

## 7.1 SIX-DIGIT CODES FOR REPORTABLE BIRTH DEFECTS

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)

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## TABLE OF CONTENTS

<b>Table of Contents</b> .....	<b>2</b>
<b>Introduction To Birth Defect Diagnosis Coding</b> .....	<b>7</b>
Purpose .....	7
BPA coding system .....	7
Description of the BPA code .....	8
Problems with the BPA coding system .....	8
<b>Instructions</b> .....	<b>10</b>
Inclusion status .....	10
Description of the birth defect .....	13
Coding the birth defect .....	14
Laterality .....	14
Possible/Probable .....	14
<b>Congenital Anomalies</b> .....	<b>15</b>
<b>740 Anencephalus and Similar Anomalies</b> .....	<b>15</b>
740.0 Anencephalus .....	15
740.1 Craniorachischisis .....	15
740.2 Iniencephaly.....	15
<b>741 Spina Bifida</b> .....	<b>15</b>
741.0 Spina Bifida with hydrocephalus .....	15
741.9 Spina bifida without mention of hydrocephalus .....	16
<b>742 Other Congenital Anomalies of Nervous System</b> .....	<b>17</b>
742.0 Encephalocele- .....	17
742.1 Microcephalus .....	17
742.2 Reduction deformities of brain.....	17
742.3 Congenital hydrocephalus.....	18
742.4 Other specified anomalies of brain.....	18
742.5 Other specified anomalies of spinal cord .....	19
742.8 Other specified anomalies of nervous system .....	19
742.9 Unspecified anomalies of brain, spinal cord and nervous systems .....	19
<b>743 Congenital Anomalies of Eye</b> .....	<b>20</b>
743.0 Anophthalmos.....	20
743.1 Microphthalmos .....	20
743.2 Buphthalmos.....	20
743.3 Congenital cataract and lens anomalies .....	20
743.4 Coloboma and other anomalies of anterior segments .....	20
743.5 Congenital anomalies of posterior segment.....	21
743.6 Congenital anomalies of eyelids, lacrimal system, and orbit .....	21
743.8 Other specified anomalies of eye .....	22
743.9 Unspecified anomalies of eye .....	23
<b>744 Congenital Anomalies of Ear, Face, and Neck</b> .....	<b>23</b>
744.0 Anomalies of ear causing impairment of hearing .....	23
744.1 Accessory auricle .....	23
744.2 Other specified anomalies of ear.....	23
744.3 Unspecified anomalies of ear .....	24
744.4 Branchial cleft, cyst, or fistula; preauricular sinus .....	24

744.5	Webbing of neck.....	24
744.8	Other unspecified anomalies of face and neck .....	25
744.9	Unspecified anomalies of face and neck.....	25
<b>745</b>	<b>Bulbus Cordis Anomalies and Anomalies of Cardiac Septal Closure.....</b>	<b>26</b>
745.0	Common truncus (See 747.200 for pseudotruncus) .....	26
745.1	Transposition of great vessels.....	26
745.2	Tetralogy of Fallot.....	26
745.3	Single ventricle .....	27
745.4	Ventricular septal defect.....	28
745.5	Ostium secundum type atrial septal defect .....	28
745.6	Endocardial cushion defects .....	29
745.7	Cor biloculare .....	29
745.8	Other specified defects of septal closure .....	29
745.9	Unspecified defect of septal closure.....	29
<b>746</b>	<b>Other Congenital Anomalies of Heart.....</b>	<b>29</b>
746.0	Anomalies of pulmonary valve.....	29
746.1	Anomalies of the tricuspid valve .....	30
746.2	Ebstein's anomaly.....	30
746.3	Congenital stenosis of aortic valve.....	30
746.4	Congenital insufficiency of aortic valve .....	30
746.5	Congenital mitral stenosis .....	31
746.6	Mitral valve insufficiency or regurgitation, congenital.....	31
746.7	Hypoplastic left heart syndrome .....	31
746.8	Other specified anomalies of the heart.....	32
746.9	Unspecified anomalies of heart.....	33
<b>747</b>	<b>Other Congenital Anomalies of Circulatory System.....</b>	<b>33</b>
747.0	Patent ductus arteriosus.....	33
747.1	Coarctation of aorta .....	33
747.2	Other anomalies of aorta .....	33
747.3	Anomalies of pulmonary artery.....	34
747.4	Anomalies of great veins .....	35
747.5	Absence or hypoplasia of umbilical artery.....	35
747.6	Other anomalies of peripheral vascular system .....	36
747.8	Other specified anomalies of circulatory system.....	36
747.9	Unspecified anomalies of circulatory system .....	36
<b>748</b>	<b>Congenital Anomalies of Respiratory System.....</b>	<b>37</b>
748.0	Choanal atresia .....	37
748.1	Other anomalies of nose .....	37
748.2	Web of larynx.....	37
748.3	Other anomalies of larynx, trachea, and bronchus .....	37
748.4	Congenital cystic lung.....	38
748.5	Agenesis or aplasia of lung .....	38
748.6	Other anomalies of lung .....	38
748.8	Other specified anomalies of respiratory system .....	39
748.9	Unspecified anomalies of respiratory system.....	39
<b>749</b>	<b>Cleft Palate and Cleft Lip .....</b>	<b>39</b>
749.0	Cleft palate alone.....	39
749.1	Cleft lip alone.....	39
749.2	Cleft lip with cleft palate.....	40
<b>750</b>	<b>Other Congenital Anomalies of Upper Alimentary Tract.....</b>	<b>40</b>
750.0	Tongue tie.....	40
750.1	Other anomalies of tongue .....	40

750.2	Other specified anomalies of mouth and pharynx.....	40
750.3	Tracheoesophageal (T-E) fistula, esophageal atresia and stenosis.....	41
750.4	Other specified anomalies of esophagus.....	41
750.5	Congenital hypertrophic pyloric stenosis.....	41
750.6	Congenital hiatus hernia.....	41
750.7	Other specified anomalies of stomach.....	42
750.8	Other specified anomalies of upper alimentary tract.....	42
750.9	Unspecified anomalies of upper alimentary tract.....	42
<b>751</b>	<b>Other Congenital Anomalies of Digestive System.....</b>	<b>42</b>
751.0	Meckel's diverticulum.....	42
751.1	Atresia and stenosis of small intestine.....	42
751.2	Atresia and stenosis of large intestine, rectum, and anal canal.....	42
751.3	Hirschsprung's disease and other congenital functional disorders of the colon.....	43
751.4	Anomalies of intestinal fixation.....	43
751.5	Other anomalies of intestine.....	43
751.6	Anomalies of gallbladder, bile ducts, and liver.....	44
751.7	Anomalies of pancreas.....	44
751.8	Other specified anomalies of digestive system.....	45
751.9	Unspecified anomalies of digestive system.....	45
<b>752</b>	<b>Congenital Anomalies of Genital Organs.....</b>	<b>45</b>
752.0	Anomalies of ovaries.....	45
752.1	Anomalies of fallopian tubes and broad ligaments.....	45
752.2	Doubling of uterus.....	46
752.3	Other anomalies of uterus.....	46
752.4	Anomalies of cervix, vagina, and external female genitalia.....	46
752.5	Undescended testicle.....	47
752.6	Hypospadias and epispadias.....	47
752.7	Indeterminate sex and pseudohermaphroditism.....	48
752.8	Other specified anomalies of male genital organs.....	48
752.9	Unspecified anomalies of genital organs.....	49
<b>753</b>	<b>Congenital Anomalies of Urinary System.....</b>	<b>49</b>
753.0	Renal agenesis and dysgenesis.....	49
753.1	Cystic kidney disease.....	49
753.2	Obstructive defects of renal pelvis and ureter.....	50
753.3	Other specified anomalies of kidney.....	51
753.4	Other specified anomalies of ureter.....	51
753.5	Exstrophy of urinary bladder.....	52
753.6	Atresia and stenosis of urethra and bladder neck.....	52
753.7	Anomalies of urachus.....	52
753.8	Other specified anomalies of bladder and urethra.....	52
753.9	Unspecified anomalies of urinary system.....	53
<b>754</b>	<b>Certain Congenital Musculoskeletal Anomalies.....</b>	<b>53</b>
754.0	Of skull, face, and jaw.....	53
754.1	Anomalies of sternocleidomastoid muscle.....	54
754.2	Certain congenital musculoskeletal deformities of spine.....	54
754.3	Congenital dislocation of hip.....	54
754.4	Congenital genu recurvatum and bowing of long bones of leg.....	55
754.5	Varus (inward) deformities of feet.....	55
754.6	Valgus (outward) deformities of feet.....	55
754.7	Other deformities of feet.....	55
754.8	Other specified congenital musculoskeletal deformities.....	56
<b>755</b>	<b>Other Congenital Anomalies of Limbs.....</b>	<b>56</b>
755.0	Polydactyly.....	56

755.1	Syndactyly .....	57
755.2	Reduction defects of upper limb .....	57
755.3	Reduction defects of lower limb .....	58
755.4	Reduction defects, unspecified limb .....	60
755.5	Other anomalies of upper limb, including shoulder girdle .....	60
755.6	Other anomalies of lower limb, including pelvic girdle .....	62
755.8	Other specified anomalies of unspecified limb .....	63
755.9	Unspecified anomalies of unspecified limb .....	64
<b>756</b>	<b>Other Congenital Musculoskeletal Anomalies.....</b>	<b>64</b>
756.0	Anomalies of skull and face bones .....	64
756.1	Anomalies of spine .....	66
756.2	Cervical rib.....	67
756.3	Other anomalies of ribs and sternum .....	67
756.4	Chondrodystrophy .....	68
756.5	Osteodystrophies.....	68
756.6	Anomalies of diaphragm.....	69
756.7	Anomalies of abdominal wall .....	69
756.8	Other specified anomalies of muscle, tendon, fascia and connective tissue.....	69
756.9	Unspecified anomalies of musculoskeletal system.....	70
<b>757</b>	<b>Congenital Anomalies of the Integument.....</b>	<b>70</b>
757.0	Hereditary edema of legs .....	70
757.1	Ichthyosis congenita.....	70
757.2	Dermatoglyphic anomalies .....	70
757.3	Other specified anomalies of skin .....	70
757.4	Specified anomalies of hair .....	71
757.5	Specified anomalies of nails.....	72
757.6	Specified anomalies of breast .....	72
757.8	Other specified anomalies of the integument.....	72
757.9	Unspecified anomalies of the integument .....	72
<b>758</b>	<b>Chromosomal Anomalies .....</b>	<b>73</b>
758.0	Down syndrome.....	73
758.1	Patau syndrome / Trisomy 13.....	73
758.2	Edwards syndrome / Trisomy 18.....	73
758.3	Autosomal deletion syndromes .....	74
758.4	Balanced autosomal translocation in normal individual .....	74
758.5	Other conditions due to autosomal anomalies .....	74
758.6	Gonadal dysgenesis.....	75
758.7	Klinefelter syndrome.....	75
758.8	Other conditions due to sex chromosome anomalies .....	75
758.9	Conditions due to anomaly of unspecified chromosomes.....	76
<b>759</b>	<b>Other and Unspecified Congenital Anomalies .....</b>	<b>76</b>
759.0	Anomalies of spleen .....	76
759.1	Anomalies of adrenal gland.....	76
759.2	Anomalies of other endocrine glands.....	77
759.3	Situs inversus .....	77
759.4	Conjoined twins .....	77
759.5	Tuberous sclerosis .....	78
759.6	Other hamartoses, not elsewhere classified .....	78
759.7	Multiple congenital anomalies .....	78
759.8	Other specified anomalies and syndromes .....	78
759.9	Congenital anomaly, unspecified .....	80
	<b>Other Specified Codes.....</b>	<b>82</b>

<b>Exclusion List.....</b>	<b>88</b>
<b>Appendix A: Flowchart For Deciding When To Code Patent Foramen Ovale (PFO) .....</b>	<b>93</b>
<b>Appendix B: Flowchart For Deciding When To Code Patent Ductus Arteriosus (PDA).....</b>	<b>95</b>
<b>Appendix C: Flowchart For Deciding When To Code Peripheral Pulmonic Stenosis (PPS).....</b>	<b>95</b>
<b>Appendix C: Flowchart For Deciding When To Code Peripheral Pulmonic Stenosis (PPS).....</b>	<b>96</b>
<b>Appendix D: Coding Plagiocephaly And Related Defects .....</b>	<b>97</b>

# 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, 9<sup>th</sup> 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:

<b>Diagnosis</b>	<b>ICD-9 Code</b>	<b>BPA Code</b>
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:

<b>Diagnosis</b>	<b>ICD-9 Code</b>	<b>BPA Code</b>
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 <sup>1</sup>
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*Special instructions:*

<sup>1</sup> *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 <sup>1</sup>

*Special instructions:*

<sup>1</sup> *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
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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
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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, supra-valvular 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 all 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, hemiccephaly
- 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 hydrocephalus  
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 hydrocephalus  
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 hydrocephalus  
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
- Sacroccygeal myelocele, myelomeningocele, meningomyelocele, and meningocele without hydrocephalus
- 741.985 Lipomyelomeningocele  
Lipomeningocele
- 741.990 Spina bifida, site unspecified, without hydrocephalus (myelocele, 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
- 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
  - Pachygyria
  - Schizencephaly
  - Pontine hypoplasia
  - Hypoplastic thalamus
  - Reduction defect of brainstem
  - Hypoplastic brainstem
  - Small brainstem
- 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 <sup>1</sup>
  - Special instructions:*
  - <sup>1</sup> *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 <sup>1</sup>
  - Macrocephaly <sup>1</sup>
  - Special instructions:*
  - <sup>1</sup> *Never code in the presence of hydrocephaly.*
- \* 742.410 Porencephaly
  - Includes: Porencephalic cysts <sup>1</sup>
  - Special instructions:*
  - <sup>1</sup> *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 <sup>1</sup>
  - Excludes: Arachnoid cysts
  - Special instructions:*
  - <sup>1</sup> *Code only if there are multiple cysts that were diagnosed postnatally.*
- 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
  - Walker-Warburg syndrome

### 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  
Hypoplasia of eye  
Dysplasia 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 <sup>1</sup>  
                  *Special instructions:*  
                  <sup>1</sup> *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 <sup>1</sup>  
                  Long eyelashes <sup>1</sup>  
                  Weakness of eyelids <sup>1</sup>  
                  Fused eyelids <sup>2</sup>  
                  Absent eyelid  
                  Ankyloblepharon

Symblepharon

Epiblepharon

Ablepharon

*Special instructions:*

<sup>1</sup> Code only if another reportable defect is present.

<sup>2</sup> 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  
                         Small or narrow palpebral fissures  
                         Thin palpebral fissure
- 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
  - Enophthalmia
  - 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 <sup>1</sup>  
                    *Special instructions:*  
                            <sup>1</sup> 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
- 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
- L    \* 744.230    Other misshapen ear  
                    Absent or decreased cartilage <sup>2</sup>  
                    Cauliflower ear <sup>1</sup>  
                    Cleft ear <sup>1</sup>  
                    Crumbled ears  
                    Dysplastic ears

Elfin <sup>1</sup>  
 Hypoplastic ear (not microtia)  
 Lop ear <sup>1</sup>  
 Malformed ear <sup>1</sup>  
 Pinnae folded <sup>1</sup>  
 Pinnae hypoplastic  
 Pixie-like <sup>1</sup>  
 Pointed ear <sup>1</sup>  
 Simplified ear  
 Small ear (not microtia)

*Special instructions:*

<sup>1</sup> Code only if another reportable defect is present.

<sup>2</sup> 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 744.210)

**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
  - Branchial cleft remnant
- 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 unspecified 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
- L    \*    744.880    Other specified anomalies of face/neck
  - Abnormal mouth shape
  - Absent eyebrow
  - Absent face
  - Arched eyebrows
  - Asymmetry mouth
  - Bowed lip
  - Carp shaped mouth
  - Downturned mouth
  - Facial cleft
  - Horner syndrome
  - Hypoplastic angularis oris <sup>1</sup>
  - Hypoplastic cheek
  - Hypoplastic obicularis oris <sup>1</sup>
  - Lip lag <sup>1</sup>
  - Synophrys
  - Tented lip

*Special instructions:*  
<sup>1</sup> 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 <sup>1</sup>
      - Dysmorphic features
      - Low anterior hairline
      - Small face
      - Triangular face
    - Excludes: Low posterior hairline (Use 744.900)

*Special instructions:*

<sup>1</sup> 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  
Absent septum between aorta and pulmonary artery
- 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 745.480)
- 745.120 Corrected transposition of great vessels,  
L-transposition, ventri in version  
Ventricular inversion  
Excludes: Dextrocardia (Use 746.800)
- 745.180 Other specified transposition of great vessels  
Includes: Double outlet right ventricle  
Double outlet left ventricle
- 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 (a.k.a. membranous, perimembraneous, 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: Membranous VSD
  - Perimembranous VSD
  - Muscular VSD
  - Mid-muscular VSD
  - Cystalline VSD
  - Sub-cystalline VSD
  - Type I VSD
  - Type II VSD
  - Apical VSD
  - Septal VSD
  - Malalignment VSD
- 745.490 Ventricular septal defect (VSD), NOS
  - Excludes: Common atrioventricular canal type (Use 745.620)

### 745.5 Ostium secundum type atrial septal defect

- \* 745.500 Nonclosure of foramen ovale, NOS <sup>1</sup>
  - Patent foramen ovale (PFO) <sup>1</sup>
  - Special instructions:*
    - <sup>1</sup> *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 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 <sup>1</sup>
  - PFO vs secundum ASD <sup>1</sup>

*Special instructions:*

<sup>1</sup> *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.*

## **745.6 Endocardial cushion defects**

- \* 745.600 Ostium primum defects  
Primum ASD <sup>1</sup>

*Special instructions:*

<sup>1</sup> *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 <sup>1</sup>  
Includes: Complete AV canal defect <sup>1</sup>

*Special instructions:*

<sup>1</sup> *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  
Small pulmonary valve  
See 746.995 if valve not specified (e.g., "pulmonary stenosis")  
Excludes: Pulmonary infundibular stenosis (Use 746.830)
- # 746.020 Pulmonary valve insufficiency or regurgitation, congenital

- 746.080 Pulmonary insufficiency or regurgitation
- 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, stenosis, or hypoplasia
  - Right atrioventricular (AV) atresia, 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
- # 746.105 Tricuspid valve insufficiency or regurgitation, congenital
  - Right atrioventricular (AV) valve insufficiency or regurgitation, congenital
  - Tricuspid valve incompetence
  - Excludes: Ebstein's anomaly (Use 746.200)
- 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

**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 bicuspid aortic valve  
Aortic valve incompetence
- 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  
Left atrioventricular (AV) valve insufficiency or regurgitation, congenital

#### 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 <sup>1</sup>  
 Cardiomyopathy, congenital  
 Cardiomyopathy, hypertrophic  
 Rhabdomyoma (heart)  
 Ventricular hypertrophy, bilateral <sup>2</sup>  
 Ventricular septal thickening  
 Ventricular septal hypertrophy  
*Special instructions:*  
<sup>1</sup> 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.  
<sup>2</sup> 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 <sup>1</sup>  
*Special instructions:*

- <sup>1</sup> 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) <sup>1</sup>  
*Special instructions:*  
<sup>1</sup> 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
- 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) <sup>1</sup>  
*Special instructions:*  
<sup>1</sup> 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

## 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
- 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 <sup>1</sup>
  - Pseudocoarctation of aorta
  - Elongation of aorta
  - Special instructions:*
    - <sup>1</sup> Code only if another reportable defect is present.
- 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 not 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 <sup>1</sup>

Includes: peripheral pulmonic stenosis (PPS) <sup>1</sup>  
 pulmonary artery branch stenosis <sup>1</sup>  
 peripheral pulmonic stenosis (PPS) murmur <sup>2</sup>

*Special instructions:*

<sup>1</sup> 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.

<sup>2</sup> 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) <sup>1</sup>
- Pulmonary vascular or artery sling

*Special instructions:*

<sup>1</sup> 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
- 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) <sup>1</sup>  
Excludes: Congenital aneurysm:  
Coronary (746.880)  
Peripheral (747.640)  
Pulmonary (747.330)  
Retinal (743.510)  
Ruptured cerebral arteriovenous  
Aneurysm  
Ruptured cerebral aneurysm

*Special instructions:*

<sup>1</sup> 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 <sup>1</sup>  
*Special instructions:*  
<sup>1</sup> *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
  - 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
- \* 748.360 Congenital laryngeal stridor, NOS <sup>1</sup>
  - Special instructions:*
  - <sup>1</sup> *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 <sup>1</sup>
  - Special instructions:*
  - <sup>1</sup> *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

**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**

(If description of condition includes Pierre Robin sequence, use additional code, 524.080)

- 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)
  - 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
      - Absent uvula
    - 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  
 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

- 750.260 Lip fistulae or pits
- 750.270 Other lip anomalies
  - Includes: Notched lip
  - Prominent philtrum
  - Long philtrum
  - Smooth philtrum
  - Smooth lip
  - Anomalies of frenulum of upper lip
  - Excludes: Cleft lip (See 749)
- 750.280 Other specified anomalies of mouth and pharynx
  - Hypertrophy gum
  - Prominent gum
  - 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
- 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
  - 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 Megalogastric
- 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
- # 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
- 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
- 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
- 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)
- 751.495 Malrotation of small intestine alone
  - Malrotation small bowel
  - Midgut malrotation

### 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
- 751.540 Congenital anal fistula
  - Rectal fistula
- 751.550 Persistent cloaca
  - Cloacal extrophy (Also assign code 756.790)
- 751.560 Duodenal web
- \* 751.580 Other specified anomalies of intestine
  - Includes: Rectal fissures <sup>1</sup>
  - Jejunal web

*Special instructions:*

<sup>1</sup> 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
- 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

## 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 <sup>1</sup>  
Hydrocolpos <sup>1</sup>  
Hydrometrocolpos  
*Special instructions:*  
<sup>1</sup> Code only if another reportable defect is present.
- \* 752.440 Absence or other anomaly of vulva  
Fusion of vulva <sup>1</sup>  
Hypoplastic labia majora <sup>2</sup>  
Large labia (majora or minora) <sup>2</sup>  
Prominent labia (majora or minora) <sup>2</sup>  
Absent external female genitalia  
*Special instructions:*  
<sup>1</sup> Code only if another reportable defect is present.  
<sup>2</sup> 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 <sup>1</sup>  
Enlarged clitoris <sup>2</sup>  
Clitoral hypertrophy <sup>2</sup>  
Prominent clitoris <sup>2</sup>  
*Special instructions:*  
<sup>1</sup> 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.

- <sup>2</sup> 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 <sup>1</sup>  
 Hymenal tags <sup>1</sup>  
 Fourchette fistula <sup>1</sup>  
 Doubling of cervix  
 Doubling of vagina  
 Vaginocele
- Special instructions:*  
<sup>1</sup> Code only if another reportable defect is present.
- 752.490 Unspecified anomalies of cervix, vagina, or external female genitalia

### 752.5 Undescended testicle

- Excludes: Retractable testicle
- L \* 752.500 Undescended testicle, unilateral <sup>1</sup>  
 Includes: Cryptorchidism, unilateral <sup>1</sup>  
 Unpalpable testicle, unilateral <sup>1</sup>
- Special instructions:*  
<sup>1</sup> 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 <sup>1</sup>  
 Includes: Cryptorchidism, bilateral <sup>1</sup>  
 Unpalpable testicle, bilateral <sup>1</sup>
- Special instructions:*  
<sup>1</sup> 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 <sup>1</sup>  
 Includes: Cryptorchidism, NOS <sup>1</sup>  
 Unpalpable testicle, NOS <sup>1</sup>
- Special instructions:*  
<sup>1</sup> 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 1°, glandular, coronal  
 Balantic hypospadias  
 Distal hypospadias  
 Mild hypospadias  
 Primary hypospadias

- Subcoronal hypospadias
- Tip hypospadias
- 752.606 2°, penile
- Mid-shaft hypospadias
- Grade II hypospadias
- 752.607 3°, 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 1°, coronal hypospadias
- Balantic hypospadias with chordee
- Subcoronal hypospadias with chordee
- 752.626 Cong. chordee with 2°, penile hypospadias
- 752.627 Cong. chordee with 3°, perineal, scrotal hypospadias

### 752.7 Indeterminate sex and pseudohermaphroditism

Excludes: Pseudohermaphroditism:  
 Female, with adrenocortical disorder (See 255.200)  
 Male, with gonadal disorder with specified chromosomal anomaly (See 758)

- 752.700 True hermaphroditism
- Ovotestis
- 752.710 Pseudohermaphroditism, male
- 752.720 Pseudohermaphroditism, female
- Pure gonadal dysgenesis
- Excludes: gonadal agenesis (758.690)
- 752.730 Pseudohermaphrodite, NOS
- \* 752.790 Indeterminate sex, NOS
- Ambiguous genitalia <sup>1</sup>
- Special instructions:*
- <sup>1</sup> Coding of “ambiguous genitalia” should only be used when no further description is available.
- 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 <sup>1</sup>
- Small scrotum <sup>1</sup>
- Small testicle <sup>1</sup>
- Atrophy testicle
- Underdeveloped/undeveloped scrotum
- Special instructions:*
- <sup>1</sup> Code only if another reportable defect is present.
- 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
- Concealed penis
- Penile torsion
- Disappearing penis syndrome
- Penoscrotal web
- Buried penis
- Adhesions penis
- Penis palmae
- L 752.865 Small penis, hypoplastic penis, or micropenis
- 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
- 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 <sup>1</sup>  
Pelvicaliectasis - NOS  
Pelviectasis  
Pyelectasis  
Dilation of upper renal collecting system  
Dilation of lower renal collecting system  
Dilation of central renal collecting system <sup>1</sup>  
*Special instructions:*  
<sup>1</sup> 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  
Cross fused renal ectopia
- L 753.330 Ectopic kidney  
Malrotated 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 <sup>1</sup>  
            Enlarged renal pelvis - NOS <sup>1</sup>  
            Congenital nephrotic syndrome  
Excludes: Dilation of upper/central/lower renal collecting system (use 753.290)  
*Special instructions:*  
            <sup>1</sup> 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 753.380)
- 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
- 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
- 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  
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 <sup>1</sup>  
Elongated head and skull <sup>1</sup>

*Special instructions:*

<sup>1</sup> 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 <sup>1</sup>  
Flattened side of head  
*Special instructions:*  
<sup>1</sup> Never code in the presence of craniosynostosis. Plagiocephaly is unilateral only. 'Bilateral plagiocephaly' should be coded as brachycephaly (use 754.080). See Appendix D for more information on coding plagiocephaly and related defects.
- 754.055 Asymmetric head  
Asymmetric skull  
Asymmetry calvarium
- \* 754.060 Scaphocephaly, no mention of craniosynostosis (fused sutures) <sup>1</sup>  
*Special instructions:*  
<sup>1</sup> 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 Trionocephaly, no mention of craniosynostosis (fused sutures)  
Triangular head shape
- 754.080 Other specified skull deformity, no mention of craniosynostosis (fused sutures)  
Includes: Brachycephaly  
Acrocephaly  
Turriccephaly  
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
- L # 754.310 Unstable hip
  - Preluxation of hip
  - Subluxation of hip
  - Predislocation status of hip at birth
  - Positive Barlow hip
  - Dislocatable hip
  - Subluxable hip
  - Ortolani positive hip
  - Excludes: Hip click
    - Loose hip
    - Hip laxity
    - Hip crepitus

## 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 or metatarsus adductus
- L 754.530 Complex varus deformities
- L 754.590 Unspecified varus deformities of feet
  - Inversion foot
  - Foot turns inward
  - Adducted foot

Pes varus

**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  
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: Deformed chest  
Deformed thorax  
Barrel chest  
Barrel thorax  
Narrow chest  
Narrow thorax  
Small chest  
Small thorax  
Asymmetric chest  
Asymmetric thorax  
Bell shaped chest  
Bell shaped thorax  
Pectus, NOS

Curved sternum

- 754.825 Shield chest
- 754.825 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 Accessory fingers (postaxial polydactyly, Type A)
- Excludes: Accessory index finger (use 755.010)
- L \* 755.006 Skin tag (postaxial polydactyly, Type B) <sup>1</sup>
- Special instructions:*
- <sup>1</sup> Do not code in Blacks unless another reportable defect is present. Always code for all other races.
- L 755.007 Unspecified finger or skin tag (postaxial polydactyly, NOS)
- Postaxial extra digit of hand
- 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 <sup>1</sup>
- Special instructions:*
- <sup>1</sup> 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: Acrodactyilia  
Arachnodactyly of fingers  
Brachydactyly  
Camptodactyly  
Clenched hand or fist  
Clinodactyly  
Digitalized thumb  
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
		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 <sup>1</sup>
		<i>Special instructions:</i>
		<sup>1</sup> 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
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
  - Flexion deformity toe
  - Hammer toes
  - Hyperextension toe
  - Incurving toes
  - Long 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
  - 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 <sup>1</sup>
  - Anomalies of tibia and fibula
  - Short fibula
  - Short tibia
  - Short lower leg
  - Hypoplastic tibia
  - Hypoplastic fibula
  - Special instructions:*
  - <sup>1</sup> *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
  - Hypoplastic hip, NOS
- L 755.666 Unilateral hip dysplasia
  - Hypoplastic hip, unilateral
- 755.667 Bilateral hip dysplasia
  - Hypoplastic hip, bilateral
- 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 <sup>1</sup>
  - Special instructions:*
  - <sup>1</sup> *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  
Hyperextended joints, NOS  
Short extremity  
Rhizomelia extremity  
Excludes: Hyperextended knees (Use 755.640)  
Overlapping toes (Use 755.600)

### 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
- 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
- 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  
Bathocephaly  
Biparietal narrowing  
Bitemporal narrowing  
Cleft mandible  
Close set eyes  
Flat facial profile  
Flat facies  
Flat midface  
Flat occiput <sup>1</sup>  
Forehead anomalies  
Hypoplastic supraorbital ridges  
Hypotelorism  
Localized skull defects  
Malar hypoplasia  
Maxillary hypoplasia  
Mid-facial hypoplasia  
Pointed chin  
Prominent maxilla  
Prominent occiput <sup>1</sup>  
Short occiput  
Temporal narrowing  
Excludes: Macrocephaly (Use 742.400)  
Small chin (See 524.0)  
Pierre Robin sequence (Use 524.080)
- Special instructions:*  
<sup>1</sup> 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)
  - Bifid vertebrae (cervical)
  - Butterfly vertebrae (cervical)
  - Cleft vertebrae (cervical)
  - Spinal dysraphism (cervical) <sup>1</sup>
  - Special instructions:*
    - <sup>1</sup> *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) <sup>1</sup>
  - Special instructions:*
    - <sup>1</sup> *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) <sup>1</sup>

*Special instructions:*

<sup>1</sup> *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) <sup>1</sup>
  - Includes: Agenesis of sacrum
  - Excludes: Pilonidal sinus (See 685.100)

*Special instructions:*

<sup>1</sup> *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 <sup>1</sup>
  - Platyspondyly

*Special instructions:*

<sup>1</sup> *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
- 756.445 Diastrophic dwarfism
- 756.446 Metatrophic dwarfism
- 756.447 Thanatophoric dwarfism
- 756.450 Metaphyseal dysostosis
- 756.460 Spondyloepiphyseal dysplasia
- 756.470 Exostosis
  - Excludes: Gardner syndrome (See 759.630)
- 756.480 Other specified chondrodystrophy
  - Achondrogenesis type I
  - Achondrogenesis type II
  - Camptomelic 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: Lipochoondrodystrophy (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
- 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
- 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

Cloacal extrophy (Also assign code 751.550)  
756.795 Epigastric 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  
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  
Jacobsen 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
- 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 216.900)  
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
- 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

### 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) <sup>1</sup>  
Special instructions:  
<sup>1</sup> *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 (758.380)  
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
- 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
- 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  
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
- 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 <sup>1</sup>  
*Special instructions:*  
<sup>1</sup> 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  
Splénomegaly  
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
  - 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 <sup>1</sup>
  - Absent thymus
  - Hypoplastic thymus
  - Small thymus
  - Enlarged thymus <sup>1</sup>

*Special instructions:*  
<sup>1</sup> 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 <sup>1</sup>
  - Situs ambiguous <sup>1</sup>
  - Heterotaxy syndrome <sup>1</sup>
  - Excludes: Dextrocardia not associated with complete situs inversus (746.800)

*Special instructions:*  
<sup>1</sup> 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 Tuberos sclerosi**

- 759.500 Tuberos sclerosi  
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  
 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)  
 Oto-palato-digital syndrome  
 Rieger syndrome  
 Smith-Magenis syndrome  
 Waardenburg syndrome  
 Whistling face syndrome (Freeman Sheldon syndrome)  
 Williams syndrome  
 XK aprosencephaly  
 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
  - Schinz-Giedion syndrome
  - Stickler syndrome
- 759.870 Cong malformation syndromes with metabolic disturbances
  - Alagille syndrome
  - Alport 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
  - 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

## OTHER SPECIFIED CODES

- # 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 <sup>1</sup>  
*Special instructions:*  
<sup>1</sup> 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 <sup>1</sup>  
*Special instructions:*  
<sup>1</sup> 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 <sup>1</sup>  
*Special instructions:*  
<sup>1</sup> 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 <sup>1</sup>  
*Special instructions:*  
<sup>1</sup> Always code regardless of size, type, or number.
- L \* 228.030 Hemangioma, retinal <sup>1</sup>  
*Special instructions:*  
<sup>1</sup> Always code regardless of size, type, or number.
- L \* 228.040 Hemangioma, intra-abdominal <sup>1</sup>  
Hemangioendothelioma liver <sup>1</sup>  
*Special instructions:*  
<sup>1</sup> Always code regardless of size, type, or number.
- L \* 228.090 Hemangioma, of other sites <sup>1</sup>  
*Special instructions:*  
<sup>1</sup> Always code regardless of size, type, or number.
- 228.100 Cystic hygroma
- Lymphangioma, any site
- 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: Lipochoondrodystrophy
# 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 Sphrintzen 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 Crossed eyes
	#	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 <sup>1</sup> <i>Special instructions:</i> <sup>1</sup> <i>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.</i>
L	*	550.000	Inguinal hernia with mention of gangrene <sup>1</sup> <i>Special instructions:</i>

<sup>1</sup> 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 <sup>1</sup>

Special instructions:

<sup>1</sup> 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 <sup>1</sup>

Special instructions:

<sup>1</sup> 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

# 771.280 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.

Abduction hip  
Absent right superior vena cava  
Acne, neonatal  
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

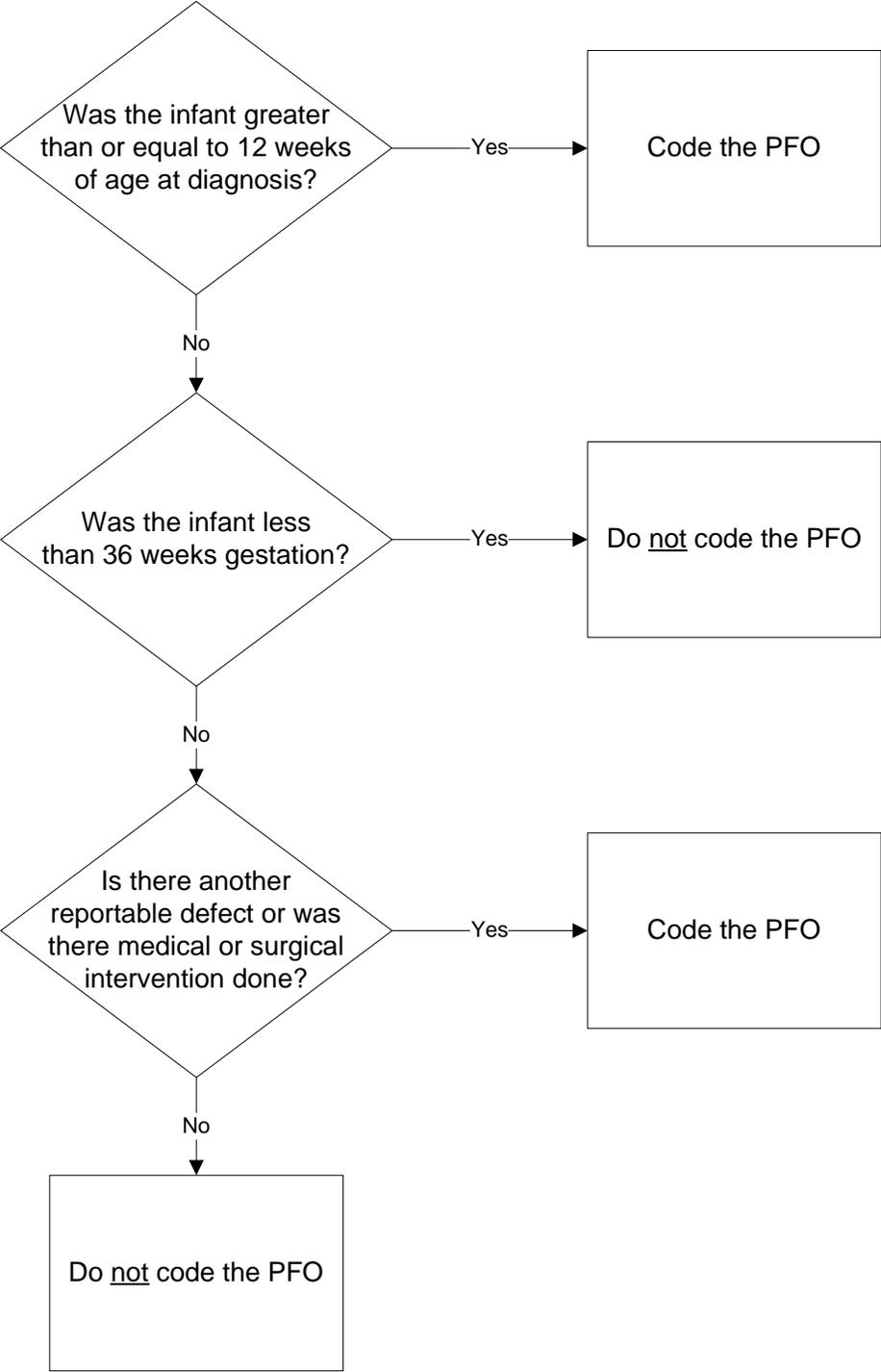
Cupped tongue  
Cutis marmorata  
Cyst, arachnoid  
Cyst, choroid plexus, unilateral  
Cyst, gum  
Cyst, tongue  
Cyst, Wharton duct  
Diaphragm, elevated  
Diastasis recti  
Dilatation/dilated/dilation atrium  
Dilatation/dilated/dilation ventricle (heart)  
Dimple, chin  
Dislocation, shoulder  
Duodenal 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  
Gastric volvulus  
Gastroesophageal reflux (GER)  
Gluteal cleft, asymmetric  
Gum cyst  
Gum, hypoplastic  
Hand, hyperflexion  
Hand, narrow  
Heart murmur  
Hemorrhage, central nervous system (CNS)  
Hepatic vein occlusion (Budd-Chiari)  
Hip, abduction  
Hip click  
Hip crepitus  
Hip laxity  
Hip, loose

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  
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  
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  
Murmur, heart  
Myopia  
Nails, long  
Nails, meconium stained  
Nails, short  
Narrow hand  
Nasal obstruction  
Neonatal acne  
Neurogenic bladder  
Nevus flammeus  
Nipple, inverted  
Obstruction, biliary  
Obstruction, duodenal  
Obstruction, ileal

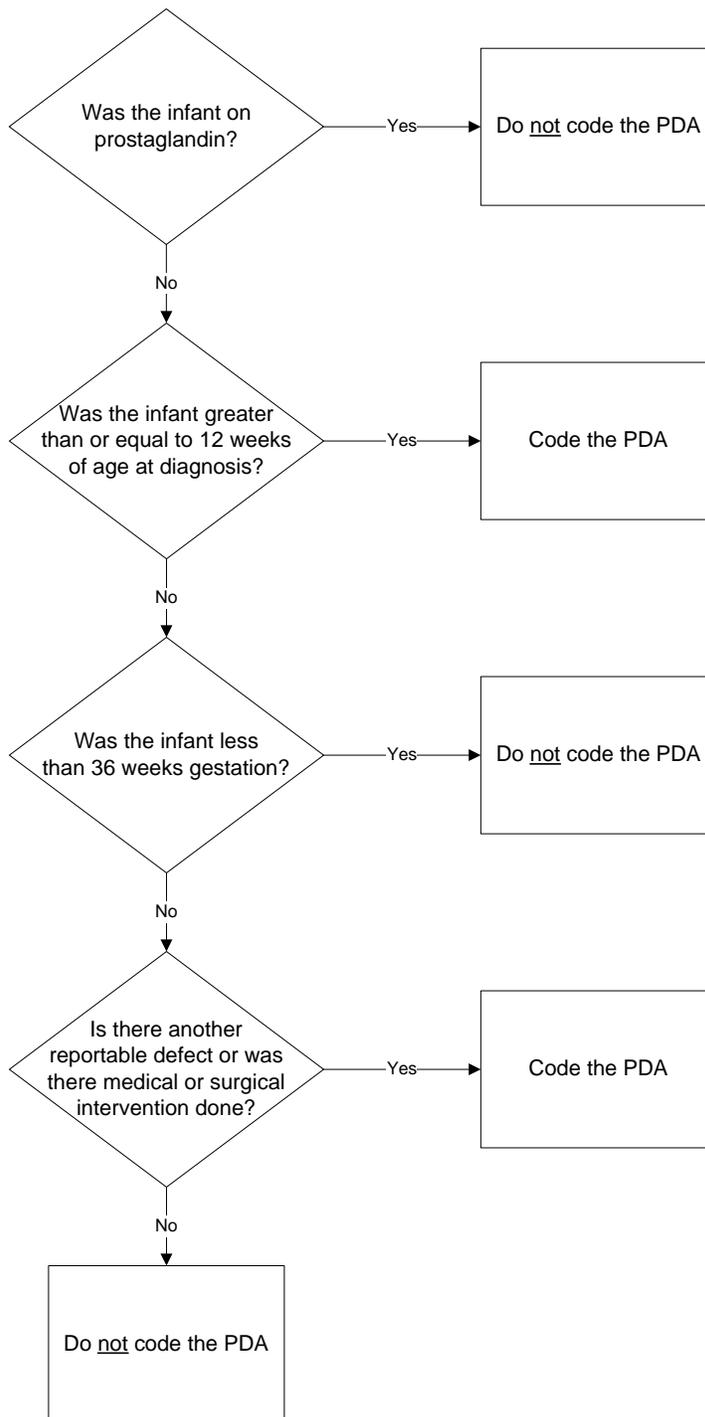
Obstruction, intestinal  
Obstruction, jejunal  
Obstruction, nasal  
Ondine's Curse syndrome  
Osteopenia  
Osteoporosis  
Overlapping sutures  
Overriding sutures  
Palpebral fissure, thick  
Paralysis, vocal cord  
Perforation, bowel  
Perforation, colon  
Perforation, duodenum  
Perforation, ileum  
Perforation, intestine  
Perforation, small intestine  
Petechiae  
Phimosis  
Pneumothorax  
Premature atrial contractions (PACs)  
Primary pulmonary artery hypertension  
Prominent prepuce of clitoris  
Prominent tongue  
Prominent xiphoid 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 pulmonary artery  
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  
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

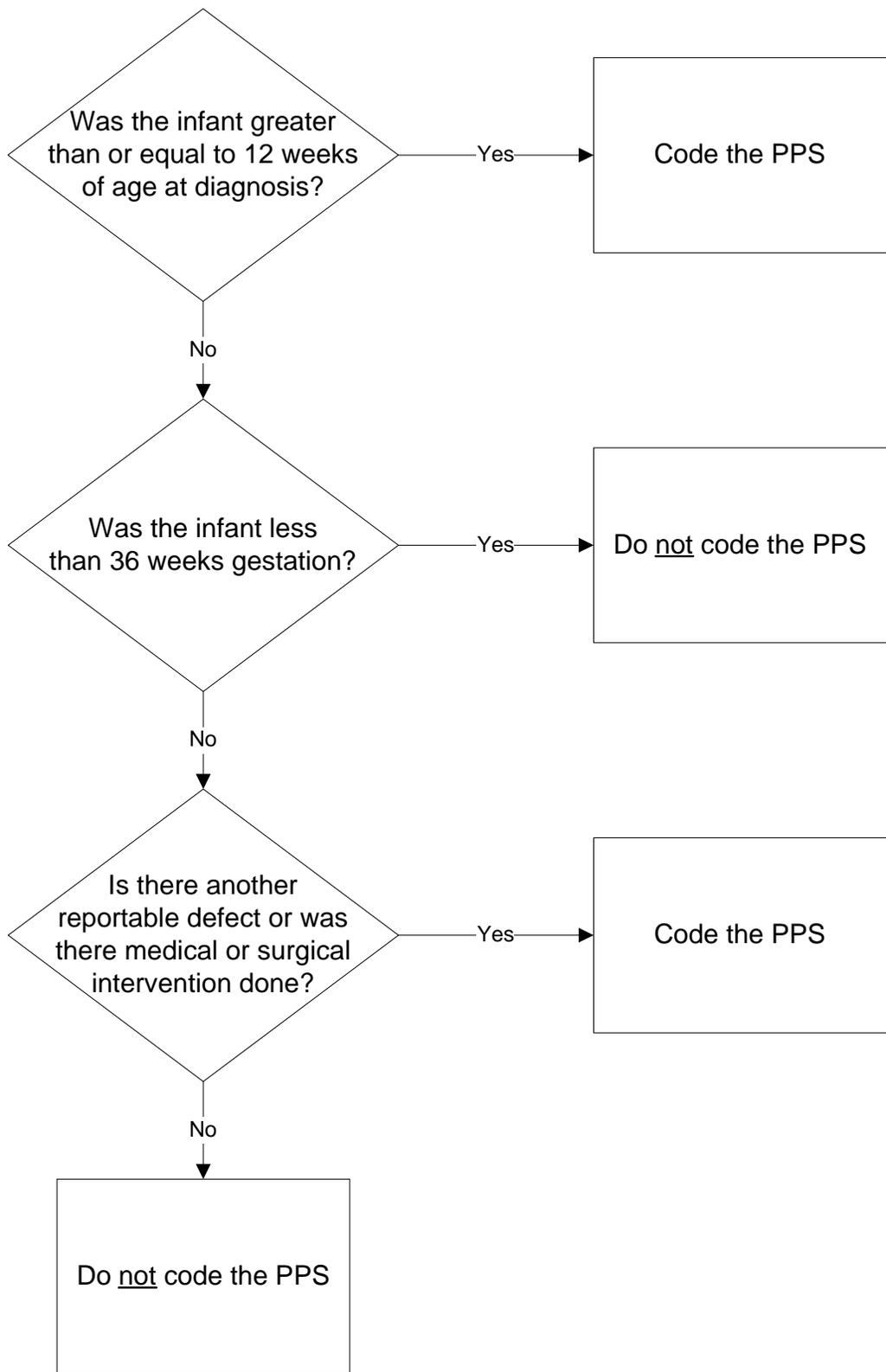
**APPENDIX A: FLOWCHART FOR DECIDING WHEN TO CODE PATENT FORAMEN OVALE (PFO)**



## 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?
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
Forehead bossing	Yes	No
Occiput flattening and laterality	Yes	No
Open/closed fontanelle	Yes, if stated abnormal	Yes, if stated abnormal
Posterior towering	Yes	No
Scaphocephaly	Yes	Yes, unless synostosis is diagnosed
Synostosis	Yes	Yes
Temporal/parietal bulging	Yes	Yes
Torticollis and laterality	Yes	Yes