>Caudal agenesis (incomplete development of lower spine)<br>Congenital absence of vertebra, NOS<br>Congenital fusion of spine, NOS<br>Congenital malformation of lumbosacral (joint) (region), NOS<br>Congenital malformation of spine, NOS<br>Dysgenesis, odontoid<br>Dysraphism, congenital<br>Dysraphysm, cervical (neck) spine<br>Fusion of atlantooccipital spine<br>Hemivertebra, NOS<br>Malformation of spine, NOS<br>Occipitoatlantoaxial spine ankylosis<br>Platyspondylisis, NOS<br>Sacral agenesis<br>Supernumerary vertebra, NOS<br>Agenesis of cervical vertebrae<br>Agenesis of lumbar vertebrae<br>Agenesis of thoracic vertebrae<br>Hemivertebrae, cervical vertebrae<br>Hemivertebrae, thoracic vertebrae<br>Hemivertebrae, lumbar vertebrae<br>Sacral mass, NOS<br>Sacrococcygeal anomaly | 756.19<br>756.13<br>756.15<br>756.14 |
| Q76.5 | *(m) Cervical rib*<br>*(m) Supernumerary rib in cervical region* | 756.2 |
| Q76.6 | Other congenital malformations of ribs (must specify)<br>Accessory rib<br>Congenital absence of rib<br>Congenital fusion of ribs<br>Congenital malformation of ribs, NOS<br>**Excludes 1:** short rib syndrome (Q77.2) | 756.3 |
| Q76.7 | Congenital malformations of sternum (must specify)<br>Congenital absence of sternum<br>Sternum bifidum | 756.3 |

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| | Anomalies of thoracic cage, unspecified<br>Congenital fissure of sternum<br>Double ossification center in the manubrium<br>Short sternum | |
| 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**<br>**Excludes 1:** mucopolysaccharidosis (E76.0-E76.3)<br>**Excludes 2:** congenital myotonic chondrodystrophy (G71.13) | |
| Q77.0 | Achondrogenesis<br>Hypochondrogenesis | 756.4 |
| Q77.1 | Thanatophoric short stature<br>Thanatophoric dysplasia | 756.4 |
| Q77.2 | Short rib syndrome<br>Asphyxiating thoracic dysplasia (Jeune)<br>Short rib dysplasia | 756.3 |
| Q77.3 | Chondrodysplasia punctate<br>Chondrodysplasia punctata (stippled epiphyses) group<br>Conradi syndrome<br>**Excludes 1:** Rhizomelic chondrodysplasia punctata (E71.43) | 756.59 |
| Q77.4 | Achondroplasia<br>Hypochondroplasia<br>Osteosclerosis congenital | 756.4 |
| Q77.5 | Diastrophic dysplasia | 756.4 |
| Q77.6 | Chondroectodermal dysplasia<br>Ellis-van Creveld syndrome | 756.55 |
| Q77.7 | Spondyloepiphyseal dysplasia<br>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**<br>**Excludes 2:** congenital myotonic chondrodystrophy (G71.13) | |
| Q78.0 | Osteogenesis imperfecta<br>Brittle bones disease<br>Fragilitas ossium<br>Osteopsathyrosis | 756.51 |
| Q78.1 | Polyostotic fibrous dysplasia<br>Albright (McCune) (Sternberg) syndrome<br>Osteitis fibrosa disseminata | 756.54 |
| Q78.2 | Osteopetrosis<br>Albers-Schönberg syndrome<br>Osteosclerosis, NOS<br>Marble bones | 756.52 |
| Q78.3 | Progressive diaphyseal dysplasia (multiple epiphyseal dysplasia)<br>Camurati-Engelmann syndrome<br>Engelmann syndrome | 756.56 |
| Q78.4 | Enchondromatosis<br>Maffucci's syndrome<br>Multiple enchondromatosis<br>Ollier's disease | 756.4 |
| Q78.5 | Metaphyseal dysplasia<br>Pyle metaphyseal dysplasia<br>Pyle's syndrome | 756.59 |
| Q78.6 | Multiple congenital exostoses<br>Diaphyseal aclasis | 756.6 |
| Q78.8 | Other specified osteochondrodysplasia<br>Dysplasia of epiphysis | 756.59 |

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| | Epiphyseal dysplasia<br>Homozygous Leri Weill dyschondrosteosis syndrome<br>Langer mesomelic dysplasia syndrome<br>Osteopoikilosis<br>Melnick-needles syndrome | |
| Q78.9 | Osteochondrodysplasia, unspecified<br>Chondrodysplasia<br>Chondrodystrophy, NOS<br>Osteodystrophy, NOS | 756.50 |
| Q79 | **Congenital malformations of musculoskeletal system, NEC**<br>**Excludes 2:** congenital (sternomastoid) torticollis (Q68.0) | |
| Q79.0 | Congenital diaphragmatic hernia<br>**Excludes 1:** congenital hiatus hernia (Q40.1) | 756.6 |
| Q79.1 | Other congenital malformations of diaphragm (must specify)<br>Absence of diaphragm<br>Congenital malformation of diaphragm, NOS<br>Eventration of diaphragm | 756.6 |
| Q79.2 | Exomphalos<br>Omphalocele<br>**Excludes 1:** umbilical hernia (K42.-) | 756.72 |
| Q79.3 | Gastroschisis | 756.73 |
| Q79.4 | Prune belly syndrome<br>Congenital prolapse of bladder mucosa<br>Eagle-Barrett syndrome<br>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)<br>*(m) Diastasis recti*<br>*(m) Unspecified anomaly of abdominal wall* | 756.79 |
| Q79.6 | Ehlers-Danlos syndromes<br>Cutis laxa hyperelastica | |
| Q79.60 | Ehlers-Danlos syndrome, unspecified | 756.83 |
| Q79.61 | Classical Ehlers-Danlos syndrome<br>Classical EDS (cEDS) | 756.83 |
| Q79.62 | Hypermobile Ehlers-Danlos syndrome<br>Hypermobile EDS (hEDS) | 756.83 |
| Q79.63 | Vascular Ehlers-Danlos syndrome<br>Vascular EDS (vEDS) | 756.83 |
| Q79.69 | Other Ehlers-Danlos syndromes (must specify) | 756.83 |
| Q79.8 | Other congenital malformations of musculoskeletal system (must specify)<br>Absence of muscle<br>Absence of pectoral muscle, congenital<br>Absence of tendon<br>Accessory muscle<br>Amyoplasia, disruptive sequence<br>Amyotrophia congenital<br>Congenital constricting bands<br>Congenital shortening of tendon<br>Contracture of gastrocnemius, congenital<br>Hereditary osteoonychodysplasia<br>Nail-patella syndrome<br>Poland anomalad<br>Poland syndrome<br>Popliteal pterygium syndrome<br>Schwartz Jampel syndrome<br>Waardenburg syndrome | 756.9<br>756.81<br>756.82 |

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| | Hypoplastic Depressor Angularis Muscle (face)<br>Other absent or hypoplastic muscle<br>Other specified anomaly muscle, tendon, fascia, connective tissue | |
| **Q79.9** | Congenital malformations of musculoskeletal system, unspecified (try not to use; must specify)<br>Unspecified anomaly of bone<br>Unspecified anomaly of cartilage<br>Unspecified anomaly of connective tissue<br>Unspecified anomaly of muscle<br>Unspecified anomaly of musculoskeletal system<br>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<br>Placental sulfatase deficiency (X-linked steryl-sulfatase deficiency) in a female | 757.1 |
| Q80.2 | Lamellar ichthyosis<br>Collodion baby | 757.1 |
| Q80.3 | Congenital bullous ichthyosiform erythroderma<br>Ichthyosiform erythroderma | 757.1 |
| Q80.4 | Harlequin fetus | 757.1 |
| Q80.8 | Other congenital ichthyosis | 757.1 |
| Q80.9 | Congenital ichthyosis, unspecified<br>Keratitis ichthyosis and deafness (KID) syndrome<br>Netherton syndrome | 757.1 |
| Q81 | **Epidermolysis bullosa** | |
| Q81.0 | Epidermolysis bullosa simplex<br>**Excludes 1:** Cockayne's syndrome (Q87.19) | 757.39 |
| Q81.1 | Epidermolysis bullosa letalis<br>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**<br>**Excludes 1:** acrodermatitis enteropathica (E83.2), congenital erythropoietic porphyria E80.0), pilonidal cyst or sinus (L05.-), Sturge-Weber (-Dimitri) syndrome (Q85.8) | |
| Q82.0 | Hereditary lymphedema<br>Hereditary edema of leg(s)<br>Meige syndrome<br>Primary congenital lymphedema<br>Hereditary trophedema<br>Milroy's disease | 757.0 |
| Q82.1 | Xeroderma pigmentosum | 757.33 |
| Q82.2 | Congenital cutaneous mastocytosis<br>Congenital diffuse cutaneous mastocytosis<br>Congenital maculopapular cutaneous mastocytosis<br>Congenital urticaria pigmentosa<br>Urticaria pigmentosa<br>**Excludes 1:** cutaneous mastocytosis NOS (D47.01), diffuse cutaneous mastocytosis (with onset after newborn period) (D47.01),<br>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)<br>Hay Wells syndrome of ectodermal dysplasia<br>X-linked type ectodermal dysplasia<br>Other specified ectodermal dysplasias<br>**Excludes 1:** Ellis-van Creveld syndrome (Q77.6) | 757.31 |
| Q82.5 | *(m) Congenital non-neoplastic nevus*<br>*(m) Birthmark, NOS*<br>*(m) Flammeus nevus*<br>*(m) Nevus anemicus*<br>*(m) Portwine nevus*<br>*(m) Sanguineous nevus*<br>*(m) Strawberry nevus*<br>*(m) Vascular nevus, NOS* | 757.32 |

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| | *(m) Verrucous nevus*<br>*(m) Angel bite or kiss*<br>*(m) Salmon patch*<br>*(m) Stork bites*<br>*(m) Vascular hamartomas*<br>**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*<br>*(m) Parasacral dimple*<br>**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)<br>Acrokeratosis verruciformis<br>Benign familial pemphigus [Hailey-Hailey disease]<br>Bloom syndrome<br>Congenital poikiloderma<br>Cutis laxa (hyperelastica)<br>Goltz syndrome<br>Goltz-Gorlin (dermal hypoplasia) syndrome<br>Howel Evans syndrome<br>Inherited keratosis palmaris et plantaris<br>Keratosis follicularis [Darier-White]<br>Absence of skin<br>Cutis aplasia<br>*(m) Abnormal palmar creases*<br>*(m) Darier disease*<br>*(m) Mongolian spot*<br>*(m) Porokeratosis*<br>*(m) Cutis marmorata*<br>*(m) Cutis marmorata telangiectatica congenital (CMTC)*<br>*(m) Depigmented or hyperpigmented spot*<br>*(m) Extra or absent creases*<br>*(m) Hyperpigmented areas*<br>*(m) Sidney line*<br>*(m) Simian creases, transverse palmar creases*<br>*(m) Single transverse palmar crease*<br>*Congenital scars and non-ear skin tags are not reportable*<br>**Excludes 1:** Ehlers-Danlos syndrome (Q79.6-) | 757.39 |
| **Q82.9** | Congenital malformation of skin, unspecified | 757.39 |
| **Q83** | **Congenital malformations of breast**<br>**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*<br>*(m) Supernumerary breast* | 757.6 |
| **Q83.2** | Absent nipple | 757.6 |
| **Q83.3** | *(m) Accessory nipple (congenital)*<br>*(m) Supernumerary nipple* | 757.6 |
| **Q83.8** | Other congenital malformations of breast (must specify)<br>Congenital absence of breast<br>Hypoplastic breast with hypoplastic nipple<br>*(m) Asymmetry of breasts, congenital*<br>*(m) Widely spaced nipples*<br>*(m) Small nipple (hypoplastic)* | 757.6 |
| **Q83.9** | Congenital malformation of breast, unspecified (try not to use) | 757.6 |

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| **Q84** | **Other congenital malformations of integument** | |
| **Q84.0** | *(m) Congenital alopecia*<br>*(m) Congenital atrichosis* | 757.4 |
| **Q84.1** | *(m) Congenital morphological disturbances of hair, NEC*<br>*(m) Beaded hair*<br>*(m) Monilethrix*<br>*(m) Pili annulati*<br>*(m) Pili torti*<br>*(m) Twisted hair*<br>*(m) Taenzer's hair*<br>**Excludes 1:** Menkes' kinky hair syndrome (E83.0) | 757.4 |
| **Q84.2** | *(m) Other congenital malformations of hair (must specify)*<br>*(m) Congenital hypertrichosis*<br>*(m) Congenital malformation of hair, NOS*<br>*(m) Persistent lanugo*<br>*(m) Hirsutism*<br>*(m) Aberrant scalp hair patterning*<br>*(m) Low hair line*<br>*(m) Other specified anomaly of hair* | 757.4 |
| **Q84.3** | *(m) Anonychia*<br>*(m) Absent nails*<br>**Excludes 1:** nail patella syndrome (Q87.2) | 757.5 |
| **Q84.4** | *(m) Congenital leukonychia* | 757.5 |
| **Q84.5** | *(m) Enlarged and hypertrophic nails*<br>*(m) Congenital onychauxis*<br>*(m) Pachyonychia*<br>*(m) Hyperconvex or thickened toenails* | 757.5 |
| **Q84.6** | Hypoplastic, hyperconvex, or duplicated fingernails<br>*(m) Other congenital malformations of nails (must specify)*<br>*(m) Congenital clubnail*<br>*(m) Congenital koilonychias*<br>*(m) Congenital malformations of nail, NOS*<br>*(m) Hypoplastic (small) fingernails and/or toenails* | 757.5 |
| **Q84.8** | Other specified congenital malformations of integument (must specify)<br>Aplasia cutis congenital<br>*(m) Scalp defects* | 757.8 |
| **Q84.9** | *(m) Congenital malformation of integument, unspecified*<br>*(m) Congenital anomaly of integument, NOS*<br>*(m) Congenital deformity of integument, NOS*<br>*(m) Unspecified anomaly of hair*<br>*(m) Unspecified anomaly of nail* | 757.9 |
| **Q85** | **Phakomatoses, NEC**<br>**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<br>Von Recklinghausen disease | 237.71 |
| **Q85.02** | Neurofibromatosis, Type 2<br>Acoustic neurofibromatosis | 237.72 |
| **Q85.03** | Schwannomatosis | 237.73 |
| **Q85.09** | Other neurofibromatosis | 237.79 |
| **Q85.1** | Tuberous sclerosis<br>Bourneville's disease<br>Epiloia | 759.5 |
| **Q85.8** | Other phakomatoses, NEC (must specify)<br>Peutz-Jeghers Syndrome<br>Sturdge-Weber (-Dimitri) syndrome | 759.6 |

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| | Von Hippel-Lindau syndrome<br>Neuro-cutaneous syndrome<br>**Excludes 1:** Meckel-Gruber syndrome (Q61.9) | |
| **Q85.9** | Phakomatosis, unspecified<br>Hamartosis, NOS<br>Hamartoma of retina<br>Vascular hamartoma of skin | 759.6 |
| **Q86** | **Congenital malformations syndromes due to known exogenous causes, NEC**<br>**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) | 760.71 |
| **Q86.1** | Fetal hydantoin syndrome<br>Meadow's syndrome | 760.77 |
| **Q86.2** | Dysmorphism due to warfarin | 760.79 |
| **Q86.8** | Other congenital malformation syndromes due to known exogenous causes | 760.76 |
| **Q87** | **Other specified congenital malformation syndromes affecting multiple systems**<br>****Use Additional code(s) to identify all associated manifestations** | |
| **Q87.0** | Congenital malformation syndromes predominantly affecting facial appearance (must specify)<br>Acrocephalopolysyndactyly<br>Acrocephalosyndactyly [Apert syndrome]<br>Branchio oto renal syndrome [Melnick-Fraser syndrome]<br>Carpenter's syndrome<br>Cryptophthalmos syndrome<br>Cyclopia<br>Freeman Sheldon syndrome<br>Goldenhar syndrome (oculo-auriculovertebral)<br>Gorlin-Chaudhry-Moss syndrome<br>Hallerman-Streiff syndrome<br>Moebius syndrome<br>Oro-facial-digital syndrome<br>Oto palato digital (OPD) syndrome<br>Pfeiffer syndrome<br>Pierre Robin syndrome<br>Robin syndrome<br>Saethre Chotzen syndrome<br>Whistling face<br>Antley-Bixler<br>Branchio-oculo-facial syndrome | 755.55<br>756.0<br>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)<br>Aarskog syndrome<br>Bonnevie-Ullrich syndrome<br>Cornelia de Lange syndrome<br>Cockayne syndrome<br>De Lange syndrome<br>Dubowitz syndrome<br>Noonan syndrome<br>Robinow-Silverman-Smith syndrome<br>Russell-Silver syndrome<br>Seckel syndrome<br>Sjogren-Larsson syndrome | 759.89 |
| **Q87.2** | Congenital malformation syndromes predominantly involving limbs (must specify)<br>Holt-Oram syndrome<br>Klippel-Trenaunay-Weber syndrome | 759.89 |

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| | Nail patella syndrome<br>Rubinstein-Taybi syndrome<br>Sirenomelia syndrome<br>Thrombocytopenia with absent radius [TAR] syndrome<br>VATER syndrome | |
| Q87.3 | Congenital malformation syndromes involving early overgrowth (must specify)<br>Beckwith-Wiedemann syndrome<br>Proteus syndrome<br>Sotos syndrome<br>Weaver syndrome | 759.89 |
| Q87.4 | **Marfan's syndrome** | |
| Q87.40 | Marfan's syndrome, unspecified | 759.82 |
| Q87.41 | **Marfan's syndrome with cardiovascular manifestations** | |
| Q87.410 | Marfan's syndrome with aortic dilation | 759.82 |
| Q87.418 | Marfan's syndrome with other cardiovascular manifestations | 759.82 |
| Q87.42 | Marfan's syndrome with ocular manifestations | 759.82 |
| Q87.43 | Marfan's syndrome with skeletal manifestation | 759.82 |
| Q87.5 | Other congenital malformation syndromes with other skeletal changes | 759.89 |
| Q87.8 | **Other specified congenital malformation syndromes, NEC**<br>**Excludes 1:** Zellweger syndrome (E71.510) | |
| Q87.81 | Alport syndrome<br>Hereditary nephritis | 759.89 |
| Q87.82 | Arterial tortuosity syndrome | 759.89 |
| Q87.89 | Other specified congenital malformation syndromes, NEC (must specify)<br>Basal cell nevus syndrome<br>Gorlin syndrome<br>Laurence-Moon (-Bardet)-Biedl syndrome | 759.89 |
| Q89 | **Other congenital malformations, NEC** | |
| Q89.0 | **Congenital absence and malformations of spleen**<br>**Excludes 1:** isomerism of atrial appendages (with asplenia or polysplenia) (Q20.6) | |
| Q89.01 | Asplenia (congenital) | 759.0 |
| Q89.09 | Congenital malformations of spleen<br>Congenital splenomegaly | 759.0 |
| Q89.1 | Congenital malformations of adrenal gland<br>**Excludes 1:** adrenogenital disorders (E25.-), congenital adrenal hyperplasia (E25.0) | 759.1 |
| Q89.2 | Congenital malformations of other endocrine glands<br>Congenital malformation of parathyroid or thyroid gland<br>Persistent thyroglossal duct<br>Thyroglossal cyst<br>**Excludes 1:** congenital goiter (E03.0), congenital hypothyroidism (E03.1) | 759.2 |
| Q89.3 | Situs inversus<br>Dextrocardia with situs inversus<br>Mirror-image atrial arrangement with situs inversus<br>Situs inversus or transversus abdominalis<br>Situs inversus or transversus thoracis<br>Transposition of abdominal viscera<br>Transposition of thoracic viscera<br>**Excludes 1:** dextrocardia NOS (Q24.0) | 759.3 |
| Q89.4 | Conjoined twins<br>Craniopagus<br>Cicephaly<br>Pygopagus<br>Thoracopagus | 759.4 |
| Q89.7 | Multiple congenital malformations, NEC (try not to code)<br>Multiple congenital anomalies, NOS<br>Multiple congenital deformities, NOS<br>**Excludes 1:** congenital malformation syndromes affecting multiple systems (Q87.-) | 759.7 |

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| **Q89.8** | Other specified congenital malformations (must specify)<br>Charge syndrome<br>Kabuki syndrome<br>Stickler syndrome<br>Williams syndrome<br>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** | |
| Q90.0 | Trisomy 21, nonmosaicism (meiotic nondisjunction)<br>Karyotype trisomy 21 (47,XX or XY,+21) | 758.0 |
| Q90.1 | Trisomy 21, mosaicism (mitotic nondisjunction)<br>(46,XX/47,XX+21 or 46,XY/47,XY+21) | 758.0 |
| Q90.2 | Trisomy 21, translocation<br>t(13:21), t(14:21), t(15:21), t(21:22)<br>Translocation trisomy duplication of a 21[t(21:21)] | 758.0 |
| Q90.9 | Down syndrome, unspecified<br>Trisomy 21, NOS | 758.0 |
| Q91 | **Trisomy 18 and Trisomy 13** | |
| Q91.0 | Trisomy 18, nonmosaicism (meiotic nondisjunction)<br>Edwards syndrome, karyotype trisomy 18(47,XX or XY+18) | 758.2 |
| Q91.1 | Trisomy 18, mosaicism (mitotic nondisjunction)<br>(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<br>Trisomy 18, complete<br>Edwards syndrome, NOS | 758.2 |
| Q91.4 | Trisomy 13, nonmosaicism (meiotic nondisjunction)<br>Patau syndrome, karyotype trisomy 13(47,XX or XY,+13) | 758.1 |
| Q91.5 | Trisomy 13, mosaicism nondisjunction)<br>(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<br>Patau syndrome, NOS | 758.1 |
| Q92 | **Other trisomies and partial trisomies of the autosome, NEC**<br>**Includes:** unbalanced transolocations and insertions<br>**Excludes 1:** trisomies of chromosomes 13, 18, 21 (Q90-Q91) | |
| Q92.0 | Whole chromosomes trisomy, nonmosaicism (meiotic nondisjunction)<br>• Trisomy 1<br>• Trisomy 2<br>• Trisomy 3<br>• Trisomy 4<br>• Trisomy 5<br>• Trisomy 6<br>• Trisomy 7<br>• Trisomy 8<br>• Trisomy 9<br>• Trisomy 10<br>• Trisomy 11<br>• Trisomy 12<br>• Trisomy 14<br>• Trisomy 15<br>• Trisomy 16<br>• Trisomy 17<br>• Trisomy 19<br>• Trisomy 20<br>• Trisomy 22 | 758.5 |
| Q92.1 | Whole chromosome trisomy, mosaicism (mitotic nondisjunction)<br>Mosaic trisomy of any chromosome except 21, 13 or 18 | 758.5 |
| Q92.2 | Partial trisomy<br>Less than whole arm duplicated<br>Whole arm or more duplicated<br>**Excludes 1:** partial trisomy due to unbalanced translocation (Q92.5) | 758.2 |
| Q92.5 | Duplications with other complex rearrangements<br>Partial trisomy due to unbalanced translocations<br>Partial trisomy syndromes (unbalanced, not defined)<br>**Also code any associated deletions due to unbalanced translocations, inversions and insertions | 758.5 |
| Q92.6 | **Marker chromosomes**<br>Trisomies due to dicentrics | |

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| | Trisomies due to extra rings<br>Trisomies due to isochromosomes<br>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<br>Triploidy, 69 chromosomes<br>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)<br>Duplication identified by fluorescence in situ hybridization (FISH)<br>Duplications identified by in situ hybridization (ISH)<br>Duplications seen only at prometaphase | 758.5 |
| Q92.9 | Trisomy and partial trisomy of autosomes, unspecified<br>Trisomy, NOS | 758.5 |
| **Q93** | **Monosomies and deletions from the autosomes, NEC** | |
| Q93.0 | Whole chromosomes monosomy, nonmosaicism (meiotic nondisjunction)<br>E.g. (45, XX, -21) or (45,XY,-21) for any chromosome #1-22 | 758.5 |
| Q93.1 | Whole chromosome monosomy, mosaicism (mitotic nondisjunction)<br>Monosomy 21 mosaicism: e.g.: (46,XX/45,XX,-21) or (46,XY/45,XY,-21)<br>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<br>4p deletion syndrome<br>Del4p15<br>Deletion 4p15-16<br>Wolff-Hirschorn syndrome | 758.39 |
| Q93.4 | Deletion of short arm of chromosome 5<br>Cri-du-chat syndrome (5p-) | 758.31 |
| **Q93.5** | **Other deletions of part of a chromosome** | |
| Q93.51 | Angelman syndrome | |
| Q93.59 | Other deletions of part of a chromosome<br>Antimongolism syndrome: partial or total deletion of 21 [del (21)]<br>Deletion of long arm 13: (del(13q)) or 13q-<br>Deletion of long arm 17: (del(17q)) or 17q-<br>Deletion of long arm 18: (del(18q)) or 18q-<br>Deletion of short arm 17: (del(17p)) or 17p-<br>Deletion of short arm 18: (del(18p)) or 18p- | |
| Q93.7 | Deletions with other complex rearrangements<br>Deletions due to unbalanced translocations, inversions and insertions<br>**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<br>Deletion 22q11.2 | 758.32 |
| Q93.82 | Williams syndrome | |
| Q93.88 | Other microdeletions (must specify)<br>Miller-Dieker syndrome<br>Smith-Magenis syndrome | 758.33 |
| Q93.89 | Other deletions from the autosomes (chromosomes 1-22) (must specify)<br>Deletions identified by fluorescence in situ hybridization (FISH)<br>Deletions identified by in situ hybridization (IS)<br>Deletions seen only at prometaphase | 758.39 |
| Q93.9 | Deletion from autosomes, unspecified (try not to use)<br>Autosomal deletion syndrome<br>Chromosomal deletion | 758.39 |

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| Q95 | **Balanced rearrangements and structural markers, NEC**<br>**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<br>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**<br>**XO syndrome**<br>**Bonnevie-Ullrich syndrome**<br>**[Only applicable to female patients.]**<br>**Excludes 1:** Noonan syndrome (Q87.19), pure gonadal dysgenesis (Q99.1) | |
| Q96.0 | Karyotype 45,X<br>Turner's phenotype, karyotype 45,X | 758.6 |
| Q96.1 | Karyotype 46, X iso (Xq)<br>Karyotype 46, isochromosome Xq | 758.6 |
| Q96.2 | Karyotype 46, X with abnormal sex chromosome, except iso (Xq)<br>Karyotype 46, X with abnormal sex chromosome, except isochromosome Xq<br>Turner syndrome variant, [46,X,r(X)]<br>Turner syndrome variant, [46,Xdel(Xq)]<br>Turner syndrome variant, [46,X,del(Xp)]<br>**[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<br>Turner Syndrome, karyotype unspecified | 758.6 |
| Q97 | **Other sex chromosome abnormalities, female phenotype, NEC**<br>**[Only applicable to female patients.]**<br>**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**<br>**[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<br>48,XXXY<br>48,XXYY<br>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<br>Klinefelter syndrome mosaic (46,XY/47,XXY)<br>Mosaic X/XY, 45X/46XY<br>Mosaic XY/XXY, 46XY/47XXY<br>Mosaic including XXXXY,49XXXXY | 758.81 |

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

## Other Selected Reportable Conditions

| ICD-10 | Notes & Description | ICD-9 |
|---|---|---|
| D82 | **Immunodeficiency associated with other major defects**<br>**Excludes 1:** ataxia telangiectasia [Louis-Bar] (G11.3) | |
| D82.1 | Di George's syndrome Also code 22q deletion separately if reported<br>Pharyngeal pouch syndrome<br>Thymic alymphoplasia<br>Thymic aplasia or hypoplasia with immunodeficiency | 279.11 |
| E25 | **Adrenogenital disorders**<br>Please report cytogenetic diagnosis<br>**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)<br>**Excludes 1:** indeterminate sex and pseudohermaphroditism (Q56), chromosomal abnormalities (Q90-Q99) | |
| E25.0 | Congenital adrenogenital disorders associated with enzyme deficiency<br>Congenital adrenal hyperplasia (CAH)<br>21-Hydroxylase deficiency<br>Salt-losing congenital adrenal hyperplasia | 255.2 |
| E25.8 | Other adrenogenital disorders idiopathic adrenogenital disorder | 255.2 |
| E25.9 | Adrenogenital disorder, unspecified<br>Adrenogenital syndrome NOS | 255.2 |
| E34 | **Other endocrine disorders ** only report if congenital or genetic**<br>**Excludes 1:** pseudohypoparathyroidism (E20.1) | |
| E34.3 | Short stature due to endocrine disorder<br>Laron-type short stature<br>**Excludes 1:** achondroplastic short stature (Q77.4), hypochondroplastic short stature (Q77.4), nutritional short stature (E45), pituitary short stature (E23.0), progeria (E34.8), renal short stature (N25.0), Russell-Silver syndrome (Q87.19), short-limbed stature with immunodeficiency (D82.2), short stature in specific dysmorphic syndromes - code to syndrome, short stature, NOS (R62.52) | 259.4 |
| E34.5 | **Androgen insensitivity syndrome** | |
| E34.50 | Androgen insensitivity syndrome, unspecified | 259.5 |
| E34.51 | Complete androgen insensitivity syndrome<br>Complete androgen insensitivity<br>de Quervain syndrome<br>Goldberg-Maxwell syndrome<br>Testicular feminization | 259.5 |
| E34.52 | Partial androgen insensitivity syndrome<br>Partial androgen insensitivity<br>Reifenstein syndrome | 259.5 |
| E78 | Disorders of lipoprotein metabolism and other lipidemias<br>**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<br>Werdnig-Hoffman disease | 335.0 |
| G12.1 | Other inherited spinal muscular atrophy (must specify)<br>Adult form spinal muscular atrophy<br>Childhood form, type II spinal muscular atrophy<br>Distal spinal muscular atrophy<br>Juvenile form, type III spinal muscular atrophy [Kugelberg-Welander]<br>Progressive bulbar palsy of childhood [Fazio-Londe]<br>Scapuloperoneal form spinal muscular atrophy | 335.11 |

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|---|---|---|
| G12.9 | Spinal muscular atrophy, unspecified | 335.10 |
| G60 | **Hereditary and idiopathic neuropathy** | |
| G60.0 | Hereditary motor and sensory neuropathy<br>Charcot-Marie-Tooth disease<br>Déjérine-Sottas disease<br>Hereditary motor and sensory neuropathy, types I-IV<br>Hypertrophic neuropathy of infancy<br>Peroneal muscular atrophy (axonal type) (hypertrophic type)<br>Roussy-Levy syndrome | 356.0 |
| G60.1 | Refsum disease<br>Infantile Refsum disease | 356.3 |
| G60.2 | Neuropathy in association with hereditary ataxia | 356.0 |
| G71 | **Primary disorders of muscles**<br>**Excludes 2:** arthrogryposis multiplex congenita (Q74.3), metabolic disorders (E70-E88), myositis (M60.-) | |
| G71.0 | Muscular dystrophy | |
| G71.00 | Muscular dystrophy, unspecified (must specify) | 359.1 |
| G71.01 | Duchenne or Becker muscular dystrophy<br>Autosomal recessive, childhood type, muscular dystrophy resembling Duchenne or Becker muscular dystrophy<br>Benign [Becker] muscular dystrophy<br>Severe [Duchenne] muscular dystrophy | 359.1 |
| G71.02 | Facioscapulohumeral muscular dystrophy<br>Scapulohumeral muscular dystrophy | 359.1 |
| G71.09 | Other specified muscular dystrophies (must specify)<br>Benign scapuloperoneal muscular dystrophy with early contractures [Emery-Dreifuss]<br>Congenital muscular dystrophy NOS<br>Congenital muscular dystrophy with specific morphological abnormalities of the muscle fiber<br>Distal muscular dystrophy<br>Limb-girdle muscular dystrophy<br>Ocular muscular dystrophy<br>Oculopharyngeal muscular dystrophy<br>Scapuloperoneal muscular dystrophy | 359.1 |
| G71.1 | **Myotonic disorders** | |
| G71.11 | Myotonic muscular dystrophy<br>Dystrophia myotonica [Steinert]<br>Myotonia atrophica<br>Myotonic dystrophy<br>Proximal myotonic myopathy (PROMM)<br>Steinert disease | 359.21 |
| G71.12 | Myotonia congenita<br>Acetazolamide responsive myotonia congenita<br>Dominant myotonia congenita [Thomsen disease]<br>Myotonia levior<br>Recessive myotonia congenita [Becker disease] | 359.22 |
| G71.13 | Myotonic chondrodystrophy<br>Chondrodystrophic myotonia<br>Congenital myotonic chondrodystrophy<br>Schwartz-Jampel disease | 359.23 |
| G71.19 | Other specified myotonic disorders (must specify)<br>Isaac-Mertens syndrome<br>Myotonia fluctuans<br>Myotonia permanens<br>Neuromyotonia [Isaacs]<br>Paramyotonia congenita (of von Eulenburg)<br>Pseudomyotonia | 359.29 |

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|---|---|---|
| | Symptomatic myotonia | |
| G71.2 | Congenital myopathies<br>Central core disease<br>Fiber-type disproportion<br>Minicore disease<br>Multicore disease<br>Myotubular (centronuclear) myopathy<br>Nemaline myopathy<br>Walker Warburg congenital muscular dystrophy<br>**Excludes 1**: arthrogryposis multiplex congenita (Q74.3) | 359.0 |
| G71.3 | Mitochondrial myopathy, not elsewhere classified<br>**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<br>Congenital hemihypertrophy | 359.89 |
| G71.9 | Primary disorder of muscle, unspecified<br>Hereditary myopathy, NOS | 359.9 |
| G80 | **Cerebral Palsy is Not Reportable** | |
| K40 | **Inguinal Hernia (report for female only)**<br>**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<br>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<br>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<br>Bilateral inguinal hernia, NOS | 550.92 |
| K40.3 | **Unilateral inguinal hernia, with obstruction, without gangrene (female only)**<br>• Inguinal hernia (unilateral) causing obstruction without gangrene<br>• Incarcerated inguinal hernia (unilateral) without gangrene<br>• Irreducible inguinal hernia (unilateral) without gangrene<br>• Strangulated inguinal hernia (unilateral) without gangrene<br>Indicate right, left, or unspecified | |
| K40.30 | Unilateral inguinal hernia, with obstruction, without gangrene, not specified as recurrent<br>Inguinal hernia, with obstruction, NOS<br>Unilateral inguinal hernia, with obstruction, without gangrene, NOS | 550.1 |
| K40.40 | Unilateral inguinal hernia, with gangrene, not specified as recurrent<br>Inguinal hernia with gangrene, NOS<br>Unilateral inguinal hernia with gangrene, NOS | 550 |
| K40.9 | **Unilateral inguinal hernia, without obstruction or gangrene (female only)**<br>Indicate right, left, or unspecified | |
| K40.90 | Unilateral inguinal hernia, without obstruction or gangrene, not specified as recurrent<br>Direct inguinal hernia<br>Indirect inguinal hernia<br>Inguinal hernia, NOS<br>Left direct inguinal hernia<br>Left indirect inguinal hernia<br>Left inguinal hernia<br>Right direct inguinal hernia<br>Right indirect inguinal hernia<br>Right inguinal hernia | 550.9 |
| K41 | **Femoral hernia (female only)** | |
| K41.0 | **Bilateral femoral hernia, with obstruction, without gangrene (female only)**<br>• Femoral hernia (bilateral) causing obstruction, without gangrene | |

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|---|---|---|
| | • Incarcerated femoral hernia (bilateral), without gangrene<br>• Irreducible femoral hernia (bilateral), without gangrene<br>• Strangulated femoral hernia (bilateral), without gangrene | |
| K41.00 | Bilateral femoral hernia, with obstruction, without gangrene, not specified as recurrent<br>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<br>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<br>Bilateral femoral hernia, NOS | 553.02 |
| K41.3 | **Unilateral femoral hernia, with obstruction, without gangrene (female only)**<br>• Femoral hernia (unilateral) causing obstruction, without gangrene<br>• Incarcerated femoral hernia (unilateral), without gangrene<br>• Irreducible femoral hernia (unilateral), without gangrene<br>• Strangulated femoral hernia (unilateral), without gangrene<br>Indicate right, left, or unspecified | |
| K41.30 | Unilateral femoral hernia, with obstruction, without gangrene, not specified as recurrent<br>Femoral hernia, with obstruction NOS<br>Unilateral femoral hernia, with obstruction NOS | 552 |
| K41.4 | **Unilateral femoral hernia, with gangrene (female only)**<br>Indicate right, left, or unspecified | |
| K41.40 | Unilateral femoral hernia, with gangrene, not specified as recurrent<br>Femoral hernia, with gangrene NOS<br>Unilateral femoral hernia, with gangrene NOS | 551 |
| K41.9 | **Unilateral femoral hernia, without obstruction or gangrene (female only)**<br>Indicate right, left, or unspecified | |
| K41.90 | Unilateral femoral hernia, without obstruction or gangrene, not specified as recurrent<br>Femoral hernia, NOS<br>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*<br>*(m) Pilonidal dimple*<br>*(m) Pilonidal cyst NOS*<br>*(m) Postanal dimple*<br>**Excludes 2:** congenital sacral dimple (Q82.6), parasacral dimple (Q82.6) | 685.1 |
| L05.92 | *(m) Pilonidal sinus without abscess*<br>*(m) Coccygeal fistula*<br>*(m) Coccygeal sinus without abscess*<br>*(m) Pilonidal fistula* | 685.1 |
| L81 | ***(m) Other disorders of pigmentation***<br>**Excludes 1:** birthmark NOS (Q82.5), Peutz-Jeghers syndrome (Q85.8)<br>**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***<br>**Excludes 1**: acromegaly (E22.0), Robin's syndrome (Q87.0) | |
| M26.01 | *(m) Maxillary hyperplasia*<br>*(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 |

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|---|---|---|
| M26.06 | *(m) Microgenia* | 524.06 |
| M26.09 | Micrognathia<br>Macrognathia<br>*(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)<br>Retrognathia<br>*(m) Congenital prognathism*<br>*(m) Mandibular prognathia*<br>*(m) Mandibular prognathism*<br>*(m) Maxillary retrognathia*<br>*(m) Maxillary retrognathism*<br>*(m) Prognathism* | 524.19 |
| **P02** | **Newborn (suspected to be) affected by complications of placenta, cord and membranes**<br>**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<br>Code first any current condition in newborn | |
| P02.8 | Newborn (suspected to be) affected by other abnormalities of membranes<br>Amniotic band syndrome<br>Amniotic bands (constricting bands)<br>Streeter's (Simonart's) bands<br>P02.8 is only applicable to newborns of age 0 years. | 762.8 |
| **P35** | **Congenital viral diseases**<br>**Includes:** infections acquired in utero or during birth<br>Only applicable to newborns of age 0 years. | |
| P35.0 | Congenital Rubella Syndrome<br>Rubella, congenital<br>Congenital rubella pneumonitis | 771.0 |
| P35.1 | Congenital cytomegalovirus infection (CMV)<br>Cytomegalovirus infection, congenital | 771.1 |
| P35.2 | Congenital herpesviral [herpes simplex] infection<br>Herpes simplex<br>Encephalitis herpes<br>Meningoencephalitis herpes | 771.2 |
| P35.3 | Congenital viral hepatitis | 771.2 |
| P35.4 | Congenital Zika virus disease<br>**Use Additional Code** to identify manifestations of congenital Zika virus disease | |
| P35.8 | Congenital infection, other specified<br>Congenital varicella [chickenpox] | 771.2 |
| P35.9 | Congenital viral disease, unspecified | 771.2 |
| **P37** | **Other congenital infectious and parasitic diseases**<br>**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<br>P37.0 is only applicable to newborns of age 0 years. | 771.2 |
| P37.1 | Congenital toxoplasmosis<br>Hydrocephalus due to congenital toxoplasmosis<br>P37.1 is only applicable to newborns of age 0 years. | 771.2 |

## Stillbirth Codes (O36.4, Z37)

| ICD-10 | Notes & Description | ICD-9 |
|---|---|---|
| **O36.4** | **Maternal care for intrauterine death**<br>• **Maternal care for intrauterine fetal death NOS**<br>• **Maternal care for intrauterine fetal death after completion of 20 weeks of gestation**<br>• **Maternal care for late fetal death**<br>• **Maternal care for missed delivery**<br>**Only applicable to female maternity patients aged 12 - 55 years inclusive.**<br>**Should only be used on the maternal record - not on the newborn record.**<br>**Use a code under Z3A to document the exact week during the pregnancy.**<br>**Excludes 1:** missed abortion (O02.1), stillbirth (P95) | |
| **O36.4XX0** | Maternal care for intrauterine death, not applicable or unspecified<br>Dead fetus in utero<br>Fetal death from asphyxia and/or anoxia, not clear if noted before or after onset of labor<br>Fetal death from asphyxia or anoxia<br>Pregnancy loss | 656.40<br>656.41<br>656.43 |
| **O36.4XX1** | Maternal care for intrauterine death, fetus 1 | 656.41<br>656.43 |
| **O36.4XX2** | Maternal care for intrauterine death, fetus 2 | 656.41<br>656.43 |
| **O36.4XX3** | Maternal care for intrauterine death, fetus 3 | 656.41<br>656.43 |
| **O36.4XX4** | Maternal care for intrauterine death, fetus 4 | 656.41<br>656.43 |
| **O36.4XX5** | Maternal care for intrauterine death, fetus 5 | 656.41<br>656.43 |
| **O36.4XX9** | Maternal care for intrauterine death, other fetus | 656.41<br>656.43 |
| **P95** | Stillbirth<br>Fetal death of unspecified cause<br>**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 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 | Q51.21 | Q93.51 | G71.0 | G71.02 |
| Q51.2 | Q51.28 | Q93.59 | G71.00 | G71.09 |
| Q51.20 | Q93.5 | Q93.82 | G71.01 | 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 | |

**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 | |

**Changes effective October 2018**

| Code | Defect | Previous classification | Current classification |
|---|---|---|---|
| Q25.6 | Peripheral pulmonic stenosis (PPS) | Minor | Major, only report if ≥2500g at birth |
| Q55.63 | Congenital torsion of penis | Major | Minor |
| Q55.63 | Phimosis | Major | Not Reportable |

- P95 moved to stillbirth section from ‘other selected reportable conditions’ section.

**Additional changes effective October 2019**

| Code | Defect | Previous classification | Current classification |
|---|---|---|---|
| Q55.64 | Hidden penis | Major | Minor |
| Q55.64 | Buried penis | Major | Minor |
| Q04.6 | Congenital choroid plexus cyst | Major | Minor |