CASEFINDING

A. General Casefinding Criteria

Casefinding is the process used to identify potential cases for inclusion in the Birth Defects Registry (BDR). Cases must always meet the following criteria to be included in the BDR.

A. Any infant/fetus born on or after January 1, 1992. (See county inclusion list by year of birth below.)

B. Mother is an Oklahoma resident at time of the infant's birth. (See county inclusion list by year of birth below.)

C. Infant/fetus must be born in or identified as a case through an Oklahoma hospital.

D. Infant/fetus must have an adverse birth outcome that is included of the abstractable diagnoses/condition list. The diagnosis must be made or signs and symptoms must be present prior to the child's second birthday.

E. A case must be abstracted by the child's sixth birthday.

F. The infant must have a gestational age of at least 20 weeks. (NOTE: A fetus of any gestational age or weight from an induced/spontaneous abortion should be abstracted when identified to have a malformation that meets the case definition.

RESIDENT COUNTIES FOR BDR CASE DEFINITION

1992 Births

<table>
<thead>
<tr>
<th>County</th>
<th>Count</th>
</tr>
</thead>
<tbody>
<tr>
<td>Oklahoma</td>
<td>55</td>
</tr>
</tbody>
</table>

1994 Births

<table>
<thead>
<tr>
<th>County</th>
<th>Count</th>
</tr>
</thead>
<tbody>
<tr>
<td>All Oklahoma Counties</td>
<td></td>
</tr>
</tbody>
</table>

1993 Births

<table>
<thead>
<tr>
<th>County</th>
<th>Count</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cleveland</td>
<td>14</td>
</tr>
<tr>
<td>Oklahoma</td>
<td>55</td>
</tr>
<tr>
<td>Tulsa</td>
<td>72</td>
</tr>
<tr>
<td>Code</td>
<td>Description</td>
</tr>
<tr>
<td>--------</td>
<td>-----------------------------------------------------------------------------</td>
</tr>
<tr>
<td>155.000</td>
<td>Neoplasms of the liver <em>(exclude if over 1 year of age)</em></td>
</tr>
<tr>
<td></td>
<td>Includes: hepatoblastoma</td>
</tr>
<tr>
<td></td>
<td>hemangio-epithelioma</td>
</tr>
<tr>
<td>189.000</td>
<td>Wilms' tumor (nephroblastoma) <em>(exclude if over 1 year of age)</em></td>
</tr>
<tr>
<td>190.500</td>
<td>Retinoblastoma <em>(exclude if over 1 year of age)</em></td>
</tr>
<tr>
<td>191.000</td>
<td>Neoplasms of the CNS <em>(exclude if over 1 year of age)</em></td>
</tr>
<tr>
<td></td>
<td>Includes: gliomas</td>
</tr>
<tr>
<td></td>
<td>medulloblastoma</td>
</tr>
<tr>
<td></td>
<td>tectal tumor</td>
</tr>
<tr>
<td>194.000</td>
<td>Neuroblastoma <em>(exclude if over 1 year of age)</em></td>
</tr>
<tr>
<td>208.000</td>
<td>Leukemia, congenital, NOS <em>(exclude if over 1 year of age)</em></td>
</tr>
<tr>
<td>214</td>
<td><strong>Lipoma</strong></td>
</tr>
<tr>
<td>214.000</td>
<td>Lipoma, skin and subcutaneous tissue of face</td>
</tr>
<tr>
<td>214.100</td>
<td>Lipoma, other skin and subcutaneous tissue</td>
</tr>
<tr>
<td>214.200</td>
<td>Lipoma, intrathoracic organs</td>
</tr>
<tr>
<td>214.300</td>
<td>Lipoma, intra-abdominal organs</td>
</tr>
<tr>
<td>214.400</td>
<td>Lipoma, spermatic cord</td>
</tr>
<tr>
<td>214.800</td>
<td>Lipoma, other specified sites</td>
</tr>
<tr>
<td>214.810</td>
<td>Lipoma, lumbar or sacral lipoma, paraspinal lipoma</td>
</tr>
<tr>
<td>214.900</td>
<td>Lipoma, unspecified site</td>
</tr>
<tr>
<td>216</td>
<td><strong>Benign neoplasm of skin (exclude if isolated)</strong></td>
</tr>
<tr>
<td></td>
<td>Includes: papilloma</td>
</tr>
<tr>
<td></td>
<td>dermatofibroma</td>
</tr>
<tr>
<td></td>
<td>dermoid cyst</td>
</tr>
<tr>
<td></td>
<td>syringoma</td>
</tr>
<tr>
<td></td>
<td>hydrocystoma</td>
</tr>
<tr>
<td></td>
<td>mastocytoma or mastcell tumor</td>
</tr>
<tr>
<td></td>
<td>syringoadenoma</td>
</tr>
<tr>
<td></td>
<td>female genital organs</td>
</tr>
<tr>
<td></td>
<td>male genital organs</td>
</tr>
<tr>
<td>216.000</td>
<td>Skin of lip</td>
</tr>
<tr>
<td></td>
<td>Excludes: vermilion border of lip</td>
</tr>
<tr>
<td>216.100</td>
<td>Eyelid, including canthus</td>
</tr>
<tr>
<td></td>
<td>Excludes: cartilage of eyelid</td>
</tr>
<tr>
<td>216.200</td>
<td>Ear and external auditory canal</td>
</tr>
<tr>
<td></td>
<td>Includes: auricle ear</td>
</tr>
<tr>
<td></td>
<td>external meatus</td>
</tr>
<tr>
<td></td>
<td>auricular canal</td>
</tr>
<tr>
<td></td>
<td>external canal</td>
</tr>
<tr>
<td></td>
<td>pinna</td>
</tr>
<tr>
<td></td>
<td>Excludes: cartilage of ear</td>
</tr>
<tr>
<td>216.300</td>
<td>Benign neoplasm of skin of other and unspecified parts of face</td>
</tr>
<tr>
<td></td>
<td>Includes: cheek, external</td>
</tr>
<tr>
<td></td>
<td>nose, external</td>
</tr>
<tr>
<td></td>
<td>eyebrow</td>
</tr>
<tr>
<td></td>
<td>temple</td>
</tr>
<tr>
<td>216.400</td>
<td>Benign neoplasm skin, scalp and skin of neck</td>
</tr>
<tr>
<td>Code</td>
<td>Description</td>
</tr>
<tr>
<td>--------</td>
<td>-----------------------------------------------------------------------------</td>
</tr>
<tr>
<td>216.500</td>
<td>Benign neoplasm of skin, trunk, except scrotum</td>
</tr>
<tr>
<td></td>
<td>Includes: axillary fold, perianal skin, skin of chest wall, abdominal wall,</td>
</tr>
<tr>
<td></td>
<td>groin, buttock, anus, perineum, back, umbilicus, breast</td>
</tr>
<tr>
<td></td>
<td>Excludes: anal canal, skin of scrotum</td>
</tr>
<tr>
<td>216.600</td>
<td>Benign neoplasm of skin, upper limb, shoulder</td>
</tr>
<tr>
<td>216.700</td>
<td>Benign neoplasm of skin of lower limb, hip</td>
</tr>
<tr>
<td>216.800</td>
<td>Benign neoplasm of other specified sites of skin</td>
</tr>
<tr>
<td></td>
<td>Excludes: epibulbar dermoid cyst</td>
</tr>
<tr>
<td></td>
<td>(use 743.810)</td>
</tr>
<tr>
<td>216.920</td>
<td>Hairy Nevus</td>
</tr>
<tr>
<td>228.020</td>
<td>Hemangioma, intracranial</td>
</tr>
<tr>
<td>228.030</td>
<td>Hemangioma, retinal</td>
</tr>
<tr>
<td>228.040</td>
<td>Hemangioma, intra-abdominal</td>
</tr>
<tr>
<td>228.090</td>
<td>Hemangioma, of other sites (except when subcutaneous or on skin)</td>
</tr>
<tr>
<td>228.100</td>
<td>Cystic hygroma, Lymphangioma, any site</td>
</tr>
<tr>
<td>237.700</td>
<td>Neurofibromatosis</td>
</tr>
<tr>
<td>238.000</td>
<td>Teratoma, NOS</td>
</tr>
<tr>
<td>238.010</td>
<td>Teratoma, head and face</td>
</tr>
<tr>
<td>238.020</td>
<td>Teratoma, neck</td>
</tr>
<tr>
<td>238.030</td>
<td>Teratoma, abdomen</td>
</tr>
<tr>
<td>238.040</td>
<td>Teratoma, sacral, coccygeal</td>
</tr>
<tr>
<td>238.080</td>
<td>Teratoma, other specified</td>
</tr>
<tr>
<td>239.200</td>
<td>Neck cyst</td>
</tr>
<tr>
<td>253.280</td>
<td>Congenital hypopituitarism</td>
</tr>
<tr>
<td>253.820</td>
<td>Diencephalic Syndrome</td>
</tr>
<tr>
<td>255.200</td>
<td>Adrenogenital disorders (congenital adrenal hyperplasia – CAH)</td>
</tr>
<tr>
<td>Code</td>
<td>Description</td>
</tr>
<tr>
<td>-----------</td>
<td>-----------------------------------------------------------------------------</td>
</tr>
<tr>
<td>257.800</td>
<td>Testicular feminization syndrome</td>
</tr>
<tr>
<td>277.400</td>
<td>Congenital hyperbilirubinemia (not physiological)</td>
</tr>
<tr>
<td></td>
<td>Includes: Crigler-Najjar syndrome</td>
</tr>
<tr>
<td></td>
<td>Dubin-Johnson syndrome</td>
</tr>
<tr>
<td></td>
<td>Gilbert's syndrome</td>
</tr>
<tr>
<td></td>
<td>Rotor's syndrome</td>
</tr>
<tr>
<td>279.110</td>
<td>DiGeorges Syndrome</td>
</tr>
<tr>
<td>331.890</td>
<td>Familial degenerative CNS disease</td>
</tr>
<tr>
<td>335.000</td>
<td>Werndig-Hoffmann disease (Spinal muscle atrophy – SMA)</td>
</tr>
<tr>
<td>352.600</td>
<td>Moebius Syndrome (Congenital Oculofacial paralysis)</td>
</tr>
<tr>
<td>362.700</td>
<td>Retinitis pigmentosa</td>
</tr>
<tr>
<td>363.200</td>
<td>Chorioretinitis</td>
</tr>
<tr>
<td>425.300</td>
<td>Endocardial fibroelastosis</td>
</tr>
<tr>
<td>426.705</td>
<td>Congenital Wolff-Parkinson-White Syndrome</td>
</tr>
<tr>
<td>427.900</td>
<td>Cardiac arrhythmias, NEC (not fetal)</td>
</tr>
<tr>
<td></td>
<td>Includes: supraventricular tachycardia</td>
</tr>
<tr>
<td></td>
<td>AV NODE dysfunction</td>
</tr>
<tr>
<td>453.00</td>
<td>Budd-Chiari, occlusion of hepatic vein</td>
</tr>
<tr>
<td>524.000</td>
<td>Major anomalies of jaw size - micrognathia, macrognathia, hypoplasia, hyperplasia, retrognathia</td>
</tr>
<tr>
<td>524.080</td>
<td>Pierre Robin sequence</td>
</tr>
<tr>
<td>635.82</td>
<td>Abortions with defect</td>
</tr>
<tr>
<td>634.92</td>
<td></td>
</tr>
<tr>
<td>648.300</td>
<td>Drug dependence</td>
</tr>
<tr>
<td>655.400</td>
<td>Suspected damage to fetus from other disease in the mother</td>
</tr>
<tr>
<td></td>
<td>Suspected damage to fetus from maternal:</td>
</tr>
<tr>
<td></td>
<td>alcohol addiction</td>
</tr>
<tr>
<td></td>
<td>listeriosis</td>
</tr>
<tr>
<td></td>
<td>toxoplasmosis</td>
</tr>
</tbody>
</table>
655.500  Suspected damage to fetus from drugs
   Excludes:  fetal distress in labor and delivery due to drug administration

655.800  Other known or suspected fetal abnormality, NEC
   Suspected damage to fetus from:
       environmental toxins
       intrauterine contraceptive device

656.400  Intrauterine fetal death after 22 weeks gestation

658.800  Delivery problem due to amniotic bands

740  Anencephalus and Similar Anomalies

740.0  Anencephalus
    740.000  Absence of brain
        Amyelencephalus
    740.010  Acrania
    740.020  Anencephaly
    740.030  Hemiencephaly, hemicphaly
    740.080  Other
        Exencephaly

740.1  740.100  Craniorachischisis

740.2  Iniencephaly
    740.200  Closed iniencephaly
    740.210  Open iniencephaly
    740.290  Unspecified iniencephaly

741  Spina bifida
   Includes:  Spina bifida aperta (open lesions)
           myeloecele
           rachischisis
           Spina bifida cystica (closed lesions)
           meningocele
           meningomyelocele
           myelomeningocele
   Excludes:  Spina bifida occulta (see 756.100)
           craniorachischisis (see 740.100)

741.0  Spina Bifida with Hydrocephalus
    741.000  Spina bifida aperta, any site, with hydrocephalus
    741.010  Spina bifida cystica, any site, with hydrocephalus and Arnold-Chiari malformation
Arnold-Chiari malformation, NOS
741.020 Spina bifida cystica, any site, with stenosed aqueduct of Sylvius

741.030 Spina bifida cystica, cervical, with unspecified hydrocephalus
    Spina bifida cystica, cervical, with hydrocephalus but without mention of Arnold-Chiari malformation or aqueduct stenosis.

741.040 Spina bifida cystica, thoracic, with unspecified hydrocephalus, no mention of Arnold-Chiari.

741.050 Spina bifida cystica, lumbar, with unspecified hydrocephalus, no mention of Arnold-Chiari.

741.060 Spina bifida cystica, sacral, with unspecified hydrocephalus, no mention of Arnold-Chiari

741.070 Spina bifida of any site with hydrocephalus of late onset

741.080 Other spina bifida, meningocele of specified site with hydrocephalus

741.085 Spina bifida, meningocele, cervicothoracic, with hydrocephalus

741.086 Spina bifida, meningocele thoracolumbar, with hydrocephalus

741.087 Spina bifida, meningocele, lumbosacral with hydrocephalus

741.090 Spina bifida, of any unspecified type with hydrocephalus

741.9 Spina Bifida without mention of hydrocephalus

741.900 Spina bifida (aperta), without hydrocephalus

741.910 Spina bifida (cystica), cervical, without hydrocephalus

741.920 Spina bifida (cystica), thoracic, without hydrocephalus

741.930 Spina bifida (cystica), lumbar, without hydrocephalus

741.940 Spina bifida (cystica) sacral, without hydrocephalus

741.980 Spina bifida, other specified site, without hydrocephalus
    Includes: cervicothoracic, thoracolumbar, lumbosacral

741.985 Lipomyelomeningocele

741.990 Spina bifida, unspecified site, without hydrocephalus
    (myelocele, myelomeningocele, meningomyelocele)

742 Other Congenital Anomalies of Nervous System

742.0 Encephalocele

742.000 Occipital encephalocele

742.080 Other encephalocele of specified site (includes midline defects)

742.085 Frontal encephalocele

742.086 Parietal encephalocele

742.090 Unspecified encephalocele

742.1 742.100 Microcephalus

742.2 Reduction deformities of brain

742.200 Anomalies of cerebrum
742.210  Anomalies of corpus callosum
742.220  Anomalies of hypothalamus
742.230  Anomalies of cerebellum
   Includes: vermian hypoplasia
742.240  Agyria and lissencephaly
   Includes: Walker-Warburg Syndrome
   Miller-Dieker Syndrome, (17p-add 758.350)
742.250  Microgyria, polymicrogyria, pachygyria
742.260  Holoprosencephaly
742.270  Arrhinencephaly
742.280  Other specified reduction defect of brain
   Includes: corpus pellucidum hypoplastic brain stem
   agenesis of septum pellucidum Aicardia syndrome
   absence of Pons absence of ventricle
742.290  Unspecified reduction defect of brain

742.3  
**Congenital hydrocephalus**
   Excludes: hydrocephalus with any condition in 741.9 (Use 741.0)
742.300  Anomalies of aqueduct of Sylvius
742.310  Atresia of foramina of Magendie and Luschka
   Dandy-Walker syndrome
742.320  Hydranencephaly
742.380  Other specified hydrocephaly
   Includes: communicating hydrocephaly
742.390  Unspecified hydrocephaly, NOS

742.4  
**Other specific anomalies of the brain**
742.400  Enlarged brain and/or head
   megalencephaly macrocephaly
   ventriculomegaly (large head)
   macrocrania (cranium)
742.410  Porencephaly
   Includes: porencephalic cysts schizencephaly
742.420  Cerebral cysts
742.480  Other specified anomalies of brain
   Includes: Arnold-Chiari malformation w/o mention of spina bifida
   bifid cranium pineal cyst
   cavum septum pellucidum posterior fossa cyst
   colpocephaly retrocerebellar cyst
   cortical atrophy septo-optic dysplasia (same as
   cortical dysplasia de Morsier’s
   syndrome)
   cranial nerve defects small lateral ventricles
<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>742.485</td>
<td>Ventricular cysts</td>
</tr>
<tr>
<td></td>
<td>Dandy Walker B variant (no hydrocephalus)</td>
</tr>
<tr>
<td></td>
<td>Excludes: arachnoid cysts</td>
</tr>
<tr>
<td>742.486</td>
<td>Small brain</td>
</tr>
<tr>
<td></td>
<td>microencephaly</td>
</tr>
<tr>
<td>742.5</td>
<td><strong>Other specified anomalies of spinal cord</strong></td>
</tr>
<tr>
<td></td>
<td>Excludes: syringomyelia</td>
</tr>
<tr>
<td>742.500</td>
<td>Amyelia</td>
</tr>
<tr>
<td>742.510</td>
<td>Hypoplasia and dysplasia of spinal cord</td>
</tr>
<tr>
<td></td>
<td>atelomyelia</td>
</tr>
<tr>
<td></td>
<td>myelodysplasia</td>
</tr>
<tr>
<td>742.520</td>
<td>Diastematomyelia</td>
</tr>
<tr>
<td>742.530</td>
<td>Other cauda equina anomalies</td>
</tr>
<tr>
<td>742.540</td>
<td>Hydromyelia</td>
</tr>
<tr>
<td></td>
<td>Hydrorhachis</td>
</tr>
<tr>
<td>742.580</td>
<td>Other specified anomalies of spinal cord and membranes</td>
</tr>
<tr>
<td></td>
<td>Includes: congenital tethered, cord</td>
</tr>
<tr>
<td>742.8</td>
<td><strong>Other specified anomalies of nervous system</strong></td>
</tr>
<tr>
<td></td>
<td>Excludes: congenital oculofacial paralysis</td>
</tr>
<tr>
<td></td>
<td>Moebius syndrome (use 352.600)</td>
</tr>
<tr>
<td>742.800</td>
<td>Jaw-winking syndrome</td>
</tr>
<tr>
<td></td>
<td>Marcus Gunn Syndrome</td>
</tr>
<tr>
<td>742.810</td>
<td>Familial dysautonomia</td>
</tr>
<tr>
<td></td>
<td>Riley-Day Syndrome</td>
</tr>
<tr>
<td>742.880</td>
<td>Other specified anomalies of nervous system</td>
</tr>
<tr>
<td></td>
<td>Includes: cerebral dysgenesis</td>
</tr>
<tr>
<td>742.9</td>
<td><strong>Unspecified anomalies of brain, spinal cord and nervous system</strong></td>
</tr>
<tr>
<td>742.900</td>
<td>Brain, unspecified anomalies</td>
</tr>
<tr>
<td>742.910</td>
<td>Spinal cord, unspecified anomalies</td>
</tr>
<tr>
<td>742.990</td>
<td>Nervous system, unspecified anomalies</td>
</tr>
<tr>
<td>743</td>
<td><strong>Congenital Anomalies of Eye</strong> (OD=Right; OS=Left; OU=Bilateral,Each Eye)</td>
</tr>
<tr>
<td>743.0</td>
<td><strong>Anophthalmos</strong></td>
</tr>
<tr>
<td></td>
<td>agenesis of eye</td>
</tr>
<tr>
<td></td>
<td>cryptophthalmos (Fraser syndrome)</td>
</tr>
<tr>
<td>743.1</td>
<td><strong>Microphthalmos, small eyes</strong></td>
</tr>
<tr>
<td></td>
<td>aplasia of eye</td>
</tr>
<tr>
<td></td>
<td>hypoplasia of eye</td>
</tr>
<tr>
<td></td>
<td>dysplasia of eye</td>
</tr>
<tr>
<td></td>
<td>rudimentary eye</td>
</tr>
</tbody>
</table>
**Inclusion ICD-9-CM Diagnostic Index - Numeric**

**743.2 Buphthalmos**
- 743.200 Buphthalmos
  - congenital glaucoma
  - hydrophthalmos
- 743.210 Enlarged eye, NOS
- 743.220 Enlarged cornea
  - keratoglobus
  - congenital megalocornea

**743.3 Congenital cataract and lens anomalies**
- 743.300 Absence of lens
  - congenital aphakia
- 743.310 Spherical lens
  - Spherophakia
- 743.320 Cataract, NOS
- 743.325 Cataract, anterior polar
- 743.326 Cataract, other specified
  - Includes: nuclear cataract
- 743.330 Displaced lens
- 743.340 Coloboma of lens
- 743.380 Other specified lens anomalies
- 743.390 Unspecified lens anomalies

**743.4 Coloboma and other anomalies of anterior segments**
- 743.400 Corneal opacity/clouding
- 743.410 Other corneal anomalies
  - Includes: microcornea
  - Excludes: megalocornea (use 743.220)
- 743.420 Absence of iris
  - aniridia
- 743.430 Coloboma of iris
- 743.440 Other anomalies of iris
  - polycoria
  - etopic pupil
  - Peter's anomaly
- 743.480 Other specified colobomas and anomalies of anterior segments
  - Rieger's anomaly
- 743.490 Unspecified colobomas and anomalies of anterior eye segments

**743.5 Congenital anomalies of the posterior segment**
- 743.500 Specified anomalies of vitreous humour
- 743.510 Specified anomalies of retina
  - Includes: congenital retinal aneurysm
    - Lebers Congenital Amaurosis
    - absence of retinal blood vessels
  - Excludes: Stickler Syndrome (use 759.860)
- 743.520 Specified anomalies of optic disc
Includes: hypoplastic optic nerve
          optic dysplasia
          coloboma of optic nerve/disc

743.530 Specified anomalies of choroid
743.535 Coloboma of choroid
743.580 Other specified anomalies of posterior segment of eye
cortical blindness
743.590 Unspecified anomalies of posterior segment of eye

743.6 Congenital anomalies of eyelids, lacrimal system and orbit
743.600 Blepharoptosis
          congenital ptosis
743.610 Ectropion
743.620 Entropion
743.635 Blepharophimosis
          small or narrow palpebral fissures
743.636 Coloboma of the eyelids (cleft eyelids)
743.640 Absence or agenesis of lacrimal apparatus
          absence of punctum lacrimale
743.660 Other anomalies of lacrimal apparatus (e.g., cyst, dacryostocele)
743.670 Anomalies of orbit
          Includes: cleft orbit

743.8 Other specified anomalies of eye
743.810 Epibulbar dermoid cyst
          Includes: dermoid of cornea

743.9 743.900 Unspecified anomalies of eye
          congenital: of eye (any part)
          anomaly, NOS
          deformity, NOS

744 Congenital Anomalies of Ear, Face, and Neck

744.0 Anomalies of ear causing hearing impairment
744.000 Absence or stricture of auditory canal
744.010 Absence of auricle (pinna)
          absence of ear, NOS
744.020 Anomaly of middle ear
          fusion of ossicles
744.030 Anomaly of inner ear
          Includes: congenital anomaly of membranous labyrinth
          organ of Corti
744.090 Unspecified anomalies of ear with hearing impairment
          Includes: congenital deafness, NOS
<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>744.2</td>
<td><strong>Other specified anomalies of ear</strong></td>
</tr>
<tr>
<td>744.200</td>
<td>Macrotia (enlarged pinna)</td>
</tr>
<tr>
<td>744.210</td>
<td>Microtia (hypoplastic pinna and absence or stricture of external auditory meatus)</td>
</tr>
<tr>
<td>744.220</td>
<td>Bat ear <em>(if isolated – exclude)</em></td>
</tr>
<tr>
<td>744.230</td>
<td>Other misshapen ear, <em>(if isolated-exclude)</em></td>
</tr>
<tr>
<td>744.240</td>
<td>Misplaced ears - <em>(exclude low set)</em></td>
</tr>
<tr>
<td>744.250</td>
<td>Absence or anomaly of eustachian tube</td>
</tr>
<tr>
<td>744.280</td>
<td>Other specified anomalies of ear <em>(see also 744.230)</em></td>
</tr>
<tr>
<td></td>
<td>Excludes: Darwin's tubercle</td>
</tr>
<tr>
<td>744.3</td>
<td>744.300</td>
</tr>
<tr>
<td></td>
<td>congenital ear (any part)</td>
</tr>
<tr>
<td></td>
<td>anomaly, deformity, NOS</td>
</tr>
<tr>
<td>744.4</td>
<td><strong>Brachial cleft, cyst, or fistula; preauricular sinus</strong></td>
</tr>
<tr>
<td>744.400</td>
<td>Brachial cleft, sinus, fistula cyst, or pit</td>
</tr>
<tr>
<td>744.480</td>
<td>Other brachial cleft anomalies</td>
</tr>
<tr>
<td></td>
<td>Includes: dermal sinus of head</td>
</tr>
<tr>
<td></td>
<td>brachial cleft remnant</td>
</tr>
<tr>
<td>744.8</td>
<td><strong>Other unspecified anomalies of face and neck</strong></td>
</tr>
<tr>
<td>744.800</td>
<td>Macrostomia (large mouth)</td>
</tr>
<tr>
<td>744.810</td>
<td>Microstomia (small mouth)</td>
</tr>
<tr>
<td>744.880</td>
<td>Other specified anomalies of face/neck</td>
</tr>
<tr>
<td>744.9</td>
<td><strong>Unspecified anomalies of face and neck</strong></td>
</tr>
<tr>
<td>744.910</td>
<td>Congenital anomaly of face, NOS</td>
</tr>
<tr>
<td></td>
<td>Abnormal facies (dysmorphic features-list)</td>
</tr>
<tr>
<td>745</td>
<td><strong>Bulbus Cordis Anomalies and Anomalies of Cardiac Septal Closure</strong></td>
</tr>
<tr>
<td>745.0</td>
<td><strong>Common truncus</strong> <em>(see 747.200 for pseudotruncus)</em></td>
</tr>
<tr>
<td>745.000</td>
<td>Persistent truncus arteriosus</td>
</tr>
<tr>
<td></td>
<td>absent septum between aorta and pulmonary artery</td>
</tr>
<tr>
<td>Code</td>
<td>Description</td>
</tr>
<tr>
<td>--------</td>
<td>-----------------------------------------------------------------------------</td>
</tr>
<tr>
<td>745.010</td>
<td>Aortic septal defect</td>
</tr>
<tr>
<td></td>
<td>Includes: aortopulmonary window</td>
</tr>
<tr>
<td></td>
<td>Excludes: atrial septal defect (use 745.590)</td>
</tr>
<tr>
<td>745.1</td>
<td><strong>Transposition of great vessels</strong></td>
</tr>
<tr>
<td>745.100</td>
<td>Transposition of great vessels, complete (no VSD)</td>
</tr>
<tr>
<td>745.110</td>
<td>Transposition of great vessels, incomplete (w/ VSD)</td>
</tr>
<tr>
<td></td>
<td>Taussig-Bing syndrome</td>
</tr>
<tr>
<td>745.120</td>
<td>Corrected transposition of great vessels, L-transposition, ventri in version</td>
</tr>
<tr>
<td></td>
<td>Excludes: dextrocardia (use 746.800)</td>
</tr>
<tr>
<td>745.180</td>
<td>Other specified transposition of great vessels</td>
</tr>
<tr>
<td></td>
<td>Includes: double outlet right ventricle</td>
</tr>
<tr>
<td></td>
<td>D Transposition</td>
</tr>
<tr>
<td>745.190</td>
<td>Unspecified transposition of great vessels</td>
</tr>
<tr>
<td>745.2</td>
<td><strong>Tetralogy of Fallot</strong></td>
</tr>
<tr>
<td>745.200</td>
<td>Fallot's Tetralogy</td>
</tr>
<tr>
<td>745.210</td>
<td>Fallot's pentalogy</td>
</tr>
<tr>
<td></td>
<td>Fallot's tetralogy plus ASD</td>
</tr>
<tr>
<td>745.3</td>
<td><strong>Single ventricle</strong></td>
</tr>
<tr>
<td>745.300</td>
<td>Common ventricle</td>
</tr>
<tr>
<td></td>
<td>Cor triloculare biaatriatum</td>
</tr>
<tr>
<td>745.4</td>
<td><strong>Ventricular Septal Defect</strong></td>
</tr>
<tr>
<td>745.400</td>
<td>Roger's disease</td>
</tr>
<tr>
<td>745.410</td>
<td>Eisenmenger's syndrome</td>
</tr>
<tr>
<td>745.420</td>
<td>Gerbode defect</td>
</tr>
<tr>
<td>745.480</td>
<td>Other specified ventricular septal defect</td>
</tr>
<tr>
<td></td>
<td>Includes: membranous perimembranous</td>
</tr>
<tr>
<td></td>
<td>muscular midmuscular</td>
</tr>
<tr>
<td></td>
<td>crystalline sub-crystalline</td>
</tr>
<tr>
<td></td>
<td>subarterial inlet conoreentricular</td>
</tr>
<tr>
<td>745.490</td>
<td>VSD (ventricular septal defect), NOS</td>
</tr>
<tr>
<td></td>
<td>Excludes: common atroventricular canal type (use 745.620)</td>
</tr>
<tr>
<td>745.498</td>
<td>Probable VSD</td>
</tr>
<tr>
<td>745.5</td>
<td><strong>Ostium secundum type atrial septal defect</strong></td>
</tr>
<tr>
<td>745.510</td>
<td>Ostium (septum) secundum defect</td>
</tr>
<tr>
<td>745.520</td>
<td>Lutembacher's syndrome</td>
</tr>
<tr>
<td>745.580</td>
<td>Other specified atrial septal defect</td>
</tr>
</tbody>
</table>
Inclusion ICD-9-CM Diagnostic Index - Numeric

745.590  ASD (atrial septal defect), NOS
    Auricular septal defect, NOS
    PFO vs. ASD

745.6  Endocardial Cushion Defects
   745.600  Ostium primum defects
   745.610  Single common atrium, cor triloculare biventriculare
   745.620  Common atroventricular canal with ventricular septal defect (VSD)
   745.630  Common atroventricular canal
   745.680  Other specified cushion defect
   745.690  Endocardial cushion defect, NOS

745.7  Cor biloculare

745.8  Other specified defects of septal closure
   Includes: aneurysm of the forame ovale
   septal aneurysm

745.9  Unspecified defect of septal closure

746  Other congenital anomalies of the heart

746.0  Anomalies of pulmonary valve
   746.000  Atresia, hypoplasia of pulmonary valve
     See 746.995 if valve is not specified (e.g. "pulmonary artresia")
   746.010  Stenosis of pulmonary valve
     See 746.995 if valve is not specified (e.g., "pulmonary stenosis");
     Excludes: pulmonary infundibular stenosis (use 746.830)
   746.020  Pulmonary valve insufficiency or regurgitation, congenital
     Excludes: case designated as "mild", "minimal", "trivial" or
     "physiologic" if isolated
   746.080  Other specified anomalies of pulmonary valve
     Includes: bicuspid pulmonary valve
     Excludes: pulmonary infundibular stenosis (use 746.830)
   746.090  Unspecified anomaly of pulmonary valve

746.1  Tricuspid atresia and stenosis
   746.100  Tricuspid atresia, stenosis, hypoplasia
   746.105  Tricuspid valve insufficiency or regurgitation, congenital
     Excludes: Ebstein's anomaly (746.200)
     Case designated as "mild", "minimal", "trivial" or
     "physiologic" if isolated

746.2  Ebstein's anomaly
746.3  746.300  **Congenital stenosis of aortic valve**
   Includes:  congenital aortic stenosis
              subvalvular aortic stenosis
   Excludes:  supravalvular aortic stenosis (747.220)

746.4  746.400  **Congenital insufficiency of aortic valve**
   Aortic valve insufficiency or regurgitation, congenital
   bicuspid aortic valve
   Excludes:  case designated as "mild", "minimal", "trivial" or
              "physiologic" if isolated

746.480  Other specified anomalies of the aortic valves
   Includes:  aortic valve atresia
   Excludes:  supravalvular aortic stenosis (747.220)

746.490  Unspecified anomalies of the aortic valves

746.5  746.500  **Congenital mitral stenosis**
   Congenital mitral stenosis
   Absence, atresia, or hypoplasia of mitral valve

746.6  746.600  **Mitral insufficiency or regurgitation**
   Excludes:  case designated as "mild", "minimal", "trivial" or
              "physiologic" if isolated

746.7  746.700  **Hypoplastic left heart syndrome**
   Atresia, or marked hypoplasia of the ascending aorta and
   defective development of left ventricle (with mitral valve atresia)

746.8  746.800  **Other specified anomalies of the heart**
   Dextrocardia without situs inversus (situs solitus)
   Dextrocardia with no mention of situs inversus
   Excludes:  dextrocardia with situs inversus (use 759.300)

746.810  Levocardia
746.820  Cor triatriatum
746.830  Pulmonary infundibular (subvalvular) stenosis
746.840  Trilogy of Fallot
746.850  Anomalies of pericardium
746.870  Congenital heart block
746.880  Other specified anomalies of heart
   Includes:  acardia
dual chamber right ventricle
   cardiac tumor
ectopia (ectopic) cordis
   coronary aneurysm
   hypoplastic heart
   double outlet left ventricle
   (mesocordia) conduction

15
conduction defects, NOS  defects, NOS

746.881 Hypoplastic left ventricle
   Excludes: hypoplastic left heart syndrome (746.700)

746.882 Hypoplastic right heart (ventricle)
   Uhl's disease

746.883 Hypoplastic ventricle, NOS

746.885 Anomalies of coronary artery or sinus

746.886 Ventricular hypertrophy (left or right)

746.887 Other defects of the atria
   Includes: dilated atria  small atria
   atrial aneurysm  abnormal atrial septum
   Excludes: congenital Wolff-Parkinson-White use (426.705)
   rhythm anomalies (use 427.900)

746.9  Unspecified anomalies of the heart

746.900 Unspecified anomalies of heart valves
   Includes: cleft mitral valve  bifid truncal valve
   single AV valve with single ventricle (double inlet ventricle)
   truncal valve stenosis  Shone complex

746.910 Anomalous bands of heart

746.920 Acyanotic congenital heart disease, NOS

746.930 Cyanotic congenital heart disease, NOS
   Blue baby

746.990 Unspecified anomaly of heart
   Includes: congenital heart disease (CHD)

746.995 "Pulmonic" or "pulmonary" atresia, stenosis, or hypoplasia, NOS (no mention
   of valve or artery) (make certain "pulmonary" refers to heart not lungs,
   look for cardioechogram)

747  Other congenital anomalies of the circulatory system

747.1 Coarctation of aorta

747.100 Preductal (proximal) coarctation of aorta

747.110 Postductal (distal) coarctation of aorta

747.190 Unspecified coarctation of aorta

747.2 Other anomalies of the aorta

747.200 Atresia of aorta
   absence of aorta
   pseudotruncus arteriosus

747.210 Hypoplasia of aorta
   tubular hypoplasia of aorta

747.215 Interrupted aortic arch

747.220 Supra-aortic stenosis (supravalvular)
   Excludes: aortic stenosis, congenital (see 746.300)
Inclusion ICD-9-CM Diagnostic Index - Numeric

747.230  Persistent right aortic arch
747.240  Aneurysm of sinus of Valsalva
747.250  Vascular ring (aorta)
   double aortic arch
   Includes: vascular ring compression of trachea
747.260  Overriding aorta
dextroposition of aorta
747.270  Congenital aneurysm of aorta
congenital dilatation of aorta
747.280  Other specified anomalies of aorta
747.290  Unspecified anomalies of aorta

747.3  Anomalies of the pulmonary artery
747.300  Pulmonary artery atresia, absence or agenesis
   Use 746.995 if artery or valve is not specified
747.310  Pulmonary artery atresia with septal defect (ASD or VSD)
747.320  Pulmonary artery stenosis
   Use 746.995 if artery or valve is not specified
747.325  Peripheral pulmonary artery stenosis
   Includes: peripheral pulmonic stenosis (PPS)
pulmonary artery branch stenosis
   Excludes: peripheral pulmonic stenosis murmur
747.330  Aneurysm of pulmonary artery
dilatation of pulmonary artery
747.340  Pulmonary arteriovenous malformation or aneurysm
747.380  Other specified anomaly of pulmonary artery
   Includes: pulmonary artery hypoplasia
747.390  Unspecified anomaly of pulmonary artery

747.4  Anomalies of the great veins
747.400  Stenosis of vena cava (inferior or superior)
747.410  Persistent left superior vena cava
747.420  Total anomalous pulmonary venous return (TAPVR)
747.430  Partial anomalous pulmonary venous return (PAPVR)
747.440  Anomalous portal vein termination
747.450  Portal vein – hepatic artery fistula
747.480  Other specified anomalies of great veins
   Includes: “crisscross” atrioventricular connection
   bilateral superior vena cava  Scimitor Syndrome
747.490  Unspecified anomalies of great veins

747.6  Other anomalies of peripheral vascular system
747.600  Stenosis of renal artery
747.610  Other anomalies of renal artery
747.620  Arteriovenous malformation (peripheral)
747.630 Congenital phlebectasia
congenital varix

747.640 Other anomalies of peripheral arteries
   Includes: aberrant subclavian artery
   aneurysm of peripheral arteries

747.650 Other anomalies of peripheral veins
   Includes: pulmonary vein stenosis or hypoplasia
   Excludes: Budd-Chiari - occlusion of hepatic vein (use 453.000)

747.680 Other anomalies of peripheral vascular system
   Includes: primary pulmonary artery
   Hypertension – (if isolated, exclude)

747.690 Unspecified anomalies of peripheral vascular system

747.8 Other specified anomalies of circulatory system

747.800 Arteriovenous (malformation) aneurysm of brain

747.880 Other specified anomalies of circulatory system
   Excludes: congenital aneurysm:
   coronary (746.880)
   peripheral (747.640)
   pulmonary (747.330)
   retinal (743.510)
   ruptured cerebral arteriovenous malformation (747.800)

747.900 Unspecified anomalies of circulatory system

748 Congenital Anomalies of Respiratory System

748.000 Choanal atresia
   atresia of nares, anterior or posterior
   congenital stenosis

748.1 Other anomalies of the nose

748.100 Agenesis or underdevelopment of nose
748.110 Accessory nose
748.120 Fissured, notched, or cleft nose
748.130 Sinus wall anomalies
748.140 Perforated nasal septum
748.185 Tubular nose, single nostril, proboscis
748.190 Unspecified anomalies of nose
748.2  Web of larynx
  748.205  Web of larynx-glottic
  748.206  Web of larynx-subglottic
  748.209  Web of larynx-NOS

748.3  Other anomalies of larynx, trachea, and bronchus
  748.300  Anomalies of larynx and supporting cartilage
    Includes:  Laryngomalacia
  748.310  Congenital subglottic stenosis
  748.330  Other anomalies of trachea
    Includes:  Tracheomalacia
    Excludes:  vascular ring compression of the trachea (use 747.250)
  748.340  Stenosis of bronchus
  748.350  Other anomalies of bronchus
  748.360  Congenital laryngeal stridor, NOS

  748.380  Other specified anomalies of larynx and bronchus
  748.385  Cleft larynx, laryngotracheoesophageal cleft
  748.390  Unspecified anomalies of larynx, trachea, and bronchus

748.4  Congenital cystic lung
  748.400  Single cyst, lung or lung cyst
    Includes:  cystic adenomatoid malformation (CAM)
  748.410  Multiple cysts, lung
    Polycystic lung
  748.420  Honeycomb lung
  748.480  Other specified congenital cystic lung

748.5  Agenesis or aplasia of lung
  748.500  Agenesis or aplasia of lung
  748.510  Hypoplasia of lung; Pulmonary hypoplasia
    (Exclude if isolated defect in infants less than 36 wks gestation)
  748.520  Sequestration of lung
  748.580  Other specified dysplasia of lung
    Fusion of lobes of lung
  748.590  Unspecified dysplasia of lung

748.6  Other anomalies of lung
  748.600  Ectopic tissues in lung
  748.610  Bronchiectasis
  748.620  Accessory lobe of lung
  748.625  Bilobar right lung or right lung with left lung bronchial pattern
  748.690  Other and unspecified anomalies of lung
**748.8 Other specified anomalies of respiratory system**
- 748.800 Anomaly of pleura
- 748.810 Congenital cyst of mediastinum
- 748.880 Other specified respiratory system anomalies
  - Includes: congenital lobar emphysema
  - lymphangiectasia of lungs

**748.9 748.900 Unspecified anomalies of respiratory system**
- Absence of respiratory organ, NOS
- Anomaly of respiratory system, NOS

**749 Cleft palate and cleft lip**

**749.0 Cleft palate alone**
(If description of condition includes Pierre Robin sequence, use additional code, 524.080)

- 749.000 Cleft hard palate, unilateral
- 749.010 Cleft hard palate, bilateral
- 749.020 Cleft hard palate, central
- 749.030 Cleft hard palate, NOS
- 749.035 Cleft hard & soft palate (uranostaphyloschisis) 11/98
- 749.040 Cleft soft palate, alone unilateral
- 749.050 Cleft soft palate, alone bilateral
- 749.060 Cleft soft palate, alone central
- 749.070 Cleft soft palate, alone, NOS
  - Includes: submucous cleft palate
- 749.080 Cleft uvula or bifid uvula
- 749.090 Cleft palate, NOS
  - palatoschisis

**749.1 Cleft lip alone**
- Includes: alveolar ridge cleft
  - cleft gum
  - harelip
- 749.100 Cleft lip, unilateral
- 749.110 Cleft lip, bilateral
- 749.120 Cleft lip, central
- 749.190 Cleft lip, NOS, cleft gum

**749.2 Cleft lip with cleft palate**
- 749.200 Cleft lip, unilateral, with any cleft palate
- 749.210 Cleft lip, bilateral, with any cleft palate
- 749.220 Cleft lip, central, with any cleft palate
749.290  Cleft lip, NOS, with any cleft palate

750  **Other Congenital Anomalies of Upper Alimentary Tract**

### 750.1 Other anomalies of tongue

- Excludes: protruding tongue (never a defect)
  - 750.100  Aglossia  
    Absence of tongue
  - 750.110  Hypoglossia of tongue (small tongue)  
    Microglossia  
    Includes: Oromandibular limb hypoplasia
  - 750.120  Macroglossia (large tongue)
  - 750.130  Dislocation or displacement of tongue  
    Glossoptosis
  - 750.140  Cleft tongue or split tongue
  - 750.180  Other specified anomalies of tongue
  - 750.190  Unspecified anomalies of tongue

### 750.2 Other specified anomalies of mouth and pharynx

- 750.200  Pharyngeal pouch
- 750.210  Other pharyngeal anomalies
- 750.230  Other anomalies of salivary glands or ducts
- 750.250  Other anomalies of palate
- 750.260  Lip fistulæ or pits
- 750.270  Other lip anomalies  
    Includes: notched lip, prominent philtrum, long philtrum  
    Excludes: cleft lip (see 749)
- 750.280  Other specified anomalies of mouth and pharynx  
    Includes: absence of muscles around mouth  
    absent uvula w/o cleft palate  
    Excludes: receding jaw (see 524.0)  
    large and small mouth (see 744.8)

### 750.3 Tracheoesophageal (T-E) fistula, esophageal atresia and stenosis

- 750.300  Esophageal atresia without mention of T-E fistula
- 750.310  Esophageal atresia with mention of T-E fistula
- 750.320  Tracheoesophageal fistula (T-E fistula) without mention of esophageal atresia
- 750.325  Tracheoesophageal fistula (T-E fistula) - "H" type
- 750.330  Bronchoesophageal fistula with or without mention of esophageal atresia
- 750.340  Stenosis or stricture of esophagus
- 750.350  Esophageal web
- 750.380  Other tracheoesophageal anomalies
<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>750.4</td>
<td><strong>Other specified anomalies of esophagus</strong></td>
</tr>
<tr>
<td></td>
<td>750.400 Congenital dilatation of esophagus</td>
</tr>
<tr>
<td></td>
<td>750.410 Displacement of esophagus</td>
</tr>
<tr>
<td></td>
<td>750.420 Diverticulum of esophagus</td>
</tr>
<tr>
<td></td>
<td>750.430 Duplication of esophagus</td>
</tr>
<tr>
<td></td>
<td>750.480 Other specified anomalies of esophagus</td>
</tr>
<tr>
<td>750.5</td>
<td><strong>Congenital hypertrophic pyloric stenosis</strong></td>
</tr>
<tr>
<td></td>
<td>750.510 Congenital hypertrophic pyloric stenosis</td>
</tr>
<tr>
<td></td>
<td>750.580 Other congenital pyloric obstruction</td>
</tr>
<tr>
<td>750.6</td>
<td><strong>Congenital hiatus hernia</strong></td>
</tr>
<tr>
<td></td>
<td>750.600 Cardia displacement through esophageal hiatus</td>
</tr>
<tr>
<td></td>
<td>Partial thoracic stomach</td>
</tr>
<tr>
<td></td>
<td>Excludes: congenital diaphragmatic hernia (756.610)</td>
</tr>
<tr>
<td>750.7</td>
<td><strong>Other specified anomalies of stomach</strong></td>
</tr>
<tr>
<td></td>
<td>750.700 Microgastria</td>
</tr>
<tr>
<td></td>
<td>750.710 Megalogastria</td>
</tr>
<tr>
<td></td>
<td>750.720 Cardiospasm</td>
</tr>
<tr>
<td></td>
<td>achalasia of cardia, congenital</td>
</tr>
<tr>
<td></td>
<td>750.730 Displacement or transposition of stomach</td>
</tr>
<tr>
<td></td>
<td>750.740 Diverticulum of stomach</td>
</tr>
<tr>
<td></td>
<td>750.750 Duplication of stomach</td>
</tr>
<tr>
<td></td>
<td>750.780 Other specified anomalies of stomach</td>
</tr>
<tr>
<td></td>
<td>Includes: gastric antrum web</td>
</tr>
<tr>
<td>750.8</td>
<td><strong>Other specified anomalies of upper alimentary tract</strong></td>
</tr>
<tr>
<td>750.9</td>
<td><strong>Unspecified anomalies of upper alimentary tract</strong></td>
</tr>
<tr>
<td></td>
<td>750.900 Unspecified anomalies of mouth and pharynx</td>
</tr>
<tr>
<td></td>
<td>750.910 Unspecified anomalies of esophagus</td>
</tr>
<tr>
<td></td>
<td>750.920 Unspecified anomalies of stomach</td>
</tr>
<tr>
<td></td>
<td>750.990 Unspecified anomalies of upper alimentary tract</td>
</tr>
<tr>
<td>751</td>
<td><strong>Other Congenital Anomalies of Digestive System</strong></td>
</tr>
<tr>
<td>751.0</td>
<td>751.000 Persistent omphalomesenteric duct</td>
</tr>
<tr>
<td></td>
<td>persistent vitelline duct</td>
</tr>
<tr>
<td>751.1</td>
<td><strong>Atresia and stenosis of small intestine</strong></td>
</tr>
<tr>
<td></td>
<td>751.100 Stenosis, atresia or absence of duodenum</td>
</tr>
</tbody>
</table>
### Inclusion ICD-9-CM Diagnostic Index - Numeric

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>751.110</td>
<td>Stenosis, atresia or absence of jejunum</td>
</tr>
<tr>
<td>751.120</td>
<td>Stenosis, atresia or absence of ileum</td>
</tr>
<tr>
<td>751.190</td>
<td>Stenosis, atresia or absence of small intestine</td>
</tr>
<tr>
<td></td>
<td>Includes: Apple-Peel Syndrome</td>
</tr>
<tr>
<td>751.195</td>
<td>Stenosis, atresia or absence of small intestine with fistula</td>
</tr>
<tr>
<td>751.2</td>
<td>Atresia and stenosis of large intestine, rectum and anal canal</td>
</tr>
<tr>
<td>751.200</td>
<td>Stenosis, atresia, or absence of large intestine</td>
</tr>
<tr>
<td></td>
<td>Stenosis, atresia or absence of appendix</td>
</tr>
<tr>
<td>751.210</td>
<td>Stenosis, atresia or absence of rectum with fistula</td>
</tr>
<tr>
<td>751.220</td>
<td>Stenosis, atresia or absence of rectum without mention of fistula</td>
</tr>
<tr>
<td>751.230</td>
<td>Stenosis, atresia or absence of anus with fistula</td>
</tr>
<tr>
<td></td>
<td>Includes: imperforate anus with fistula</td>
</tr>
<tr>
<td>751.240</td>
<td>Stenosis, atresia or absence of anus without mention of fistula</td>
</tr>
<tr>
<td></td>
<td>Includes: imperforate anus without fistula</td>
</tr>
<tr>
<td></td>
<td>Note: anterior anus coded as 751.240 &amp; 751.530</td>
</tr>
<tr>
<td>751.3</td>
<td>Hirschsprung's disease and other congenital functional disorders of the colon</td>
</tr>
<tr>
<td>751.300</td>
<td>Total intestinal aganglionosis</td>
</tr>
<tr>
<td>751.310</td>
<td>Long-segment Hirschsprung's disease; aganglionosis beyond the rectum</td>
</tr>
<tr>
<td>751.320</td>
<td>Short-segment Hirschsprung's disease; aganglionosis involving no more than the anal sphincter and the rectum.</td>
</tr>
<tr>
<td>751.330</td>
<td>Hirschsprung's disease, NOS</td>
</tr>
<tr>
<td>751.340</td>
<td>Congenital megacolon, not aganglionic</td>
</tr>
<tr>
<td>751.4</td>
<td>Anomalies of intestinal fixation</td>
</tr>
<tr>
<td>751.400</td>
<td>Malrotation of cecum and/or colon</td>
</tr>
<tr>
<td>751.410</td>
<td>Anomalies of mesentery</td>
</tr>
<tr>
<td>751.420</td>
<td>Congenial adhesions or bands of omentum and peritoneum; Ladd's bands</td>
</tr>
<tr>
<td>751.490</td>
<td>Other specified and unspecified malrotation</td>
</tr>
<tr>
<td>751.495</td>
<td>Malrotation of small intestine alone</td>
</tr>
<tr>
<td>751.5</td>
<td>Other anomalies of the intestine</td>
</tr>
<tr>
<td>751.500</td>
<td>Duplication of anus, appendix, cecum, or intestine enterogenous cyst</td>
</tr>
<tr>
<td>751.510</td>
<td>Transposition of appendix, colon, or intestine</td>
</tr>
<tr>
<td>751.520</td>
<td>Microcolon</td>
</tr>
<tr>
<td>751.530</td>
<td>Ectopic (displaced) anus</td>
</tr>
<tr>
<td>751.540</td>
<td>Congenital anal fistula</td>
</tr>
<tr>
<td>751.550</td>
<td>Persistent cloaca</td>
</tr>
<tr>
<td>Code</td>
<td>Description</td>
</tr>
<tr>
<td>--------</td>
<td>--------------------------------------------------</td>
</tr>
<tr>
<td>751.560</td>
<td>Duodenal web</td>
</tr>
<tr>
<td>751.590</td>
<td>Unspecified anomalies of intestine</td>
</tr>
<tr>
<td>751.6</td>
<td><strong>Anomalies of gallbladder, bile ducts and liver</strong></td>
</tr>
<tr>
<td>751.600</td>
<td>Absence or agenesis of liver, total or partial</td>
</tr>
<tr>
<td>751.610</td>
<td>Cystic or fibrocystic disease of liver</td>
</tr>
<tr>
<td>751.630</td>
<td>Agenesis or hypoplasia of gallbladder</td>
</tr>
<tr>
<td>751.640</td>
<td>Other anomalies of gallbladder</td>
</tr>
<tr>
<td></td>
<td>duplication of gallbladder</td>
</tr>
<tr>
<td>751.650</td>
<td>Agenesis or atresia of hepatic ducts or bile ducts</td>
</tr>
<tr>
<td></td>
<td>Includes: biliary atresia</td>
</tr>
<tr>
<td></td>
<td>Excludes: congenital or neonatal hepatitis</td>
</tr>
<tr>
<td></td>
<td>(use 774.480 or 774.490)</td>
</tr>
<tr>
<td>751.660</td>
<td>Choledochal cysts</td>
</tr>
<tr>
<td>751.670</td>
<td>Other anomalies of hepatic or bile ducts</td>
</tr>
<tr>
<td></td>
<td>Includes: Alagille Syndrome</td>
</tr>
<tr>
<td>751.680</td>
<td>Anomalies of biliary tract, NEC</td>
</tr>
<tr>
<td>751.7</td>
<td><strong>Anomalies of pancreas</strong></td>
</tr>
<tr>
<td></td>
<td>Excludes: fibrocystic disease of pancreas</td>
</tr>
<tr>
<td></td>
<td>diabetes mellitus, congenital and neonatal</td>
</tr>
<tr>
<td>751.700</td>
<td>Absence, agenesis or hypoplasia of pancreas</td>
</tr>
<tr>
<td>751.710</td>
<td>Accessory pancreas</td>
</tr>
<tr>
<td>751.720</td>
<td>Annular pancreas</td>
</tr>
<tr>
<td>751.730</td>
<td>Ectopic pancreas</td>
</tr>
<tr>
<td>751.740</td>
<td>Pancreatic cyst</td>
</tr>
<tr>
<td>751.780</td>
<td>Other specified anomalies of pancreas</td>
</tr>
<tr>
<td>751.790</td>
<td>Unspecified anomalies of pancreas</td>
</tr>
<tr>
<td>751.8</td>
<td><strong>Other specified anomalies of digestive system</strong></td>
</tr>
<tr>
<td>751.800</td>
<td>Absence of alimentary tract, NOS</td>
</tr>
<tr>
<td></td>
<td>(complete or partial)</td>
</tr>
<tr>
<td>751.810</td>
<td>Duplication of alimentary tract</td>
</tr>
<tr>
<td>751.820</td>
<td>Ectopic digestive organs, NOS</td>
</tr>
<tr>
<td>751.880</td>
<td>Other specified anomalies of digestive system</td>
</tr>
<tr>
<td>751.9</td>
<td><strong>Unspecified anomalies of digestive system</strong></td>
</tr>
<tr>
<td>751.900</td>
<td>congenital of digestive system, NOS</td>
</tr>
<tr>
<td></td>
<td>anomaly, NOS</td>
</tr>
<tr>
<td></td>
<td>deformity, NOS</td>
</tr>
<tr>
<td></td>
<td>obstruction, NOS</td>
</tr>
<tr>
<td>752</td>
<td><strong>Congenital Anomalies of Genital Organs</strong></td>
</tr>
<tr>
<td></td>
<td>Excludes: congenital hydrocele</td>
</tr>
<tr>
<td>Code</td>
<td>Description</td>
</tr>
<tr>
<td>--------</td>
<td>-----------------------------------------------------------------------------------------------</td>
</tr>
<tr>
<td>257.800</td>
<td>Testicular feminization syndrome</td>
</tr>
<tr>
<td>758</td>
<td>Syndromes associated with anomalies in number and form of chromosomes</td>
</tr>
<tr>
<td>752.0</td>
<td><strong>Anomalies of ovaries</strong></td>
</tr>
<tr>
<td>752.000</td>
<td>Absence or agenesis of ovaries</td>
</tr>
<tr>
<td>752.010</td>
<td>Streak ovary</td>
</tr>
<tr>
<td>752.020</td>
<td>Accessory ovary</td>
</tr>
<tr>
<td>752.080</td>
<td>Other specified anomalies of ovaries</td>
</tr>
<tr>
<td>752.085</td>
<td>Multiple ovarian cysts (single ovarian cysts)</td>
</tr>
<tr>
<td>752.090</td>
<td>Unspecified anomalies of ovaries</td>
</tr>
<tr>
<td>752.1</td>
<td><strong>Anomalies of fallopian tubes and broad ligaments</strong></td>
</tr>
<tr>
<td>752.100</td>
<td>Absence of fallopian tube or broad ligament</td>
</tr>
<tr>
<td>752.110</td>
<td>Cyst of mesenteric remnant</td>
</tr>
<tr>
<td></td>
<td>epoophoron cyst</td>
</tr>
<tr>
<td></td>
<td>cyst of Gartner's duct</td>
</tr>
<tr>
<td>752.120</td>
<td>Fimbrial cyst</td>
</tr>
<tr>
<td></td>
<td>parovarian cyst</td>
</tr>
<tr>
<td>752.190</td>
<td>Other and unspecified anomalies of fallopian tube and broad ligaments</td>
</tr>
<tr>
<td>752.2</td>
<td><strong>Doubling of uterus</strong></td>
</tr>
<tr>
<td>752.200</td>
<td>Doubling of uterus (any degree) or associated with doubling of cervix and vagina</td>
</tr>
<tr>
<td>752.3</td>
<td><strong>Other anomalies of uterus</strong></td>
</tr>
<tr>
<td>752.300</td>
<td>Absence of agenesis of uterus</td>
</tr>
<tr>
<td>752.310</td>
<td>Displaced uterus</td>
</tr>
<tr>
<td>752.320</td>
<td>Fistulae involving uterus with digestive or urinary tract</td>
</tr>
<tr>
<td></td>
<td>Includes: uterointestinal fistula</td>
</tr>
<tr>
<td></td>
<td>uterovesical fistula</td>
</tr>
<tr>
<td>752.380</td>
<td>Other anomalies of uterus</td>
</tr>
<tr>
<td></td>
<td>bicornuate uterus</td>
</tr>
<tr>
<td></td>
<td>unicornsis uterus</td>
</tr>
<tr>
<td>752.390</td>
<td>Unspecified anomalies of uterus</td>
</tr>
<tr>
<td>752.4</td>
<td><strong>Anomalies of cervix, vagina, and external female genitalia</strong></td>
</tr>
<tr>
<td>752.400</td>
<td>Absence, atresia or agenesis of cervix</td>
</tr>
<tr>
<td>752.410</td>
<td>Absence or atresia of vagina, complete or partial</td>
</tr>
<tr>
<td>752.420</td>
<td>Congenital rectovaginal fistula</td>
</tr>
<tr>
<td>752.470</td>
<td>Other cyst of vagina, vulva, or canal of Nuck</td>
</tr>
<tr>
<td>752.490</td>
<td>Unspecified anomalies of cervix, vagina, or external female genitalia</td>
</tr>
</tbody>
</table>
752.5  752.530  Ectopic testis, unilateral and bilateral

752.6  Hypospadias and epispadias
  752.600  Hypospadias (alone), NOS
  752.605  Hypospadias 1°, glandular, coronal (distal shaft)
  752.606  Hypospadias 2°, penile
  752.607  Hypospadias 3°, perineal, scrotal
  752.610  Epispadias
  752.620  Congenital chordee (with hypospadias), NOS
  752.621  Congenital chordee alone (chordee without hypospadias)
  752.625  Congenital chordee with 1°, coronal hypospadias
  752.626  Congenital chordee with 2°, penile hypospadias
  752.627  Congenital chordee with 3°, perineal, scrotal hypospadias

752.7  Indeterminate sex and pseudohermaphroditism
  Excludes: pseudohermaphroditism:
  female, with adrenocortical disorder (see 255.200)
  male, with gonadal disorder with specified
  chromosomal anomaly (see 758)
  752.700  True hermaphroditism
  ovotestis
  752.710  Pseudohermaphroditism, male

  752.720  Pseudohermaphroditism, female
  pure gonadal dysgenesis
  Excludes: gonadal agenesis (see specific: ovaries(752.000),
  testes(752.800))
  752.730  Pseudohermaphroditite, NOS
  752.790  Indeterminate sex, NOS
  ambiguous genitalia

752.8  Other specified anomalies of male genital organs
  752.800  Absence of testis
  monorchidism, NOS
  752.820  Other anomalies of testis and scrotum
  Includes: bifid scrotum  penoscrotal web
  germ cell tumor  penoscrotal transposition
  polyorchidism  shawl scrotum
  Excludes: torsion of the testes or spermatic cord
  752.830  Atresia of vas deferens
  752.840  Other anomalies of vas deferens and prostate
  752.850  Absence or aplasia of penis
  752.865  Small penis, hypoplastic penis, micropenis or concealed penis
  752.870  Cysts of embryonic remnants
cyst:     hydatid of Morgagni
        Wolffian duct
        appendix testis

752.880  Other specified anomalies of genital organs
         Includes:  microgenitalia  agenesis of external genitalia
                     macrogenitalia  testes in genetic female
                     ovaries, fallopian tube, uterus or vagina in genetic male

752.9  752.900  **Unspecified anomalies of genital organs**
         Congenital:  of genital organ, NEC
                     anomaly, NOS or deformity, NOS

753  **Congenital Anomalies of Urinary System**

753.0  **Renal agenesis and dysgenesis**

    753.000  Bilateral absence, agenesis, dysplasia, or hypoplasia of kidneys
             Potter's syndrome
    753.009  Renal agenesis, NOS
    753.010  Unilateral absence, agenesis, dysplasia or hypoplasia of kidneys

753.1  **Cystic kidney disease**

    753.100  Renal cyst (single)
    753.110  Polycystic kidneys, infantile type
    753.120  Polycystic kidneys, adult type

    753.130  Polycystic kidneys, NOS
    753.140  Medullary cystic disease, juvenile type
    753.150  Medullary cystic disease, adult type
             Medullary sponge kidney
    753.160  Multicystic renal dysplasia
             Multicystic kidney
    753.180  Other specified cystic disease
             Includes:  cystic kidneys, NOS

753.2  **Obstructive Defects of Renal Pelvis and Ureter**

    753.200  Congenital hydronephrosis
    753.210  Atresia, stricture, or stenosis of ureter
             Includes:  ureteropelvic junction obstruction/stenosis
                         ureterovesical junction obstruction/stenosis
                         hypoplastic ureter
    753.220  Megaloureter, NOS
             Includes:  megaureter
                         hydroureter
    753.290  Other and unspecified obstructive defects of renal pelvis and ureter
             Includes:  extra renal pelvis
753.3 Other specified anomalies of kidney
   753.300 Accessory kidney
   753.310 Double or triple kidney and pelvis
              pyelon duplex or triplex
   753.320 Lobulated, fused, or horseshoe kidney
   753.330 Ectopic kidney (pelvic kidney)
   753.340 Enlarged, hyperplastic or giant kidney
              Includes: hypertrophied kidney
   753.350 Congenital renal calculi
   753.380 Other specified anomalies of kidney

753.4 Specified anomalies of ureter
   753.400 Absence of ureter
   753.410 Accessory ureter
              double ureter, duplex collecting system
   753.420 Ectopic ureter
   753.480 Other specified anomalies of ureter
              Includes: ureterocele
   753.485 Variations of vesicoureteral reflux
   753.500 Exstrophy of urinary bladder
              ectopia vesicae
              extroversion of bladder

753.6 Atresia and Stenosis of Urethra and Bladder Neck
   753.600 Congenital posterior urethral valves or posterior
              urethral obstruction
   753.610 Other atresia, or stenosis of bladder neck
              Includes: anterior urethral valves
   753.620 Obstruction, atresia or stenosis of anterior urethra
   753.630 Obstruction, atresia or stenosis of urinary meatus
              Includes: meatal stenosis
   753.690 Other and unspecified atresia and stenosis of urethra and urinary
              bladder neck

753.7 Anomalies of urachus
   753.710 Cyst of urachus
   753.790 Other and unspecified anomaly of urachus

753.8 Other specified anomalies of bladder and urethra
   753.800 Absence of urinary bladder or urethra
   753.810 Ectopic urinary bladder
   753.820 Congenital diverticulum or hernia of urinary bladder
<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>753.830</td>
<td>Congenital prolapse of urinary bladder (mucosa)</td>
</tr>
<tr>
<td>753.840</td>
<td>Double urethra or urinary meatus</td>
</tr>
<tr>
<td>753.850</td>
<td>Ectopic urethra or urethral orifice</td>
</tr>
<tr>
<td>753.860</td>
<td>Congenital digestive-urinary tract fistulae</td>
</tr>
<tr>
<td></td>
<td>rectovesical fistula</td>
</tr>
<tr>
<td>753.870</td>
<td>Urethral fistula, NOS</td>
</tr>
<tr>
<td>753.880</td>
<td>Other specified anomalies of urinary bladder and urethra</td>
</tr>
<tr>
<td></td>
<td>Includes: urogenital sinus/fistulabifid bladder</td>
</tr>
<tr>
<td></td>
<td>megameatus</td>
</tr>
<tr>
<td></td>
<td>duplicated bladder</td>
</tr>
<tr>
<td>753.9</td>
<td><strong>Unspecified anomalies of urinary system</strong></td>
</tr>
<tr>
<td>753.900</td>
<td>Unspecified anomaly of kidney</td>
</tr>
<tr>
<td>753.910</td>
<td>Unspecified anomaly of ureter</td>
</tr>
<tr>
<td>753.920</td>
<td>Unspecified anomaly of urinary bladder</td>
</tr>
<tr>
<td>753.930</td>
<td>Unspecified anomaly of urethra</td>
</tr>
<tr>
<td>753.990</td>
<td>Unspecified anomaly of urinary system, NOS</td>
</tr>
<tr>
<td>754</td>
<td><strong>Certain Congenital Musculoskeletal Anomalies</strong></td>
</tr>
<tr>
<td>754.0</td>
<td><strong>Of skull, face, and jaw</strong></td>
</tr>
<tr>
<td></td>
<td>Excludes: dentofacial anomalies (524.0)</td>
</tr>
<tr>
<td></td>
<td>Pierre Robin Sequence (524.080)</td>
</tr>
<tr>
<td></td>
<td>syphilitic saddle nose</td>
</tr>
<tr>
<td>754.000</td>
<td>Asymmetry of face, <em>(if isolated-exclude)</em></td>
</tr>
<tr>
<td>754.010</td>
<td>Compression (Potter's) facies</td>
</tr>
<tr>
<td>754.030</td>
<td>Dolichocephaly, <em>(if isolated-exclude)</em></td>
</tr>
<tr>
<td>754.050</td>
<td>Plagiocephaly</td>
</tr>
<tr>
<td>754.055</td>
<td>Asymmetric head – <em>(if isolated, exclude)</em></td>
</tr>
<tr>
<td>754.060</td>
<td>Scaphocephaly, no mention of craniosynostosis</td>
</tr>
<tr>
<td>754.070</td>
<td>Trigonocephaly, no mention of craniosynostosis</td>
</tr>
<tr>
<td>754.080</td>
<td>Other specified skull deformity, no mention of craniosynostosis</td>
</tr>
<tr>
<td></td>
<td>Includes: brachycephaly</td>
</tr>
<tr>
<td></td>
<td>acrocephaly</td>
</tr>
<tr>
<td></td>
<td>turricephaly</td>
</tr>
<tr>
<td></td>
<td>oxycephaly</td>
</tr>
<tr>
<td>754.090</td>
<td>Deformity of skull, NOS</td>
</tr>
<tr>
<td>754.1</td>
<td><strong>Anomalies of sternocleidomastoid muscle</strong></td>
</tr>
<tr>
<td>754.100</td>
<td>Includes: absent or hypoplastic sternocleidomastoid</td>
</tr>
<tr>
<td></td>
<td>contracture of sternocleidomastoid muscle</td>
</tr>
<tr>
<td></td>
<td>sternomastoid tumor</td>
</tr>
<tr>
<td></td>
<td>Excludes: congenital sternocleidomastoid torticollis (use 756.860)</td>
</tr>
</tbody>
</table>
754.2  **Musculoskeletal deformities of spine**
754.200  Congenital postural scoliosis
754.210  Congenital postural lordosis
754.220  Congenital postural curvature of spine, NOS

754.3  **Congenital hip dislocation**
754.300  Congenital dislocation of hip / Developmental dislocation of hip
754.310  Unstable hip
  - preluxation of hip
  - subluxation of hip
  - predislocation status of hip at birth

754.4  **Congenital genu recurvatum and bowing of long bones of leg**
754.400  Bowing, femur
754.410  Bowing, tibia, and/or fibula
754.420  Bow legs, NOS
754.430  Genu recurvatum
754.440  Dislocation of knee, congenital
754.490  Deformity of leg, NOS

754.5  **Varus (inward) deformities of feet**
754.500  Talipes equinovarus
754.510  Talipes calcaneovarus
754.530  Complex varus deformities
754.590  Unspecified varus deformities of feet

754.6  **Valgus (outward) deformities of feet**
754.600  Talipes calcaneovalgus
754.610  Congenital pes planus
754.615  Pes valgus
754.680  Other specified valgus deformities of foot
754.690  Unspecified valgus deformities of foot

754.7  **Other deformities of feet**
754.700  Pes cavus
  - Claw foot (use 755.350 for claw foot)
754.720  Short Achilles tendon
754.730  Club foot, NOS
talipes, NOS
754.735  Congenital deformities of foot, NOS
754.780  Other specified deformities of ankle and/or toes
  - Excludes:  widespread 1st and 2nd toes
754.8 Other specified congenital musculoskeletal deformities

754.800 Pigeon chest (pectus carinatum)
754.810 Funnel chest (pectus excavatum)
754.820 Other anomalies of chest wall
    Includes: deformed chest, barrel chest
    Pentology of Cantrell
754.825 Shield chest
754.830 Dislocation of elbow
754.840 Club hand or fingers
754.850 Spade-like hand
754.880 Other specified deformity of hands
    (see 755.500 for specified anomalies of fingers)

755 Other congenital anomalies of limbs

755.0 Polydactyly – Type A = well formed finger
    Type B = tag (exclude)
755.005 Accessory fingers (postaxial polydactyly, Type A)
755.007 Unspecified finger or skin tag (postaxial polydactyly, NOS)
755.010 Accessory thumbs (preaxial polydactyly)
755.020 Accessory toes (postaxial)
755.030 Accessory big toe (preaxial) (Bifid big toe)
755.090 Accessory digits, NOS (hand/foot not specified)
755.095 Accessory digits hand, NOS (preaxial, postaxial not specified)
755.096 Accessory digits foot, NOS (preaxial, postaxial not specified)
    Excludes: polydactyly Type B – skin tag

755.1 Syndactyly
755.100 Fused fingers
755.110 Webbed fingers
755.120 Fused toes
755.130 Webbed toes (exclude if isolated: webbing of the second and third toes only)
755.190 Unspecified syndactyly (see below for specified site)
755.191 Unspecified syndactyly thumb and/or fingers unilateral
755.192 Unspecified syndactyly thumb and/or fingers bilateral
755.193 Unspecified (webbed vs. fused) syndactyly thumb and/or fingers, NOS
755.194 Unspecified syndactyly toes unilateral
755.195 Unspecified syndactyly toes bilateral
755.196 Unspecified syndactyly toes, NOS
755.199 Unspecified syndactyly (i.e., webbed vs. fused) digits not known

755.2 Reduction defects of upper limb
Excludes:  shortening of upper limb (use 755.580) or hypoplasia of upper limb (use 755.585)

755.200 Absence of upper limb
  Absent: humerus (total or partial), radius, ulna and hand
  Includes:  amelia of upper limb, NOS
             infants with rudimentary or nubbin fingers
             attached to stump of humerus or shoulder girdle

755.210 Absence of upper arm and forearm
  Absent: humerus (total or partial), radius and ulna (total or partial)
  Present: hand (total or partial)
  Includes:  phocomelia of upper limb, NOS;
             Intercalary reduction defect of upper limb, NOS

755.220 Absence of forearm only or upper arm only
  Absent: radius and ulna
  Present: humerus, hand (total or partial)
  or
  Absent: humerus
  Present: radius, ulna and hand

755.230 Absence of forearm and hand
  Absent: radius and ulna (total or partial) and hand
  Includes:  infants with rudimentary or nubbin fingers
             attached to stump of forearm or elbow

755.240 Absence of hand or fingers
  Absent: hand or fingers (total or partial) not in conjunction with
          ray or long bone reduction
  Includes:  rudimentary or nubbin fingers
             absent individual phalanges
             absent or missing fingers, NOS
  Excludes:  isolated, absent or hypoplastic thumb (use 755.260)

755.250 Split-hand malformation
  Absent: central fingers (third with or without second, fourth) and metacarpals (total or partial)
  Includes:  monodactyly
             lobster-claw hand
  Excludes:  isolated absent central fingers without
             metacarpal defects (use 755.240)

755.260 Preaxial longitudinal reduction defect of upper limb
  Absent: radius (total or partial) and/or thumb with or without second finger (total or partial)
  Includes:  isolated absent or hypoplastic thumb;
             radial ray defect, NOS

755.265 Longitudinal reduction defect of upper limb, NOS
  Includes:  absent forearm long bone with absent fingers, NOS

755.270 Postaxial longitudinal reduction defects of upper limb
Includes:  isolated absent ulna (total or partial);  
absent fifth with or without fourth finger (total or partial)  
only if ulna or fifth \( \pm \) fourth metacarpal also totally  
or partially absent  
ulnar ray defect, NOS  

755.280  Other specified reduction defect of upper limb  
755.285  Transverse reduction defect of upper limb, NOS  
    Includes: congenital amputation of upper limb, NOS  
755.290  Unspecified reduction defect of upper limb, NOS  

755.3  Reduction defects of lower limb  
Excludes:  shortening of lower limb (use 755.680) and  
hypoplasia of lower limb (use 755.685)  

755.300  Absence of lower limb  
    Absent:  femur (total or partial), tibia, fibula, and foot  
    Includes:  amelia of lower limb, NOS  
    Infants with rudimentary or nubbin toes attached  
to stump of femur or pelvic girdle  

755.310  Absence of thigh and lower leg  
    Absent:  femur (total or partial), tibia and fibula  
    (total or partial)  
    Present:  foot (total or partial)  
    Includes:  phocomelia of lower limb, NOS  
    Intercalary reduction defect of lower limb, NOS  

755.320  Absence of lower leg only or femur only  
    Absent:  tibia and fibula  
    Present:  femur, foot (total or partial) or  
    Absent:  femur  
    Present:  tibia, fibula, and foot  

755.330  Absence of lower leg and foot  
    Absent:  tibia and fibula (total or partial), foot  
    Includes:  infants with rudimentary or nubbin toes attached  
to stump of leg or knee  

755.340  Absence of foot or toes  
    Absent:  foot or toes (total or partial) not in conjunction  
    with ray or long bone reduction  
    Includes:  rudimentary or nubbin toes  
    absent individual phalanges  
    absent or missing toes, NOS  
    Excludes:  isolated, absent or hypoplastic great toe (use 755.365)  

755.350  Split-foot malformation  
    Absent:  central toes (third with or without second, fourth)  
    And metatarsals (total or partial)
Includes: monodactyly
Lobster claw foot
Excludes: isolated absent central toes without metatarsal defects (use 755.340)
755.360 Longitudinal reduction defect of lower limb, NOS
Includes: absent long bone of leg with absent toes, NOS
755.365 Preaxial longitudinal reduction defect of lower limb
Absent: tibia (total or partial) and/or great toe with
Or without second toe (total or partial)
Includes: isolated absent or hypoplastic great toe
tibial ray defect, NOS
755.366 Postaxial longitudinal reduction defect of lower limb
Includes: isolated absent fibula (total or partial);
Absent fifth with or without fourth toe (total
or partial) only if fibula or fifth + fourth
metatarsal also totally or partially absent;
fibular ray defect, NOS
755.380 Other specified reduction defect of lower limb
755.385 Transverse reduction defect of lower limb, NOS
Includes: congenital amputation of lower limb, NOS
755.390 Unspecified reduction defect of lower limb

755.4 Reduction defects, unspecified limb
755.400 Absence limb, NOS
Includes: amelia, NOS
755.410 Phocomelia, NOS
Includes: intercalary reduction defect, NOS
755.420 Transverse reduction defect, NOS
Includes: congenital amputation of unspecified limb
755.430 Longitudinal reduction defect, NOS
Includes: preaxial or postaxial reduction defect, NOS
755.440 Absent digits, not specified whether fingers or toes
755.480 Other specified reduction defect of unspecified limb
755.490 Unspecified reduction defect of unspecified limb

755.5 Other anomalies of upper limb, including shoulder girdle
Includes: complex anomalies involving all or part of upper limb
755.510 Anomalies of hand
Includes: trigger thumb or finger
cleft hand (all fingers & metacarpals present)
Excludes: simian crease
Cleft hand with absent fingers and/or metacarpals
(Use 755.250)
755.520 Anomalies of wrist
Includes: dislocation of radial head

755.525 Accessory carpal bones
755.526 Madelung's deformity
755.530 Anomalies of forearm, NOS
755.535 Radioulnar dysostosis
755.536 Radioulnar synostosis
755.540 Anomalies of elbow and upper arm
755.550 Anomalies of shoulder
   Includes: anomalies of the clavicle
755.555 Cleidocranial dysostosis
755.556 Sprengel's deformity
755.560 Other anomalies of whole arm
755.580 Other specified anomalies of upper limb
   Includes: hyperextensibility of upper limb
   shortening of arm
   ulnar deviation
755.585 Hypoplasia of upper limb
   Includes: hypoplasia of fingers, hands, or arms
   Excludes: aplasia or absent upper limb (see 755.2)
755.590 Unspecified anomalies of upper limb

755.6 Other anomalies of lower limb, including pelvic girdle
   Includes: complex anomalies involving all or part of lower limb

755.605 Hallux valgus
755.606 Hallux varus
755.610 Anomalies of foot
   Includes: plantar furrow
   Excludes: lobster claw foot (use 755.350)
755.620 Anomalies of ankle
   Astragaloscaphoid synostosis
755.640 Anomalies of knee
   hyperextended knee

755.645 Genu valgum
755.646 Genu varum
755.647 Absent patella or rudimentary patella
755.650 Anomalies of upper leg
   antversion of femur, (exclude if over 1 year of age at time of diagnosis)
755.660 Anomalies of hip
   Includes: coxa vara
   coxa valga
   other abnormalities of hips
755.665 Hip dysplasia, NOS
755.666 Unilateral hip dysplasia
755.667 Bilateral hip dysplasia
755.670  Anomalies of pelvis
  fusion of sacroiliac joint
755.680  Other specified anomalies of lower limb
  hyperextended legs
  shortening of legs
755.685  Hypoplasia of lower limb
  Includes: hypoplasia of toes, feet, legs
  Excludes: tibia hemimelia (see 755.365)
  aplasia or absent lower limb (see 755.3)
755.690  Unspecified anomalies of legs

755.8  Other specified anomalies of unspecified limb

755.800  Arthrogryposis multiplex congenital
  Includes: amyoplasia congenita
  distal arthrogryposis syndrome
  Temporarily includes: flexion contractures of individual joints
755.810  Larsen's syndrome
755.880  Other specified anomalies of unspecified limb
  Includes: hyperextended joints, NOS
  Excludes: hyperextended knees (use 755.640)

755.9  755.900  Unspecified anomalies of unspecified limb

756  Other Congenital Musculoskeletal Anomalies

756.0  Anomalies of skull and face bones
  Excludes: skull and face deformities in 754
  Pierre Robin Sequence (use 524.080)
756.000  Craniostenosis, NOS
  craniostenosis, NOS
  closed-skull sutures, NOS
756.005  Sagittal craniostenosis
756.006  Metopic craniostenosis
756.010  Coronal craniostenosis
756.020  Lambdoidal craniostenosis
756.030  Other types of craniostenosis
  Includes: basilar craniostenosis
  cloverleaf skull
  multiple craniosynostosis, NOS
756.040  Craniofacial dysostosis
  Includes: Crouzon's disease
  Pfeiffer Syndrome
756.045  Mandibulofacial dysostosis
  Includes: Franceschetti syndrome
Inclusion ICD-9-CM Diagnostic Index - Numeric

756.046 Treacher-Collins syndrome
Other craniofacial syndromes
Includes; oculomandibulofacial syndrome
Hallerman-Streif syndrome
756.050 Acrocephalosyndactyly, NOS
756.055 Acrocephalosyndactyly types I or II
Apert syndrome
756.056 Acrocephalosyndactyly type III (Saethre-Chotzen Syndrome)
756.057 Other specified acrocephalosyndactylies
Includes: Jackson Weiss syndrome
756.060 Goldenhar syndrome
oculoauriculo-vertebral dysplasia
756.065 Hemifacial microsomia
Includes: facio-auriculo-vertebral syndrome
756.080 Other specified skull and face bone anomalies
Includes: Adam-Oliver Syndrome
hypotelorism
localized skull defects
midfacial hypoplasia, (if isolated-exclude)
prominent maxilla
small or stenosis of foramen magnum
cranio-metaphyseal dysplasia
Excludes: macrocephaly (use 742.400)
small chin (see 524.0)
Pierre Robin sequence (use 524.080)
756.085 Hypertelorism, telecanthus, wide set eyes
756.090 Unspecified skull and face bone anomalies
Excludes: dentofacial anomalies (524.0)
skull defects associated with brain anomalies such as:
anencephalus (740.0)
encephalocele (742.0)
hydrocephalus (742.3)
microcephalus (742.100)

756.1 Anomalies of spine
756.100 Spina bifida occulta
756.110 Klippel-Feil syndrome
Wildervanck syndrome
756.120 Kyphosis
kyphoscoliosis
756.130 Congenital spondylolisthesis
756.140 Anomalies of cervical vertebrae
756.145 Hemivertebrae (cervical)
756.146 Agenesis (cervical)
<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>756.150</td>
<td>Anomalies of thoracic vertebrae</td>
</tr>
<tr>
<td>756.155</td>
<td>Hemivertebrae of thoracic vertebrae</td>
</tr>
<tr>
<td>756.156</td>
<td>Agenesis of thoracic vertebrae</td>
</tr>
<tr>
<td>756.160</td>
<td>Anomalies of lumbar vertebrae</td>
</tr>
<tr>
<td>756.165</td>
<td>Hemivertebrae of lumbar vertebrae</td>
</tr>
<tr>
<td>756.166</td>
<td>Agenesis of lumbar vertebrae</td>
</tr>
<tr>
<td>756.170</td>
<td>Sacrococcygeal anomalies</td>
</tr>
<tr>
<td></td>
<td>Includes: agenesis of sacrum (also caudal regression syndrome)</td>
</tr>
<tr>
<td></td>
<td>sacral dysraphism</td>
</tr>
<tr>
<td></td>
<td>Excludes: pilonidal sinus</td>
</tr>
<tr>
<td>756.179</td>
<td>Sacral mass, NOS</td>
</tr>
<tr>
<td>756.180</td>
<td>Other specified vertebral anomalies</td>
</tr>
<tr>
<td>756.185</td>
<td>Hemivertebrae, NOS</td>
</tr>
<tr>
<td>756.190</td>
<td>Unspecified anomalies of spine</td>
</tr>
</tbody>
</table>

### 756.3

**Other anomalies of ribs and sternum**

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>756.300</td>
<td>Absence of ribs</td>
</tr>
<tr>
<td>756.310</td>
<td>Misshapen ribs</td>
</tr>
<tr>
<td>756.320</td>
<td>Fused ribs</td>
</tr>
<tr>
<td>756.330</td>
<td>Extra ribs</td>
</tr>
<tr>
<td>756.340</td>
<td>Other anomalies of ribs</td>
</tr>
<tr>
<td>756.350</td>
<td>Absence of sternum</td>
</tr>
<tr>
<td>756.360</td>
<td>Misshapen sternum</td>
</tr>
<tr>
<td>756.380</td>
<td>Other anomalies of sternum</td>
</tr>
<tr>
<td></td>
<td>Includes: double ossification center in the manubrium, bifid sternum, short sternum</td>
</tr>
<tr>
<td>756.390</td>
<td>Anomalies of thoracic cage, unspecified</td>
</tr>
<tr>
<td></td>
<td>Excludes: deformed chest (use 754.820)</td>
</tr>
</tbody>
</table>

### 756.4

**Chondrodystrophy**

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>756.400</td>
<td>Asphyxiating thoracic dystrophy</td>
</tr>
<tr>
<td></td>
<td>Jeune syndrome</td>
</tr>
<tr>
<td></td>
<td>thoracic-pelvic-phalangeal dysplasia</td>
</tr>
<tr>
<td></td>
<td>Excludes: homozygous achondroplasia (756.430)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>756.410</td>
<td>Chondrodysplasia</td>
</tr>
<tr>
<td></td>
<td>Ollier syndrome, enchondromatosis</td>
</tr>
<tr>
<td>756.420</td>
<td>Chondrodysplasia with hemangioma</td>
</tr>
<tr>
<td></td>
<td>Kast syndrome</td>
</tr>
<tr>
<td></td>
<td>Maffucci syndrome</td>
</tr>
<tr>
<td>756.430</td>
<td>Achondroplastic dwarfism</td>
</tr>
<tr>
<td></td>
<td>Includes: achondroplasia, heterozygous or homozygous</td>
</tr>
<tr>
<td>756.440</td>
<td>Other specified chondrodystrophies</td>
</tr>
<tr>
<td></td>
<td>Includes: hypochondroplasia</td>
</tr>
</tbody>
</table>
Excludes: Conradi's (use 756.575)

756.445 Diastrophic dwarfism
756.446 Metatrophic dwarfism
756.447 Thanatophoric dwarfism
756.450 Metaphyseal dysostosis
756.460 Spondyloepiphyseal dysplasia
756.470 Exostosis
Excludes: Gardner syndrome (see 759.630)
756.480 Other specified chondrodystrophy (same as 756.440)
756.490 Unspecified chondrodystrophy
Excludes: Lipochondrodystrophy

756.5 Osteodystrophies

756.500 Osteogenesis imperfecta
756.505 Osteopsathyrosis
756.506 Fragilitas ossium
756.510 Polyostotic fibrous dysplasia
Albright-McCune-Sternberg syndrome
756.520 Chondroectodermal dysplasia
756.525 Ellis-van Creveld syndrome
756.530 Infantile cortical hyperostosis
Caffey syndrome
756.540 Osteopetrosis
Albers-Schonberg syndrome
marble bones
756.550 Progressive diaphyseal dysplasia
Engelmann syndrome
Camurati-Englemann disease
756.560 Osteopoikilosis
756.570 Multiple epiphyseal dysplasia
756.575 Conradi syndrome
chondrodysplasia punctata
Excludes: warfarin embryopathy
756.580 Other specified osteodystrophies
Includes: Trevor disease
Dysplasia epiphysealis hemimelica
756.590 Unspecified osteodystrophies

756.6 Anomalies of diaphragm

756.600 Absence of diaphragm
756.610 Congenital diaphragmatic hernia
756.615 Diaphragmatic hernia (Bochdalek)
756.616 Diaphragmatic hernia (Morgagni)
756.617 Hemidiaphragm
756.620 Eventration of diaphragm
756.680 Other specified anomalies of diaphragm
756.690  Unspecified anomalies of diaphragm

756.7  **Anomalies of abdominal wall**
756.700  Exomphalos, omphalocele
756.710  Gastrostomia - excludes: umbilical hernia
756.720  Prune belly syndrome (Eagle-Barrett syndrome)
756.790  Other and unspecified anomalies of abdominal wall
    Includes: ventral hernia
    Abdominal cyst, NOS
756.795  Epigastric hernia

756.8  **Other specified anomalies of muscle, tendon, fascia and connective tissue**
756.800  Poland syndrome or anomaly
756.810  Other absent or hypoplastic abdominal wall muscle
    Includes: absent pectoralis major
    Excludes: prune belly syndrome (use 756.720)
756.820  Absent tendon
756.830  Nail-patella syndrome
756.840  Amyotrophy congenital
756.850  Ehlers-Danlos syndrome
756.860  Congenital torticollis
    (see also 754.100, anomalies of sternocleidomastoid muscle)
756.880  Other specified anomalies of muscle, tendon, fascia and connective tissue
    Includes: myopathy, congenital NOS
    congenital myotonic dystrophy
    myotonia congenita

756.9  **Unspecified anomalies of musculoskeletal system**
756.900  Unspecified anomalies of muscle
756.910  Unspecified anomalies of tendon
756.920  Unspecified anomalies of bone
756.930  Unspecified anomalies of cartilage
756.940  Unspecified anomalies of connective tissue
756.990  Unspecified anomalies of musculoskeletal system

757  **Congenital Anomalies of the Integument**
757.0  757.000  Hereditary edema of legs
    Hereditary trophedema
    Milroy's disease
757.1  **Ichthyosis congenita**
757.100  Harlequin fetus
757.110 Collodion baby
757.115 Bullous type
757.120 Sjogren-Larsson syndrome
757.190 Other and unspecified ichthyosis congenital
757.195 Ichthyosis vulgaris
757.196 X-linked ichthyosis
757.197 Ichthysiform erythroderma

757.3 Other specified anomalies of skin
   Excludes: pigmented mole
            hemangioma (see 228.0)
757.300 Specified syndromes, not elsewhere classified, involving skin anomalies
757.320 Urticaria pigmentosa
757.330 Epidermolysis bullosa
757.340 Ectodermal dysplasia
   Exclude: Ellis-van Creveld syndrome (756.525)
757.345 X-linked type ectodermal dysplasia
757.346 Other specified ectodermal dysplasia
757.350 Incontinentia pigmenti
757.360 Xeroderma pigmentosum
757.370 Cutes laxa hyperelastica
757.380 Linear Sebaceous Nevus syndrome
757.395 Absence of skin
   Includes: aplasia cutis

757.4 Specified anomalies of hair
   Excludes: kinky hair syndrome (use 759.870)
757.400 Congenital alopecia
   Excludes: ectodermal dysplasia (use 757.340)
757.410 Beaded hair
   Monilethrix
757.420 Twisted hair
   Pili torti
757.430 Taenzer's hair
757.480 Other specified anomalies of hair
   Includes: Trichothiodystrophy - ichthyosis
            Tay Syndrome

757.5 Specific anomalies of nails
757.500 Congenital anonychia
   Absent nails
757.510 Enlarged or hypertrophic nails
757.515 Onychauxis
757.516 Pachyonychia
757.520  Congenital koilonychia
757.530  Congenital leukonychia
757.540  Club nail
757.580  Other specified anomalies of nails

**757.6  Specified anomalies of breast**
- 757.600  Absent breast with absent nipple
- 757.610  Hypoplastic breast with hypoplastic nipple
- 757.620  Accessory (ectopic) breast with nipple
- 757.630  Absent nipple

**757.8  Other specified anomalies of the integument**
- 757.800  Other specified anomalies of the integument
  - Includes: scalp defects
  - For specified anomalies of skin see 757.390
  - For specified anomalies of hair see 757.480
  - For specified anomalies of nails see 757.580

**757.9  Unspecified anomalies of the integument**
- 757.900  Unspecified anomalies of skin
- 757.910  Unspecified anomalies of hair, NOS
- 757.920  Unspecified anomalies of nail, NOS
- 757.990  Unspecified anomalies of the integument, NOS

**758  Chromosome Anomalies**

**758.0  Down syndrome**
- 758.000  Down syndrome, karyotype trisomy 21, cytogenetics result (written nomenclature or cytogenetic report in record)
- 758.008  Down syndrome, suspected, cytogenetics pending
- 758.010  Down syndrome, karyotype trisomy G, NOS
- 758.020  Down syndrome - translocation trisomy - duplication of a 21
- 758.030  Down syndrome - translocation trisomy - duplication of G, NOS
- 758.040  Mosaic Down syndrome
- 758.090  Down syndrome, NOS (No chromosome results in record but chart states diagnosis of Trisomy 21 or Down syndrome)
- 758.098  Down syndrome suspected, cytogenetics never done

**758.1  Patau Syndrome**
- 758.100  Patau syndrome, karyotype trisomy 13, cytogenetics result (written nomenclature or cytogenetic report in record)
- 758.108  Patau syndrome suspected cytogenetics pending
- 758.110  Patau syndrome, karyotype trisomy D, NOS
- 758.120  Patau syndrome - translocation trisomy - duplication of a 13
- 758.130  Patau syndrome - translocation trisomy - duplication of a D, NOS
- 758.140  Mosaic Trisomy 13 1/98
<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>758.190</td>
<td>Patau syndrome, NOS (no chromosome results in chart, but chart states a diagnosis of Trisomy 13 or Pateau syndrome</td>
</tr>
<tr>
<td>758.198</td>
<td>Patau syndrome suspected, cytogenetics not done</td>
</tr>
<tr>
<td><strong>758.2</strong></td>
<td><strong>Edwards Syndrome</strong></td>
</tr>
<tr>
<td>758.200</td>
<td>Edwards syndrome, karyotype trisomy 18, cytogenetics result (written nomenclature or cytogenetic report in record)</td>
</tr>
<tr>
<td>758.208</td>
<td>Edwards syndrome suspected, cytogenetics pending</td>
</tr>
<tr>
<td>758.210</td>
<td>Edwards syndrome, karyotype trisomy E, NOS</td>
</tr>
<tr>
<td>758.240</td>
<td>Mosaic trisomy 18 10/04</td>
</tr>
<tr>
<td>758.220</td>
<td>Translocation trisomy - duplication of an 18</td>
</tr>
<tr>
<td>758.230</td>
<td>Translocation Trisomey-duplication of an E, NOS</td>
</tr>
<tr>
<td>758.290</td>
<td>Edwards syndrome, NOS (no chromosome results in record, but chart states Trisomy 18 or Edwards Syndrome)</td>
</tr>
<tr>
<td>758.295</td>
<td>Edwards phenotype - normal karyotype</td>
</tr>
<tr>
<td>758.298</td>
<td>Edwards syndrome suspected, cytogenetics not done</td>
</tr>
<tr>
<td><strong>758.3</strong></td>
<td><strong>Autosomal deletion syndromes</strong></td>
</tr>
<tr>
<td>758.300</td>
<td>Antimongolism syndrome</td>
</tr>
<tr>
<td></td>
<td>Clinical antimongolism syndrome:</td>
</tr>
<tr>
<td></td>
<td>karyotype - partial or total deletion of:</td>
</tr>
<tr>
<td></td>
<td>21 G, NOS</td>
</tr>
<tr>
<td>758.310</td>
<td>Cri du chat syndrome</td>
</tr>
<tr>
<td></td>
<td>Clinical Cri du chat syndrome:</td>
</tr>
<tr>
<td></td>
<td>karyotype - deletion of:</td>
</tr>
<tr>
<td></td>
<td>5 B, NOS</td>
</tr>
<tr>
<td>758.320</td>
<td>Wolff-Hirschorn syndrome</td>
</tr>
<tr>
<td></td>
<td>Clinical Wolff-Hirschorn syndrome:</td>
</tr>
<tr>
<td></td>
<td>karyotype - deletion of:</td>
</tr>
<tr>
<td></td>
<td>4 B, NOS</td>
</tr>
<tr>
<td>758.330</td>
<td>Deletion of long arm of 13</td>
</tr>
<tr>
<td></td>
<td>deletion of long arm (q) of D, NOS</td>
</tr>
<tr>
<td>758.340</td>
<td>Deletion of long arm (q) of E</td>
</tr>
<tr>
<td></td>
<td>deletion of long arm of 17 or 18</td>
</tr>
<tr>
<td>758.350</td>
<td>Deletion of short arm (p) of E</td>
</tr>
<tr>
<td></td>
<td>deletion of short arm of 17 or 18</td>
</tr>
<tr>
<td></td>
<td>Includes: Miller Dicker Syndrome (17p-add 742.204)</td>
</tr>
<tr>
<td>758.360</td>
<td>Monosomy G mosaicism</td>
</tr>
<tr>
<td>758.370</td>
<td>Deletion of band 11 of long arm(q) of 22 (22q11 deletion)</td>
</tr>
<tr>
<td></td>
<td>To be used on births after 4/1/2001</td>
</tr>
<tr>
<td>758.380</td>
<td>Other loss of autosomal material</td>
</tr>
</tbody>
</table>
758.390  Unspecified autosomal deletion syndromes
    Includes: microdeletions of autosomes

758.4  758.400  **Balanced autosomal translocation in normal individual**
    Balanced translocation = reciprocal translocation

758.5  **Other conditions due to autosomal anomalies**
758.500  Trisomy 8
758.510  Other trisomy C syndromes
    Trisomy: 6, 7, 9, 10, 11, 12, or C, NOS
758.520  Other total trisomy syndromes
    Trisomy 22  Trisomy 15
    Trisomy, NOS
758.530  Partial trisomy syndromes
    Includes: autosomal duplications
758.540  Other translocations
    Includes: balanced autosomal translocation with abnormal phenotype
    Excludes: balanced translocation in normal individual (use 758.400)
758.580  Other specified anomalies of autosomes, inversions, NOS
    Includes: marker autosome
    Pallester Killian Syndrome
    Ring chromosomes
758.585  Polyploidy
758.586  Triploidy
758.590  Unspecified anomalies of autosomes

758.6  **Gonadal Dysgenesis**
    Excludes: pure gonadal dysgenesis (752.720)
    Noonan syndrome (759.800)
758.600  Turner's phenotype, karyotype 45, X [XO]
758.610  Turner's phenotype, variant karyotypes
    karyotype characterized by:
    isochromosome
    mosaic, including XO
    partial X deletion
    ring chromosome
    Excludes: Turner's phenotype, karyotype normal
    XX (use 759.800, Noonan syndrome)
758.690  Turner syndrome, karyotype unspecified, NOS (no chromosome results)
    Bonneville-Ullrich syndrome, NOS

758.7  **Klinefelter Syndrome**
758.700  Klinefelter's phenotype, karyotype 47, XXY
758.710  Klinefelter's phenotype, other karyotype with additional X chromosomes
XX
XXXY
XXYY
XXXXY
758.790  Klinefelter syndrome, NOS (no chromosome results)

758.8  Other conditions due to sex chromosome anomalies

758.800  Mosaic XO/XY, 45X/46XY
   Excludes: with Turner's phenotype (758.610)

758.810  Mosaic XO/XX
   Excludes: with Turner's phenotype (758.610)

758.820  Mosaic XY/XXX, 46XY/47XXX
   Excludes: Klinefelter's phenotype (758.710)

758.830  Mosaic including XXXXY, 49XXXXY
   Excludes: with Klinefelter's phenotype (use 758.710)

758.840  XYY, male, 47XYY
   mosaic XYY male

758.850  XXX, female, 47XXX
   mosaic XXX female

758.860  Additional sex chromosomes, NOS

758.880  Other specified sex chromosomes anomaly
   Includes: fragile X
   XX/XY mosaicism
   partial deletion of Y chromosome

758.890  Unspecified sex chromosome anomaly
   (use for X, Y translocation)

758.9  Conditions due to anomaly of unspecified chromosomes

758.900  Mosaicism, NOS

758.910  Additional chromosome(s), NOS – (9qht)

758.920  Deletion of chromosome(s), NOS

758.930  Duplication of chromosome(s), NOS

758.990  Unspecified anomaly of chromosome(s)

759  Other and Unspecified Congenital Anomalies

759.0  Anomalies of spleen

759.000  Absence of spleen
   asplenia

759.005  Ivemark syndrome, Polysplenia syndrome, Asplenia syndrome

759.010  Hypoplasia of spleen

759.030  Missshapen spleen

759.040  Accessory spleen

759.050  Ectopic spleen

759.080  Other specified anomalies of spleen
Includes: spleenic cyst
759.090 Unspecified anomalies of spleen

759.1 **Anomalies of adrenal gland**
- 759.100 Absence of adrenal gland
- 759.110 Hyoplasia of adrenal gland
- 759.120 Accessory adrenal gland
- 759.130 Ectopic adrenal gland
- 759.180 Other specified anomaly of adrenal gland
  - Excludes: congenital adrenal hyperplasia (use 255.200)
- 759.190 Unspecified anomalies of adrenal gland

759.2 **Anomalies of other endocrine glands**
- 759.200 Anomalies of pituitary gland
- 759.210 Anomalies of thyroid gland
- 759.220 Thyroglossal duct anomalies
  - thyroglossal cyst
- 759.230 Anomalies of parathyroid gland
- 759.280 Other specified anomalies of endocrine gland
- 759.290 Unspecified anomaly of endocrine gland

759.3 **Situs inversus (heterotaxia)**
- 759.300 Dextrocardia with complete situs inversus (totalis)
- 759.310 Situs inversus with levocardia
- 759.320 Situs inversus thoracis
- 759.330 Situs inversus abdominis
  - Includes: abdominal heterotaxia
- 759.340 Kartagener syndrome (triad)
- 759.390 Unspecified situs inversus
  - Excludes: dextrocardia (746.800) not associated with complete situs inversus

759.4 **Conjoined twins**
- 759.400 Dicephalus (two heads)
- 759.410 Craniopagus
  - head-joined twins
- 759.420 Thoracopagus
  - thorax-joined twins
- 759.430 Xiphopagus
  - xiphoid- and pelvis-joined twins
- 759.440 Pygopagus
Inclusion ICD-9-CM Diagnostic Index - Numeric

759.480 Buttock-joined twins
759.490 Other specified conjoined twins
759.490 Unspecified conjoined twins

**759.5 759.500 Tuberous sclerosis**
Bourneville's disease
epiloia

**759.6 759.600 Other hamartoses, not elsewhere classified**
Peutz-Jegher syndrome
Encephalocutaneous angiomatosis
Kalischer's disease
Sturge-Weber syndrome
Von Hippel-Lindau syndrome
Gardner syndrome
Other specified hamartomas
Unspecified hamartomas

**759.7 759.700 Multiple congenital anomalies (Do not use if any specific anomalies stated)**
anomaly, multiple, NOS
deformity, multiple, NOS

**759.8 759.800 Other specified anomalies and syndromes**
Cong malformation syndromes affecting facial appearance
Noonan syndrome
Oral-facial-digital (OFD) syndrome, type I
Orofaciodigital syndrome, type II (Mohr syndrome)
Velo-Cardio Facial (VCF)
Waardenburg syndrome
Whistling face syndrome
Pena-Shokeir syndrome (Fetal Akinesia Sequence)
Tricho-rhino-phalangeal syndrome
Oto-palato-digital syndrome
Oculo-dental-digital syndrome
Dubowitz syndrome
Friedman-Sheldon syndrome
Coleman Hughes syndrome
Amsterdam dwarf (Cornelia de Lange syndrome)
Cockayne syndrome
Laurence-Moon-Biedl syndrome
Russell-Silver syndrome
Seckel syndrome
Smith-Lemli-Opitz syndrome
Schwartz-Jamble syndrome
Hurler’s Syndrome
Short Rib-polydactyly syndrome Type II (Majewski Syndrome)
Robinow syndrome

759.840 Congenital malformation syndromes involving limbs
Carpenter syndrome
Holt-Oram syndrome
Klippel-Trenaunay-Weber syndrome
Rubenstein-Taybi syndrome
Sirenomelia
Thrombocytopenia-absent radius (TAR) syndrome
VATER Syndrome
Charcot Marie Tooth
Beals Syndrome
Towne-Brock Syndrome

759.860 Congenital malformation syndromes with other skeletal changes
Marfan syndrome
Stickler syndrome
Jacho-Levine Syndrome (spondylocostal dysplasia)

759.870 Congenital malformation syndromes with metabolic disturbances
Alport syndrome
Beckwith (Wiedemann-Beckwith) syndrome
Leprechaunism
Menke syndrome (kinky hair syndrome)
Prader-Willi syndrome
Zellweger syndrome
Cerebro-Costo-Manibular Syndrome
Sotos Syndrome

759.890 Other specified anomalies
Includes: hemihypertrophy
Low syndrome
Meckel-Gruber syndrome
Cat Eye syndrome
Opitz syndrome
Marden-Walker syndrome
Landau-Kleffner syndrome
Fryns syndrome
Currrarino’s Triad
Joubert syndrome
Williams syndrome
Levi-Hollister syndrome
Aarskog syndrome
Rett syndrome
Hecht syndrome
Angelman syndrome

759.9 Congenital anomaly, unspecified
759.910 Embryopathia, NEC
759.990 Congenital anomaly, NOS
Inclusion ICD-9-CM Diagnostic Index - Numeric

760 760.710  **Fetal alcohol syndrome, (list stigmata)**
760.718 Fetal alcohol syndrome, probable (Includes: "facies")
760.750 Fetal hydantoin (Dilantin) syndrome

771 **Congenital infections in utero infections only**
Excludes: Congenital Syphilis
771.000 Rubella congenital
771.090 TORCH infection - unspecified
771.100 Congenital cytomegalovirus (CMV) infection
771.210 Toxoplasmosis
771.220 Herpes Simplex
    Includes: encephalitis
    meningoencephalitis
771.280 Congenital infection - other specified
    Excludes: human immunodeficiency virus (HIV)
    infection and acquired immunodeficiency
    syndrome (AIDS)

774.4 774.480  **Hepatitis neonatal - other specified**
774.490 Hepatitis neonatal NOS

779.900  **Stillbirth NEC (not elsewhere classified)**
V27.3 Twins, one liveborn and one stillborn
V27.4 Twins, both stillborn
V27.6 Other multiple birth, some liveborn
V27.7 Other multiple birth, all stillborn
V27.9 Unspecified outcome of delivery
V32 Twin, mate stillborn
V35 Other multiple, mates all stillborn
V36 Other multiple, mates liveborn and stillborn

Revised 12/04