7.1 BIRTH DEFECTS CODE LIST

Based on
the British Pediatric Association (BPA) Classification of Diseases (1979)
and the World Health Organization's International Classification of Diseases,
9th Revision, Clinical Modification (ICD-9-CM) (1979)

Code modifications developed by

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INTRODUCTION TO BIRTH DEFECT DIAGNOSIS CODING

Purpose

Any given birth defect can be described in a number of different ways. For example, ventricular septal defect can also be described as interventricular septal defect, defect of the ventricular septum, and VSD. Tracheoesophageal fistula is also called tracheal-esophageal fistula, esophageal-tracheal fistula, and TE fistula. Anal atresia and imperforate anus are different terms for the same defect.

Coding birth defects eliminates the problem of having to sort through a variety of differing descriptions. It allows for timely and efficient analyses of data and referral of cases. Coding of birth defects also enables researchers, both inside and outside Texas, to know that they are all talking about the same defect, and it allows for comparability between different birth defects registries using the same or comparable coding systems.

BPA coding system

The current birth defect coding system was provided by the Centers for Disease Control and Prevention (CDC) and is based on the British Pediatric Association (BPA) Classification of Diseases (1979). From this point on, the birth defects coding system will be referred to as the BPA coding system.

The BPA coding system was derived from the World Health Organization's International Classification of Disease, 9th Revision, Clinical Modification (ICD-9-CM)(1979). The ICD-9 system is a 4-5-digit hierarchical coding system used by hospitals nationally and internationally to code diseases, conditions, and procedures. A birth defects registry staff person is likely to encounter ICD-9 codes through hospital discharge lists and notations on the discharge summary in medical records.

In general, the BPA coding system was created from the ICD-9 coding system by the addition of a fifth and sixth digit. For example, the following table shows the corresponding ICD-9 and BPA codes for several diagnoses:

Diagnosis	ICD-9 Code	BPA Code
Anencephaly	740.0	740.020
Ventricular cysts	742.4	742.485
Sequestration of lung	748.5	748.520
Tracheoesophageal fistula	750.3	750.320
Genu varum	755.64	755.646
Sagittal craniosynostosis	756.0	756.005
Lambdoidal craniosynostosis	756.0	756.020
Omphalocele	756.7	756.700
Gastroschisis	756.7	756.710

However, there are a number of exceptions to this rule, as shown by the examples in the following table:

Diagnosis	ICD-9 Code	BPA Code
Absence of iris	743.45	743.420
Low set ears	744.29	744.245
Rocker-bottom foot	754.61	755.616
Apert syndrome	755.55	756.050

The Birth Defects Epidemiology and Surveillance (BDES) Branch uses the BPA coding system instead of the ICD-9 coding system for a couple of reasons. One reason is that the additional digits allow greater distinction between similar defects (e.g., see omphalocele and gastroschisis in the first table above). Also, the BPA coding

system is used by the CDC, as well as a number of other birth defects registries in the United States, thus allowing for comparability of data between the registries.

Description of the BPA code

The BPA code is a six-digit code with a decimal point after the third digit. The majority of the codes are between 740.000 and 759.999 because the ICD-9 range of codes for birth defects is 740.0-759.9. A few defects or other diagnoses of interest to BDES have BPA codes outside of this range. They are found at the back of the BPA coding manual in the list of "Other Specified Codes".

The BPA coding system is a hierarchical system, going from general to specific. The first five digits starting from the left are generally grouped by organ system, then by particular organs, and then by specific defects for that organ. (See the Table of Contents for the general organization of organ systems and organs.)

Examples:

743 Congenital anomalies of eye
743.3 Congenital cataract and lens anomalies
743.33 Displaced lens

748 Congenital anomalies of respiratory system
748.6 Other anomalies of lung
748.62 Accessory lobe of lung

753 Congenital anomalies of urinary system
753.4 Other specified anomalies of ureter
753.40 Absence of ureter

The sixth digit is sometimes used to indicate greater specificity for a particular defect. The sixth digits '5', '6', and '7' are used in a small number of cases where greater specificity is required than can be provided by the first five digits.

Problems with the BPA coding system

The BPA coding system does have various shortcomings. These include:

- In order to allow for very specific and detailed coding of diagnoses, the coding system is large and complex, allowing for thousands of potential codes to be used.
- It requires some knowledge of medical terminology, anatomy, and birth defects.
- It was created several decades ago, and knowledge of birth defects has changed since then, so some of the organization of defects is incomplete and/or obsolete.
- Some of the sections, such as those for the musculoskeletal system and chromosomal abnormalities, are confusing.
- Not all possible birth defects are explicitly mentioned, so there can be some question about how to code a particular defect, or even whether it should be coded at all.
- Explicit instructions on a number of diagnoses are not included.
- There are exceptions to many of the rules (e.g., laterality does not apply to all diagnoses).
- There is no guiding organization to approach if we have questions about the BPA coding system.

Steps are currently being taken to address some of these difficulties. For example, BDES has created an alphabetic cross-linked index of birth defects and their BPA codes, and is amending the BPA code manual to include additional birth defects as well as guidelines for when and how to code selected defects. Also, one of the duties of the National Birth Defects Prevention Network in the future may be to serve as an arbiter of coding questions.

In spite of its problems, the BPA coding system is the best coding system for birth defects that is currently available for the reasons outlined previously.

INSTRUCTIONS

Inclusion status

A number of guidelines need to be followed when determining whether or not a defect should be listed in the "Birth Defect Information" section of the abstraction form. Among these guidelines are:

- The defect must be described or referred to in the "Prenatal Procedures and Tests", "Postnatal Procedures", or "Abstractor Notes" section of the abstraction form. Do not list a defect in the "Birth Defect Information" section just because the hospital listed the ICD-9 code. Hospitals mainly use ICD-9 codes for billing purposes. Sometimes hospitals assign the wrong ICD-9 code or they may not code all of the defects.

- The defect must be included among those diagnoses that are reportable to BDES. In order to determine whether a given diagnosis or condition is reportable, consult the BPA coding manual or the birth defects index. Keep in mind though, that the manual and the index are not complete. There may be reportable diagnoses that are not explicitly listed in these documents. Also, not all of the defects in the manual or the index should always be listed in the "Birth Defect Information" section. The symbols "#", and "*" located to the left of selected defects are used to denote the inclusion status of those defects. If the defect is marked with any of these symbols, the BPA coding manual needs to be reviewed to determine if that particular defect should be listed.

  Note: The following instructions and examples assume that the information found in the chart meets all of the other criteria for inclusion in the Registry listed in Section 3 (e.g., mother's county of residence at delivery).

  - If one or more defects without any symbols (i.e., without a pound sign (#) or an asterisk (*)) appear in a chart, an abstraction form should always be filled out.

    Example:

    If the following defects appeared in a chart,

	758.200	Edwards syndrome, karyotype trisomy 18
L	753.200	Congenital hydronephrosis
	744.810	Microstomia (small mouth)

    an abstraction form should be filled out.

  - If a defect with a pound sign (#) appears in a chart, singly or along with other defects which all have a pound sign (#), an abstraction form should not be filled out.

    Example:

    If only the following defects appeared in a chart,

#	750.000	Tongue tie
#	757.385	Birthmark, NOS

    an abstraction form should not be filled out.

  - If one or more defects with a pound sign (#) appear in a chart along with a defect without a pound sign (#), an abstraction form should be filled out, and all defects (including those with a pound sign (#)) should be included on the abstraction form.

    Example:

If the following defects appeared in a chart,

		745.100	Transposition of great vessels, complete (no VSD)
L	#	744.245	Low set ears
	#	747.500	Single umbilical artery

an abstraction form should be filled out, and all of the defects should be included on the form.

- If a defect with an asterisk (*) appears in a chart, the special instructions for that defect must be considered in order to determine whether or not it should be abstracted.

  Example:

  If the following defect appeared alone in a chart,

  L * 755.130 Webbed toes [1]

  Special instructions:

  [1] *Code webbing of the second and third toes only if another reportable defect is present. Always code webbing of other toes regardless of whether another reportable defect is present.*

  an abstraction form should only be filled out if the webbing involved toes other than the second and third toes.

- If one or more defects with a pound sign (#) appear in a chart, along with a defect with an asterisk (*), the special instructions for the defect with the asterisk (*) must be considered first in order to determine whether or not it should be abstracted. If it should, an abstraction form should be filled out, and all conditions and defects (including those with a pound sign (#)) should be included on the abstraction form.

  Example:

  If the following defects appeared in a chart,

  L # 744.246 Posteriorly rotated ears
  L * 748.510 Hypoplasia of lung; Pulmonary hypoplasia [1]

  Special instructions:

  [1] *If the infant is less than 36 weeks gestation, code only if another reportable defect is present. Always code if greater than or equal to 36 weeks gestation.*

  L # 755.616 Rocker-bottom foot

  and the chart stated that the infant was greater than or equal to 36 weeks gestation, an abstraction form should be filled out and all of the defects should be included on the form.

- If a defect listed on the Exclusion List (Section 7.3) appears in a chart, do not include it on any abstraction form.

  Example:

  If the following defects appeared in a chart,

  L 756.610 Congenital diaphragmatic hernia
  Hydrocephalus secondary to intraventricular hemorrhage (IVH) or CNS bleed

  an abstraction form should be filled out, but the hydrocephalus should not be included on the form.

- The wording “Includes” and “Excludes” listed under some six-digit codes refers to other defects that should or should not be coded using that six-digit code. These lists are not exhaustive. They simply give some examples of other defects that should or should not be coded using that six-digit code.

  Example:

  For the following six-digit code,

  746.300 Congenital stenosis of aortic valve
  Includes: Congenital aortic stenosis
  Subvalvular aortic stenosis
  Excludes: Supravalvular aortic stenosis (747.220)

  congenital aortic stenosis and subvalvular aortic stenosis should be coded 746.300. However, supravalvular aortic stenosis should not be coded 746.300. Instead it should be coded 747.220.

- All reportable defects should be listed and coded unless explicitly stated otherwise in these procedures. Those defects that should not be listed and coded in the presence of another specific defect have special instructions stating that (e.g., the instructions for macrocephaly state that it should never be coded in the presence of hydrocephaly, the instructions for dolichocephaly state that it should never be coded in the presence of craniosynostosis, the instructions for spinal dysraphism state that it should never be coded in the presence of spina bifida, etc.). Those syndromes for which the components should not be listed and coded also have instructions stating that (e.g., the instructions for hypoplastic left heart syndrome state that, in general, the components of that syndrome should not be listed and coded separately).

- General defects (e.g., those with NOS) should not be listed if a more specific defect is known. For example, do not list congenital heart disease if the specific heart defect is known. The exception is when the general defect is confirmed but the more specific defect is not (i.e., it is possible/probable). If a general birth defect has been definitively diagnosed, but the specific type of the defect is only reported as possible or probable, code both the definitive general diagnosis and the possible/probable specific diagnosis. If the general diagnosis is not coded, then a definitive, if general, diagnosis may be excluded from analyses.

  For example, a case may be reported as hydrocephaly secondary to probable aqueductal stenosis. In this instance, the general diagnosis is hydrocephaly while the specific type of hydrocephaly is probable aqueductal stenosis. If only the probable aqueductal stenosis is coded, then a definitive diagnosis of hydrocephaly is excluded. Therefore, code both the hydrocephaly and the probable aqueductal stenosis. However, if the diagnosis of aqueductal stenosis had been definitive, then only the aqueductal stenosis would need to be coded. The more general diagnosis of hydrocephaly would not need to be coded.

- A given defect should only be listed once. Frequently a given defect will be referred to by different synonymous terms in the medical record. For example, a defect may be called an imperforate anus in one report and an anal atresia in another. Or hydrocephalus may be referred to as ventriculomegaly on a prenatal ultrasound. You may list the synonymous terms in the same description if you wish - imperforate anus (anal atresia), hydrocephalus (ventriculomegaly); however, they should not be listed as separate defects.

- Sometimes two defects are coded together. Examples of this are spina bifida and hydrocephalus; cleft lip and cleft palate; imperforate anus and a fistula; esophageal atresia and tracheoesophageal fistula; and hypospadias and chordee. If two defects can be coded together, they should be listed as a single description, not listed and coded separately. The exception is when one of the defects is definitively diagnosed and the other is possible/probable. In this case, the two diagnoses should be listed and coded separately so that the fact that one of the defects was definitively diagnosed is not lost.

  For example, if the diagnosis is hypospadias with possible chordee, do not list and code the diagnoses together. List and code them separately as hypospadias and possible chordee.

- In some cases, the infant/fetus may have several different defects that can be assigned the same code. For example, an infant may have clinodactyly and a digitalized thumb. Both of these defects are assigned the same code (755.500). Both of these defects should be listed and coded separately.

- If there is any question as to whether a particular diagnosis or condition should be listed, either list the condition, or consult your field supervisor, a clinical reviewer, or the central office.

Description of the birth defect

The BPA coding manual is used to code birth defects listed in the "Birth Defect Information" section of the abstraction form. Before a defect is coded though, a description of the defect must first be listed in the "Birth Defect Information" section. A list of BPA codes alone is not enough. The description of the defect is needed to insure that the correct BPA code has been assigned and to make it easier to reassign codes in the future if necessary. The description also allows for easier review of concise diagnostic information than would be possible if researchers had to scour the "Prenatal Procedures and Tests", "Postnatal Procedures", and "Abstractor Notes" sections of the abstraction form.

There are a number of guidelines that need to be followed when describing birth defects in the "Birth Defect Information" section of the abstraction form.

- The description must be as complete and accurate as possible.

- For some defects, you will have to combine information from multiple procedures. For example, one procedure may report that the infant had a myelomeningocele, while a second may mention a lumbar spina bifida. These should all be combined into a single description such as "lumbar myelomeningocele." Or one procedure may mention the infant had a cleft lip and palate, while second procedure notes that the cleft lip was only on the left side of the mouth. These should be combined into something like "left cleft lip and palate."

- If the laterality is reported for the defect, include the laterality in the description. Laterality should be included in the description even if an "L" is not located to the left of the code.

- If the defect is reported to be only possible, then this needs to be noted in the description.

- If there is some subclassification of the defect that is coded, include that classification in the description. For example, hypospadias is classified by its location (first degree/glandular/coronal, second degree/penile, third degree/perineal/scrotal) and the presence or absence of penile chordee. Craniosynostosis is distinguished by which cranial sutures are affected (sagittal, metopic, coronal, lambdoidal). If this information is known, it should be used in the description.

- For chromosomal abnormalities, if a cytogenetic analysis (karyotype) was performed and the result is known, the karyotype should be included in the description (e.g., 47,XX,+21; 45,X).

- Do not include in the description any information that cannot be directly derived from the prenatal or postnatal procedures unless so instructed by the clinical reviewer. For example, if the postnatal procedures simply report a cleft palate, but the clinical reviewer notes that, because of other diagnoses, the cleft palate must be bilateral and that this should be recorded, then the description should be bilateral cleft palate or some variation of this.

- The description should be unambiguous.

- Only commonly accepted abbreviations should be used.

- The description should be spelled correctly. This is especially true when abbreviations are used. Unlike the "Results and Interpretation" fields in the "Prenatal Procedures and Tests" and "Postnatal Procedures"

sections, misspellings in the medical record should be corrected when defects are listed in the "Birth Defect Information" section.

- The description should not be the same as the description listed in the BPA coding manual corresponding to the code unless that is the description derived from the prenatal or postnatal procedures. For example, if the procedures report the defect as an imperforate anus and make no mention of a fistula, do not list the defect as stenosis, atresia, or absence of anus without mention of fistula.

- As long as the description meets the other criteria listed above, it can be written any number of ways. For example,

  left cleft lip
  cleft lip, left
  cleft left lip
  left cleft of lip

  are all acceptable descriptions for the same defect.

Coding the birth defect

Once a defect is listed in the "Birth Defect Information" section, the defect needs to be coded. To do this, search a copy of the BPA coding manual or the birth defects index for the appropriate code. If the defect was assigned an ICD-9 code in the medical record, the ICD-9 code can be used as a guide to the first digits of the BPA code. However, as noted previously, BPA codes can deviate from the corresponding ICD-9 code. If searching an electronic version of the manual or index, you may use the "find" option. If you are using the birth defects index and the defect is marked with a symbol (e.g., #, *, x), be sure to consult the BPA coding manual because there may be special instructions for the particular defect.

Always double-check to make certain that the correct BPA code has been assigned to each defect. This is very important because analyses will be performed using the BPA code, and incorrect codes will decrease the usefulness of the analyses.

If there is any question as to the proper way to code a defect, consult your field supervisor, a clinical reviewer, or the central office.

Laterality

If there is an "L" located to the left of the code, be sure to record the laterality of the defect in the "Laterality" field. Note: An "L" located to the left of a code applies to <u>all</u> defects that fall under that code. This includes all of the defects that are explicitly listed under that code in the manual, as well as any defects that should be assigned that code, but that are not explicitly listed in the manual.

Possible/Probable

If the most definitive information available in the chart states that a defect is possible/probable, be sure to record that information in the "Possible/Probable" field. You should also use the "Possible/Probable" field to indicate that the defect is possible/probable when the diagnosis of the defect is based solely on a prenatal diagnosis (i.e., where there is no postnatal confirmation of the diagnosis). The only exception to this rule is a diagnosis based on genetic lab tests such as chromosomal analyses, karyotypes, or FISH.

CONGENITAL ANOMALIES

740 Anencephalus and Similar Anomalies

740.0 Anencephalus

740.000 Absence of brain
740.010 Acrania
740.020 Anencephaly
Exencephaly
Cranioschisis
740.030 Hemianencephaly, hemicephaly
740.080 Other

740.1 Craniorachischisis

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)
Myelocele
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
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
Meningomyelocele, cervical, with unspecified hydrocephalus
Myelomeningocele, cervical, with unspecified hydrocephalus
Meningomyelocele, cervicothoracic with unspecified hydrocephalus
Myelomeningocele, cervicothoracic with unspecified hydrocephalus

741.040 Spina bifida cystica, thoracic, with unspecified hydrocephalus, no mention of Arnold-Chiari
Meningomyelocele, thoracic, with unspecified hydrocephalus
Myelomeningocele, thoracic, with unspecified hydrocephlus
Meningomyelocele, thoracolumbar with unspecified hydrocephalus
Myelomeningocele, thoracolumbar with unspecified hydrocephalus

741.050 Spina bifida cystica, lumbar, with unspecified hydrocephalus, no mention of Arnold-Chiari
Meningomyelocele, lumbar, with unspecified hydrocephalus
Myelomeningocele, lumbar, with unspecified hydrocephlus
Meningomyelocele, lumbosacral with unspecified hydrocephalus
Myelomeningocele, lumbosacral with unspecified hydrocephalus

741.060 Spina bifida cystica, sacral, with unspecified hydrocephalus, no mention of Arnold-Chiari
Meningomyelocele, sacral, with unspecified hydrocephalus
Myelomeningocele, sacral, with unspecified hydrocephlus
Meningomyelocele, sacrococcygeal with unspecified hydrocephalus
Myelomeningocele, sacrococcygeal with unspecified hydrocephalus

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
Cervical meningocele

741.086 Spina bifida, meningocele thoracolumbar, with hydrocephalus
Thoracic meningocele

741.087 Spina bifida, meningocele, lumbosacral with hydrocephalus
Lumbar meningocele
Sacral meningocele

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
Includes: Cervical myelocele, myelomeningocele, meningomyelocele, and meningocele without hydrocephalus

741.920 Spina bifida (cystica), thoracic, without hydrocephalus
Includes: Thoracic myelocele, myelomeningocele, meningomyelocele, and meningocele without hydrocephalus

741.930 Spina bifida (cystica), lumbar, without hydrocephalus
Includes: Lumbar myelocele, myelomeningocele, meningomyelocele, and meningocele without hydrocephalus

741.940 Spina bifida (cystica), sacral, without hydrocephalus
Includes: Sacral myelocele, myelomeningocele, meningomyelocele, and meningocele without hydrocephalus

741.980 Spina bifida, other specified site, without hydrocephalus
Spina bifida, cervicothoracic, without hydrocephalus
Spina bifida, thoracolumbar, without hydrocephalus
Spina bifida, lumbosacral, without hydrocephalus
Spina bifida, sacrococcygeal, without hydrocephalus
Includes: Cervicothoracic myelocele, myelomeningocele, meningomyelocele, and meningocele without hydrocephalus

Thoracolumbar myelocele, myelomeningocele, meningomyelocele, and meningocele without hydrocephalus
Lumbosacral myelocele, myelomeningocele, meningomyelocele, and meningocele without hydrocephalus
Sacrococcygeal myelocele, myelomeningocele, meningomyelocele, and meningocele without hydrocephalus

741.985 Lipomyelomeningocele
Lipomeningocele

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

742 Other Congenital Anomalies of Nervous System

742.0 Encephalocele-

742.000 Occipital encephalocele
Occipital meningocele
Posterior encephalocele
Occipitocervical encephalocele

742.080 Other encephalocele of specified site (includes midline defects)
Sphenoid encephalocele

742.085 Frontal encephalocele
Frontonasal encephalocele

742.086 Parietal encephalocele

742.090 Unspecified encephalocele

742.1 Microcephalus

742.100 Microcephalus
Small head

742.2 Reduction deformities of brain

742.200 Anomalies of cerebrum
Anomalies of frontal lobes
Anomalies of cortex (brain)
Excludes: Cortical atrophy (Use 742.480)

* 742.210 Anomalies of corpus callosum
Hypoplasia of septum pellucidum [1]
Special instructions:
[1] *Never code an absent septum pellucidum in the presence of an absent corpus callosum.*

742.220 Anomalies of hypothalamus

742.230 Anomalies of cerebellum
Anomalies of inferior vermis
Cerebellar atrophy
Posterior fossa cyst (not associated with Dandy-Walker malformation)
Vermian atrophy

742.240 Agyria and lissencephaly

742.250 Microgyria, polymicrogyria

742.260 Holoprosencephaly

Fused thalami

742.270 Arrhinencephaly
Absent olfactory nerve
Hypoplastic olfactory nerve

* 742.280 Other specified reduction defect of brain
Includes: Colpocephaly [1]
Pachygyria
Schizencephaly
Pontine hypoplasia
Hypoplastic thalamus
Reduction defect of brainstem
Hypoplastic brainstem
Small brainstem
Special instructions:
[1] *Never code colpocephaly in the presence of agenesis of the corpus callosum.*

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
Includes: Aqueductal stenosis

* 742.310 Atresia of foramina of Magendie and Luschka
Dandy-Walker syndrome [1]
Special instructions:
[1] *If a diagnosis of Dandy-Walker syndrome is made, do not list and code the hypoplasia/aplasia of the cerebellar vermis or the dilated fourth ventricle separately.*

742.320 Hydranencephaly

742.380 Other specified hydrocephaly
Includes: Communicating hydrocephaly
Enlarged cisterna magna
Non-communicating hydrocephaly

742.390 Unspecified hydrocephaly, NOS
Enlarged ventricles
Ventriculomegaly
Dilation ventricles

742.4 Other specified anomalies of brain

* 742.400 Enlarged brain and/or head
Megalencephaly [1]
Macrocephaly [1]
Special instructions:
[1] *Never code in the presence of hydrocephaly.*

* 742.410 Porencephaly
Includes: Porencephalic cysts [1]
Special instructions:
[1] *Never code if secondary to intraventricular hemorrhage.*

742.420 Cerebral cysts
Subependymal cyst

Periventricular cyst
Intracranial cyst
Corpus callosum cyst
Ependymal cysts
Glioependymal cysts

742.480 Other specified anomalies of brain
Includes: Cortical atrophy
Cranial nerve defects
Anomalies of brainstem
Cerebral atrophy
Arnold-Chiari malformation without spina bifida
Cortical dysplasia (cerebral)
Excludes: Reduction defect of brainstem (Use 742.280)

* 742.485 Ventricular cysts
Choroid plexus cyst [1]
Excludes: Arachnoid cysts
Special instructions:
[1] *Code only if there are multiple cysts that were diagnosed postnatally. Never code in an infant of < 33 weeks gestation.*

742.486 Small brain

742.5 Other specified anomalies of spinal cord

742.500 Amyelia

742.510 Hypoplasia and dysplasia of spinal cord
Atelomyelia
Myelodysplasia

742.520 Diastematomyelia

742.530 Other cauda equina anomalies

742.540 Hydromyelia
Hydrorachis
Syringohydromyelia
Syringomyelia

742.580 Other specified anomalies of spinal cord and membranes
Includes: Congenital tethered cord

742.8 Other specified anomalies of nervous system

Excludes: Congenital oculofacial paralysis
Moebius syndrome (Use 352.600)

742.800 Jaw-winking syndrome
Marcus Gunn syndrome

742.810 Familial dysautonomia
Riley-Day syndrome

* 742.880 Other specified anomalies of nervous system
Septo-optic dysplasia [1]
Walker-Warburg syndrome
Special instructions:
[1] *The individual components of this defect that are present (e.g., absent septum pellucidum, optic nerve hypoplasia) should also be listed and coded separately.*

742.9 Unspecified anomalies of brain, spinal cord and nervous systems

	742.900	Brain, unspecified anomalies
	742.910	Spinal cord, unspecified anomalies
	742.990	Nervous system, unspecified anomalies

743 Congenital Anomalies of Eye

743.0 Anophthalmos

L	743.000	Anophthalmos Agenesis of eye Cryptophthalmos

743.1 Microphthalmos

L	743.100	Microphthalmos, small eyes Aplasia of eye Atrophic eye globe Dysplasia of eye Hypoplasia of eye Rudimentary eye

743.2 Buphthalmos

L	743.200	Buphthalmos Congenital glaucoma Hydrophthalmos
L	743.210	Enlarged eye, NOS
L	743.220	Enlarged cornea Keratoglobus Congenital megalocornea Macrocornea

743.3 Congenital cataract and lens anomalies

L	743.300	Absence of lens Congenital aphakia
L	743.310	Spherical lens Spherophakia
L	743.320	Cataract, NOS
L	743.325	Cataract, anterior polar
L	743.326	Cataract, other specified
L	743.330	Displaced lens
L	743.340	Coloboma of lens
L	743.380	Other specified lens anomalies Lenticonus
L	743.390	Unspecified lens anomalies

743.4 Coloboma and other anomalies of anterior segments

L		743.400	Corneal opacity Cloudy cornea Leukoma cornea
L		743.410	Other corneal anomalies Microcornea Sclerocornea Excludes: Megalocornea (Use 743.220)
L		743.420	Absence of iris Aniridia
L		743.430	Coloboma of iris
L		743.440	Other anomalies of iris Anisocoria Ectopic pupil Peter's anomaly Polycoria Microcoria Excludes: Brushfield spots (Use 743.800)
L	*	743.450	Blue sclera [1] *Special instructions:* [1] *Never code if the infant is less than 36 weeks gestation. If the infant is greater than or equal to 36 weeks gestation, code only if another reportable defect is present.*
L		743.480	Other specified colobomas and anomalies of anterior segments Rieger's anomaly
L		743.490	Unspecified colobomas and anomalies of anterior eye segments Coloboma NOS

743.5 Congenital anomalies of posterior segment

L		743.500	Specified anomalies of vitreous humour Persistent hyperplastic primary vitreous
L		743.510	Specified anomalies of retina Congenital retinal aneurysm Excludes: Stickler syndrome (Use 759.860)
L		743.520	Specified anomalies of optic disc Hypoplastic optic nerve Coloboma of the optic disc Optic nerve atrophy
L		743.530	Specified anomalies of choroid
L		743.535	Coloboma of choroid Coloboma of retina
L		743.580	Other specified anomalies of posterior segment of eye
L		743.590	Unspecified anomalies of posterior segment of eye Unspecified anomalies of the retina

743.6 Congenital anomalies of eyelids, lacrimal system, and orbit

L		743.600	Blepharoptosis Congenital ptosis
L		743.610	Ectropion

Eversion/everted eyelid

L 743.620 Entropion

L * 743.630 Other anomalies of eyelids
Absence of eyelashes [1]
Long eyelashes [1]
Weakness of eyelids [1]
Fused eyelids [2]
Absent eyelid
Ankyloblepharon
Symblepharon
Epiblepharon
Ablepharon
Special instructions:
[1] *Code only if another reportable defect is present.*
[2] *Never code if the infant is less than 25 weeks gestation. If the infant is greater than or equal to 25 weeks gestation, code only if another reportable defect is present.*

L 743.635 Blepharophimosis
Short, small, narrow, or thin palpebral fissures/eye openings

L 743.636 Coloboma of the eyelids

L 743.640 Absence or agenesis of lacrimal apparatus
Absence of punctum lacrimale

L # 743.650 Stenosis or stricture of lacrimal duct
Dacryostenosis
Lacrimal duct obstruction

L 743.660 Other anomalies of lacrimal apparatus (e.g., cyst)
Dacryocystocele

L 743.670 Anomalies of orbit
Harlequin deformity of eye

743.8 Other specified anomalies of eye

L # 743.800 Other specified anomalies of eye
Includes:
Almond shaped eye
Antimongoloid slant
Brushfield spots
Bulging eye
Deep set eyes
Enophalmia
Enophthalmos
Epicanthal folds
Exophthalmos
Flat eye
Infraorbital crease
Palpebral fissure slant
Prominent eye
Proptosis
Protruding eye
Sunken eye
Upward eye slant

Excludes: Congenital nystagmus (Use 379.500)
Retinitis pigmentosa (Use 362.700)
Ocular albinism (Use 270.200)
Wide spaced eyes, hypertelorism (Use 756.085)

L 743.810 Epibulbar dermoid cyst
Excludes: Dermoid tumors or cysts in any other part of the body

743.9 Unspecified anomalies of eye

L 743.900 Unspecified anomalies of eye
Congenital: of eye (any part)
Anomaly, NOS
Deformity, NOS
Mesodermal dysgenesis eye
Congenital blindness

744 Congenital Anomalies of Ear, Face, and Neck

744.0 Anomalies of ear causing impairment of hearing

L 744.000 Absence or stricture of auditory canal
Includes: External auditory meatal stenosis
Small auditory canal

L 744.010 Anotia (absent external ear and absent ear canal)
Absence of auricle (pinna)
Absence of ear, NOS

L 744.020 Anomaly of middle ear
Fusion of ossicles
Tympanic membrane anomalies

L 744.030 Anomaly of inner ear
Includes: Congenital anomaly of membranous labyrinth
Congenital anomaly of organ of Corti

L * 744.090 Unspecified anomalies of ear with hearing impairment
Includes: Congenital deafness, NOS [1]
Special instructions:
[1] *Do not code based solely on a failed hearing screen or failed ALGO.*

744.1 Accessory auricle

L # 744.100 Accessory auricle
Polyotia

L # 744.110 Preauricular appendage, tag, or lobule (in front of ear canal)
Skin tags on face and neck
Skin tag on nose
Branchial remnant tag or growth

L # 744.120 Other appendage, tag, or lobule include papillomas, ear tags

744.2 Other specified anomalies of ear

L 744.200 Macrotia (enlarged pinna)

L		744.210	Microtia (hypoplastic pinna and absence or stricture of external auditory meatus)
L	#	744.220	Bat ear Prominent ears
L	*	744.230	Other misshapen ear Absent or decreased cartilage [2] Cauliflower ear [1] Cleft ear [1] Crumbled ears Dysplastic ears Elfin [1] Hypoplastic ear (not microtia) Lop ear [1] Malformed ear [1] Pinnae folded [1] Pinnae hypoplastic Pixie-like [1] Pointed ear [1] Prominent ear lobes [1] Redundant ears [1] Simplified ear Small ear (not microtia) *Special instructions:* [1] *Code only if another reportable defect is present.* [2] *Never code if the infant is less than 36 weeks gestation. If the infant is greater than or equal to 36 weeks gestation, code only if another reportable defect is present.*
L		744.240	Misplaced ears Anteriorly displaced ear Synotia
L	#	744.245	Low set ears
L	#	744.246	Posteriorly rotated ears Rotated ear
L		744.250	Absence or anomaly of eustachian tube
L		744.280	Other specified anomalies of ear (See also 744.230) Auricular pit Ear crease Excludes: Darwin's tubercle Preauricular pit (Use 744.410)

744.3 Unspecified anomalies of ear

L		744.300	Unspecified anomalies of ear Congenital: ear (any part) Anomaly, deformity, NOS

744.4 Branchial cleft, cyst, or fistula; preauricular sinus

L		744.400	Branchial cleft, sinus, fistula cyst, or pit Excludes: Branchial remnant tag or growth (use 744.110)
L	#	744.410	Preauricular sinus, cyst, or pit Excludes: Auricular pit (Use 744.280)
L		744.480	Other branchial cleft anomalies

Includes: Dermal sinus of head

744.5 Webbing of neck

744.500 Webbing of neck
Includes: Pterygium colli
Redundant neck skin folds
Broad neck
Thick neck
Wide neck

744.8 Other specified anomalies of face and neck

744.800 Macrostomia (large mouth)
Lateral cleft of the mouth
Lateral cleft of the lip
744.810 Microstomia (small mouth)
Small oral cavity
744.820 Macrocheilia (large lips)
744.830 Microcheilia (small lips)
Thin lip
Hypoplastic lip
Narrow vermillion borders
L * 744.880 Other specified anomalies of face/neck
Abnormal mouth shape
Absent depressor angularis
Absent eyebrow
Absent face
Arched eyebrows
Asymmetry mouth
Bowed lip
Carp shaped mouth
Downturned mouth
Facial cleft
Horner syndrome
Hypoplastic angularis oris [1]
Hypoplastic cheek
Hypoplastic obiqularis oris [1]
Lip lag [1]
Synophrys
Tented lip
Special instructions:
[1] *Code only if another reportable defect is present.*

744.9 Unspecified anomalies of face and neck

744.900 Congenital anomaly of neck, NOS
Includes: Short neck
Low posterior hairline
Low hairline (anterior or posterior not stated)
Long neck

Absent neck
Redundant chin
Excludes: Low anterior hairline (Use 744.910)

* 744.910 Congenital anomaly of face, NOS
Abnormal facies
Includes: Broad face
Down syndrome facies [1]
Dysmorphic features
Hairy/hirsute forehead
High anterior hairline
Low anterior hairline
Small face
Triangular face
Excludes: Low posterior hairline (Use 744.900)
Special instructions:
[1] *Only code when there is a possible or real diagnosis of Down syndrome.*

745 Bulbus Cordis Anomalies and Anomalies of Cardiac Septal Closure

745.0 Common truncus (See 747.200 for pseudotruncus)

* 745.000 Persistent truncus arteriosus [1]
Absent septum between aorta and pulmonary artery
Special instructions:
[1] *When coding truncus arteriosus, do not code absent pulmonary artery or inlet VSD separately. However, if a muscular VSD is present, it should be coded separately.*

745.010 Aortic septal defect
Includes: Aortopulmonary window
Excludes: Atrial septal defect (Use 745.590)

745.1 Transposition of great vessels

745.100 Transposition of great vessels, complete (no VSD)
D-transposition with no VSD

745.110 Transposition of great vessels, incomplete (with VSD)
D-transposition with a VSD
Taussig-Bing syndrome
Transposition with inlet VSD
Transposition with perimembraneous VSD
Excludes: Transposition with muscular VSD (code as 745.100 and 745.486)

745.120 Corrected transposition of great vessels,
L-transposition, ventri in version
Ventricular inversion
Excludes: Dextrocardia (Use 746.800)

745.130 Double outlet right ventricle (DORV) with normally related great vessels

745.140 Double outlet right ventricle (DORV) with transposed great vessels

745.150 Double outlet right ventricle (DORV), relationship of great vessels not specified

745.180 Other specified transposition of great vessels, no mention of double outlet right ventricle (DORV)

745.190 Unspecified transposition of great vessels

745.2 Tetralogy of Fallot

In general, if a diagnosis of Tetralogy of Fallot is made, do not list and code the following diagnoses separately:

Ventricular septal defect - 745.4
Overriding aorta - 747.260
Right ventricular outlet obstruction defects (pulmonary valve and artery) - 746.0, 747.3, or 746.995
Right ventricular hypertrophy - 746.886

Exceptions:

If the VSD is a muscular or apical VSD, code that defect separately. The included VSD is an inlet VSD (may be called a membranous, perimembranous, or canal type).

If the right ventricular outlet obstruction defect is pulmonary atresia, code that defect separately.

745.200 Fallot's tetralogy

745.3 Single ventricle

745.300 Single ventricle
Common ventricle
Cor triloculare biatriatum
Double inlet right ventricle
Double inlet left ventricle
Excludes: "functional" single ventricle

745.4 Ventricular septal defect

745.400 Roger's disease
745.410 Eisenmenger's syndrome
745.420 Gerbode defect
745.480 Other specified ventricular septal defect (VSD)
Includes: Cystalline VSD
Sub-cystalline VSD
Subarterial
Conoventricular
745.485 Perimembranous VSD
Includes: membranous VSD
745.486 Muscular VSD
Includes: mid-muscular and apical VSDs
745.487 Inlet VSD
745.490 Ventricular septal defect (VSD), NOS
Excludes: Common atrioventricular canal type (Use 745.630)

745.5 Ostium secundum type atrial septal defect

* 745.500 Nonclosure of foramen ovale, NOS [1]
Patent foramen ovale (PFO) [1]

Special instructions:

[1] *If <36 weeks gestation at delivery, code only if >12 weeks of age when the last echo that detected the PFO was performed. If ≥36 weeks gestation at delivery and the PFO was closed by ≤12 weeks of age, code only if there is another reportable defect or surgical correction. If the PFO is documented to persist >12 weeks of age, regardless of when it was diagnosed, always code. See Appendix A for a flowchart for deciding when to code PFO.*

745.510 Ostium (septum) secundum defect
Secundum ASD
Fossa ovalis atrial septal defect
Fenestrated ASD

745.520 Lutembacher's syndrome

745.580 Other specified atrial septal defect

* 745.590 ASD (atrial septal defect), NOS
Auricular septal defect, NOS
Partial foramen ovale
PFO vs. ASD [1]
PFO vs secundum ASD [1]

Special instructions:

[1] *If the defect size by echo is <= 4mm, assume it is a PFO and follow the coding instructions for 745.500 (PFO). If the defect size by echo is > 4mm, assume it is an ASD and code as 745.590 (ASD, NOS). If an echo is done, but the defect size is not stated, assume it is a PFO and follow the coding instructions for 745.500 (PFO). If unable to determine the appropriate code based on the above criteria, code as 745.590 (PFO vs. ASD).*

745.6 Endocardial cushion defects

* 745.600 Ostium primum defects
Primum ASD [1]

Special instructions:

[1] *Never code an primum ASD if there is an atrioventricular canal (AVC).*

745.610 Single common atrium, cor triloculare biventriculare

* 745.630 Common atrioventricular canal [1]
Includes: Complete AV canal defect [1]

Special instructions:

[1] *Atrioventricular canal (AVC) by definition includes a primum ASD and an inlet VSD. (Note: An inlet VSD may also be called a membranous, perimembranous, or canal type VSD). Therefore, when coding a complete AVC, it is not necessary to also code the primum ASD or the inlet VSD separately. However, if a muscular or apical VSD is present, it should be coded separately.*

745.680 Other specified cushion defect

745.690 Endocardial cushion defect, NOS

745.7 Cor biloculare

745.700 Cor biloculare

745.8 Other specified defects of septal closure

745.800 Other specified defects of septal closure

745.9 Unspecified defect of septal closure

745.900 Unspecified defect of septal closure
"Hole in the heart"

746 Other Congenital Anomalies of Heart

746.0 Anomalies of pulmonary valve

746.000 Atresia, hypoplasia of pulmonary valve
Absent pulmonary valve
See 746.995 if valve is not specified (e.g., "pulmonary atresia")

* 746.010 Stenosis of pulmonary valve [1]
Small pulmonary valve
See 746.995 if valve not specified (e.g., "pulmonary stenosis")
Excludes: Pulmonary infundibular stenosis (Use 746.830)
Special instructions:
[1] *When coding pulmonary valve stenosis, do not code dysplastic pulmonary valve leaflets separately.*

* 746.020 Pulmonary valve insufficiency or regurgitation, congenital [1]
Pulmonary insufficiency or regurgitation [1]
Special instructions:
[1] *Never code cases designated as 'mild', 'minimal', 'trivial', or 'physiologic'. Code cases designated as 'moderate' or 'severe' and those where the degree is not specified (NOS) only if another reportable heart defect is present.*

746.080 Other specified anomalies of pulmonary valve
Thickened pulmonary valve
Dysplastic pulmonary valve
Enlarged pulmonary valve
Dilated pulmonary valve
Bicuspid pulmonary valve
Redundant pulmonary valve
Dysmorphic pulmonary valve
Excludes: Pulmonary infundibular Stenosis (Use 746.830)

746.090 Unspecified anomaly of pulmonary valve

746.1 Anomalies of the tricuspid valve

746.100 Tricuspid atresia
Right atrioventricular (AV) atresia
Excludes: tricuspid stenosis and hypoplasia

* 746.105 Tricuspid valve insufficiency or regurgitation, congenital [1]
Right atrioventricular (AV) valve insufficiency or regurgitation, congenital [1]
Tricuspid valve incompetence [1]
Excludes: Ebstein's anomaly (Use 746.200)
Special instructions:
[1] *Never code cases designated as 'mild', 'minimal', 'trivial', or 'physiologic'. Code cases designated as 'moderate' or 'severe' and those where the degree is not specified*

(NOS) only if another reportable heart defect is present. Never code tricuspid valve insufficiency or regurgitation in the presence of Ebstein's anomaly.

746.106 Tricuspid stenosis or hypoplasia
Right atrioventricular (AV) stenosis or hypoplasia
Bicuspid right atrioventricular (AV) valve
Bicuspid tricuspid valve
Cleft right atrioventricular (AV) valve
Cleft tricuspid valve
Dysplastic right atrioventricular (AV) valve
Dysplastic tricuspid valve
Small right atrioventricular (AV) valve
Small tricuspid valve

746.180 Other anomalies of the tricuspid valve
Abnormal tricuspid valve
Dilated right atrioventricular (AV) valve
Dilated tricuspid valve
Enlarged right atrioventricular (AV) valve
Enlarged tricuspid valve
Redundant tricuspid valve
Right atrioventricular (AV) valve aneurysm
Thickened right atrioventricular (AV) valve
Thickened tricuspid valve
Tricuspid valve aneurysm
Tricuspid valve prolapse

746.2 Ebstein's anomaly

746.200 Ebstein's anomaly
Atrialization of right ventricle

746.3 Congenital stenosis of aortic valve

746.300 Congenital stenosis of aortic valve
Includes: Congenital aortic stenosis
Subvalvular aortic stenosis
Small aortic valve
Excludes: Supravalvular aortic stenosis (747.220)

746.4 Congenital insufficiency of aortic valve

* 746.400 Aortic valve insufficiency or regurgitation, congenital [1]
Aortic valve incompetence [1]
Excludes: bicuspid aortic valve
Special instructions:
[1] *Never code cases designated as 'mild', 'minimal', 'trivial', or 'physiologic'. Code cases designated as 'moderate' or 'severe' and those where the degree is not specified (NOS) only if another reportable heart defect is present.*

746.470 Bicuspid aortic valve
Fusion of left and right coronary cusps

746.480 Other specified anomalies of the aortic valves
Includes: Aortic valve atresia

Aortic annulus defects
Hypoplastic aortic valve
Dysplastic aortic valve
Thickened aortic valve
Absent aortic valve
Dysmorphic aortic valve
Quadricuspid aortic valve
Narrow aortic annulus

Excludes: Supravalvular aortic stenosis (747.220)

746.490 Unspecified anomalies of the aortic valves
Abnormal aortic valve

746.5 Congenital mitral stenosis

746.500 Congenital mitral stenosis
Congenital left atrioventricular (AV) stenosis
Thickened mitral valve
Thickened left atrioventricular (AV) valve

746.505 Absence, atresia, or hypoplasia of mitral valve
Abnormal mitral valve
Absence, atresia, or hypoplasia of left atrioventricular (AV) valve
Cleft left atrioventricular (AV) valve
Cleft mitral valve
Double orifice mitral valve
Dysmorphic mitral valve
Dysplastic left atrioventricular (AV) valve
Dysplastic mitral valve
Left atrioventricular (AV) valve prolapse
Mitral valve anomaly
Mitral valve prolapse
Parachute left atrioventricular (AV) valve
Parachute mitral valve

746.6 Mitral valve insufficiency or regurgitation, congenital

* 746.600 Mitral valve insufficiency or regurgitation, congenital [1]
Left atrioventricular (AV) valve insufficiency or regurgitation, congenital [1]

Special instructions:

[1] *Never code cases designated as 'mild', 'minimal', 'trivial', or 'physiologic'. Code cases designated as 'moderate' or 'severe' and those where the degree is not specified (NOS) only if another reportable heart defect is present.*

746.7 Hypoplastic left heart syndrome

In general, if a diagnosis of hypoplastic left heart syndrome is made, do not list and code the following diagnoses separately:

Hypoplastic left ventricle - 746.881
Mitral valve anomalies - 746.5 and 746.6
Aortic valve anomalies - 746.3 and 746.4
Atresia or hypoplasia of the ascending aorta - 747.210

However, if the mitral valve or the aortic valve is absent/atretic, code that defect separately.

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

746.8 Other specified anomalies of the heart

746.800 Dextrocardia without situs inversus (situs solitus)
Dextrocardia with no mention of situs inversus
Excludes: Dextrocardia with situs inversus (Use 759.300)
Dextrocardia with left congenital diaphragmatic hernia

746.820 Cor triatriatum

746.830 Pulmonary infundibular (subvalvular) stenosis

746.840 Trilogy of Fallot

746.850 Anomalies of pericardium

* 746.860 Anomalies of myocardium
Cardiomegaly, congenital, NOS [1]
Cardiomyopathy, congenital
Cardiomyopathy, hypertrophic
Double chamber right ventricle
Left ventricular aneurysm
Rhabdomyoma (heart)
Ventricular hypertrophy, bilateral [2]
Ventricular septal thickening
Ventricular septal hypertrophy
Special instructions:
[1] *Code only if another reportable defect is present. Never code if the diagnosis is based solely on an electrocardiogram (EKG, ECG) and/or an X-ray.*
[2] *Never code if the diagnosis is based solely on an electrocardiogram (EKG, ECG).*
Excludes: Ventricular hypertrophy, unilateral (Use 746.886)

746.870 Congenital heart block

746.880 Other specified anomalies of heart
Includes: Ectopia (ectopic) cordis (mesocardia), conduction defects, NOS
Shone's complex
Tumor of the heart
Hypoplastic heart
Long Q-T syndrome
Left ventricular outflow tract obstruction - NOS
Right ventricular outflow tract obstruction – 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
Dilated coronary sinus [1]
Single coronary artery
Special instructions:
[1] *Only code when there is no indication of a left superior vena cava (LSVC).*
Excludes: Dilated coronary sinus when there is a left superior vena cava

L * 746.886 Ventricular hypertrophy, unilateral (right or left) [1]
Special instructions:
[1] *Code only if another reportable defect is present. Never code if the diagnosis is based solely on an electrocardiogram (EKG, ECG).*
Excludes: Ventricular hypertrophy, bilateral (Use 746.860)

746.887 Other defects of the atria
Hypoplastic atrium
Excludes: Congenital Wolfe-Parkinson-White (Use 426.705)
Rhythm anomalies (Use 426.-, 427.-)

746.9 Unspecified anomalies of heart

746.900 Unspecified anomalies of heart valves
Truncal valve
Truncal valve insufficiency and regurgitation
Truncal valve stenosis
Narrow truncal valve
Single atrioventricular valve
Single atrioventricular valve insufficiency and regurgitation
Atrioventricular valve regurgitation, laterality unspecified

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)

747 Other Congenital Anomalies of Circulatory System

747.0 Patent ductus arteriosus

* 747.000 Patent ductus arteriosus (PDA) [1]
Special instructions:
[1] *Never code if the infant was less than 36 weeks gestation and less than 12 weeks of age at diagnosis, or if the infant was on prostaglandin. If the infant was greater than or equal to 36 weeks gestation and less than 12 weeks of age at diagnosis, code only if there is another reportable defect present, or if there was a medical/surgical intervention for this problem. Always code if greater than or equal to 12 weeks of age at diagnosis, unless the infant was on prostaglandin. See Appendix B for a flowchart for deciding when to code PDA.*

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
Juxtaductal coarctation of aorta
Long-segment coarctation of aorta

Preductal and postductal coarctation of aorta (in the same child)

747.2 Other anomalies of aorta

747.200 Atresia of aorta
Absence of aorta
Atrophy of aorta
Pseudotruncus arteriosus
Stenosis of aorta

747.210 Hypoplasia of aorta
Tubular hypoplasia of aorta
Small aorta
Narrowing of aorta
Proximal distal transverse arch hypoplasia
Narrow aortic isthmus
Hypoplastic aortic arch

747.215 Interrupted aortic arch, Type A

747.216 Interrupted aortic arch, Type B

747.217 Interrupted aortic arch, Type C

747.220 Supra-aortic stenosis (supravalvular)
Excludes: Aortic valve stenosis, congenital (See 746.300)

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
Malaligned aorta

747.270 Congenital aneurysm of aorta
Congenital dilatation of aorta
Enlarged aorta

* 747.280 Other specified anomalies of aorta
Collateral vessel involving aorta [1]
Pseudocoarctation of aorta
Elongation of aorta
Aortopulmonary collateral vessel [1]
Special instructions:
[1] *Code only if another reportable defect is present.*

747.285 Interrupted aortic arch, NOS, type not specified

747.290 Unspecified anomalies of aorta

747.3 Anomalies of pulmonary artery

If the defect is in the main pulmonary artery, laterality does not apply. If the same defect exists in more than one part of the pulmonary artery (e.g., in the main pulmonary artery and in the left pulmonary artery, or in both the left and right pulmonary arteries), list and code those defects separately.

L 747.300 Pulmonary artery atresia, absence, or agenesis
Use 746.995 if artery or valve is <u>not</u> specified

L		747.310	Pulmonary artery atresia with septal defect
L		747.320	Pulmonary artery stenosis Supravalvular pulmonary stenosis Pulmonary artery narrowing Use 746.995 if artery or valve is not specified
L	*	747.325	Peripheral pulmonary artery stenosis [1] Includes: peripheral pulmonic stenosis (PPS) [1] pulmonary artery branch stenosis [1] peripheral pulmonic stenosis (PPS) murmur [2] *Special instructions:* [1] *Never code if the infant was less than 36 weeks gestation and less than 12 weeks of age at diagnosis. If the infant was greater than or equal to 36 weeks gestation and less than 12 weeks of age at diagnosis, code only if there is another reportable defect present, or if there was a medical/surgical intervention for this problem. Always code if greater than or equal to 12 weeks of age at diagnosis. See Appendix C for a flowchart for deciding when to code PPS.* [2] *Code only if documented by echocardiogram.*
		747.330	Aneurysm of pulmonary artery Dilatation of pulmonary artery Enlarged pulmonary artery
L		747.340	Pulmonary arteriovenous malformation or aneurysm
L	*	747.380	Other specified anomaly of pulmonary artery Includes: Pulmonary artery hypoplasia Small pulmonary artery Overriding pulmonary artery Collateral vessel involving pulmonary artery (and not aorta) [1] Pulmonary vascular or artery sling *Special instructions:* [1] *Code only if another reportable defect is present.*
L		747.390	Unspecified anomaly of pulmonary artery

747.4 Anomalies of great veins

	747.400	Stenosis of vena cava (inferior or superior) Small vena cava (inferior or superior)
	747.410	Persistent left superior vena cava Bilateral superior vena cava
	747.420	(TAPVR) Total anomalous pulmonary venous return
	747.430	Partial anomalous pulmonary venous return
	747.440	Anomalous portal vein termination
	747.450	Portal vein - hepatic artery fistula
	747.480	Other specified anomalies of great veins Enlarged vena cava (inferior or superior) Pulmonary vein atresia Pulmonary vein stenosis Dilated vena cava (inferior or superior) Absent vena cava (inferior or superior) Interrupted inferior vena cava Bilateral inferior vena cava Anomalous pulmonary venous return – total/partial not specified

Small pulmonary veins
Excludes: Absent left superior vena cava (LSVC) which would be normal

747.490 Unspecified anomalies of great veins

747.5 Absence or hypoplasia of umbilical artery

747.500 Single umbilical artery
Two-vessel cord
Umbilical artery hypoplasia

747.6 Other anomalies of peripheral vascular system

L 747.600 Stenosis of renal artery

L 747.610 Other anomalies of renal artery
Absent renal artery

L 747.620 Arteriovenous malformation (peripheral)
Excludes: Pulmonary (747.340)
Cerebral (747.800)
Retinal (743.510)

L 747.630 Congenital phlebectasia
Congenital varix

L 747.640 Other anomalies of peripheral arteries
Includes: Aberrant subclavian artery
Common brachiocephalic trunk
Aberrant innominate artery
Absent carotid artery
Anomalous carotid artery
Retroesophageal subclavian artery

L 747.650 Other anomalies of peripheral veins
Hemiazygos vein anomalies
Hypoplastic innominate vein
Hypoplastic jugular vein
Hepatic vein stenosis

L 747.680 Other anomalies of peripheral vascular system
Includes: Four vessel umbilical cord

L 747.690 Unspecified anomalies of peripheral vascular system

747.8 Other specified anomalies of circulatory system

L 747.800 Arteriovenous (malformation) aneurysm of brain

L 747.810 Other anomalies of cerebral vessels
Includes: Anomalies of Vein of Galen

L * 747.880 Other specified anomalies of circulatory system
Endothelial vessel
Collateral vessel (not involving aorta or pulmonary artery) [1]
Excludes: Congenital aneurysm:
Coronary (746.880)
Peripheral (747.640)
Pulmonary (747.330)
Retinal (743.510)
Ruptured cerebral arteriovenous

Aneurysm
Ruptured cerebral aneurysm

Special instructions:

[1] *Code only if another reportable defect is present.*

747.9 Unspecified anomalies of circulatory system

747.900 Unspecified anomalies of circulatory system

748 Congenital Anomalies of Respiratory System

748.0 Choanal atresia

L 748.000 Choanal atresia
Atresia of nares, anterior or posterior congenital stenosis
Piriform aperature atresia and stenosis

748.1 Other anomalies of nose

748.100 Agenesis or underdevelopment of nose
Absent nose
Hypoplastic nose
Absent nares

748.110 Accessory nose

748.120 Fissured, notched, or cleft nose
Bifid nose
Excludes: Nose deformed because of cleft lip

748.130 Sinus wall anomalies

748.140 Perforated nasal septum

748.180 Other specified anomalies of nose
Flat bridge of nose
Wide nasal bridge
Small nose and nostril
Absent nasal septum
Asymmetry nose
Small nares
Hypoplasia of nasal bridge
Prominent glabella

748.185 Tubular nose, single nostril, proboscis

748.190 Unspecified anomalies of nose
Excludes: Congenital deviation of the nasal septum (Use 754.020)

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
Laryngeal stenosis
Hypoplastic larynx
Epiglottis anomalies
Hypoplastic epiglottis

* 748.310 Congenital subglottic stenosis [1]
Special instructions:
[1] *Never code if the chart states that the condition was acquired secondary to endotracheal (ET) intubation or ventilation.*

748.330 Other anomalies of trachea
Tracheal atresia
Tracheal stenosis
Small trachea
Complete tracheal rings
Excludes: Vascular ring compression of the trachea (Use 747.250)

L 748.340 Stenosis of bronchus

L 748.350 Other anomalies of bronchus
Absent bronchus
Bronchopulmonary fistula
Bronchogenic cyst
Narrow distal airway

* 748.360 Congenital laryngeal stridor, NOS [1]
Special instructions:
[1] *Do not code if stridor is secondary to another known cause.*

L 748.380 Other specified anomalies of larynx and bronchus

748.385 Cleft larynx, laryngotracheoesophageal cleft
Laryngeal cleft

748.390 Unspecified anomalies of larynx, trachea, and bronchus

748.4 Congenital cystic lung

L 748.400 Single cyst, lung or lung cyst

L 748.410 Multiple cysts, lung
Polycystic lung

L 748.420 Honeycomb lung

L 748.480 Other specified congenital cystic lung
Cystic adenomatoid malformation lung

748.5 Agenesis or aplasia of lung

L 748.500 Agenesis or aplasia of lung
Lung atresia

L * 748.510 Hypoplasia of lung; Pulmonary hypoplasia [1]
Special instructions:
[1] *If the infant is less than 36 weeks gestation, code only if another reportable defect is present. Always code if greater than or equal to 36 weeks gestation.*
Small lung

L 748.520 Sequestration of lung

L 748.580 Other specified dysplasia of lung

Fusion of lobes of lung
Incomplete separation of lung lobes
One lobe lung

L 748.590 Unspecified dysplasia of lung

748.6 Other anomalies of lung

L 748.600 Ectopic tissues in lung
L 748.610 Bronchiectasis
L 748.620 Accessory lobe of lung
Three or more lobes of left lung
Four or more lobes of right lung
748.625 Bilobar right lung or right lung with left lung bronchial pattern
L 748.690 Other and unspecified anomalies of lung
Exstrophy lung
Scimitar Syndrome

748.8 Other specified anomalies of respiratory system

L 748.800 Anomaly of pleura
748.810 Congenital cyst of mediastinum
L 748.880 Other specified respiratory system anomalies
Includes: Congenital lobar emphysema
Lymphangiectasia of lungs
Lung mass, NOS

748.9 Unspecified anomalies of respiratory system

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

L 749.000 Cleft hard palate
Cleft anterior palate
749.020 Cleft hard palate, central
Cleft anterior palate, central
Submucosal cleft palate (hard/anterior palate)
Midline cleft palate extending to posterior hard palate
749.030 Absent hard palate
Absent anterior palate
L 749.040 Cleft soft palate
Cleft posterior palate
749.060 Cleft soft palate, central
Cleft posterior palate, central
Submucosal cleft palate (soft/posterior palate)
749.070 Absent soft palate

Absent posterior palate

* 749.080 Cleft uvula
Bifid uvula [1]
Absent uvula
Special instructions:
[1] *Never code bifid uvula in the presence of a cleft palate.*

749.090 Cleft palate, NOS (hard/soft not specified)
Palatoschisis
Includes: Cleft soft and hard palate
Submucosal cleft palate (hard/soft not specified)
Absent palate (hard/soft not specified)

749.1 Cleft lip alone

L 749.100 Cleft lip
Alveolar ridge cleft
Cleft gingiva
Cleft gum
Harelip

749.120 Cleft lip, central
Alveolar ridge cleft, central
Anterior midline cleft
Cleft gingival, central
Cleft gum, central
Harelip, central

749.2 Cleft lip with cleft palate

L 749.200 Cleft lip with any cleft palate

749.220 Cleft lip, central, with any cleft palate

750 Other Congenital Anomalies of Upper Alimentary Tract

750.0 Tongue tie

\# 750.000 Tongue tie
Ankyloglossia
Anterior tongue/lingual frenulum
Short tongue/lingual frenulum
Excludes: Thickening or shortening of labial (lip) frenulum

750.1 Other anomalies of tongue

750.100 Aglossia
Absence of tongue

750.110 Hypoglossia (small tongue)
Microglossia

750.120 Macroglossia (large tongue)
Thick 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

L 750.200 Pharyngeal pouch
L 750.210 Other pharyngeal anomalies
L 750.230 Other anomalies of salivary glands or ducts
\# 750.240 High arched palate
750.250 Other anomalies of palate
Small palate
Narrow palate
Ridge shaped palate
Grooved palate
750.260 Lip fistulae or pits
* 750.270 Other lip anomalies
Includes: Anomalies of frenulum of upper lip [1]
Flat philtrum [1]
Lip tie [1]
Long philtrum [1]
Multiple lip frenula
Notched lip [1]
Prominent philtrum [1]
Short lip frenulum [1]
Smooth lip [1]
Smooth philtrum [1]
Tight lip frenulum [1]
Excludes: Cleft lip (See 749)
Special instructions:
[1] *Code only if another reportable defect is present.*
750.280 Other specified anomalies of mouth and pharynx
Hypertrophy gum
Prominent gum
Saw-tooth gums
Thick lateral palatine ridges
Other gum anomalies
Includes: Abnormalities of the gum or alveolar ridge (except cleft)
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 [1]
Special instructions:
[1] *When coding esophageal atresia with T-E fistula, do not code an esophageal pouch separately.*
750.320 Tracheoesophageal fistula without mention of esophageal atresia

750.325 Tracheoesophageal 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

750.4 Other specified anomalies of esophagus

750.400 Congenital dilatation of esophagus
Giant esophagus
750.410 Displacement of esophagus
750.420 Diverticulum of esophagus
Esophageal pouch
750.430 Duplication of esophagus
750.480 Other specified anomalies of esophagus
Esophageal fistula

750.5 Congenital hypertrophic pyloric stenosis

750.500 Pylorospasm
750.510 Congenital hypertrophic pyloric stenosis
750.580 Other congenital pyloric obstruction
Antral web
Excludes: Congenital hiatal hernia – use code 750.600

750.6 Congenital hiatus hernia

750.600 Congenital hiatus hernia
Cardia displacement through esophageal hiatus
Paraesophageal hernia
Partial thoracic stomach
Excludes: Congenital diaphragmatic hernia (756.610)

750.7 Other specified anomalies of stomach

750.700 Microgastria
Small stomach
750.710 Megalogastria
750.720 Cardiospasm
Achalasia of cardia, congenital
750.730 Displacement or transposition of stomach
Right-sided stomach
Includes: Transposition of the stomach as part of heterotaxy syndrome
750.740 Diverticulum of stomach
750.750 Duplication of stomach
750.780 Other specified anomalies of stomach
Absent stomach

750.8 Other specified anomalies of upper alimentary tract

750.800 Other specified anomalies of upper alimentary tract

750.9 Unspecified anomalies of upper alimentary tract

750.900 Unspecified anomalies of mouth and pharynx
750.910 Unspecified anomalies of esophagus
750.920 Unspecified anomalies of stomach
750.990 Unspecified anomalies of upper alimentary tract

751 Other Congenital Anomalies of Digestive System

751.0 Meckel's diverticulum

751.000 Persistent omphalomesenteric duct
Persistent vitelline duct
Omphalomesenteric remnant
\# 751.010 Meckel's diverticulum

751.1 Atresia and stenosis of small intestine

751.100 Stenosis, atresia, or absence of duodenum
Pyloric atresia
751.110 Stenosis, atresia, or absence of jejunum
751.120 Stenosis, atresia, or absence of ileum
751.190 Stenosis, atresia, or absence of small intestine
Short small intestine
751.195 Stenosis, atresia, or absence of small intestine with fistula

751.2 Atresia and stenosis of large intestine, rectum, and anal canal

751.200 Stenosis, atresia, or absence of large intestine
Stenosis, atresia, or absence of appendix
Colon atresia
Colon stenosis
Absent colon
Sigmoid web
751.210 Stenosis, atresia, or absence of rectum with fistula
Note: This code covers rectoperineal fistulas. Other fistulas should be coded separately.
Rectourethral - 753.860
Rectovaginal - 752.420
Rectovesical - 753.860
751.220 Stenosis, atresia, or absence of rectum without mention of fistula
Small rectum
Short rectum
Anorectal stenosis
751.230 Stenosis, atresia, or absence of anus with fistula
Includes: Imperforate anus with fistula
Anal dysgenesis with fistula
Note: This code covers anoperineal fistulas. Other fistulas should be coded separately.
Anourethral - 753.860

Anovaginal - 752.420
Anovesical - 753.860

751.240 Stenosis, atresia, or absence of anus without mention of fistula
Includes: Imperforate anus without fistula
Anal dysgenesis without fistula

751.3 Hirschsprung's disease and other congenital functional disorders of the colon

751.300 Total intestinal aganglionosis

751.310 Long-segment Hirschsprung's disease; aganglionosis beyond the rectum

751.320 Short-segment Hirschsprung's disease; aganglionosis involving no more than the anal sphincter and the rectum

* 751.330 Hirschsprung's disease, NOS [1]
Special instructions:
[1] *When coding Hirschsprung's disease, do not code rectal stenosis separately.*

751.340 Congenital megacolon
Congenital macrocolon, not aganglionic

751.4 Anomalies of intestinal fixation

751.400 Malrotation of cecum and/or colon
Malrotation large bowel
Malrotation large intestine

751.410 Anomalies of mesentery

751.420 Congenital adhesions or bands of omentum and peritoneum; Ladd's bands

751.490 Other specified and unspecified malrotation
Malrotation bowel (large/small not specified)
Midgut malrotation

751.495 Malrotation of small intestine alone
Malrotation small bowel

751.5 Other anomalies of intestine

751.500 Duplication of anus, appendix, cecum, or intestine
Enterogenous cyst
Duplicated pylorus

751.510 Transposition of appendix, colon, or intestine

751.520 Microcolon
Small colon
Hypoplastic colon
Short colon

751.530 Ectopic (displaced) anus
Anteriorly placed anus

751.540 Congenital anal fistula
Rectal fistula

751.550 Persistent cloaca

751.555 Cloacal exstrophy

751.560 Duodenal web

* 751.580 Other specified anomalies of intestine
Includes: Rectal fissures [1]
Jejunal web

Special instructions:
[1] *Code only if another reportable defect is present.*

751.590 Unspecified anomalies of intestine

751.6 Anomalies of gallbladder, bile ducts, and liver

751.600 Absence or agenesis of liver, total or partial

751.610 Cystic or fibrocystic disease of liver

\# 751.620 Other anomalies of liver
Hepatomegaly
Hepatosplenomegaly (Also use code 759.020)
Transverse liver
Enlarged liver
Left-sided liver
Includes: Displacement of the liver as part of heterotaxy syndrome

* 751.630 Agenesis or hypoplasia of gallbladder [1]
Special instructions:
[1] *Never code absent gallbladder in the presence of biliary atresia.*

751.640 Other anomalies of gallbladder
Duplication of gallbladder

751.650 Agenesis or atresia of hepatic or bile ducts
Includes: Biliary atresia
Excludes: Congenital or neonatal hepatitis (Use 774.480 or 774.490)

751.660 Choledochal cysts

751.670 Other anomalies of hepatic or bile ducts
Biliary dysgenesis

751.680 Anomalies of biliary tract, NEC

751.7 Anomalies of pancreas

Excludes: Fibrocystic disease of pancreas (277.000)
Diabetes mellitus, congenital or neonatal

751.700 Absence, agenesis or hypoplasia of pancreas
Small pancreas

751.710 Accessory pancreas

751.720 Annular pancreas

751.730 Ectopic pancreas

751.740 Pancreatic cyst

751.780 Other specified anomalies of pancreas
Multicystic pancreas
Pancreas divisum
Heterotopia pancreas

751.790 Unspecified anomalies of pancreas

751.8 Other specified anomalies of digestive system

751.800 Absence of alimentary tract, NOS (complete or partial)

751.810 Duplication of alimentary tract

751.820 Ectopic digestive organs, NOS

751.880 Other specified anomalies of digestive system

751.9 Unspecified anomalies of digestive system

751.900 Unspecified anomalies of digestive system
Congenital of digestive system, NOS
Anomaly, NOS
Deformity, NOS

752 Congenital Anomalies of Genital Organs

Excludes: Congenital hydrocele (778.600)
Testicular feminization syndrome (257.800)
Syndromes associated with anomalies in number and form of chromosomes (758)

752.0 Anomalies of ovaries

L 752.000 Absence or agenesis of ovaries
L 752.010 Streak ovary
L 752.020 Accessory ovary
L 752.080 Other specified anomalies of ovaries
Single ovarian cyst
Torsion of ovary
Hypoplastic ovary
L 752.085 Multiple ovarian cysts
L 752.090 Unspecified anomalies of ovaries

752.1 Anomalies of fallopian tubes and broad ligaments

L 752.100 Absence of fallopian tube or broad ligament
L 752.110 Cyst of mesenteric remnant
Epoophoron cyst
Cyst of Gartner's duct
L 752.120 Fimbrial cyst
Parovarian cyst
L 752.190 Other and unspecified anomalies of fallopian tube and broad ligaments
Hypoplastic fallopian tube

752.2 Doubling of uterus

752.200 Doubling of uterus
Doubling of uterus (any degree) or associated with doubling of cervix and vagina
Uterine didelphys

752.3 Other anomalies of uterus

752.300 Absence or agenesis of uterus
752.310 Displaced uterus
752.320 Fistulae involving uterus with digestive or urinary tract
Includes: Uterointestinal fistula
Uterovesical fistula
L 752.380 Other anomalies of uterus
Bicornate uterus

Unicornis uterus
Septate uterus
Small uterus

752.390 Unspecified anomalies of uterus

752.4 Anomalies of cervix, vagina, and external female genitalia

752.400 Absence, atresia, or agenesis of cervix

752.410 Absence or atresia of vagina, complete or partial
Small vagina
Short vagina

752.420 Congenital rectovaginal fistula
Anovaginal fistula
Vesicovaginal fistula

* 752.430 Imperforate hymen [1, 2]
Hydrocolpos [1]
Hydrometrocolpos
Special instructions:
[1] *Code only if another reportable defect is present.*
[2] *Never code imperforate hymen if absence of the vagina is coded.*

* 752.440 Absence or other anomaly of vulva
Fusion of vulva [1]
Hypoplastic labia majora [2]
Large labia (majora or minora) [2]
Prominent labia (majora or minora) [2]
Absent external female genitalia
Special instructions:
[1] *Code only if another reportable defect is present.*
[2] *Never code if the infant is less than 36 weeks gestation. If the infant is greater than or equal to 36 weeks gestation, code only if another reportable defect is present.*

* 752.450 Absence or other anomaly of clitoris
Includes: Clitoromegaly [1]
Enlarged clitoris [2]
Clitoral hypertrophy [2]
Prominent clitoris [2]
Special instructions:
[1] *Never code if the infant is less than 36 weeks gestation. If the infant is greater than or equal to 36 weeks gestation, code only if another reportable defect is present.*
[2] *Code only if another reportable defect is present.*

\# 752.460 Embryonal cyst of vagina

752.470 Other cyst of vagina, vulva, or canal of Nuck

* 752.480 Other specified anomalies of cervix, vagina, or external female genitalia
Includes: Vaginal tags [1]
Hymenal tags [1]
Fourchette fistula [1]
Doubling of cervix
Doubling of vagina
Vaginocele
Special instructions:
[1] *Code only if another reportable defect is present.*

752.490 Unspecified anomalies of cervix, vagina, or external female genitalia

752.5 Undescended testicle

Excludes: Retractile testicle

L * 752.500 Undescended testicle, unilateral [1]
Includes: Cryptorchidism, unilateral [1]
Unpalpable testicle, unilateral [1]
Special instructions:
[1] *Do not code in infants less than 36 weeks gestation regardless of the presence of other defects, unless there was a medical/surgical intervention for this problem. If the infant was greater than or equal to 36 weeks gestation, code only if there is another reportable defect present, or if there was a medical/surgical intervention for this problem.*

* 752.514 Undescended testicle, bilateral [1]
Includes: Cryptorchidism, bilateral [1]
Unpalpable testicle, bilateral [1]
Special instructions:
[1] *Do not code in infants less than 36 weeks gestation unless another reportable defect is present or there was a medical/surgical intervention for this problem. Always code if greater than or equal to 36 weeks gestation.*

* 752.520 Undescended testicle, NOS [1]
Includes: Cryptorchidism, NOS [1]
Unpalpable testicle, NOS [1]
Special instructions:
[1] *Do not code in infants less than 36 weeks gestation unless another reportable defect is present or there was a medical/surgical intervention for this problem. Always code if greater than or equal to 36 weeks gestation.*

L 752.530 Ectopic testis
Intra-abdomen testes

752.6 Hypospadias and epispadias

752.600 Hypospadias (alone), NOS
752.605 1º, glandular,coronal
Balantic hypospadias
Distal hypospadias
Mild hypospadias
Primary hypospadias
Subcoronal hypospadias
Tip hypospadias
752.606 2º, penile
Mid-shaft hypospadias
Grade II hypospadias
Distal shaft hypospadias
752.607 3º, perineal, scrotal
752.610 Epispadias
752.620 Congenital chordee (with hypospadias), NOS
752.621 Congenital chordee alone (chordee w/o hypospadias)
Webbed penis
752.625 Cong. chordee with 1º, coronal hypospadias

Balantic hypospadias with chordee
Subcoronal hypospadias with chordee

752.626 Cong. chordee with 2º, penile hypospadias

752.627 Cong. chordee with 3º, 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 (758.690)

752.730 Pseudohermaphrodite, NOS

* 752.790 Indeterminate sex, NOS
Ambiguous genitalia [1]
Special instructions:
[1] *Coding of "ambiguous genitalia" should only be used when no further description is available. Do not code ambiguous genitalia if chromosomes are given.*
Absent genitalia (sex unknown)

752.8 Other specified anomalies of male genital organs

L 752.800 Absence of testis
Monorchidism, NOS
Vanishing testicle
Testicle regression

L * 752.810 Aplasia of testis and scrotum
Hypoplasia of testis and scrotum [1]
Small scrotum [1, 2]
Small testicle [1]
Atrophy testicle
Underdeveloped/undeveloped scrotum
Special instructions:
[1] *Code only if another reportable defect is present.*
[2] *Never code small scrotum in the presence of undescended testicles.*

752.820 Other anomalies of testis and scrotum
Polyorchidism
Bifid scrotum
Large testicle
Excludes: Torsion of the testes or spermatic cord (Use 608.200)

L 752.830 Atresia of vas deferens

752.840 Other anomalies of vas deferens and prostate

752.850 Absence or aplasia of penis

* 752.860 Other anomalies of penis
Absent or hooded foreskin [1]
Buried penis

Concealed penis
Disappearing penis syndrome
Penile torsion
Penis palmae
Penoscrotal web
Rotation of the penis
Twisted penis
Special instructions:
[1] *Never code foreskin anomalies in the presence of hypospadias.*

752.865 Small penis, hypoplastic penis, or micropenis

L 752.870 Cysts of embryonic remnants
Cyst: hydatid of Morgagni
Wolffian duct
Appendix testis

752.880 Other specified anomalies of genital organs
Microgenitalia
Macrogenitalia
Penoscrotal transposition
Large penis
Penoscrotal fusion

752.9 Unspecified anomalies of genital organs

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
Bilateral small kidneys
Potter's syndrome
Potter’s sequence

753.009 Renal agenesis, NOS
Renal dysplasia, NOS

L 753.010 Unilateral absence, agenesis, dysplasia, or hypoplasia of kidney
Unilateral small kidney

753.1 Cystic kidney disease

L 753.100 Renal cyst (single)

753.110 Polycystic kidneys, infantile type
Autosomal recessive polycystic kidney disease (ARPKD)

753.120 Polycystic kidneys, adult type
Autosomal dominant polycystic kidney disease (ADPKD)

753.130 Polycystic kidneys, NOS

753.140 Medullary cystic disease, juvenile type

753.150 Medullary cystic disease, adult type

Medullary sponge kidney

L 753.160 Multicystic renal dysplasia
Multicystic kidney
Cystic dysplasia kidney
Multicystic dysplastic kidney

L 753.180 Other specified cystic disease
Includes: Cystic kidneys, NOS

753.2 Obstructive defects of renal pelvis and ureter

L 753.200 Congenital hydronephrosis
Hydroureteronephrosis (Also assign code 753.220)
Lower hydronephrosis
Upper hydronephrosis

L 753.210 Atresia, stricture, or stenosis of ureter
Includes: Ureteropelvic junction obstruction/stenosis
Ureterovesical junction obstruction/stenosis
Hypoplastic ureter

L 753.220 Megaloureter, NOS
Includes: Hydroureter
Hydroureteronephrosis (Also assign code 753.200)
Dilated ureter
Ureterectasis

L * 753.290 Other and unspecified obstructive defects of renal pelvis and ureter
Obstructive uropathy
Pyelocaliectasis - NOS
Dilated renal pelvis - NOS [1]
Pelvicaliectasis - NOS
Pelviectasis
Pyelectasis
Dilation of upper renal collecting system
Dilation of lower renal collecting system
Dilation of central renal collecting system [1]
Special instructions:
[1] *Never code if described as 'mild' or 'transient'.*

753.3 Other specified anomalies of kidney

L 753.300 Accessory kidney

L 753.310 Double or triple kidney and pelvis
Pyelon duplex or triplex

* 753.320 Lobulated, fused, or horseshoe kidney [1]
Cross fused renal ectopia
Special instructions:
[1] *When coding horseshoe kidney, do not code ectopic kidney separately.*

L 753.330 Ectopic kidney
Malrotated kidney
Pelvic kidney

L 753.340 Enlarged, hyperplastic, or giant kidney
Renomegaly
Hypertrophy kidney

L 753.350 Congenital renal calculi
L * 753.380 Other specified anomalies of kidney
Includes: Extra renal pelvis
Prominent renal pelvis - NOS [1]
Enlarged renal pelvis - NOS [1]
Congenital nephrotic syndrome
Excludes: Dilation of upper/central/lower renal collecting system (use 753.290)
Special instructions:
[1] *Never code if described as 'mild' or 'transient'.*

753.4 Other specified anomalies of ureter

L 753.400 Absence of ureter
L 753.410 Accessory ureter
Double ureter, duplex collecting system
L 753.420 Ectopic ureter
L 753.480 Other specified anomalies of ureter
Includes: Ureterocele
Short ureter
Excludes: Dilation of central renal collecting system (use 753.290)
L 753.485 Variations of vesicoureteral reflux

753.5 Exstrophy of urinary bladder

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
753.620 Obstruction, atresia, or stenosis of anterior urethra
Anterior urethral valve
753.630 Obstruction, atresia, or stenosis of urinary meatus
Includes: Meatal stenosis
Small opening of penis
753.690 Other and unspecified atresia and stenosis of urethra and bladder neck
Urethral stricture
Bladder outlet obstruction, NOS

753.7 Anomalies of urachus

753.700 Patent urachus
Urachal sinus
753.710 Cyst of urachus
753.790 Other and unspecified anomaly of urachus
Urachal remnant

753.8 Other specified anomalies of bladder and urethra

753.800 Absence of bladder or urethra

753.810 Ectopic bladder
753.820 Congenital diverticulum or hernia of bladder
Cystocele bladder
Hutch diverticulum
753.830 Congenital prolapse of bladder (mucosa)
753.840 Double urethra or urinary meatus
753.850 Ectopic urethra or urethral orifice
753.860 Congenital digestive-urinary tract fistulae
Rectourethral fistula
Rectovesical fistula
Urethrorectal fistula
753.870 Urethral fistula, NOS
753.880 Other specified anomalies of bladder and urethra
Duplicated bladder
Hypoplasia bladder
Hypoplastic bladder
Megameatus
Megaurethra
Septation of bladder
Urethral diverticulum
Urogenital sinus malformation

753.9 Unspecified anomalies of urinary system

L 753.900 Unspecified anomaly of kidney
L 753.910 Unspecified anomaly of ureter
753.920 Unspecified anomaly of bladder
753.930 Unspecified anomaly of urethra
753.990 Unspecified anomaly of urinary system, NOS

754 Certain Congenital Musculoskeletal Anomalies

754.0 Of skull, face, and jaw

Excludes: Dentofacial anomalies (524.0)
Pierre Robin sequence (524.080)
Syphilitic saddle nose (090.000)

754.000 Asymmetry of face
754.010 Compression (Potter's) facies
Excludes: Potter's syndrome (See 753.000)
754.020 Congenital deviation of nasal septum
Bent nose
* 754.030 Dolichocephaly [1]
Elongated head and skull [1]
Special instructions:
[1] *Never code in the presence of craniosynostosis. Never code if the infant is less than 36 weeks gestation. If the infant is greater than or equal to 36 weeks gestation, code only if another reportable defect is present.*

754.040 Depressions in skull
Includes: Large fontanelle
Small fontanelle
Three fontanelles
Absent fontanelle

L * 754.050 Plagiocephaly [1]
Flattened side of head
Special instructions:
[1] *Code only if another reportable defect is present. Never code in the presence of lambdoidal or coronal craniosynostosis. Plagiocephaly is unilateral only. 'Bilateral plagiocephaly' should be coded as brachycephaly (use 754.080). When coding plagiocephaly, do not code asymmetric head separately. See Appendix D for more information on coding plagiocephaly and related defects.*

754.055 Asymmetric head
Asymmetric skull
Asymmetry calvarium
Cranial scoliosis
Cranioscoliosis

* 754.060 Scaphocephaly, no mention of craniosynostosis (fused sutures) [1]
Special instructions:
[1] *Never code if the infant is less than 36 weeks gestation. If the infant is greater than or equal to 36 weeks gestation, code only if another reportable defect is present.*

754.070 Trigonocephaly, no mention of craniosynostosis (fused sutures)
Triangular head shape

754.080 Other specified skull deformity, no mention of craniosynostosis (fused sutures)
Includes: Brachycephaly
Acrocephaly
Turricephaly
Oxycephaly
Frontal bossing
Square head
Square cranium
Cone shaped head
Flat head
Tower skull
Tower head
Platycephaly
Box shaped head

754.090 Deformity of skull, NOS
Misshapen head
Misshapen skull
Abnormal head shape NOS

754.1 Anomalies of sternocleidomastoid muscle

L 754.100 Anomalies of sternocleidomastoid muscle
Includes: Absent or hypoplastic sternocleidomastoid
Contracture of sternocleidomastoid muscle
Sternomastoid tumor
Fibromatosis colli

Excludes: Congenital sternocleidomastoid torticollis (Use 756.860)

754.2 Certain congenital musculoskeletal deformities of spine

Note: Do not use these codes when a diagnosis of vertebral anomalies has been made.

754.200 Congenital postural scoliosis
Cervical scoliosis
Thoracic scoliosis
Lumbar scoliosis
Sacral scoliosis
Scoliosis, NOS

754.210 Congenital postural lordosis

754.220 Congenital postural curvature of spine, NOS

754.3 Congenital dislocation of hip

L * 754.300 Congenital dislocation of hip [1]

Special instructions:

[1] *When coding hip defects, a diagnosis of hip dysplasia (755.665-755.667) overrides coding congenital hip dislocation (754.300), which overrides coding any of the diagnoses in 754.310. Only code the condition highest in the hierarchy. Code 754.300 should be used when there is no diagnosis of hip dysplasia, and it overrides any conditions in 754.310.*

L * 754.310 Unstable hip [1]
Preluxation of hip [1]
Subluxation of hip [1]
Predislocation status of hip at birth [1]
Positive Barlow hip [1]
Dislocatable hip [1]
Subluxable hip [1]
Ortolani positive hip [1]
Excludes: Hip click
Hip crepitus
Hip laxity
Immature hip
Loose hip
Physiologic hip
Physiologic immature hip

Special instructions:

[1] *Code only if another reportable defect is present. When coding hip defects, a diagnosis of hip dysplasia (755.665-755.667) overrides coding congenital hip dislocation (754.300), which overrides coding any of the diagnoses in 754.310. Only code the condition highest in the hierarchy. Code 754.310 should be used for hips that are dislocatable, but not dislocated or dysplastic.*

754.4 Congenital genu recurvatum and bowing of long bones of leg

L 754.400 Bowing, femur

L 754.410 Bowing, tibia and/or fibula
Bowed lower leg

		754.420	Bow legs, NOS
L		754.430	Genu recurvatum
L		754.440	Dislocation of knee, congenital Subluxation knee Laxity knee
L		754.490	Deformity of leg, NOS

754.5 Varus (inward) deformities of feet

L		754.500	Talipes equinovarus
L		754.510	Talipes calcaneovarus
L	*	754.520	Metatarsus varus [1] Metatarsus adductus [1, 2] *Special instructions:* [1] *Code only if another reportable defect is present.* [2] *Do not code metatarsus adductus if the infant has talipes equinovarus.*
L		754.530	Complex varus deformities
L		754.590	Unspecified varus deformities of feet Inversion foot Foot turns inward Adducted foot Pes varus Medial rotation of foot/feet

754.6 Valgus (outward) deformities of feet

L		754.600	Talipes calcaneovalgus
L		754.610	Congenital pes planus Flat foot
L		754.615	Pes valgus
L		754.680	Other specified valgus deformities of foot Talipes equinovalgus Eversion foot
L		754.690	Unspecified valgus deformities of foot Abducted foot Foot turns outward

754.7 Other deformities of feet

L		754.700	Pes cavus Claw foot (Use 755.350 for lobster claw foot)
L		754.720	Short Achilles tendon
L		754.730	Clubfoot, NOS Talipes, NOS Equinus foot
L		754.735	Congenital deformities of foot, NOS
L		754.780	Other specified deformities of ankle and/or toes Includes: Dorsiflexion of foot Positional defect foot, NOS Foot turns upward Hyperextended foot

Malpositioned feet
Excludes: Widely spaced 1st and 2nd toes (Use 755.600)

754.8 Other specified congenital musculoskeletal deformities

754.800 Pigeon chest (pectus carinatum)
Pigeon thorax (pectus carinatum)
754.810 Funnel chest (pectus excavatum)
Funnel thorax (pectus excavatum)
754.820 Other anomalies of chest wall
Includes: Curved sternum
Pectus, NOS
Chest, thorax, or rib cage that is:
Asymmetric
Barrel shaped
Bell shaped
Compressed

Deformed
Narrow
Short
Small
754.825 Shield chest
Shield thorax
L 754.830 Dislocation of elbow
L 754.840 Club hand or fingers
Abnormal hand position with mention of forearm/wrist bone abnormality
Ulnar deviation of hand or wrist with mention of ulnar defect
Radial deviation of hand or wrist with mention of radial defect
Talipomanus
L 754.850 Spade-like hand
L 754.880 Other specified deformity of hands
Flat hand
(See 755.500 for specified anomalies of fingers)

755 Other Congenital Anomalies of Limbs

755.0 Polydactyly

L 755.005 Postaxial polydactyly of the hand
Postaxial skin tag
Postaxial finger/digit
Excludes: Accessory index finger (use 755.010)
L 755.010 Accessory thumbs (preaxial polydactyly)
Accessory index finger
Bifid thumb
L 755.020 Accessory toes (postaxial)
L 755.030 Accessory big toe (preaxial)
Accessory second toe

L		755.090	Accessory digits, NOS (hand/foot not specified)
L		755.095	Accessory digits hand, NOS (preaxial, postaxial not specified)
L		755.096	Accessory digits foot, NOS (preaxial, postaxial not specified)

755.1 Syndactyly

L		755.100	Fused fingers
L		755.110	Webbed fingers
L		755.120	Fused toes
L	*	755.130	Webbed toes [1]

Special instructions:

[1] *Code webbing of the second and third toes only if another reportable defect is present. Always code webbing of other toes regardless of whether another reportable defect is present.*

L		755.190	Unspecified syndactyly (See below for specified site)
L		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
L		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

If the description of the condition includes amniotic or constricting bands, use additional code 658.800.

Excludes: Shortening of upper limb (Use 755.580) or hypoplasia of upper limb (Use 755.585).

L 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

L 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

L 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

L 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
  - Hemimelia forearm

L 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)

L 755.250 Split-hand malformation
Cleft hand
Ectrodactyly hand
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)

L 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

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

L 755.270 Postaxial longitudinal reduction defect 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 + or - fourth metacarpal also totally or partially absent
Ulnar ray defect, NOS

L 755.280 Other specified reduction defect of upper limb

L 755.285 Transverse reduction defect of upper limb, NOS
Includes: Congenital amputation of upper limb, NOS

L 755.290 Unspecified reduction defect of upper limb

755.3 Reduction defects of lower limb

If the description of the condition includes amniotic or constricting bands, use additional code 658.800.

Excludes shortening of lower limb (Use 755.680) and hypoplasia of lower limb (Use 755.685).

L 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

L 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

L 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

L 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

L 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
Absent tarsal bones
Excludes: Isolated absent or hypoplastic great toe (Use 755.365)

L 755.350 Split-foot malformation
Cleft foot
Ectrodactyly foot
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)
Note: Preaxial lower limb reductions can occur with split-hand malformations of the upper limb and these lower limb defects should be coded 755.365

L 755.360 Longitudinal reduction defect of lower limb, NOS
Includes: Absent long bone of leg with absent toes, NOS

L 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
Hemimelia tibia

L 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 + or - fourth metatarsal also totally or partially absent
Fibular ray defect, NOS
Fibular hemimelia

L 755.380 Other specified reduction defect of lower limb
Proximal femoral focal deficiency

L 755.385 Transverse reduction defect of lower limb, NOS
Includes: Congenital amputation of lower limb, NOS

L 755.390 Unspecified reduction defect of lower limb

755.4 Reduction defects, unspecified limb

If the description of the condition includes amniotic or constricting bands, use additional code 658.800

L 755.400 Absence of limb, NOS
Includes: Amelia, NOS

L 755.410 Phocomelia, NOS
Includes: Intercalary reduction defect, NOS

L 755.420 Transverse reduction defect, NOS
Includes: Congenital amputation of unspecified limb

L 755.430 Longitudinal reduction defect, NOS
Includes: Preaxial or postaxial reduction defect, NOS

L 755.440 Absent digits, not specified whether fingers or toes

Ectrodactyly NOS

755.480 Other specified reduction defect of unspecified limb

L 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

L # 755.500 Anomalies of fingers

Includes: Acrodactylia
Anomalies of the finger crease
Arachnodactyly of fingers
Brachydactyly
Camptodactyly
Clenched hand or fist
Clinodactyly
Digitalized thumb
Finger contractures
Finger-like thumb
Flexion deformity finger
Hyperextension finger
Incurving fingers
Longer finger
Overlapping finger
Short finger
Small finger, all other
Small finger, thumb
Symphalangism finger
Triphalangeal thumb

Excludes: Acrocephalosyndactyly (See 756.050)
Apert syndrome (See 756.055)

L 755.510 Anomalies of hand

Broad hand
Large hand
Long hand
Short hand
Short metacarpals

Excludes: Simian crease (Use 757.200)

L 755.520 Anomalies of wrist

Abnormal hand position without mention of forearm/wrist bone abnormality
Ulnar deviation hand or wrist with no mention of ulnar defect
Radial deviation hand or wrist with no mention of radial defect
Flexed wrist

L 755.525 Accessory carpal bones

L * 755.526 Madelung deformity [1]

Special instructions:

[1] *Never code a bowed ulna in the presence of Madelung deformity.*

L 755.530 Anomalies of forearm, NOS

Anomalies of radius and ulna
Bowed ulna with no mention of Madelung deformity
Short forearm

Short radius
Short ulna
Hypoplastic radius
Hypoplastic ulna

L 755.535 Radioulnar dysostosis

L 755.536 Radioulnar synostosis
Fused radius and ulna

L 755.540 Anomalies of elbow and upper arm
Anomalies of humerus
Rhizomelia arm
Short humerus
Cubitus valgus
Hyperextension elbow
Hypoplastic humerus
Abnormal humeral metaphysis
Radiohumeral synostosis

L 755.550 Anomalies of shoulder
Anomalies of scapula
Anomalies of clavicle

755.555 Cleidocranial dysostosis

L 755.556 Sprengel's deformity

L 755.560 Other anomalies of whole arm

L 755.580 Other specified anomalies of upper limb
Includes: Hyperextensibility of upper limb
Shortening of arm
Micromelia arm
Positional deformity arm

L 755.585 Hypoplasia of upper limb
Includes: Hypoplasia of fingers, hands, or arms
Hypomelia arm
Excludes: Aplasia or absent upper limb (See 755.2)

L 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

L # 755.600 Anomalies of toes
Includes: Acrodactylia
Arachnodactyly of toes
Brachydactyly
Camptodactyly
Clenched toes
Clinodactyly
Digitalized great toe
Displaced toe
Flexion deformity toe
Hammer toes
Hyperextension toe
Incurving toes

Long toe
Malpositioned toe
Overlapping toes
Short toe
Symphalangism toe
Triphalangeal great toe
Widely spaced first and second toes

L		755.605	Hallux valgus
L		755.606	Hallux varus
L		755.610	Anomalies of foot
			Includes: Plantar furrow
			Prominent heel
			Deep plantar crease
			Abnormal creases of foot
			Short metatarsals
			Short foot
			Large foot
			Small foot
			Long foot
			Broad foot
			Excludes: Lobster claw foot (Use 755.350)
L	#	755.616	Rocker-bottom foot
			Vertical talus foot
L		755.620	Anomalies of ankle
			Astragaloscaphoid synostosis
L	*	755.630	Anomalies of lower leg
			Angulation of tibia, tibial torsion [1]
			Anomalies of tibia and fibula
			Short fibula
			Short tibia
			Short lower leg
			Hypoplastic tibia
			Hypoplastic fibula
			Special instructions:
			[1] *Never code if clubfoot present.*
L		755.640	Anomalies of knee
			Hyperextended knee
			Webbed knee
L		755.645	Genu valgum
			Knee valgus
L		755.646	Genu varum
L		755.647	Absent patella or rudimentary patella
L		755.650	Anomalies of upper leg
			Anteversion of femur
			Anomalies of femur
			Rhizomelia leg
			Short femur
			Femur torsion
			Short thigh
			Hypoplastic femur

L 755.660 Anomalies of hip
Includes: Coxa vara
Coxa valga
Hyperextended hip
Other abnormalities of hips

* 755.665 Hip dysplasia, NOS [1]
Hypoplastic hip, NOS [1]
Special instructions:
[1] *When coding hip defects, a diagnosis of hip dysplasia (755.665-755.667) overrides coding congenital hip dislocation (754.300), which overrides coding any of the diagnoses in 754.310. Only code the condition highest in the hierarchy.*

L * 755.666 Unilateral hip dysplasia [1]
Hypoplastic hip, unilateral [1]
Special instructions:
[1] *When coding hip defects, a diagnosis of hip dysplasia (755.665-755.667) overrides coding congenital hip dislocation (754.300), which overrides coding any of the diagnoses in 754.310. Only code the condition highest in the hierarchy.*

* 755.667 Bilateral hip dysplasia [1]
Hypoplastic hip, bilateral [1]
Special instructions:
[1] *When coding hip defects, a diagnosis of hip dysplasia (755.665-755.667) overrides coding congenital hip dislocation (754.300), which overrides coding any of the diagnoses in 754.310. Only code the condition highest in the hierarchy.*

L 755.670 Anomalies of pelvis
Fusion of sacroiliac joint
Anomalies of ilium, ischium, and pubis

L * 755.680 Other specified anomalies of lower limb
Hyperextended legs
Micromelia leg
Positional deformity leg
Hemihypertrophy leg (Also assign code 759.890)
Short leg
Bifid pelvis [1]
Special instructions:
[1] *Never code in the presence of exstrophy of the cloaca/bladder.*

L 755.685 Hypoplasia of lower limb
Includes: Hypoplasia of toes, feet, legs
Excludes: Aplasia or absent lower limb (See 755.3)

L 755.690 Unspecified anomalies of legs

755.8 Other specified anomalies of unspecified limb

L 755.800 Arthrogryposis multiplex congenita
Includes: Distal arthrogryposis syndrome
Flexion contractures of individual joints
Webbed elbow
Pena-Shokier syndrome
Webbed hip

755.810 Larsen's syndrome

L * 755.880 Other specified anomalies of unspecified limb

Includes: Overlapping digits, NOS [1]
Hyperextended joints, NOS
Short extremity
Rhizomelia extremity
Excludes: Hyperextended knees (Use 755.640)
Overlapping fingers (use 755.500)
Overlapping toes (Use 755.600)
Special instructions:
[1] *Code only if another reportable defect is present.*

755.9 Unspecified anomalies of unspecified limb

L 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 Craniosynostosis, NOS
Fused sutures, NOS
Craniostenosis, NOS
Closed-skull sutures, NOS
Cloverleaf shaped head
Squamosal craniosynostosis
756.005 Sagittal craniosynostosis
Sagittal fused sutures
756.006 Metopic craniosynostosis
Metopic fused sutures
L 756.010 Coronal craniosynostosis
Coronal fused sutures
L 756.020 Lambdoidal craniosynostosis
Lambdoidal fused sutures
756.030 Other types of craniosynostosis (fused sutures)
Includes: Basilar craniosynostosis
Basilar fused sutures
756.040 Craniofacial dysostosis
Includes: Crouzon's disease
756.045 Mandibulofacial dysostosis
Includes: Franceschetti syndrome
Treacher-Collins syndrome
756.046 Other craniofacial syndromes
Includes: Oculomandibulofacial syndrome
Hallermann-Streiff syndrome
Fronto-nasal dysplasia
Nager syndrome
Antley Bixler syndrome

Rib gap syndrome
Cerebromandibular syndrome
Smith-Theiler-Schachenmann 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
Pfeiffer syndrome
Greig cephalopolysyndactyly syndrome

756.060 Goldenhar syndrome
Oculoauriculovertebral dysplasia
Facio-auriculo-vertebral syndrome
OAV syndrome

L 756.065 Hemifacial microsomia
Facial microsomia

* 756.080 Other specified skull and face bone anomalies
Includes: Asymmetric jaw
Bathrocephaly
Biparietal narrowing
Bitemporal narrowing
Cleft mandible
Close set eyes
Flat facial profile
Flat facies
Flat midface
Flat occiput [1]
Flat supraorbital ridges
Forehead anomalies
Hypoplastic supraorbital ridges
Hypotelorism
Localized skull defects
Malar hypoplasia
Maxillary hypoplasia
Mid-facial hypoplasia
Pointed chin
Prominent maxilla
Prominent occiput [1]
Short occiput
Sloping forehead
Temporal narrowing
Excludes: Macrocephaly (Use 742.400)
Small chin (See 524.0)
Pierre Robin sequence (Use 524.080)
Special instructions:
[1] *Code only if another reportable defect is present.*

756.085 Hypertelorism, telecanthus, wide set eyes

756.090 Unspecified skull and face bone anomalies
Craniofacial abnormality NOS

Craniofacial disproportion
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
Segmentation anomaly (cervical)
Fused vertebrae (cervical)
Hypoplastic vertebrae (cervical)
Hypoplastic odontoid
Bifid vertebrae (cervical)
Butterfly vertebrae (cervical)
Cleft vertebrae (cervical)
Spinal dysraphism (cervical) [1]
Special instructions:
[1] *Never code in the presence of a cervical spina bifida.*
756.145 Hemivertebrae (cervical)
756.146 Agenesis (cervical)
* 756.150 Anomalies of thoracic vertebrae
Segmentation anomaly (thoracic)
Fused vertebrae (thoracic)
Hypoplastic vertebrae (thoracic)
Bifid vertebrae (thoracic)
Butterfly vertebrae (thoracic)
Cleft vertebrae (thoracic)
Spinal dysraphism (thoracic) [1]
Special instructions:
[1] *Never code in the presence of a thoracic spina bifida.*
756.155 Hemivertebrae of thoracic vertebrae
756.156 Agenesis of thoracic vertebrae
* 756.160 Anomalies of lumbar vertebrae
Segmentation anomaly (lumbar)
Fused vertebrae (lumbar)
Hypoplastic vertebrae (lumbar)
Bifid vertebrae (lumbar)
Butterfly vertebrae (lumbar)
Cleft vertebrae (lumbar)
Spinal dysraphism (lumbar) [1]
Special instructions:

[1] *Never code in the presence of a lumbar spina bifida.*

756.165 Hemivertebrae of lumbar vertebrae

756.166 Agenesis of lumbar vertebrae

* 756.170 Sacrococcygeal anomalies
Segmentation anomaly (sacral)
Fused sacrum
Hypoplastic sacrum
Bifid sacrum
Butterfly sacrum
Cleft sacrum
Sacral hemivertebrae
Spinal dysraphism (sacral) [1]
Includes: Agenesis of sacrum
Excludes: Pilonidal sinus (See 685.100)
Special instructions:
[1] *Never code in the presence of a sacral spina bifida.*

756.179 Sacral mass, NOS

* 756.180 Other specified vertebral anomalies
Segmentation anomaly, NOS
Fused vertebrae, NOS
Hypoplastic vertebrae, NOS
Bifid vertebrae, NOS
Butterfly vertebrae, NOS
Cleft vertebrae, NOS
Spinal dysraphism, NOS [1]
Platyspondyly
Special instructions:
[1] *Never code in the presence of a spina bifida.*

756.185 Hemivertebrae, NOS

756.190 Unspecified anomalies of spine

756.2 Cervical rib

L # 756.200 Cervical rib
Supernumerary rib in cervical region

756.3 Other anomalies of ribs and sternum

L # 756.300 Absence of ribs
Less than 24 ribs
Less than 12 rib pairs

L 756.310 Misshapen ribs
Bifid rib

L 756.320 Fused ribs

L 756.330 Extra ribs
More than 24 ribs
More than 12 rib (pairs)

L 756.340 Other anomalies of ribs
Gracile rib
Short rib
Hypoplastic rib

Small rib
Thin rib
Dysplasia rib
Rudimentary rib

756.350 Absence of sternum

756.360 Misshapen sternum

756.380 Other anomalies of sternum
Includes: Double ossification center in the manubrium
Bifid sternum
Bifid xyphoid process
Short sternum
Long sternum
Wide sternum

756.390 Anomalies of thoracic cage, unspecified
Excludes: Deformed chest (Use 754.820)

756.4 Chondrodystrophy

756.400 Asphyxiating thoracic dystrophy
Jeune syndrome
Thoracic-pelvic-phalangeal dysplasia
Excludes: Homozygous achondroplasia

756.410 Chondrodysplasia
Ollier syndrome, enchondromatosis

756.420 Chondrodysplasia with hemangioma
Kast syndrome
Maffucci syndrome

* 756.430 Achondroplastic dwarfism [1]
Special instructions:
[1] *When coding dwarfism, do not code short limbs separately.*

* 756.445 Diastrophic dwarfism [1]
Special instructions:
[1] *When coding dwarfism, do not code short limbs separately.*

* 756.446 Metatrophic dwarfism [1]
Special instructions:
[1] *When coding dwarfism, do not code short limbs separately.*

* 756.447 Thanatophoric dwarfism [1]
Special instructions:
[1] *When coding dwarfism, do not code short limbs separately.*

756.450 Metaphyseal dysostosis

756.460 Spondyloepiphyseal dysplasia

756.470 Exostosis
Excludes: Gardner syndrome (See 759.630)

756.480 Other specified chondrodystrophy
Achondrogenesis type I
Achondrogenesis type II
Campomelic dysplasia
Dyssegmental dysplasia
Hypochondrogenesis
Jarcho Levin syndrome

Kyphomelic dysplasia
Short rib-polydactyly syndrome
Spondylometaphyseal dysplasia
Spondylothoracic dysplasia
Werner mesomelic dysplasia
Excludes: Conradi's (Use 756.575)

756.490 Unspecified chondrodystrophy
Osteochondrodysplasia
Skeletal dysplasia
Dwarfism, NOS
Excludes: Lipochondrodystrophy (Use 277.510)

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-Engelmann disease
756.560 Osteopoikilosis
756.570 Multiple epiphyseal dysplasia
756.575 Conradi syndrome
Chondrodysplasia punctata
Excludes: Warfarin embryopathy
756.580 Other specified osteodystrophies
Craniometaphyseal dysplasia
756.590 Unspecified osteodystrophies

756.6 Anomalies of diaphragm

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

756.7 Anomalies of abdominal wall

		756.700	Exomphalos, omphalocele
		756.710	Gastroschisis
			Excludes: Umbilical hernia (553.100)
		756.720	Prune belly syndrome
			Eagle-Barrett's syndrome
		756.790	Other and unspecified anomalies of abdominal wall
			Body stalk anomaly
		756.795	Epigastric hernia
			Supraumbilical hernia

756.8 Other specified anomalies of muscle, tendon, fascia and connective tissue

L		756.800	Poland syndrome or anomaly
			Absent pectoralis major
L		756.810	Other absent or hypoplastic muscle
			Excludes: Prune belly syndrome (Use 756.720)
L		756.820	Absent tendon
		756.830	Nail-patella syndrome
		756.840	Amyotrophia congenital
			Amyoplasia congenita
		756.850	Ehlers-Danlos syndrome
L	#	756.860	Congenital torticollis (See also 754.100, anomalies of sternocleidomastoid muscle)
L		756.880	Other specified anomalies of muscle, tendon, fascia, and connective tissue
			Includes: Myopathy, congenital NOS
			Muscle atrophy (specified muscle)

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 Hereditary edema of legs

	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 ichthyoses
Keratitis-ichthyosis-deafness syndrome
KID syndrome
Lamellar ichthyosis
757.195 Ichthyosis vulgaris
757.196 X-linked ichthyosis
757.197 Ichthyosiform erythroderma

757.2 Dermatoglyphic anomalies

L # 757.200 Abnormal palmar creases
Includes: Simian creases
Transverse palmar creases

757.3 Other specified anomalies of skin

Excludes: Pigmented mole (216.900)
Hemangioma (See 228.0)

757.300 Specified syndromes, not elsewhere classified, involving skin anomalies
Neurocutaneous melanosis syndrome
Goltz syndrome
Chediak-Higashi syndrome
757.310 Skin tags
Includes: Anal tags
Excludes: Preauricular tag (See 744.110)
Vaginal tags (See 752.480)
Skin tags on the face (see 744.110 and 744.120)
757.320 Urticaria pigmentosa
757.330 Epidermolysis bullosa
Bart syndrome
757.340 Ectodermal dysplasia
Excludes: Ellis-van Creveld syndrome (756.525)
757.345 X-linked type ectodermal dysplasia
757.346 Other specified ectodermal dysplasias
Ectrodactyly-Ectodermal Dysplasia-Clefting (EEC) Syndrome
757.350 Incontinentia pigmenti
757.360 Xeroderma pigmentosum
757.370 Cutis laxa hyperelastica
757.380 Nevus, not elsewhere classifiable
Includes: Port wine stain
Excludes: Hairy nevus (Use 216.920)
Sturge-Weber syndrome (Use 759.610)
757.385 Birthmark, NOS
757.390 Other specified anomalies of skin
Includes: Cafe au lait spots
Skin cysts
Hypoplastic dermal patterns
Hyperpigmentation
Hair whorl anomalies

Hypopigmentation
Axillary freckling

757.395 Absence of skin
Aplasia cutis not involving the scalp

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

757.450 Persistent or excessive lanugo
Includes: Hirsutism
Hypertrichosis

757.480 Other specified anomalies of hair
White forelock
Sparse hair

757.5 Specified anomalies of nails

L 757.500 Congenital anonychia
Absent nails

L 757.510 Enlarged or hypertrophic nails

757.515 Onychauxis

757.516 Pachyonychia

757.520 Congenital koilonychia

757.530 Congenital leukonychia

L 757.540 Club nail

L 757.580 Other specified anomalies of nails
Duplication nail
Hyperconvex nail
Dystrophic nail
Dysplastic nail

L 757.585 Hypoplastic (small) fingernails and/or toenails
Narrow nails

757.6 Specified anomalies of breast

L 757.600 Absent breast with absent nipple

L 757.610 Hypoplastic breast with hypoplastic nipple
Absent breast tissue

L 757.620 Accessory (ectopic) breast with nipple

L 757.630 Absent nipple

L * 757.640 Small nipple (hypoplastic) [1]
Special instructions:

[1] *Never code if the infant is less than 36 weeks gestation. If the infant is greater than or equal to 36 weeks gestation, code only if another reportable defect is present.*

L # 757.650 Accessory (ectopic) nipple, supernumerary
Polythelia

757.680 Other specified anomalies of breast
Widely spaced nipples
Asymmetry nipples

757.8 Other specified anomalies of the integument

757.800 Includes: Scalp defects
Aplasia cutis scalp
Excludes: Aplasia cutis not involving the scalp (Use 757.395)
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 Chromosomal Anomalies

If a cytogenetic analysis was performed (including amniocentesis or chorionic villus sampling) and the karyotype is known, code the specific type of chromosome anomaly (e.g., 758.020 - Translocation trisomy - duplication of a 21).

If a cytogenetic analysis was performed (including amniocentesis or chorionic villus sampling) and the karyotype is unknown, code as NOS (e.g., 758.090 - Down syndrome, NOS).

If the diagnosis was made based solely on physical exam or on the presence of other birth defects, code the diagnosis as NOS (e.g., 758.090 - Down syndrome, NOS) and check the Possible/Probable checkbox.

Note: Some clinical syndromes are inconsistently associated with chromosome problems. When both the syndrome and the chromosome problem are present, code both. Code the chromosome problem only if it has been demonstrated by cytogenetic analysis.

Examples: DiGeorge syndrome (279.110) and deletion 22q (758.370)
Beckwith-Wiedemann syndrome (759.870) and disomy 11 (758.580)
Prader-Willi syndrome (759.870) and deletion 15q (758.380) or disomy 15 (758.580)

758.0 Down syndrome

Clinical Down syndrome karyotype identified as:

758.000 Down syndrome, karyotype trisomy 21
758.010 Down syndrome, karyotype trisomy G, NOS
758.020 Translocation trisomy - duplication of a 21
758.030 Translocation trisomy - duplication of a G, NOS

758.040 Mosaic Down syndrome
758.090 Down syndrome, NOS

758.1 Patau syndrome / Trisomy 13

Clinical Patau syndrome karyotype identified as:

758.100 Patau syndrome, karyotype trisomy 13
758.110 Patau syndrome, karyotype trisomy D, NOS
758.120 Translocation trisomy - duplication of a 13
758.130 Translocation trisomy - duplication of a D, NOS
758.140 Mosaic Patau syndrome
758.190 Patau syndrome, NOS

758.2 Edwards syndrome / Trisomy 18

Clinical Edwards syndrome karyotype identified as:

758.200 Edwards syndrome, karyotype trisomy 18
758.210 Edwards syndrome, karyotype trisomy E, NOS
758.220 Translocation trisomy - duplication of an 18
758.230 Translocation trisomy - duplication of an E, NOS
758.240 Mosaic Edwards syndrome
758.290 Edwards syndrome, NOS
758.295 Edwards phenotype - normal karyotype

758.3 Autosomal deletion syndromes

758.300 Antimongolism syndrome
Clinical antimongolism syndrome:
Karyotype - partial or total deletion of:
21
G, NOS
NOS
758.310 Cri du chat syndrome
Clinical Cri du chat syndrome:
Karyotype - deletion of:
5
B, NOS
NOS
758.320 Wolff-Hirschorn syndrome
Clinical Wolff-Hirschorn syndrome:
Karyotype - deletion of:
4
B, NOS
NOS
758.330 Deletion of long arm of 13
Deletion of long arm of D, NOS
758.340 Deletion of long arm of E
Deletion of long arm of 17 or 18
758.350 Deletion of short arm of E

Deletion of short arm of 17 or 18

758.360 Monosomy G mosaicism

758.370 Deletion in band 11 of long arm of 22 (22q11 deletions)

758.380 Other loss of autosomal material
Monosomy

758.390 Unspecified autosomal deletion syndromes

758.4 Balanced autosomal translocation in normal individual

758.400 Balanced autosomal translocation in normal individual

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, NOS
Trisomy: 1, 2, 3, 4, 5, 14, 15, 16, 17, 19, or 20

758.530 Partial trisomy syndromes

758.540 Other translocations
Excludes: Balanced translocation in normal Individual (Use 758.400)

758.580 Other specified anomalies of autosomes, NOS
Includes: Marker autosome
Cat eye syndrome
Tetrasomy

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 phenotype, karyotype 45, X [XO]

758.610 Turner phenotype, variant karyotypes
Karyotype characterized by:
Isochromosome
Mosaic, including XO
Partial X deletion
Ring chromosome
Excludes: Turner phenotype, karyotype normal XX (Use 759.800, Noonan syndrome)

758.690 Turner syndrome, karyotype unspecified, NOS
Bonneville-Ullrich syndrome, NOS

758.7 Klinefelter syndrome

758.700 Klinefelter phenotype, karyotype 47, XXY
758.710 Klinefelter phenotype, other karyotype with additional X chromosomes
XXXY
XXYY
XXXXY
758.790 Klinefelter syndrome, NOS

758.8 Other conditions due to sex chromosome anomalies

758.800 Mosaic XO/XY, 45X/46XY
Monosomy X mosaicism
Excludes: with Turner phenotype (758.610)
758.810 Mosaic XO/XX
Excludes: with Turner phenotype (758.610)
758.820 Mosaic XY/XXY,46XY/47XXY
Excludes: Klinefelter phenotype (758.710)
758.830 Mosaic including XXXXY,49XXXXY
Excludes: with Klinefelter phenotype (Use 758.710)
758.840 XYY, male, 47XYY
Mosaic XYY male
758.850 XXX female,47XXX
758.860 Additional sex chromosomes, NOS
758.880 Other specified sex chromosome anomaly
Includes: Fragile X
46,XX male
46,XY female, without a diagnosis of androgen insensitivity
Xq duplication
758.890 Unspecified sex chromosome anomaly

758.9 Conditions due to anomaly of unspecified chromosomes

758.900 Mosaicism, NOS
758.910 Additional chromosome(s), NOS
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 [1]
Special instructions:
[1] *Code only if absence of spleen is demonstrated by radiology or surgery.*
759.005 Ivemark syndrome
759.010 Hypoplasia of spleen
Small spleen

#	759.020	Hyperplasia of spleen
		Splenomegaly
		Hepatosplenomegaly (Also use code 751.620)
		Enlarged spleen
	759.030	Misshapen spleen
		Lobulated spleen
	759.040	Accessory spleen
		Polysplenia
	759.050	Ectopic spleen
		Spleen on right in heterotaxy syndrome
	759.080	Other specified anomalies of spleen
		Spleen cyst
	759.090	Unspecified anomalies of spleen

759.1 Anomalies of adrenal gland

L	759.100	Absence of adrenal gland
L	759.110	Hypoplasia of adrenal gland
L	759.120	Accessory adrenal gland
L	759.130	Ectopic adrenal gland
L	759.180	Other specified anomaly of adrenal gland
		Enlarged adrenal gland
		Fused adrenal glands
		Dysgenesis of adrenal gland
		Disc shaped adrenals
		Excludes: Congenital adrenal hyperplasia (Use 255.200)
L	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
		Congenital goiter
	759.220	Thyroglossal duct anomalies
		Thyroglossal cyst
	759.230	Anomalies of parathyroid gland
*	759.240	Anomalies of thymus
		Thymic hypertrophy [1]
		Absent thymus
		Hypoplastic thymus
		Small thymus
		Enlarged thymus [1]
		Special instructions:
		[1] *Code only if another reportable defect is present.*
	759.280	Other specified anomalies of endocrine gland
	759.290	Unspecified anomaly of endocrine gland

759.3 Situs inversus

	759.300	Dextrocardia with complete situs inversus
		Complete, mirror reversal of all organs

759.310 Situs inversus with levocardia
759.320 Situs inversus thoracis
Complete, mirror reversal of thoracic organs with normal abdominal organs
759.330 Situs inversus abdominis
Complete, mirror reversal of abdominal organs with normal thoracic organs
759.340 Kartagener syndrome (triad)
Immotile cilia syndrome
Sinusitis, bronchitis and situs inversus
* 759.390 Unspecified situs inversus [1]
Situs ambiguous [1]
Heterotaxy syndrome [1]
Excludes: Dextrocardia not associated with complete situs inversus (746.800)
Special instructions:
[1] *When coding this defect, also code all components separately.*

759.4 Conjoined twins

For all conjoined twins, fill out an abstraction form for each of the twins. For each twin, only code the birth defects specific to that twin, including the diagnosis of conjoined twin.

759.400 Dicephalus
Two heads
759.410 Craniopagus
Head-joined twins
Cephalopagus conjoined twins
759.420 Thoracopagus
Thorax-joined twins
759.430 Xiphopagus
Xiphoid-joined twins
759.440 Pygopagus
Buttock-joined twins
759.480 Other specified conjoined twins
Acardiac twins
Ischiopagus conjoined twins
Pelvis-joined twins
759.490 Unspecified conjoined twins

759.5 Tuberous sclerosis

759.500 Tuberous sclerosis
Bourneville's disease
Epiloia

759.6 Other hamartoses, not elsewhere classified

759.600 Peutz-Jegher syndrome
759.610 Encephalocutaneous angiomatosis
Kalischer's disease
Sturge-Weber syndrome
759.620 Von Hippel-Lindau syndrome
759.630 Gardner syndrome

759.680 Other specified hamartomas
Infantile myofibromatosis
Rhabdomyoma (organs other than the heart)
759.690 Unspecified hamartomas

759.7 Multiple congenital anomalies

759.700 Multiple congenital anomalies
Anomaly, multiple, NOS
Deformity, multiple, NOS

759.8 Other specified anomalies and syndromes

* 759.800 Cong malformation syndromes affecting facial appearance
Aarskog syndrome
Agnathia formation complex
Blepharophimosis syndrome
BOR syndrome
Branchial arch syndrome
Cebocephaly
Coffin-Siris syndrome
Costello syndrome
Cyclops
Ethmocephaly
Facio-auricular-digital syndrome
Frasier syndrome
Fused eyes (eyes fused together)
Hypertelorism-hypospadias syndrome
Jacobsen syndrome [1]
Kabuki syndrome
Melnick-Fraser syndrome
Miller-Dieker syndrome
Noonan syndrome
Ochoa syndrome
Opitz G/BBB syndrome
Oral-facial-digital (OFD) syndrome, type I
Orofaciodigital syndrome, type II (Mohr syndrome)
Otocephaly
Oto-palato-digital syndrome
Rieger syndrome
Smith-Magenis syndrome
Waardenburg syndrome
Whistling face syndrome (Freeman Sheldon syndrome)
Williams-Beuren syndrome
Williams syndrome
XK aprosencephaly
Special instructions:
[1] *If a diagnosis of Jacobsen syndrome and an 11q23-24 deletion are found, use code 759.800 for Jacobsen syndrome and code 758.380 for the chromosome deletion. If a diagnosis of Jacobsen syndrome is found, but an 11q23-24 deletion is not found, use only code 759.800 for Jacobsen syndrome. If an 11q23-24 deletion is found, but a*

diagnosis of Jacobsen syndrome is not found or is based only on the chromosome report, use only code 758.380 for the chromosome deletion.

759.820 Cong malformation syndromes associated with short stature
Amsterdam dwarf (Cornelia de Lange syndrome)
Cockayne syndrome
Laurence-Moon-Biedl syndrome
Russell-Silver syndrome
Seckel syndrome
Smith-Lemli-Opitz syndrome

759.840 Cong malformation syndromes involving limbs
Adams-Oliver syndrome
Baller-Gerold syndrome
Carpenter syndrome
Caudal dysplasia
Caudal regression syndrome
Du Pan syndrome
Escobar syndrome
Femoral fibular hypoplasia – unusual facies syndrome
Femoral hypoplasia – unusual facies syndrome
Femur-fibula-ulna syndrome
Fetal akinesia deformation sequence
Fryn syndrome
Holt-Oram syndrome
Hypoglossia-hypodactylia syndrome
Klippel-Trenaunay-Weber syndrome
Lethal multiple pterygium syndrome
Limb-body wall complex
Mermaid syndrome
Multiple pterygium syndrome
Oro-mandibular-limb hypogenesis syndrome
Pena-Shokeir II syndrome
Roberts phocomelia syndrome
Rubinstein-Taybi syndrome
Single lower extremity (fused lower extremities)
Sirenomelia
Thrombocytopenia-absent radius (TAR) syndrome

759.860 Cong malformation syndromes with other skeletal changes
Beals syndrome
Beemer Langer syndrome
Congenital contractural arachnodactyly syndrome
Marfan syndrome
Schinzel-Giedion syndrome
Stickler syndrome

759.870 Cong malformation syndromes with metabolic disturbances
Alagille syndrome
Alport syndrome
Barth syndrome
Beckwith (Wiedemann-Beckwith) syndrome
Johansen-Blizzard syndrome
Leprechaunism

Lowe syndrome
Menkes syndrome (kinky hair syndrome)
Pearson syndrome
Prader-Willi syndrome
Schwachman Diamond syndrome
Zellweger syndrome

759.890 Other specified anomalies

Includes: Acrocallosal syndrome
Aicardi syndrome
Angelman syndrome
Bloom syndrome
Cardio-splenic syndrome
Cerebro-oculo-facial-skeletal syndrome
CHARGE association
COFS syndrome
FG syndrome
Fukuyama congenital muscular dystrophy
Hemihypertrophy
Hydrolethalus syndrome
Jadassohn-Lewandasky syndrome
Meckel-Gruber syndrome
Muscle-eye-brain disease
Myotonic dystrophy
Neu-Laxova syndrome
Norrie disease
Oeis syndrome
Pentalogy of Cantrell
PHACE syndrome
Proteus syndrome
Sotos syndrome
Townes-Brock syndrome
Twin reversed arterial perfusion (TRAP) sequence
VACTERL association
VATER association
Weaver syndrome

759.9 Congenital anomaly, unspecified

759.900 Anomalies of umbilicus
Low-lying umbilicus
Umbilical cord atrophy
Small umbilical cord
Short umbilical cord

759.910 Embryopathia, NEC

759.990 Congenital anomaly, NOS
Abdominal cyst NOS
Abdominal mass NOS

052.000 Varicella, congenital (in utero infections with structural malformations or a diagnosis of congenital varicella syndrome)
090.000 Syphilis, congenital (in utero infections with structural malformations or a diagnosis of congenital syphilis syndrome)

214 Lipoma

214.000 Lipoma, skin and subcutaneous tissue of face
214.100 Lipoma, other skin and subcutaneous tissue
L # 214.200 Lipoma, intrathoracic organs
L # 214.300 Lipoma, intra-abdominal organs
214.400 Lipoma, spermatic cord
L # 214.800 Lipoma, other specified sites
214.810 Lipoma, lumbar or sacral lipoma
Paraspinal lipoma
214.900 Lipoma, unspecified site

216 Benign neoplasm of skin

Includes: Blue nevus
Papilloma
Syringoadenoma
Hydrocystoma
Excludes: Skin of female genital organs (Use 221.000),
Skin of male genital organs (Use 222.000)

216.000 Skin of lip
Excludes: Vermillion border of lip
L # 216.100 Eyelid, including canthus
Excludes: Cartilage of eyelid
L # 216.200 Ear and external auditory canal
Includes: Auricle ear
External meatus
Auricular canal
External canal
Pinna
Excludes: Cartilage of ear
216.300 Skin of other and unspecified parts of face
Includes: Cheek, external
Nose, external
Eyebrow
Temple
216.400 Scalp and skin of neck
216.500 Skin of trunk, except scrotum
Includes: Axillary fold
Perianal skin
Skin of chest wall, abdominal wall, groin, buttock, anus, perineum, back, umbilicus, or breast

Excludes: Anal canal
Anus, NOS
Skin of scrotum

L # 216.600 Skin of upper limb, shoulder
L # 216.700 Skin of lower limb, hip
L # 216.800 Other specified sites of skin
Excludes: Epibulbar dermoid cyst (Use 743.810)
216.900 Site unspecified
216.910 Sebaceous cyst
* 216.920 Hairy nevus [1]
Special instructions:
[1] *Code only if greater than four centimeters in diameter.*
221.000 Benign skin neoplasm of female genital organs
222.000 Benign skin neoplasm of male genital organs

228.0 Hemangioma

* 228.000 Hemangioma, of unspecified site [1]
Special instructions:
[1] *Only code if a) multiple hemangiomas of any size are present, b) if one or more cavernous hemangiomas of any size are present, or c) if a single hemangioma measuring greater than or equal to 4 centimeters in diameter or described as large, huge, or of medical significance is present.*
* 228.010 Hemangioma, skin & subcutaneous, NOS [1]
Special instructions:
[1] *Only code if a) multiple hemangiomas of any size are present, b) if one or more cavernous hemangiomas of any size are present, or c) if a single hemangioma measuring greater than or equal to 4 centimeters in diameter or described as large, huge, or of medical significance is present.*
* 228.020 Hemangioma, intracranial [1]
Special instructions:
[1] *Always code regardless of size, type, or number.*
L * 228.030 Hemangioma, retinal [1]
Special instructions:
[1] *Always code regardless of size, type, or number.*
L * 228.040 Hemangioma, intra-abdominal [1]
Hemangioendothelioma liver [1]
Special instructions:
[1] *Always code regardless of size, type, or number.*
L * 228.090 Hemangioma, of other sites [1]
Special instructions:
[1] *Always code regardless of size, type, or number.*
228.100 Cystic hygroma
Lymphangioma, any site
Multiloculated cyst of neck
237.700 Neurofibromatosis
Neurofibromatosis-Noonan syndrome
238.000 Teratoma, NOS
238.010 Teratoma, head and face
238.020 Teratoma, neck

	238.030	Teratoma, abdomen
	238.040	Teratoma, sacral, coccygeal
	238.080	Teratoma, other specified
	239.200	Neck cyst
	243.990	Hypothyroidism, congenital
#	244.800	Hypothyroidism, secondary/tertiary
#	251.200	Hypoglycemia, idiopathic
#	252.100	Hypoparathyroidism, congenital
#	253.280	Hypopituitarism, congenital
	253.820	Diencephalic syndrome
#	255.200	Adrenal hyperplasia, congenital, classical (salt) waster
#	255.210	Adrenal hyperplasia, congenital, classical (simple virilizer)
#	255.240	Adrenal hyperplasia, congenital, other than 21-OHP deficiency
#	255.290	Adrenal hyperplasia, congenital, NOS
		Adrenogenital syndrome
	257.800	Testicular feminization syndrome
		Androgen insensitivity syndrome
		46,XY female, with a diagnosis of androgen insensitivity
#	270.100	Phenylketonuria (PKU), classic
#	270.110	Phenylketonuria (PKU), hyperphenylalaninemia variant
#	270.190	Phenylketonuria (PKU), NOS
#	270.200	Albinism
#	270.210	Tyrosinemia
#	270.300	Maple syrup urine disease (MSUD)
#	270.400	Homocystinuria
#	270.600	Disorders of urea cycle metabolism
#	270.800	Disorders of amino acid metabolism NEC
#	270.900	Organic acidemias NEC
#	271.000	Glycogen storage diseases
#	271.100	Galactosemia, classic
#	271.110	Galactokinase deficiency
#	271.190	Galactosemia, NOS
#	271.800	Disorders of carbohydrate metabolism NEC
#	272.800	Disorders of lipid metabolism NEC
#	275.330	Hypophosphatemic rickets
		Hypophosphatasia, congenital
#	277.000	Cystic fibrosis, no mention of meconium ileus
#	277.010	Cystic fibrosis, with mention of meconium ileus
#	277.400	Disorders of bilirubin excretion
	277.510	Hurler syndrome
		Includes: Lipochondrodystrophy
#	277.800	Other specified inborn errors of metabolism NEC
#	277.850	Disorders of fatty acid oxidation
#	277.860	Peroxisomal disorders
#	277.900	Inborn error of metabolism NOS
	279.110	DiGeorge syndrome
		Shprintzen syndrome
		Velocardiofacial syndrome (VCFS)
#	282.600	Sickle cell disease, SS
#	282.630	Sickle cell disease, SC

	#	282.690	Sickle cell disease, other
	#	284.000	Red cell aplasia
	#	286.000	Hemophilia (all types)
	#	286.400	Von Willebrand disease
	#	330.100	Cerebral lipidoses
			Includes: Tay-Sachs disease
			Gangliosidosis
		335.000	Werdnig-Hoffman disease
			Infantile spinal muscular atrophy
		345.600	Infantile spasms, congenital
L	#	351.000	Facial palsy
			Asymmetric crying facies
			Bell's palsy
		352.600	Moebius syndrome
		362.600	Retinal degeneration, peripheral
		362.700	Retinitis pigmentosa
	#	363.200	Chorioretinitis
	*	368.000	Esotropia [1]
			Crossed eyes [1]
			Special instructions:
			[1] *Code only if another reportable defect is present. When coding esotropia, do not code strabismus separately.*
	#	378.000	Exotropia
			Duane syndrome
			Brown syndrome
	#	378.900	Strabismus, NOS
	#	379.500	Nystagmus
		425.300	Endocardial fibroelastosis
			Myocardial fibrosis
			Myofibroma (cardiac)
		426.705	Congenital Wolfe-Parkinson-White syndrome
		427.900	Cardiac arrhythmias, NEC
			Excludes: Premature atrial contractions (PACs)
	#	457.800	Other specified disorders of lymphatics (including chylothorax)
	#	520.600	Natal teeth
		524.000	Abnormalities of jaw size
			(Note: This code is only for abnormalities of jaw SIZE. Abnormalities of jaw SHAPE should be coded 756.080.)
			Micrognathia
			Macrognathia
			Recessed chin
			Mandibular hypoplasia
			Hypognathia
			Receding chin
			Small chin
			Retrognathia
			Agnathia
	*	524.080	Pierre Robin sequence [1]
			Special instructions:

[1] *Pierre Robin sequence should only be coded when all components are present - small jaw, displaced tongue and cleft soft palate. When coding this defect, the components should not be coded separately.*

L * 550.000 Inguinal hernia with mention of gangrene [1]
Special instructions:
[1] *If the infant is greater than or equal to 36 weeks gestation, and the infant is male, code only if another reportable defect is present. If the infant is greater than or equal to 36 weeks gestation, and the infant is female, always code. If the infant is less than 36 weeks gestation, never code.*

L * 550.100 Inguinal hernia with obstruction, (incarcerated) with no mention of gangrene [1]
Special instructions:
[1] *If the infant is greater than or equal to 36 weeks gestation, and the infant is male, code only if another reportable defect is present. If the infant is greater than or equal to 36 weeks gestation, and the infant is female, always code. If the infant is less than 36 weeks gestation, never code.*

L * 550.900 Inguinal hernia with no obstruction with no mention of gangrene [1]
Special instructions:
[1] *If the infant is greater than or equal to 36 weeks gestation, and the infant is male, code only if another reportable defect is present. If the infant is greater than or equal to 36 weeks gestation, and the infant is female, always code. If the infant is less than 36 weeks gestation, never code.*

\# 553.100 Umbilical hernia

L # 608.200 Torsion of testes or spermatic cord

\# 658.800 Amniotic bands (constricting bands, amniotic cyst)
Streeter syndrome/dysplasia
Constriction band syndrome

\# 685.100 Pilonidal sinus (sacrodermal), sacral sinus, sacral dimple
Dermal sinus spine

760.710 Fetal alcohol syndrome (FAS) or other alcohol-related birth defects (diagnosis of fetal alcohol syndrome, alcohol-related birth defects, or fetal alcohol spectrum disorder)

760.750 Fetal hydantoin (Dilantin) syndrome

760.760 Fetal Accutane (Isoretinoin) syndrome

L # 767.600 Erb's palsy
Brachial plexus palsy

771 Congenital infections (in utero infections only)

Excludes: Congenital syphilis (Use 090.000)

771.000 Rubella, congenital (in utero infections with structural malformations or a diagnosis of congenital rubella syndrome)

\# 771.090 TORCH infection, unspecified

\# 771.100 Cytomegalovirus (CMV)

\# 771.210 Toxoplasmosis

\# 771.220 Herpes simplex
Includes: Encephalitis
Meningoencephalitis

771.230 Congenital Zika virus infection (in utero infections with structural malformations or a diagnosis of congenital Zika syndrome)

	#	771.280	Congenital infection, other specified Congenital parvovirus infection Excludes: Human immunodeficiency virus (HIV) infection Acquired immunodeficiency syndrome (AIDS)
	#	774.480	Hepatitis, neonatal, other specified
	#	774.490	Hepatitis, neonatal, NOS
	#	777.100	Meconium plug syndrome
	#	777.600	Meconium peritonitis
	#	778.000	Ascites, congenital Includes: Hydrops fetalis Anasarca
L	#	778.600	Hydrocele, congenital

EXCLUSION LIST

The following conditions are considered to be excludable or non-reportable conditions. These conditions should **never** be listed in the "Birth Defects Diagnosis Information" section of the abstraction form.

Abducted thumb
Abduction hip
Absence of heterozygosity
Absent right superior vena cava
Acne, neonatal
Adhesions penis
Anal fissure
Aneurysm of atrial septum
Angel kisses
Anteverted uterus
Arachnoid cyst
Arm, long
Asymmetric gluteal cleft
Asymmetry brain
Asymmetry ears
Asymmetry eyes
Asymmetry of cerebral ventricles
Atrial dilatation
Atrial septum aneurysm
Atrium, enlarged
Benign external hydrocephaly
Biliary obstruction
Bladder, enlarged
Bladder hypertrophy
Bladder, neurogenic
Bladder, small
Bladder, thickened
Bladder, trabeculated
Blueberry muffin spots
Bowel perforation
Bradycardia
Brain, asymmetry
Breast hypertrophy
Breast, small
Bronchomalacia
Bronchopulmonary dysplasia
Budd-Chiari
Cavum septum pellucidum
Central nervous system (CNS) hemorrhage
Cephalohematoma
Cerebral ventricle asymmetry
Chalasia
Chin cleft
Chin dimple
Choroid plexus cyst, unilateral
Cleft chin
Click, hip, with no follow-up or therapy
Clifford's syndrome
Clitoris, prominent prepuce
Colon perforation

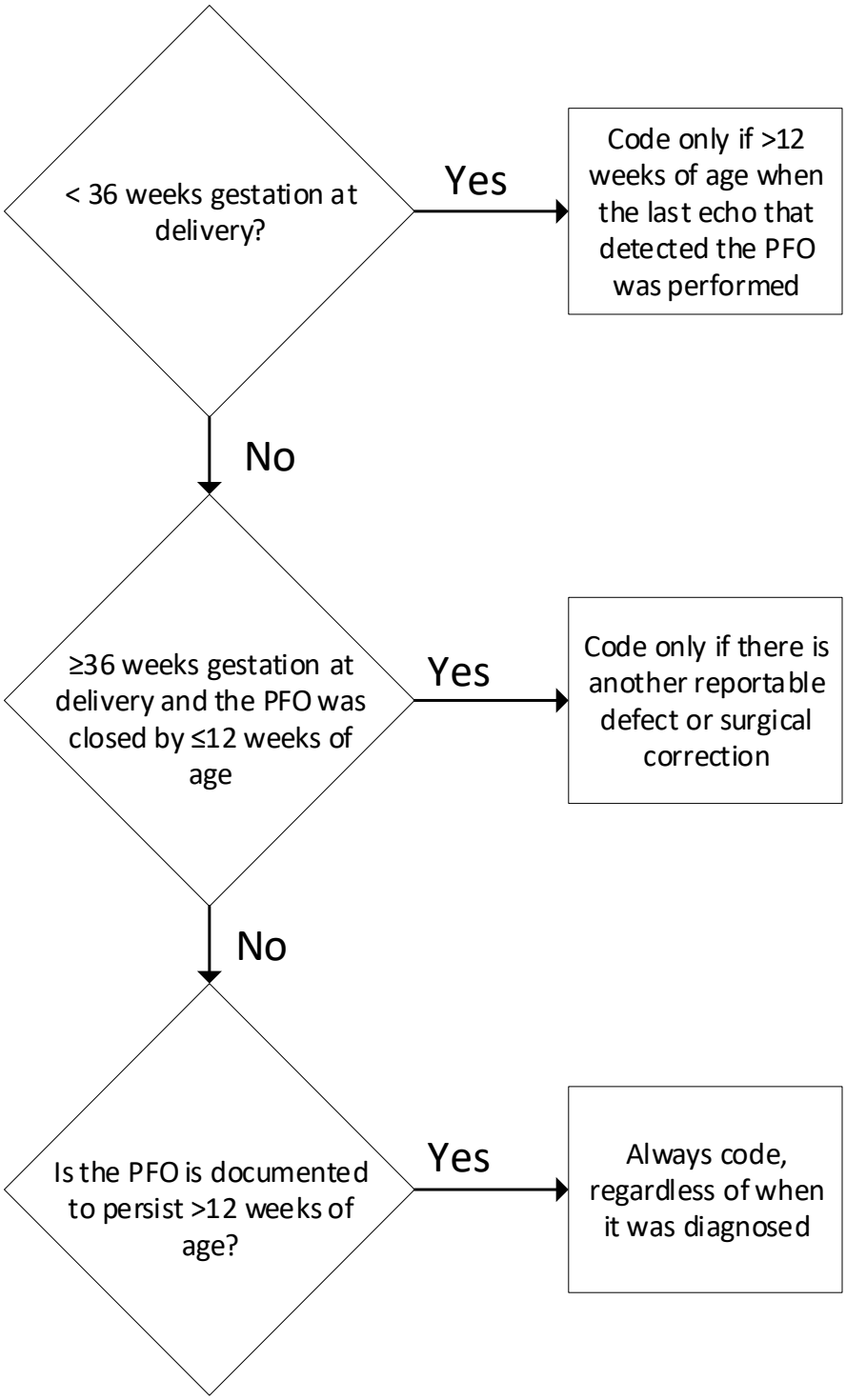
Congenital encephalopathy
Conjunctivitis
Craniotabes
Cupped tongue
Cutis marmorata
Cyst, arachnoid
Cyst, choroid plexus, unilateral
Cyst, gum
Cyst, tongue
Cyst, Wharton duct
Diaphragm, elevated
Diastsis recti
Dilatation/dilated/dilation atrium
Dilatation/dilated/dilation ventricle (heart)
Dimple, chin
Dislocation, shoulder
Ductal shelf
Duodenal obstruction
Duodenal perforation
Dysplasia, bronchopulmonary
Ears, asymmetry
Echogenic kidney
Edema (not of legs)
Elevated diaphragm
Encephalopathy, congenital
Enlarged atrium
Enlarged bladder
Enlarged septum pellucidum
Enlarged urethra
Enlarged uvula
Enlarged ventricle (heart)
Epstein’s pearls
Epulis
Esophagus, short
Eustacian valve
Eyes, asymmetry
Eyes, sun-setting
Fat filum terminale
Filum terminale, fat
Finger, cortical (thumb)
Fissure, anal
Foreskin, incomplete
Foreskin, redundant
Frenulum, thick
Fused scrotum
Gallbladder, small
Gallbladder wall, thickened
Gastric volvulus
Gastroesophageal reflux (GER)
Gluteal cleft, asymmetric
Gum cyst
Gum, hypoplastic
Hand, hyperflexion
Hand, narrow
Heart murmur
Heart valves, doming
Heart valves, floppy

Heart valves, thin
Hemorrhage, central nervous system (CNS)
Hepatic vein occlusion (Budd-Chiari)
Heterozygosity, absence or loss of
Hip, abduction
Hip click
Hip crepitus
Hip, immature
Hip laxity
Hip, loose
Hip, physiologic
Hip, physiologic immature
Human immunodeficiency virus (HIV)
Hyaline membrane disease
Hydrocephaly, benign external
Hydrocephaly secondary to intraventricular hemorrhage (IVH) or CNS bleed
Hydrocephaly, ex-vacuo
Hyperextended thigh
Hyperflexion hand
Hyperplasia, lung
Hyperplasia, pulmonary
Hypertrophic bladder
Hypertrophic breast
Hypertrophic urethra
Hypoplastic gum
Icterus
Ileal obstruction
Ileal perforation
Immature hip
Incomplete foreskin
Intestinal obstruction
Intestinal perforation
Intussusception
Inverted nipple
Jejunal obstruction
Kawasaki disease
Lagophthalmos
Large bladder
Large atrium
Large septum pellucidum
Large urethra
Large ventricle (heart)
Laryngomalacia
Laryngotracheomalacia
Levocardia
Long arm
Long nails
Loss of heterozygosity
Lung hyperplasia
Lung, left, two lobes
Lung, right, three lobes
Lymphedema (not of legs)
Meconium stained skin
Meconium stained nails
Mitral valve, redundant
Mongolian blue spot
Mucocele

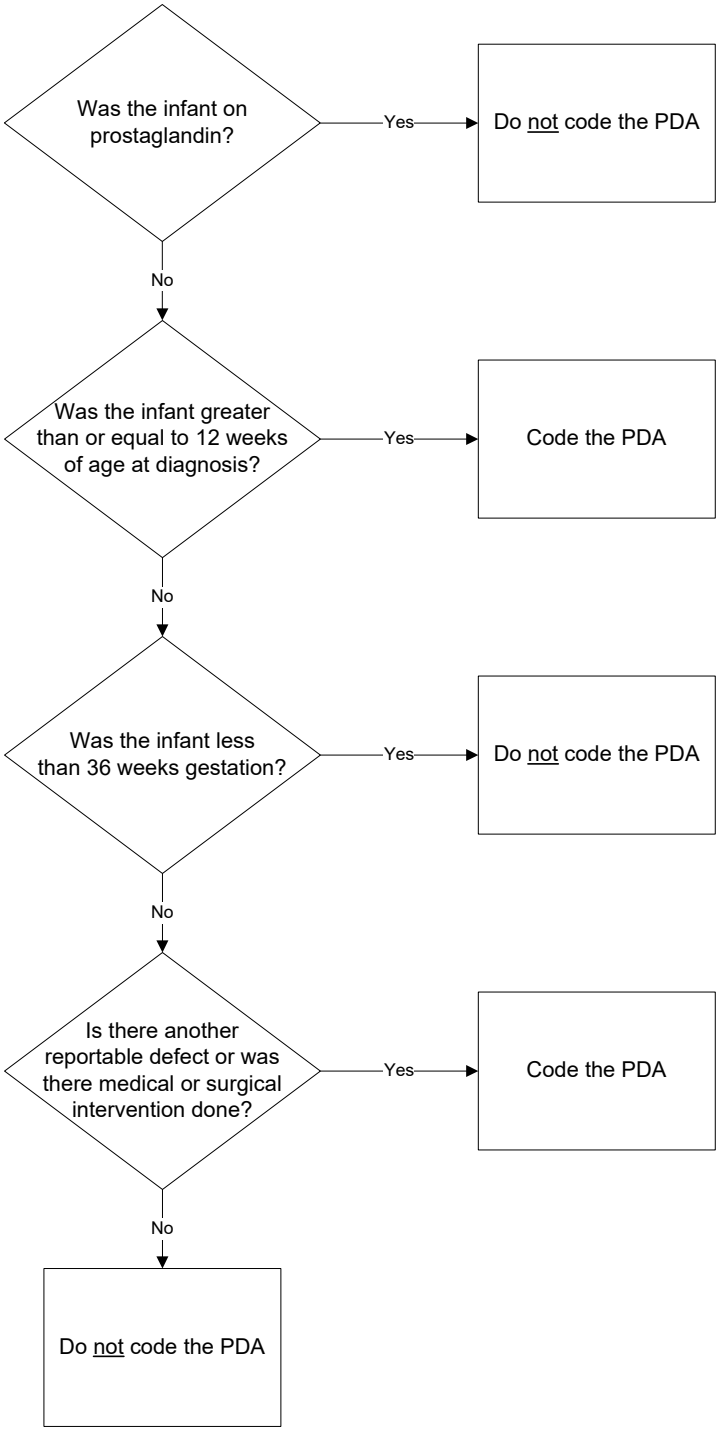
Mucous fistula
Murmur, heart
Myopia
Nails, long
Nails, meconium stained
Nails, short
Narrow hand
Nasal obstruction
Neonatal acne
Neurogenic bladder
Nevus flammeus
Nevus simplex
Nipple, inverted
Obstruction, biliary
Obstruction, duodenal
Obstruction, ileal
Obstruction, intestinal
Obstruction, jejunal
Obstruction, nasal
Ondine’s Curse syndrome
Osteopenia
Osteoporosis
Overlapping sutures
Overriding sutures
Palpebral fissure, thick
Paralysis, vocal cord
Perforation, bowel
Perforation, colon
Perforation, duodenum
Perforation, ileum
Perforation, intestine
Perforation, small intestine
Petechiae
Phimosis
Physiologic hip
Physiologic immature hip
Pneumothorax
Polymorphism
Premature atrial contractions (PACs)
Primary pulmonary artery hypertension
Prominent prepuce of clitoris
Prominent tongue
Prominent xyphoid process
Protruding/protuberant tongue
Pseudocircumcision
Pulmonary, hyperplasia
Ranula
Redundant foreskin
Redundant mitral valve
Reflux, gastroesophageal (GER)
Retractile testicle
Retroverted uterus
Right superior vena cava, absent
Sacral hair tuft
Salmon patches
Scaphoid abdomen
Scrotum, fused

Septum pellucidum, enlarged
Septum pellucidum, cavum
Short esophagus
Short nails
Shoulder dislocation
Skin, meconium stained
Small bladder
Small breast
Small gallbladder
Small intestine perforation
Small uvula
Stork bite
Sun-setting eyes
Superior vena cava, right, absent
Suture, overriding
Suture, overlapping
Syndrome, Clifford's
Syndrome, Ondine's Curse
Syndrome, Wilson-Mikity
Testicle/testis, retractile
Thick/thickened bladder
Thick frenulum
Thick palpebral fissure
Thick/thickened urethra
Thigh, hyperextended
Three lobes right lung
Thumb, abducted
Thumb tucked into palm
Tongue, cupped
Tongue cyst
Tongue, prominent
Tongue, protruding
Trabeculated bladder
Tracheomalacia
Two lobes left lung
Ulna, bowed, with Madelung deformity
Urethra, enlarged
Urethra, thickened
Urethral hypertrophy
Uterus, anteverted
Uterus, retroverted
Uvula, enlarged
Uvula, small
Ventricle, enlarged (heart)
Ventricular dilatation (heart)
Vesiculobullous dermatosis
Vocal cord paralysis
Volvulus, gastric
Volvulus, intestinal
Wharton duct cyst
Wilson-Mikity syndrome
Xyphoid process, prominent

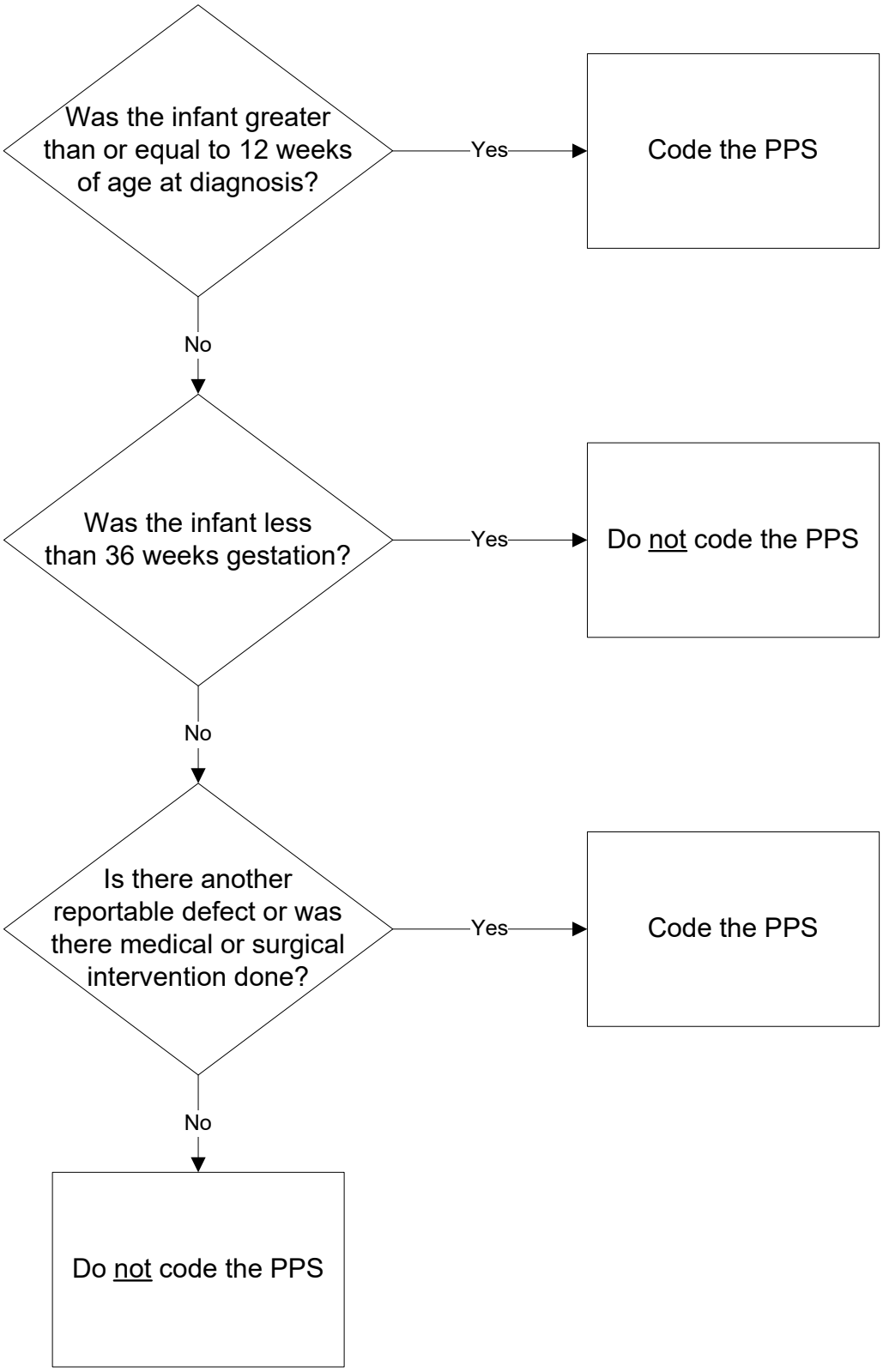
APPENDIX A: FLOWCHART FOR DECIDING WHEN TO CODE PATENT FORAMEN OVALE (PFO)



APPENDIX B: FLOWCHART FOR DECIDING WHEN TO CODE PATENT DUCTUS ARTERIOSUS (PDA)



APPENDIX C: FLOWCHART FOR DECIDING WHEN TO CODE PERIPHERAL PULMONIC STENOSIS (PPS)



APPENDIX D: CODING PLAGIOCEPHALY AND RELATED DEFECTS

Note: Plagiocephaly should only be coded as unilateral. If 'bilateral plagiocephaly' is listed in the medical record, it should be coded as brachycephaly. Also, it is possible to have brachycephaly and either left or right plagiocephaly in the same child.

If plagiocephaly is diagnosed and the following is listed in the medical record:	Abstract from the medical record?	Code?
Asymmetric head	Yes	No
Brachycephaly	Yes	Yes, unless synostosis is diagnosed
Coronal ridging or other suture ridging	Yes	No
Decreased neck range of motion (ROM)	Yes	No
Ear anteriorly displaced and laterality	Yes	Yes
Facial asymmetry	Yes	Yes
Flat/short/broad forehead	Yes	No
Forehead bossing	Yes	No
Frontal bossing	Yes	No
Metopic craniosynostosis	Yes	Yes
Occiput flattening and laterality	Yes	No
Open/closed fontanelle	Yes, if stated abnormal	Yes, if stated abnormal
Posterior towering	Yes	No
Sagittal craniosynostosis	Yes	Yes
Scaphocephaly	Yes	Yes, unless sagittal craniosynostosis is diagnosed
Temporal/parietal bulging	Yes	Yes
Temporoparietal bossing	Yes	Yes
Torticollis and laterality	Yes	Yes