

BIRTH DEFECTS AND GENETIC DISEASES BRANCH 6-DIGIT CODE

For Reportable Congenital Anomalies

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

Code modifications developed by Division of Birth Defects and Developmental Disabilities, National Center on Birth Defects and Developmental Disabilities, Centers for Disease Control and Prevention, Public Health Service, U.S. Department of Health and Human Services, Atlanta, Georgia 30333

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Version 08/07

Replaces Versions 06/04, 05/07, and 06/07

## Explanation of 6-Digit Code

### 6th Digit Code - Master

- .000 Blank
- .001 Left Only
- .002 Right Only
- .003 Unilateral Unspecified
- .004 Bilateral
- .005
- .006
- .007
- .008 Possible, Probable, Borderline, or Rule Out;  
Defects only diagnosed prenatally should be coded with the last digit 8  
when the prenatal diagnosis is not definitive.
- .009 Not Otherwise Specified (NOS)

### Notes:

An asterisk (\*) beside a disease code indicates that the code was created by CDC.

A pound symbol (#) beside a disease code indicates that the condition or defect is listed on the MACDP Exclusion List.

A check (T) beside a disease code indicates that an addition/revision was made since the last printing of the Procedure Manual. Use of the code should be according to the exclusion list criteria.

The abbreviations NEC and NOS used in this code are defined as not elsewhere classified and as not otherwise specified, respectively.

## CONGENITAL ANOMALIES

### **Anencephalus and Similar Anomalies**

#### **740.0 Anencephalus**

- 740.000 Absence of brain
- 740.010 Acrania
- 740.020 Anencephaly
- 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  
Arnold-Chiari malformation, NOS
- 741.020 Spina bifida cystica, any site, with stenosed aqueduct of Sylvius
- 741.030 Spina bifida cystica, cervical, with unspecified hydrocephalus  
Spina bifida cystica, cervical, with hydrocephalus but without mention of Arnold-Chiari malformation or aqueduct stenosis
- 741.040 Spina bifida cystica, thoracic, with unspecified hydrocephalus, no mention of Arnold-Chiari
- 741.050 Spina bifida cystica, lumbar, with unspecified hydrocephalus, no mention of Arnold-Chiari
- 741.060 Spina bifida cystica, sacral, with unspecified hydrocephalus, no mention of Arnold-Chiari
- 741.070 Spina bifida of any site with hydrocephalus of late onset

- 741.080 Other spina bifida, meningocele of specified site with hydrocephalus
- 741.085 Spina bifida, meningocele, cervicothoracic, with hydrocephalus
- 741.086 Spina bifida, meningocele thoracolumbar, with hydrocephalus
- 741.087 Spina bifida, meningocele, lumbosacral with hydrocephalus
- 741.090 Spina bifida of any unspecified type with hydrocephalus

**741.9 Spina bifida without mention of hydrocephalus**

- 741.900 Spina bifida (aperta), without hydrocephalus
- 741.910 Spina bifida (cystica), cervical, without hydrocephalus
- 741.920 Spina bifida (cystica), thoracic, without hydrocephalus
- 741.930 Spina bifida (cystica), lumbar, without hydrocephalus
- 741.940 Spina bifida (cystica), sacral, without hydrocephalus
- 741.980 Spina bifida, other specified site, without hydrocephalus  
Includes: cervicothoracic, thoracolumbar, lumbosacral
- 741.985 Lipomyelomeningocele
- 741.990 Spina bifida, site unspecified, without hydrocephalus (myelocoele, myelomeningocele, meningomyelocoele)

**742 Other Congenital Anomalies of Nervous System**

**742.0 Encephalocele**

- 742.000 Occipital encephalocele
- 742.080 Other encephalocele of specified site (includes midline defects)
- 742.085 Frontal encephalocele
- 742.086 Parietal encephalocele
- 742.090 Unspecified encephalocele

**742.1 Microcephalus**

- 742.100 Microcephalus

**742.2 Reduction deformities of brain**

- 742.200 Anomalies of cerebrum
- 742.210 Anomalies of corpus callosum
- 742.220 Anomalies of hypothalamus
- 742.230 Anomalies of cerebellum
- 742.240 Agyria and lissencephaly
- 742.250 Microgyria, polymicrogyria
- 742.260 Holoprosencephaly
- 742.270 Arrhinencephaly
- 742.280 Other specified reduction defect of brain
- 742.290 Unspecified reduction defect of brain

**742.3 Congenital hydrocephalus**

Excludes: hydrocephalus with any condition in 741.9 (use 741.0)

- 742.300 Anomalies of aqueduct of Sylvius
- 742.310 Atresia of foramina of Magendie and Luschka

- Dandy-Walker syndrome
- 742.320 Hydranencephaly
- 742.380 Other specified hydrocephaly
  - Includes: communicating hydrocephaly
- # 742.385 Hydrocephalus secondary to intraventricular hemorrhage (IVH) or CNS bleed
- 742.390 Unspecified hydrocephaly, NOS

**742.4 Other specified anomalies of brain**

- 742.400 Enlarged brain and/or head
  - megalencephaly
  - macrocephaly
- 742.410 Porencephaly
  - Includes: porencephalic cysts
- 742.420 Cerebral cysts
- 742.480 Other specified anomalies of brain
  - Includes: cortical atrophy
  - cranial nerve defects
- 742.485 Ventricular cysts
  - Excludes: arachnoid cysts
- 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
- 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

**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**

- 743.000 Anophthalmos  
agenesis of eye  
cryptophthalmos
- 743.100 Microphthalmos, small eyes  
aplasia of eye  
hypoplasia of eye  
dysplasia of eye  
rudimentary eye

**743.2 Buphthalmos**

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

**743.3 Congenital cataract and lens anomalies**

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

**743.4 Coloboma and other anomalies of anterior segments**

- 743.400 Corneal opacity
- 743.410 Other corneal anomalies  
Excludes: megalocornea (use 743.220)
- 743.420 Absence of iris  
aniridia
- 743.430 Coloboma of iris
- 743.440 Other anomalies of iris  
polycoria  
ectopic pupil  
Peter's anomaly  
# Excludes: brushfield spots (use 743.800)
- 743.450 Blue sclera  
# If <36 weeks gestation, code only if another reportable defect is present.  
Always code if ≥36 weeks gestation.
- 743.480 Other specified colobomas and anomalies of anterior segments  
Rieger's anomaly
- 743.490 Unspecified colobomas and anomalies of anterior eye segments

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**743.5 Congenital anomalies of posterior segment**

- 743.500 Specified anomalies of vitreous humour
- 743.510 Specified anomalies of retina  
congenital retinal aneurysm  
Excludes: Stickler syndrome (use 759.860)
- 743.520 Specified anomalies of optic disc  
hypoplastic optic nerve  
coloboma of the optic disc
- 743.530 Specified anomalies of choroid
- 743.535 Coloboma of choroid
- 743.580 Other specified anomalies of posterior segment of eye
- 743.590 Unspecified anomalies of posterior segment of eye

**743.6 Congenital anomalies of eyelids, lacrimal system, and orbit**

- 743.600 Blepharoptosis  
congenital ptosis
- 743.610 Ectropion
- 743.620 Entropion
- # 743.630 Other anomalies of eyelids  
absence of eyelashes  
long eyelashes  
weakness of eyelids
- T # fused eyelids (exclude if <25 weeks gestation unless another reportable defect is present)
- 743.635 Blepharophimosis  
small or narrow palpebral fissures
- 743.636 Coloboma of the eyelids
- 743.640 Absence or agenesis of lacrimal apparatus  
absence of punctum lacrimale
- # 743.650 Stenosis or stricture of lacrimal duct
- 743.660 Other anomalies of lacrimal apparatus (e.g., cyst)
- 743.670 Anomalies of orbit

**743.8 Other specified anomalies of eye**

- # 743.800 Other specified anomalies of eye  
Includes: exophthalmos  
epicanthal folds  
antimongoloid slant  
upward eye slant  
Brushfield spots  
Excludes: congenital nystagmus (use 379.500)  
retinitis pigmentosa (use 362.700)  
ocular albinism (use 270.200)  
wide spaced eyes, hypertelorism (use 756.085)
- \* 743.810 Epibulbar dermoid cyst

**743.9 Unspecified anomalies of eye**

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

**744 Congenital Anomalies of Ear, Face, and Neck**

**744.0 Anomalies of ear causing impairment of hearing**

- 744.000 Absence or stricture of auditory canal
- 744.010 Absence of auricle (pinna)  
absence of ear, NOS
- 744.020 Anomaly of middle ear  
fusion of ossicles
- 744.030 Anomaly of inner ear  
Includes: congenital anomaly of membranous  
labyrinth organ of Corti
- 744.090 Unspecified anomalies of ear with hearing impairment  
Includes: congenital deafness, NOS

**744.1 Accessory auricle**

- # 744.100 Accessory auricle  
Polyotia
- # 744.110 Preauricular appendage, tag, or lobule  
(in front of ear canal)
- # 744.120 Other appendage, tag, or lobule include papillomas,  
ear tags

**744.2 Other specified anomalies of ear**

- 744.200 Macrotia (enlarged pinna)
- 744.210 Microtia (hypoplastic pinna and absence or  
stricture of external auditory meatus)
- 744.220 Bat ear
- T # 744.230 Other misshapen ear  
pointed ear  
elfin  
pixie-like  
lop ear  
cauliflower ear  
cleft in ear  
malformed ear  
absent or decreased cartilage
- 744.240 Misplaced ears
- # 744.245 Low set ears
- # 744.246 Posteriorly rotated ears
- 744.250 Absence or anomaly of eustachian tube
- 744.280 Other specified anomalies of ear (see also 744.230)
- # Excludes: Darwin's tubercle

**744.3 Unspecified anomalies of ear**

- 744.300 Unspecified anomalies of ear  
Congenital: ear (any part)  
anomaly, deformity, NOS

**744.4 Branchial cleft, cyst, or fistula; preauricular sinus**

- 744.400 Branchial cleft, sinus, fistula cyst, or pit
- # 744.410 Preauricular sinus, cyst, or pit

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- 744.480 Other branchial cleft anomalies  
Includes: dermal sinus of head
- # 744.500 Webbing of neck  
Includes: pterygium colli,  
redundant neck skin folds

**744.8 Other unspecified anomalies of face and neck**

- 744.800 Macrostomia (large mouth)
- 744.810 Microstomia (small mouth)
- # 744.820 Macrocheilia (large lips)
- # 744.830 Microcheilia (small lips)
- 744.880 Other specified anomalies of face/neck

**744.9 Unspecified anomalies of face and neck**

- # 744.900 Congenital anomaly of neck, NOS  
Includes: short neck
- 744.910 Congenital anomaly of face, NOS  
Abnormal facies

**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)
- 745.110 Transposition of great vessels, incomplete (w/ VSD)  
Taussig-Bing syndrome
- 745.120 Corrected transposition of great vessels,  
L-transposition, ventri in version  
Excludes: dextrocardia (use 746.800)
- N 745.130 Double outlet right ventricle (DORV) with normally  
related great vessels
- N 745.140 Double outlet right ventricle (DORV) with transposed  
great vessels
- N 745.150 Double outlet right ventricle (DORV), relationship of great  
vessels not specified
- N 745.180 Other specified transposition of great vessels,  
no mention of double outlet right ventricle (DORV)
- 745.190 Unspecified transposition of great vessels

**745.2 Tetralogy of Fallot**

- 745.200 Fallot's tetralogy
- 745.210 Fallot's pentalogy  
Fallot's tetralogy plus ASD

**745.3 Single ventricle**

- 745.300 Single ventricle  
Common ventricle  
Cor triloculare biatriatum

**745.4 Ventricular septal defect**

- N 745.400 Roger's disease  
Note: This is an outdated term and the code is no longer  
used. If this diagnostic term is encountered in  
the medical record, code it as a ventricular septal  
defect.
- 745.410 Eisenmenger's syndrome
- 745.420 Gerbode defect
- T 745.480 Other specified ventricular septal defect  
Includes: crystalline  
sub-crystalline  
subarterial  
conoventricular
- N 745.485 Perimembranous VSD  
Includes: membranous VSD

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- N 745.486 Muscular VSD  
Includes: mid-muscular and apical VSDs
- N 745.487 Inlet VSD  
Includes: common atrioventricular (AV) canal type VSD  
Note: Code common atrioventricular (AV) canal as  
745.630  
Code common atrioventricular (AV) canal with  
muscular VSD as 745.620
- 745.490 Ventricular septal defect, NOS  
Excludes: common atrioventricular canal type (use  
745.620)
- 745.498 Probable VSD

**745.5 Ostium secundum type atrial septal defect**

- N # 745.500 Nonclosure of foramen ovale, NOS  
Patent foramen ovale (PFO)  
1) Always code if  $\geq 36$  weeks of gestation at birth and defect  
last noted at  $\geq 6$  weeks of age.  
2) If  $\geq 36$  weeks gestation at birth and defect last noted  
< 6 weeks of age, code only if another reportable heart  
defect is present.  
3) Never code if < 36 weeks gestation at birth regardless  
of presence of other defects.
- S 745.510 Ostium (septum) secundum defect  
Note: If the defect size by echo is  $\leq 4$ mm, assume it  
is a PFO and follow the coding instructions for  
745.500, even if the record says secundum ASD.
- N 745.520 Lutembacher's syndrome  
Note: This is an outdated term and the code is no  
longer used. If this diagnostic term is  
encountered in the medical record, code the  
individual components, not the syndrome.
- S 745.570 PFO vs. ASD  
1) If the defect size by echo is  $\leq 4$ mm, assume it is a  
PFO and follow the coding instructions for 745.500.  
2) If the defect size by echo is  $> 4$ mm, assume it is an  
atrial septal defect and code as 745.590 ASD, NOS.  
3) If an echo is done but the defect size is not stated,  
assume it is a PFO and follow the coding instructions  
for 745.500.  
4) If unable to determine the appropriate code based on  
above criteria, use code 745.570.
- 745.580 Other specified atrial septal defect
- S 745.590 ASD (atrial or auricular septal defect), NOS  
Excludes: PFO vs. ASD (see 745.570).  
Note: If the defect size by echo is  $\leq 4$ mm, assume it  
is a PFO and follow the coding instructions for  
745.500, even if the record says ASD.

**745.6 Endocardial cushion defects**

- 745.600 Ostium primum defects
- 745.610 Single common atrium, cor triloculare biventriculare
- N 745.620 Common atrioventricular canal with ventricular  
septal defect (VSD)  
Includes: Common AV canal with muscular VSD

Excludes: Inlet VSD or common AV canal type VSD (code as 745.487)

745.630 Common atrioventricular canal  
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

**746 Other Congenital Anomalies of Heart**

**746.0 Anomalies of pulmonary valve**

N 746.000 Atresia, hypoplasia of pulmonary valve  
Note: Code pulmonary artery atresia as 747.300  
Code pulmonary artery hypoplasia as 747.380  
Code "pulmonic" or "pulmonary" atresia or  
hypoplasia, NOS (no mention of valve or  
artery) as 746.995

N # 746.010 Stenosis of pulmonary valve  
# Excludes: pulmonary infundibular  
stenosis (use 746.830)  
Note: Code pulmonary artery stenosis as 747.320  
Code "pulmonic" or "pulmonary" stenosis, NOS (no  
mention of valve or artery) as 746.995

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

# 746.080 Other specified anomalies of pulmonary valve  
# Excludes: pulmonary infundibular  
stenosis (use 746.830)

746.090 Unspecified anomaly of pulmonary valve

**746.1 Tricuspid atresia and stenosis**

N 746.100 Tricuspid atresia only  
Excludes: tricuspid stenosis and hypoplasia

N # 746.105 Tricuspid valve insufficiency or regurgitation,  
congenital  
Never code cases designated as 'mild', 'minimal', 'trivial',  
or 'physiologic'.  
Code cases designated as 'moderate' or 'severe' and those

- where the degree is not specified (NOS) only if another reportable heart defect is present.
- N 746.106 Tricuspid stenosis or hypoplasia
- 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  
Excludes: supravalvular aortic stenosis (747.220)
- 746.4 Congenital insufficiency of aortic valve**
- N # 746.400 Aortic valve insufficiency or regurgitation, congenital  
Excludes: bicuspid aortic valve.  
Never code cases designated as 'mild', 'minimal', 'trivial', or 'physiologic'.  
Code cases designated as 'moderate' or 'severe' and those where the degree is not specified (NOS) only if another reportable heart defect is present.
- N 746.470 Bicuspid aortic valve
- \* 746.480 Other specified anomalies of the aortic valves  
Includes: aortic valve atresia  
Excludes: supravalvular aortic stenosis (747.220)
- \* 746.490 Unspecified anomalies of the aortic valves
- 746.5 Congenital mitral stenosis**
- 746.500 Congenital mitral stenosis  
746.505 Absence, atresia, or hypoplasia of mitral valve
- 746.6 Mitral valve insufficiency or regurgitation, congenital**
- N # 746.600 Mitral valve insufficiency or regurgitation, congenital  
Never code cases designated as 'mild', 'minimal', 'trivial', or 'physiologic'.  
Code cases designated as 'moderate' or 'severe' and those where the degree is not specified (NOS) only if another reportable heart defect is present.
- 746.7 Hypoplastic left heart syndrome**
- 746.700 Hypoplastic left heart syndrome  
Atresia, or marked hypoplasia of the ascending aorta and defective development of left ventricle (with mitral valve atresia)
- 746.8 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)

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- N 746.810 Levocardia  
 Note: This condition has been moved to the never code list.
- 746.820 Cor triatriatum  
 746.830 Pulmonary infundibular (subvalvular) stenosis  
 746.840 Trilogy of Fallot  
 746.850 Anomalies of pericardium
- N # 746.860 Anomalies of myocardium  
 cardiomegaly, congenital, NOS  
 cardiomyopathy, congenital  
 cardiomyopathy, hypertrophic  
 Note: Do not code cardiomyopathy of any type in a newborn of a diabetic mother (either gestational or pre-existing diabetes).
- 746.870 Congenital heart block
- 746.880 Other specified anomalies of heart  
 Includes: ectopia (ectopic) cordis (mesocardia),  
 conduction defects, NOS
- 746.881 Hypoplastic left ventricle  
 Excludes: hypoplastic left heart syndrome (746.700)
- 746.882 Hypoplastic right heart (ventricle)  
 Uhl's disease
- \* 746.883 Hypoplastic ventricle, NOS
- 746.885 Anomalies of coronary artery or sinus
- N 746.886 Ventricular hypertrophy (right or left)  
 Note: Do not code ventricular hypertrophy of any type in a newborn of a diabetic mother (either gestational or pre-existing diabetes).
- 746.887 Other defects of the atria  
 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  
 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)
- N 746.995 "Pulmonic" or "pulmonary" atresia, stenosis, or hypoplasia, NOS (no mention of valve or artery)  
 Note: Code pulmonary valve atresia or hypoplasia as 746.000  
 Code pulmonary valve stenosis as 746.010  
 Code pulmonary artery atresia as 747.300  
 Code pulmonary artery stenosis as 747.320  
 Code pulmonary artery hypoplasia as 747.380

#### 747 Other Congenital Anomalies of Circulatory System

- N # 747.000 Patent ductus arteriosus (PDA)  
 Note: 1) Always code if  $\geq 36$  weeks of gestation at birth and defect last noted at  $\geq 6$  weeks of age.

2) If  $\geq 36$  weeks gestation at birth and defect last noted  $< 6$  weeks of age, code only if the PDA was treated (e.g. by ligation or indomethacin) or if another reportable heart defect is present.

3) Never code if  $< 36$  weeks gestation at birth or if treated with prostaglandins regardless of gestational age. (See PDA Tree Appendix)

747.008 Probable 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

#### 747.2 Other anomalies of aorta

747.200 Atresia of aorta  
absence of aorta  
pseudotruncus arteriosus

747.210 Hypoplasia of aorta  
tubular hypoplasia of aorta

N 747.215 Interrupted aortic arch, Type A

N 747.216 Interrupted aortic arch, Type B

N 747.217 Interrupted aortic arch, Type C

747.220 Supra-aortic stenosis (supravalvular)  
Excludes: aortic 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

747.270 Congenital aneurysm of aorta  
congenital dilatation of aorta

747.280 Other specified anomalies of aorta

N 747.285 Interrupted aortic arch, NOS, type not specified

747.290 Unspecified anomalies of aorta

#### 747.3 Anomalies of pulmonary artery

N 747.300 Pulmonary artery atresia, absence or agenesis  
Note: Code pulmonary valve atresia as 746.000  
Code "pulmonic" or "pulmonary" atresia, NOS (no  
mention of valve or artery) as 746.995

747.310 Pulmonary artery atresia with septal defect

N 747.320 Pulmonary artery stenosis  
Includes: Stenosis of the main pulmonary artery or of  
the right or left main branches  
Note: Code pulmonary valve stenosis as 746.010  
Code "pulmonic" or "pulmonary" stenosis, NOS (no  
mention of valve or artery) as 746.995

N 747.325 Peripheral pulmonary artery stenosis  
Includes: Stenosis of a pulmonary artery peripheral to

the main right or left main branches  
Peripheral pulmonic stenosis (PPS), NOS,  
documented by echocardiogram

- # Excludes: Peripheral pulmonic stenosis (PPS) murmur only  
(not documented by echocardiogram)  
Note: 1) Always code if  $\geq 36$  weeks of gestation at birth and  
defect last noted at  $\geq 6$  weeks of age.  
2) If  $\geq 36$  weeks gestation at birth and defect last noted  
< 6 weeks of age, code only if another reportable heart  
defect is present.  
3) Never code if < 36 weeks gestation at birth.  
(See PPS Tree Appendix)

- 747.330 Aneurysm of pulmonary artery  
dilatation of pulmonary artery  
747.340 Pulmonary arteriovenous malformation or aneurysm  
747.380 Other specified anomaly of pulmonary artery  
Includes: pulmonary artery hypoplasia  
Note: Code pulmonary valve hypoplasia as 746.000  
Code "pulmonic" or "pulmonary" hypoplasia, NOS  
(no mention of valve or artery) as 746.995  
747.390 Unspecified anomaly of pulmonary artery

#### 747.4 Anomalies of great veins

- 747.400 Stenosis of vena cava (inferior or superior)  
747.410 Persistent left 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  
747.490 Unspecified anomalies of great veins

#### 747.5 Absence or hypoplasia of umbilical artery

- # 747.500 Single umbilical artery

#### 747.6 Other anomalies of peripheral vascular system

- 747.600 Stenosis of renal artery  
747.610 Other anomalies of renal artery  
747.620 Arteriovenous malformation (peripheral)  
Excludes: pulmonary (747.340)  
cerebral (747.800)  
retinal (743.510)  
747.630 Congenital phlebectasia  
congenital varix  
747.640 Other anomalies of peripheral arteries  
Includes: aberrant subclavian artery  
747.650 Other anomalies of peripheral veins  
Excludes: Budd-Chiari - occlusion of hepatic vein (use  
453.000)  
N 747.680 Other anomalies of peripheral vascular system  
# Includes: primary pulmonary artery hypertension ONLY if  
it is present in an infant at  $\geq 7$  days of age  
747.690 Unspecified anomalies of peripheral vascular system

**747.8 Other specified anomalies of circulatory system**

- 747.800 Arteriovenous (malformation) aneurysm of brain
- 747.810 Other anomalies of cerebral vessels
  - Includes: vein of Galen
- 747.880 Other specified anomalies of circulatory system
  - Excludes: congenital aneurysm:
    - coronary (746.880)
    - peripheral (747.640)
    - pulmonary (747.330)
    - retinal (743.510)
    - ruptured cerebral arteriovenous aneurysm (430.000)
    - ruptured cerebral aneurysm (430.000)

**747.9 Unspecified anomalies of circulatory system**

- 747.900 Unspecified anomalies of circulatory system

**748 Congenital Anomalies of Respiratory System**

**748.0 Choanal atresia**

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

**748.1 Other anomalies of nose**

748.100 Agenesis or underdevelopment of nose  
748.110 Accessory nose  
748.120 Fissured, notched, or cleft nose  
748.130 Sinus wall anomalies  
748.140 Perforated nasal septum  
# 748.180 Other specified anomalies of nose  
flat bridge of nose  
wide nasal bridge  
small nose and nostril  
absent nasal septum  
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  
T 748.310 Congenital subglottic stenosis - Never code if chart states  
the condition was acquired or secondary to endotracheal (ET)  
intubation or ventilation  
748.330 Other anomalies of trachea  
# Excludes: vascular ring compression of the  
trachea (use 747.250)  
748.340 Stenosis of bronchus  
748.350 Other anomalies of bronchus  
748.360 Congenital laryngeal stridor, NOS  
748.380 Other specified anomalies of larynx and bronchus  
748.385 Cleft larynx, laryngotracheoesophageal cleft  
748.390 Unspecified anomalies of larynx, trachea, and bronchus

**748.4 Congenital cystic lung**

748.400 Single cyst, lung or lung cyst  
748.410 Multiple cysts, lung  
Polycystic lung  
748.420 Honeycomb lung  
748.480 Other specified congenital cystic lung

**748.5 Agenesis or aplasia of lung**

- 748.500 Agenesis or aplasia of lung
- T 748.510 Hypoplasia of lung; Pulmonary hypoplasia  
# Exclude if isolated defect in infants <36 weeks gestation.
- 748.520 Sequestration of lung
- 748.580 Other specified dysplasia of lung  
Fusion of lobes of lung
- \* 748.590 Unspecified dysplasia of lung

**748.6 Other anomalies of lung**

- 748.600 Ectopic tissues in lung
- 748.610 Bronchiectasis
- 748.620 Accessory lobe of lung
- 748.625 Bilobar right lung or right lung with left lung bronchial  
pattern
- 748.690 Other and unspecified anomalies of lung

**748.8 Other specified anomalies of respiratory system**

- 748.800 Anomaly of pleura
- 748.810 Congenital cyst of mediastinum
- 748.880 Other specified respiratory system anomalies  
Includes: congenital lobar emphysema  
lymphangiectasia of lungs

**748.9 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)

749.000 Cleft hard palate, unilateral  
749.010 Cleft hard palate, bilateral  
749.020 Cleft hard palate, central  
749.030 Cleft hard palate, NOS  
749.040 Cleft soft palate, alone unilateral  
749.050 Cleft soft palate, alone bilateral  
749.060 Cleft soft palate, alone central  
749.070 Cleft soft palate, alone, NOS  
749.080 Cleft uvula  
749.090 Cleft palate, NOS  
palatoschisis

**749.1 Cleft lip alone**

Includes: alveolar ridge cleft  
cleft gum  
harelip

749.100 Cleft lip, unilateral  
749.110 Cleft lip, bilateral  
749.120 Cleft lip, central  
749.190 Cleft lip, NOS (fused lip)  
cleft gum

**749.2 Cleft lip with cleft palate**

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

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

- # 750.000 Tongue tie  
Ankyloglossia

**750.1 Other anomalies of tongue**

Excludes: protruding tongue (never a defect)

- 750.100 Aglossia  
Absence of tongue
- 750.110 Hypoglossia (small tongue)  
Microglossia
- 750.120 Macroglossia (large tongue)
- 750.130 Dislocation or displacement of tongue  
Glossoptosis
- 750.140 Cleft tongue or split tongue
- 750.180 Other specified anomalies of tongue
- 750.190 Unspecified anomalies of tongue

**750.2 Other specified anomalies of mouth and pharynx**

- 750.200 Pharyngeal pouch
- 750.210 Other pharyngeal anomalies
- 750.230 Other anomalies of salivary glands or ducts
- # 750.240 High arched palate
- 750.250 Other anomalies of palate
- 750.260 Lip fistulae or pits
- 750.270 Other lip anomalies  
Includes: notched lip, prominent philtrum,  
long philtrum  
Excludes: cleft lip (see 749)
- 750.280 Other specified anomalies of mouth and pharynx  
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

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**750.5 Congenital hypertrophic pyloric stenosis**

- # 750.500 Pylorospasm
- 750.510 Congenital hypertrophic pyloric stenosis
- 750.580 Other congenital pyloric obstruction

**750.6 Congenital hiatus hernia**

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

**750.7 Other specified anomalies of stomach**

- 750.700 Microgastria
- 750.710 Megalogastrica
- 750.720 Cardiospasm
  - achalasia of cardia, congenital
- 750.730 Displacement or transposition of stomach
- 750.740 Diverticulum of stomach
- 750.750 Duplication of stomach
- 750.780 Other specified anomalies of 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
- 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
- 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
- 751.210 Stenosis, atresia or absence of rectum with fistula
- 751.220 Stenosis, atresia or absence of rectum without mention of  
fistula
- 751.230 Stenosis, atresia or absence of anus with fistula  
Includes: imperforate anus with fistula
- 751.240 Stenosis, atresia or absence of anus without mention of fistula  
Includes: imperforate anus 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
- 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
- 751.495 Malrotation of small intestine alone

**751.5 Other anomalies of intestine**

- 751.500 Duplication of anus, appendix, cecum, or intestine  
enterogenous cyst
- 751.510 Transposition of appendix, colon, or intestine
- 751.520 Microcolon

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- 751.530 Ectopic (displaced) anus
- 751.540 Congenital anal fistula
- 751.550 Persistent cloaca
- R 751.555 **Exstrophy of cloaca**  
Excludes exstrophy of urinary bladder not associated with imperforate anus (use 753.500)
- \* 751.560 Duodenal web
- # 751.580 Other specified anomalies of intestine  
Includes: rectal fissures
- 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)  
Excludes: Budd-Chiari (use 453.000)
- 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
- 751.680 Anomalies of biliary tract, NEC

**751.7 Anomalies of pancreas**

- Excludes: fibrocystic disease of pancreas (277.000)  
diabetes mellitus,  
congenital  
neonatal
- 751.700 Absence, agenesis or hypoplasia of pancreas
- 751.710 Accessory pancreas
- 751.720 Annular pancreas
- 751.730 Ectopic pancreas
- 751.740 Pancreatic cyst
- 751.780 Other specified anomalies of 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  
obstruction, 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**

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

**752.1 Anomalies of fallopian tubes and broad ligaments**

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

**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  
752.380 Other anomalies of uterus  
bicornuate uterus  
unicornis 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  
752.420 Congenital rectovaginal fistula  
# 752.430 Imperforate hymen  
# 752.440 Absence or other anomaly of vulva  
fusion of vulva  
hypoplastic labia majora - Always code if ≥36weeks gestation. If  
<36 weeks gestation, code only if another reportable defect is  
present.

- # 752.450 Absence or other anomaly of clitoris  
Includes: clitoromegaly  
enlarged clitoris  
clitoral hypertrophy  
prominent clitoris
- # 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  
hymenal tags
- 752.490 Unspecified anomalies of cervix, vagina, or external female genitalia

### 752.5 Undescended testicle

- # 1)If < 36 weeks gestation, code only if there is a medical/surgical intervention for this problem;  
2)If ≥36 weeks gestation and defect last noted at <1 year of age, code only if there was a medical/surgical intervention for this problem or if another reportable defect is present  
3)Always code if ≥36 weeks gestation and defect first noted at ≥1 of age.
- # 752.500 Undescended testicle, unilateral undescended, unpalpable
- # 752.501 Left undescended testicle
- # 752.502 Right undescended testicle
- # 752.514 Undescended testicle, bilateral
- # 752.520 Undescended testicle, NOS (Cryptorchidism)
- 752.530 Ectopic testis, unilateral and bilateral

### 752.6 Hypospadias and epispadias

- 752.600 Hypospadias (alone), NOS
- 752.605 1°, glandular, coronal
- 752.606 2°, penile
- 752.607 3°, perineal, scrotal
- 752.610 Epispadias
- 752.620 Congenital chordee (with hypospadias), NOS
- 752.621 Congenital chordee alone (chordee w/o hypospadias)
- 752.625 Cong. chordee with 1°, coronal hypospadias
- 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

**752.8 Other specified anomalies of male genital organs**

752.800 Absence of testis  
           monorchidism, NOS  
 # 752.810 Aplasia or hypoplasia of testis and scrotum  
 752.820 Other anomalies of testis and scrotum  
           polyorchidism  
           bifid scrotum  
           Excludes: torsion of the testes or spermatic  
                           cord (use #608.200)  
 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  
 #           redundant foreskin (never a defect)  
 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

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

**753.1 Cystic kidney disease**

- 753.100 Renal cyst (single)
- 753.110 Polycystic kidneys, infantile type
- 753.120 Polycystic kidneys, adult type
- 753.130 Polycystic kidneys, NOS
- 753.140 Medullary cystic disease, juvenile type
- 753.150 Medullary cystic disease, adult type  
Medullary sponge kidney
- 753.160 Multicystic renal dysplasia  
Multicystic kidney
- 753.180 Other specified cystic disease  
Includes: cystic kidneys, NOS

**753.2 Obstructive defects of renal pelvis and ureter**

- 753.200 Congenital hydronephrosis
- 753.210 Atresia, stricture, or stenosis of ureter  
Includes: ureteropelvic junction obstruction/stenosis  
ureterovesical junction obstruction/stenosis  
hypoplastic ureter
- 753.220 Megaloureter, NOS  
Includes: hydroureter
- 753.290 Other and unspecified obstructive defects of renal pelvis and ureter

**753.3 Other specified anomalies of kidney**

- 753.300 Accessory kidney
- 753.310 Double or triple kidney and pelvis  
pyelon duplex or triplex
- 753.320 Lobulated, fused, or horseshoe kidney
- 753.330 Ectopic kidney
- 753.340 Enlarged, hyperplastic or giant kidney
- 753.350 Congenital renal calculi
- 753.380 Other specified anomalies of kidney

**753.4 Other specified anomalies of ureter**

- 753.400 Absence of ureter
- 753.410 Accessory ureter  
double ureter, duplex collecting system
- 753.420 Ectopic ureter
- 753.480 Other specified anomalies of ureter  
Includes: ureterocele

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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  
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

**753.7 Anomalies of urachus**

T # 753.700 Patent urachus  
753.710 Cyst of urachus  
753.790 Other and unspecified anomaly of urachus

**753.8 Other specified anomalies of bladder and urethra**

753.800 Absence of bladder or urethra  
753.810 Ectopic bladder  
753.820 Congenital diverticulum or hernia of 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  
rectovesical fistula  
753.870 Urethral fistula, NOS  
753.880 Other specified anomalies of bladder and urethra

**753.9 Unspecified anomalies of urinary system**

753.900 Unspecified anomaly of kidney  
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
- # 754.020 Congenital deviation of nasal septum  
bent nose
- T 754.030 Dolichocephaly  
Always code if  $\geq 36$  weeks gestation  
# If  $< 36$  weeks gestation, code only if another reportable  
defect is present
- # 754.040 Depressions in skull  
Includes: large fontanelle  
small fontanelle
- 754.050 Plagiocephaly
- 754.055 Asymmetric head
- T # \* 754.060 Scaphocephaly, no mention of craniosynostosis
- \* 754.070 Trigenocephaly, no mention of craniosynostosis  
Always code if  $\geq 36$  weeks gestation  
# If  $< 36$  weeks gestation, code only if another reportable  
defect is present
- \* 754.080 Other specified skull deformity, no mention of  
craniosynostosis  
Includes: brachycephaly  
acrocephaly  
turriccephaly  
oxycephaly
- \* 754.090 Deformity of skull, NOS

**754.1 Anomalies of sternocleidomastoid muscle**

- \* 754.100 Anomalies of sternocleidomastoid muscle  
Includes: absent or hypoplastic sternocleidomastoid  
contracture of sternocleidomastoid muscle  
sternomastoid tumor  
Excludes: congenital sternocleidomastoid torticollis  
(use 756.860)

**754.2 Certain congenital musculoskeletal deformities of spine**

- 754.200 Congenital postural scoliosis
- 754.210 Congenital postural lordosis
- 754.220 Congenital postural curvature of spine, NOS

**754.3 Congenital dislocation of hip**

- 754.300 Congenital dislocation of hip
- 754.310 Unstable hip  
preluxation of hip  
subluxation of hip  
predislocation status of hip at birth

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**754.4 Congenital genu recurvatum and bowing of long bones of leg**

754.400 Bowing, femur  
754.410 Bowing, tibia and/or fibula  
754.420 Bow legs, NOS  
754.430 Genu recurvatum  
754.440 Dislocation of knee, congenital  
754.490 Deformity of leg, NOS

**754.5 Varus (inward) deformities of feet**

754.500 Talipes equinovarus  
754.510 Talipes calcaneovarus  
# 754.520 Metatarsus varus or metatarsus adductus  
754.530 Complex varus deformities  
754.590 Unspecified varus deformities of feet

**754.6 Valgus (outward) deformities of feet**

754.600 Talipes calcaneovalgus  
754.610 Congenital pes planus  
754.615 Pes valgus  
754.680 Other specified valgus deformities of foot  
754.690 Unspecified valgus deformities of foot

**754.7 Other deformities of feet**

754.700 Pes cavus  
Claw foot (use 755.350 for claw foot)  
754.720 Short Achilles tendon  
754.730 Clubfoot, NOS  
talipes, NOS  
754.735 Congenital deformities of foot, NOS  
754.780 Other specified deformities of ankle and/or toes  
Includes: dorsiflexion of foot  
Excludes: widely spaced 1st and 2nd toes (use 755.600)

**754.8 Other specified congenital musculoskeletal deformities**

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

## 755 Other Congenital Anomalies of Limbs

### 755.0 Polydactyly

- 755.005 Accessory fingers (postaxial polydactyly, Type A)
- # 755.006 Skin tag (postaxial polydactyly, Type B)  
Exclude: Do not code in black infants.
- 755.007 Unspecified finger or skin tag (postaxial polydactyly, NOS)
- 755.010 Accessory thumbs (preaxial polydactyly)
- 755.020 Accessory toes (postaxial)
- 755.030 Accessory big toe (preaxial)
- 755.090 Accessory digits, NOS (hand/foot not specified)
- 755.095 Accessory digits hand, NOS (preaxial, postaxial not specified)
- 755.096 Accessory digits foot, NOS (preaxial, postaxial not specified)

### 755.1 Syndactyly

- 755.100 Fused fingers
- 755.110 Webbed fingers
- 755.120 Fused toes
- T # 755.130 Webbed toes  
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
- 755.190 Unspecified syndactyly (see below for specified site)
- 755.191 Unspecified syndactyly thumb and/or fingers, unilateral
- 755.192 Unspecified syndactyly thumb and/or fingers, bilateral
- 755.193 Unspecified (webbed vs. fused) syndactyly thumb and/or fingers, NOS
- 755.194 Unspecified syndactyly toes unilateral
- 755.195 Unspecified syndactyly toes bilateral
- 755.196 Unspecified syndactyly toes, NOS
- 755.199 Unspecified syndactyly (i.e., webbed vs. fused) digits not known

### 755.2 Reduction defects of upper limb

- T If description of the condition includes amniotic or constricting bands use additional code, 658.800 (Only use 658.800 if another reportable defect is present)  
Excludes shortening of upper limb (use 755.580) or hypoplasia of upper limb (use 755.585)
- 755.200 Absence of upper limb  
Absent: humerus (total or partial), radius, ulna and hand  
Includes: amelia of upper limb, NOS  
infants with rudimentary or nubbin fingers attached to stump of humerus or shoulder girdle
- 755.210 Absence of upper arm and forearm  
Absent: humerus (total or partial), radius and ulna (total or partial)  
Present: hand (total or partial)  
Includes: phocomelia of upper limb, NOS;  
intercalary reduction defect of upper limb, NOS

- 755.220 Absence of forearm only or upper arm only  
 Absent: radius and ulna  
 Present: humerus, hand (total or partial)  
 or  
 Absent: humerus  
 Present: radius, ulna, and hand
- 755.230 Absence of forearm and hand  
 Absent: radius and ulna (total or partial) and hand  
 Includes: infants with rudimentary or nubbin fingers  
 attached to stump of forearm or elbow
- 755.240 Absence of hand or fingers  
 Absent: hand or fingers (total or partial) not in  
 conjunction with ray or long bone reduction  
 Includes: rudimentary or nubbin fingers;  
 absent individual phalanges;  
 absent or missing fingers, NOS  
 Excludes: isolated absent or hypoplastic thumb  
 (use 755.260)
- 755.250 Split-hand malformation  
 Absent: central fingers (third with or without second,  
 fourth) and metacarpals (total or partial)  
 Includes: monodactyly;  
 lobster-claw hand  
 Excludes: isolated absent central fingers without  
 metacarpal defects (use 755.240)
- 755.260 Preaxial longitudinal reduction defect of upper limb  
 Absent: radius (total or partial) and/or thumb with or  
 without second finger (total or partial)  
 Includes: isolated absent or hypoplastic thumb;  
 radial ray defect, NOS
- 755.265 Longitudinal reduction defect of upper limb, NOS  
 Includes: absent forearm long bone with absent fingers,  
 NOS
- 755.270 Postaxial longitudinal reduction 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 ±  
 fourth metacarpal also totally or partially  
 absent;  
 ulnar ray defect, NOS
- 755.280 Other specified reduction defect of upper limb
- 755.285 Transverse reduction defect of upper limb, NOS  
 Includes: congenital amputation of upper limb, NOS
- 755.290 Unspecified reduction defect of upper limb

### 755.3 Reduction defects of lower limb

**T** If description of condition includes amniotic or constricting bands  
 use additional code, 658.800 (Only use this code if another  
 reportable defect is present)

Excludes shortening of lower limb (use 755.680) and hypoplasia of  
 lower limb (use 755.685)

- 755.300 Absence of lower limb  
Absent: femur (total or partial), tibia, fibula, and foot  
Includes: amelia of lower limb, NOS  
infants with rudimentary or nubbin toes attached to stump of femur or pelvic girdle
- 755.310 Absence of thigh and lower leg  
Absent: femur (total or partial), tibia and fibula (total or partial)  
Present: foot (total or partial)  
Includes: phocomelia of lower limb, NOS;  
intercalary reduction defect of lower limb, NOS
- 755.320 Absence of lower leg only or femur only  
Absent: tibia and fibula  
Present: femur, foot (total or partial)  
or  
Absent: femur  
Present: tibia, fibula, and foot
- 755.330 Absence of lower leg and foot  
Absent: tibia and fibula (total or partial), foot  
Includes: infants with rudimentary or nubbin toes attached to stump of leg or knee
- 755.340 Absence of foot or toes  
Absent: foot or toes (total or partial) not in conjunction with ray or long bone reduction  
Includes: rudimentary or nubbin toes;  
absent individual phalanges;  
absent or missing toes, NOS  
Excludes: isolated absent or hypoplastic great toe (use 755.365)
- 755.350 Split-foot malformation  
Absent: central toes (third with or without second, fourth) and metatarsals (total or partial)  
Includes: monodactyly;  
lobster claw foot  
Excludes: isolated absent central toes without metatarsal defects (use 755.340)  
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
- 755.360 Longitudinal reduction defect of lower limb, NOS  
Includes: absent long bone of leg with absent toes, NOS
- 755.365 Preaxial longitudinal reduction defect of lower limb  
Absent: tibia (total or partial) and/OR great toe with or without second toe (total or partial)  
Includes: isolated absent or hypoplastic great toe;  
tibial ray defect, NOS
- 755.366 Postaxial longitudinal reduction defect of lower limb  
Includes: isolated absent fibula (total or partial);  
absent fifth with or without fourth toe (total or partial) only if fibula or fifth ± fourth metatarsal also totally or partially absent;  
fibular ray defect, NOS
- 755.380 Other specified reduction defect of lower limb
- 755.385 Transverse reduction defect of lower limb, NOS  
Includes: congenital amputation of lower limb, NOS
- 755.390 Unspecified reduction defect of lower limb

#### 755.4 Reduction defects of unspecified limb

T If description of condition includes amniotic or constricting bands use additional code, 658.800 (note: 658.00 should only be used with another reportable defect)

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

#### 755.5 Other anomalies of upper limb, including shoulder girdle

Includes: complex anomalies involving all or part of upper limb

- # 755.500 Anomalies of fingers  
Includes: camptodactyly  
clinodactyly  
macroductyilia  
brachydactyly  
triphalaingeal thumb  
incurving fingers  
Excludes: acrocephalosyndactyly (see 756.050)  
Apert's syndrome (see 756.055)
- 755.510 Anomalies of hand  
Excludes: simian crease (use 757.200)
- 755.520 Anomalies of wrist
- 755.525 Accessory carpal bones
- 755.526 Madelung's deformity
- 755.530 Anomalies of forearm, NOS
- 755.535 Radioulnar dysostosis
- 755.536 Radioulnar synostosis
- 755.540 Anomalies of elbow and upper arm
- 755.550 Anomalies of shoulder
- 755.555 Cleidocranial dysostosis
- 755.556 Sprengel's deformity
- 755.560 Other anomalies of whole arm
- 755.580 Other specified anomalies of upper limb  
Includes: hyperextensibility of upper limb  
shortening of arm
- 755.585 Hypoplasia of upper limb  
Includes: hypoplasia of fingers, hands, or arms  
Excludes: aplasia or absent upper limb (see 755.2)
- 755.590 Unspecified anomalies of upper limb

**755.6 Other anomalies of lower limb, including pelvic girdle**

Includes: complex anomalies involving all  
or part of lower limb

- # 755.600 Anomalies of toes
  - Includes: overlapping toes
  - hammer toes
  - widely spaced first and second toes
- 755.605 Hallux valgus
- 755.606 Hallux varus
- 755.610 Anomalies of foot
  - Includes: plantar furrow
  - Excludes: lobster claw foot (use 755.350)
- # 755.616 Rocker-bottom foot
- 755.620 Anomalies of ankle
  - astragaloscaphoid synostosis
- # 755.630 Anomalies of lower leg
  - angulation of tibia, tibial torsion
  - (exclude if clubfoot present)
- 755.640 Anomalies of knee
  - hyperextended knee
- 755.645 Genu valgum
- 755.646 Genu varum
- 755.647 Absent patella or rudimentary patella
- 755.650 Anomalies of upper leg
  - anteversion of femur
- 755.660 Anomalies of hip
  - Includes: coxa vara
  - coxa valga
  - other abnormalities of hips
- 755.665 Hip dysplasia, NOS
- 755.666 Unilateral hip dysplasia
- 755.667 Bilateral hip dysplasia
- 755.670 Anomalies of pelvis
  - fusion of sacroiliac joint
- 755.680 Other specified anomalies of lower limb
  - hyperextended legs
  - shortening of legs
- 755.685 Hypoplasia of lower limb
  - Includes: hypoplasia of toes, feet, legs
  - Excludes: aplasia or absent lower limb (see 755.3)
- 755.690 Unspecified anomalies of legs

**755.8 Other specified anomalies of unspecified limb**

- 755.800 Arthrogryposis multiplex congenita
  - Includes: distal arthrogryposis syndrome
- 755.810 Larsen's syndrome
- 755.880 Other specified anomalies of unspecified limb
  - Includes: overlapping digits, NOS
  - hyperextended joints, NOS
  - Excludes: hyperextended knees (use 755.640)

**755.9 Unspecified anomalies of unspecified limb**

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T = Rev. 6/04  
\* = code created by CDC  
# = on the MACDP Excl List

755.900 Unspecified anomalies of unspecified limb

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**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  
craniostenosis, NOS  
closed-skull sutures, NOS
- 756.005 Sagittal craniosynostosis
- 756.006 Metopic craniosynostosis
- 756.010 Coronal craniosynostosis
- 756.020 Lambdoidal craniosynostosis
- 756.030 Other types of craniosynostosis  
Includes: basilar craniosynostosis
- 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  
Hallermand-Streiff syndrome
- 756.050 Acrocephalosyndactyly, NOS
- 756.055 Acrocephalosyndactyly types I or II  
Apert syndrome
- 756.056 Acrocephalosyndactyly type III
- 756.057 Other specified acrocephalosyndactylies
- 756.060 Goldenhar syndrome  
oculoauriculovertebral dysplasia
- 756.065 Hemifacial microsomia
- 756.080 Other specified skull and face bone anomalies  
Includes: localized skull defects  
# flat occiput  
mid-facial hypoplasia  
# prominent occiput  
prominent maxilla  
hypotelorism  
Excludes: macrocephaly (use 742.400)  
small chin (see 524.0)  
Pierre Robin sequence (use 524.080)
- 756.085 Hypertelorism, telecanthus, wide set eyes
- 756.090 Unspecified skull and face bone anomalies  
Excludes: dentofacial anomalies (524.0)  
skull defects associated with brain anomalies  
such as:  
anencephalus (740.0)  
encephalocele (742.0)  
hydrocephalus (742.3)  
microcephalus (742.100)

**756.1 Anomalies of spine**

- 756.100 Spina bifida occulta
- 756.110 Klippel-Feil syndrome  
Wildervanck syndrome

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- 756.120 Kyphosis  
kyphoscoliosis
- 756.130 Congenital spondylolisthesis
- 756.140 Anomalies of cervical vertebrae
- 756.145 Hemivertebrae (cervical)
- 756.146 Agenesis (cervical)
- 756.150 Anomalies of thoracic vertebrae
- 756.155 Hemivertebrae of thoracic vertebrae
- 756.156 Agenesis of thoracic vertebrae
- 756.160 Anomalies of lumbar vertebrae
- 756.165 Hemivertebrae of lumbar vertebrae
- 756.166 Agenesis of lumbar vertebrae
- 756.170 Sacrococcygeal anomalies  
Includes: agenesis of sacrum  
Excludes: pilonidal sinus (see 685.100)
- 756.179 Sacral mass, NOS
- 756.180 Other specified vertebral anomalies
- 756.185 Hemivertebrae, NOS
- 756.190 Unspecified anomalies of spine

**756.2 Cervical rib**

- # 756.200 Cervical rib  
supernumerary rib in cervical region

**756.3 Other anomalies of ribs and sternum**

- 756.300 Absence of ribs
- 756.310 Misshapen ribs
- 756.320 Fused ribs
- 756.330 Extra ribs
- 756.340 Other anomalies of ribs
- 756.350 Absence of sternum
- 756.360 Misshapen sternum
- 756.380 Other anomalies of sternum  
Includes: double ossification center in the manubrium,  
bifid sternum, short 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
- 756.440 Other specified chondrodystrophies  
Excludes: Conradi's (use 756.575)
- 756.445 Diastrophic dwarfism
- 756.446 Metatrophic dwarfism
- 756.447 Thanatophoric dwarfism

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- 756.450 Metaphyseal dysostosis
- 756.460 Spondyloepiphyseal dysplasia
- 756.470 Exostosis
  - Excludes: Gardner syndrome (see 759.630)
- 756.480 Other specified chondrodystrophy
- 756.490 Unspecified chondrodystrophy
  - Excludes: lipocondrodystrophy (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**

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

**756.7 Anomalies of abdominal wall**

- 756.700 Exomphalos, omphalocele
- 756.710 Gastroschisis
  - Excludes: umbilical hernia (553.100)
- 756.720 Prune belly syndrome
- 756.790 Other and unspecified anomalies of abdominal wall
- 756.795 Epigastric hernia

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

- 756.800 Poland syndrome or anomaly
- 756.810 Other absent or hypoplastic muscle

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Includes: absent pectoralis major  
 Excludes: prune belly syndrome (use 756.720)

756.820 Absent tendon  
 756.830 Nail-patella syndrome  
 756.840 Amyotrophia congenita  
 756.850 Ehlers-Danlos syndrome  
 756.860 Congenital torticollis  
 (see also 754.100, anomalies of sternocleidomastoid muscle)  
 756.880 Other specified anomalies of muscle, tendon, fascia and  
 connective tissue  
 Includes: myopathy, congenital NOS

**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**

757.000 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  
757.195 Ichthyosis vulgaris  
757.196 X-linked ichthyosis  
757.197 Ichthyosiform erythroderma

**757.2 Dermatoglyphic anomalies**

# 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  
# 757.310 Skin tags  
Includes: anal tags  
Excludes: preauricular tag (see 744.110)  
vaginal tags (see 752.480)  
757.320 Urticaria pigmentosa  
757.330 Epidermolysis bullosa  
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 or nevus flammeus  
T Excludes: hairy nevus (use 216.920)  
Sturge-Weber syndrome (use 759.610)  
# 757.385 Birthmark, NOS  
# 757.386 Mongolian blue spot  
# 757.390 Other specified anomalies of skin  
Includes: cafe au lait spots  
hyperpigmented areas  
skin cysts  
hypoplastic dermal patterns  
757.395 Absence of skin

**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
- 757.480 Other specified anomalies of hair

**757.5 Specified anomalies of nails**

- 757.500 Congenital onychia  
Absent nails
- 757.510 Enlarged or hypertrophic nails
- 757.515 Onychauxis
- 757.516 Pachyonychia
- 757.520 Congenital koilonychia
- 757.530 Congenital leukonychia
- 757.540 Club nail
- 757.580 Other specified anomalies of nails
- 757.585 Hypoplastic (small) fingernails and/or toenails

**757.6 Specified anomalies of breast**

- 757.600 Absent breast with absent nipple
- 757.610 Hypoplastic breast with hypoplastic nipple
- 757.620 Accessory (ectopic) breast with nipple
- 757.630 Absent nipple
- T # 757.640 Small nipple (hypoplastic)  
Always code if ≥36 weeks gestation  
# If <36 weeks gestation, code only if another reportable defect is present
- # 757.650 Accessory (ectopic) nipple, supernumerary
- # 757.680 Other specified anomalies of breast  
Widely spaced nipples  
Excludes: inverted nipples (never a defect)

**757.8 Other specified anomalies of the integument**

- 757.800 Includes: scalp defects  
For specified anomalies of skin see 757.390  
For specified anomalies of hair see 757.480  
For specified anomalies of nails see 757.580

**757.9 Unspecified anomalies of the integument**

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

**758 Chromosomal Anomalies**

**758.0 Down syndrome**

Clinical Down syndrome karyotype identified as:

- T 758.000 Down syndrome, karyotype trisomy 21, cytogenetics result in record
- T 758.008 Down syndrome suspected, cytogenetics pending
- 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
- T 758.090 Down syndrome, NOS (i.e. chart states a diagnosis of Trisomy 21 or Downs syndrome, but no cytogenetics result in record)
- T 758.098 Down syndrome suspected, cytogenetics never done

**758.1 Patau syndrome**

Clinical Patau syndrome karyotype identified as:

- T 758.100 Patau syndrome, karyotype trisomy 13, cytogenetics result in record
- 758.108 Patau syndrome suspected, cytogenetics pending
- 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
- T 758.190 Patau syndrome, NOS (i.e. chart states a diagnosis of Trisomy 13 or Patau syndrome, but no cytogenetics result in record)
- T 758.198 Patau syndrome suspected, cytogenetics pending

**758.2 Edwards syndrome**

Clinical Edwards syndrome karyotype identified as:

- T 758.200 Edwards syndrome, karyotype trisomy 18, cytogenetics result in record
- T 758.208 Edwards syndrome suspected, cytogenetics pending
- 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
- 758.290 Edwards syndrome, NOS (i.e. chart states a diagnosis of Trisomy 18 or Edwards syndrome, but no cytogenetics result in record)
- T 758.295 Edwards phenotype - normal karyotype
- T 758.298 Edwards syndrome suspected, cytogenetics pending

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### 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)  
Note: Code added for use with births on or after 4/1/2001
- 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
- 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
- 758.585 Polyploidy
- 758.586 Triploidy
- 758.590 Unspecified anomalies of autosomes

**758.6 Gonadal Dysgenesis**

Excludes: pure gonadal dysgenesis (752.720)  
Noonan syndrome (759.800)

- 758.600 Turner's phenotype, karyotype 45, X [XO]
- 758.610 Turner's phenotype, variant karyotypes  
karyotype characterized by:  
isochromosome  
mosaic, including XO  
partial X deletion  
ring chromosome  
Excludes: Turner's phenotype, karyotype normal XX  
(use 759.800, Noonan syndrome)
- 758.690 Turner syndrome, karyotype unspecified, NOS  
Bonnevill-Ullrich syndrome, NOS

**758.7 Klinefelter syndrome**

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

**758.8 Other conditions due to sex chromosome anomalies**

- 758.800 Mosaic XO/XY, 45X/46XY  
Excludes: with Turner's phenotype (758.610)
- 758.810 Mosaic XO/XX  
Excludes: with Turner's phenotype (758.610)
- 758.820 Mosaic XY/XXY, 46XY/47XXY  
Excludes: Klinefelter's phenotype (758.710)
- 758.830 Mosaic including XXXXY, 49XXXXY  
Excludes: with Klinefelter's phenotype (use 758.710)
- 758.840 XYY, male, 47XYY  
mosaic XYY male
- 758.850 XXX female, 47XXX
- 758.860 Additional sex chromosomes, NOS
- 758.880 Other specified sex chromosome anomaly  
Includes: fragile X
- 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)

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**759 Other and Unspecified Congenital Anomalies**

**759.0 Anomalies of spleen**

- 759.000 Absence of spleen  
asplenia
- 759.005 Ivemark syndrome
- 759.010 Hypoplasia of spleen
- # 759.020 Hyperplasia of spleen  
splenomegaly  
hepatosplenomegaly (also use code 751.620)
- 759.030 Misshapen spleen
- 759.040 Accessory spleen
- 759.050 Ectopic spleen
- 759.080 Other specified anomalies of spleen
- 759.090 Unspecified anomalies of spleen

**759.1 Anomalies of adrenal gland**

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

**759.2 Anomalies of other endocrine glands**

- 759.200 Anomalies of pituitary gland
- 759.210 Anomalies of thyroid gland
- 759.220 Thyroglossal duct anomalies  
thyroglossal cyst
- 759.230 Anomalies of parathyroid gland
- # 759.240 Anomalies of thymus  
thymic hypertrophy  
absent thymus
- 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
- 759.310 Situs inversus with levocardia
- 759.320 Situs inversus thoracis
- 759.330 Situs inversus abdominis
- 759.340 Kartagener syndrome (triad)
- 759.390 Unspecified situs inversus  
Excludes: dextrocardia (746.800) not  
associated with complete situs inversus

**759.4 Conjoined twins**

- 759.400 Dicephalus  
two heads
- 759.410 Craniopagus

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- head-joined twins
- 759.420 Thoracopagus  
thorax-joined twins
- 759.430 Xiphopagus  
xiphoid- and pelvis-joined twins
- 759.440 Pygopagus  
buttock-joined twins
- 759.480 Other specified conjoined twins
- 759.490 Unspecified conjoined twins

**759.5 Tuberos sclerosis**

- 759.500 Tuberos sclerosis  
Bourneville's disease  
epiloia

**759.6 Other hamartoses, not elsewhere classified**

- 759.600 Peutz-Jeghers 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
- 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  
cyclops  
Noonan syndrome  
oral-facial-digital (OFD) syndrome, type I  
Orofaciodigital syndrome, type II (Mohr syndrome)  
Waardenburg syndrome  
whistling face syndrome
- 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  
Carpenter syndrome  
Holt-Oram syndrome  
Klippel-Trenaunay-Weber syndrome  
Rubinstein-Taybi syndrome  
sirenomelia  
thrombocytopenia-absent radius (TAR) syndrome
- 759.860 Cong malformation syndromes with other skeletal changes

Marfan syndrome  
 Stickler syndrome  
 759.870 Cong malformation syndromes with metabolic disturbances  
 Alport syndrome  
 Beckwith (Wiedemann-Beckwith) syndrome  
 leprechaunism  
 Menkes syndrome (kinky hair syndrome)  
 Prader-Willi syndrome  
 Zellweger syndrome  
 759.890 Other specified anomalies  
 Includes:       hemihypertrophy  
                   Meckel-Gruber syndrome

**759.9 Congenital anomaly, unspecified**

# 759.900 Anomalies of umbilicus  
           low-lying umbilicus  
           umbilical cord atrophy  
 759.910 Embryopathia, NEC  
 759.990 Congenital anomaly, NOS



## Continued: Other Specified Codes Used in Metro Atlanta Congenital Defects Program

T	#	216.600	Benign neoplasm of skin, upper limb, shoulder
	#	221.000	Benign skin neoplasm of female genital organs
	#	222.000	Benign skin neoplasm of male genital organs
		453.000	Budd-Chiari, occlusion of hepatic vein
		427.900	Cardiac arrhythmias, NEC. Never code premature atrial contractions, PACs.
	#	330.100	Cerebral lipidoses Includes: Tay-Sachs disease, gangliosidosis
		363.200	Chorioretinitis
		279.200	Combined immunodeficiency syndrome
		771.280	Congenital infection, other specified Excludes: human immunodeficiency virus (HIV) infection and acquired immunodeficiency syndrome (AIDS)
	#	277.000	Cystic fibrosis No mention of meconium ileus
	#	277.010	Cystic fibrosis With mention of meconium ileus
		228.100	Cystic hygroma Lymphangioma, any site
		771.100	Cytomegalovirus (CMV) (in utero infections only)
		253.820	Diencephalic syndrome
		279.110	DiGeorge syndrome
		277.400	Disorders of bilirubin excretion
		425.300	Endocardial fibroelastosis
		553.200	Epigastric hernia
	#	767.600	Erb's palsy
	#	368.000	Esotropia
	#	378.000	Exotropia
	#	351.000	Facial palsy
		331.890	Familial degenerative CNS disease
		760.710	Fetal alcohol syndrome
		760.718	Fetal alcohol syndrome, probable Includes: "facies"
		760.750	Fetal hydantoin (Dilantin) syndrome
	#	282.200	Glucose-6-phosphate dehydrogenase (G-6-PD) deficiency
	#	271.000	Glycogen storage diseases
T		216.920	Hairy nevus
T		228.0	Hemangioma Include if greater than 4-inches diameter, if multiple hemangiomas, or if cavernous hemangioma
		228.040	Hemangioma, intra-abdominal (Always code regardless of size, type or number)
		228.020	Hemangioma, intracranial (Always code regardless of size, type or number)
		228.090	Hemangioma, of other sites (Always code regardless of size, type or number)
	#	228.000	Hemangioma, of unspecified site. Always code if multiple hemangiomas of any size are present, if one or more cavernous hemangiomas of any size are present, or if a single hemangioma measuring $\geq 4$ cm in diameter or described as large, huge, or of medical significance is present.
		228.030	Hemangioma, retinal (Always code regardless of size, type or number)

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**Continued: Other Specified Codes Used in Metro Atlanta Congenital Defects Program**

- # 228.010 Hemangioma, skin & subcutaneous, NOS Always code if multiple hemangiomas of any size are present, if one or more cavernous hemangiomas of any size are present, or if a single hemangioma measuring ≥ 4cm in diameter or described as large, huge, or of medical significance is present.
  
- # 286.000 Hemophilia (all types)
- 774.490 Hepatitis, neonatal, NOS
- 774.480 Hepatitis, neonatal, other specified
- # 282.100 Hereditary elliptocytosis
- # 282.000 Hereditary spherocytosis
  
- 771.220 Herpes simplex (in utero infections only)  
Includes: encephalitis  
                  meningoencephalitis
- 202.300 Histiocytosis, malignant
- 277.510 Hurler syndrome  
Includes: lipocondrodystrophy
- # 778.600 Hydrocele, congenital
- # 270.700 Hyperglycinemia
- # 251.200 Hypoglycemia, idiopathic
- # 252.100 Hypoparathyroidism, congenital
- # 275.330 Hypophosphatemic rickets
- 253.280 Hypopituitarism, congenital
- # 243.990 Hypothyroidism, congenital (Exclude even if other defects are present only if the record specifies hypothyroidism of prematurity. Other types of hypothyroidism or hypothyroidism NOS should continue to be on the routine exclusion list.)
- 345.600 Infantile spasms, congenital
- # 550.000 Inguinal hernia or patent processus vaginalis never  
-550.900 code in infants if <36 weeks gestation regardless of the presence of a reportable defect.  
NOTE: for those ≥36 weeks:  
Code in **males** only if another reportable defect is present;  
Code in **females**, always code even if found in isolation
- 208.000 Leukemia, congenital, NOS
- 214 Lipoma
- 214.300 Lipoma, intra-abdominal organs
- 214.200 Lipoma, intrathoracic organs
- 214.810 Lipoma, lumbar or sacral lipoma  
                  paraspinal lipoma
- 214.100 Lipoma, other skin and subcutaneous tissue
- 214.800 Lipoma, other specified sites
- 214.000 Lipoma, skin and subcutaneous tissue of face
- 214.400 Lipoma, spermatic cord
- 214.900 Lipoma, unspecified site
- # 457.800 Lymphatics - other specified disorders of (including chylothorax)
- 524.000 Macrognathia
- # 270.300 Maple syrup urine disease
- # 777.600 Meconium peritonitis
- # 777.100 Meconium plug syndrome
- 524.000 Micrognathia
- 352.600 Moebius syndrome

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# 520.600 Natal teeth  
 239.200 Neck cyst  
 774.490 Neonatal hepatitis, NOS

**Continued: Other Specified Codes Used in Metro Atlanta Congenital Defects Program**

774.480 Neonatal hepatitis, other specified  
 159.800 Neoplasms of the abdomen, other specified  
 191.000 Neoplasms of the CNS  
 Includes: medulloblastoma, gliomas  
 171.800 Neoplasms of the connective tissue  
 Includes: Ewing's sarcoma  
 fibrosarcoma  
 155.000 Neoplasms of the liver  
 Includes: hepatoblastoma  
 hemangio-epithelioma  
 162.800 Neoplasms of the lung  
 186.000 Neoplasms of the testes  
 194.000 Neuroblastoma  
 237.700 Neurofibromatosis  
 # 379.500 Nystagmus  
 # 270.100 Phenylketonuria (PKU)  
 \* 524.080 Pierre Robin sequence  
 # 685.100 Pilonidal sinus (sacrodermal), sacral sinus, sacral dimple  
 # 277.630 Pseudocholinesterase enzyme deficiency  
 # 284.000 Red cell aplasia  
 362.600 Retinal degeneration, peripheral  
 362.700 Retinitis pigmentosa  
 190.500 Retinoblastoma  
 771.000 Rubella, congenital (in utero infections only)  
 # 685.100 Sacral dimple  
 T # 216.910 Sebaceous cyst  
 # 282.600 Sickle cell anemia  
 # 090.000 Syphilis, congenital (in utero infections only)  
 238.030 Teratoma, abdomen  
 238.010 Teratoma, head and face  
 238.020 Teratoma, neck  
 238.000 Teratoma, NOS  
 238.080 Teratoma, other specified  
 238.040 Teratoma, sacral, coccygeal  
 257.800 Testicular feminization syndrome  
 771.090 TORCH infection, unspecified (in utero infections only)  
 # 608.200 Torsion of the testes or spermatic cord  
 771.210 Toxoplasmosis (in utero infections only)  
 # 553.100 Umbilical hernia  
 # 286.400 von Willebrand disease  
 335.000 Werdnig-Hoffman disease  
 189.000 Wilms tumor (nephroblastoma)  
 426.705 Wolfe-Parkinson-White syndrome, congenital

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## Other Specified Codes Used in Metro Atlanta Congenital Defects Program

List ordered by 6-digit code number

- # 090.000 Syphilis, congenital (in utero infections only)
- 155.000 Neoplasms of the liver
  - Includes: hepatoblastoma
  - hmangio-epithelioma
- 159.800 Neoplasms of the abdomen
- 162.800 Neoplasms of the lung
- 171.800 Neoplasms of connective tissue
  - Includes: Ewing's sarcoma
  - fibrosarcoma
- 186.000 Neoplasms of the testes
- 189.000 Wilms tumor (nephroblastoma)
- 190.500 Retinoblastoma
- 191.000 Neoplasms of the CNS
  - Includes: gliomas
  - mdulloblastoma
- 194.000 Neuroblastoma
- 202.300 Histiocytosis, malignant
- 208.000 Leukemia, congenital, NOS
  
- 214 Lipoma
  - 214.000 Lipoma, skin and subcutaneous tissue of face
  - 214.100 Lipoma, other skin and subcutaneous tissue
  - 214.200 Lipoma, intrathoracic organs
  - 214.300 Lipoma, intra-abdominal organs
  - 214.400 Lipoma, spermatic cord
  - 214.800 Lipoma, other specified sites
  - 214.810 Lipoma, lumbar or sacral lipoma
  - paraspinal lipoma
  - 214.900 Lipoma, unspecified site
  
- T 216 Benign neoplasm of skin
  - (NOTE: All benign neoplasms should be coded ONLY if another reportable code is present)**
  - Includes: blue nevus   pigmented nevus
  - papilloma    dermatofibroma
  - syringoadenoma
  - \*dermoid cyst
  - hydrocystoma
  - syringoma
  - 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
- # 216.100 Eyelid, including canthus
  - Excludes: cartilage of eyelid
- # 216.200 Ear and external auditory canal
  - Includes: auricle ear
  - external meatus
  - auricular canal
  - external canal
  - pinna
  - Excludes: cartilage of ear

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# 216.300 Skin of other and unspecified parts of face  
Includes: cheek, external nose,  
external eyebrow temple

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**Continued: Other Specified Codes Used in Metro Atlanta Congenital Defects Program**

- # 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
  - breast
  - Excludes: anal canal
  - anus, NOS
  - skin of scrotum
- # 216.600 Skin of upper limb, shoulder
- # 216.700 Skin of lower limb, hip
- # 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
- # 221.000 Benign skin neoplasm of female genital organs
- # 222.000 Benign skin neoplasm of male genital organs

- T 228.0 Hemangioma
  - Include if greater than 4-inches diameter, if multiple hemangiomas, or if cavernous hemangioma
- # 228.000 Hemangioma, of unspecified site
  - Always code if multiple hemangiomas of any size are present, if one or more cavernous hemangiomas of any size are present, or if a single hemangioma measuring  $\geq$  4cm in diameter or described as large, huge, or of medical significance is present.
- # 228.010 Hemangioma, skin & subcutaneous, NOS
  - Always code if multiple hemangiomas of any size are present, if one or more cavernous hemangiomas of any size are present, or if a single hemangioma measuring  $\geq$  4cm in diameter or described as large, huge, or of medical significance is present.
- 228.020 Hemangioma, intracranial (Always code regardless of size, type or number)
- 228.030 Hemangioma, retinal (Always code regardless of size, type or number)
- 228.040 Hemangioma, intra-abdominal (Always code regardless of size, type or number)
- 228.090 Hemangioma, of other sites (Always code regardless of size, type or number)
- 228.100 Cystic hygroma
- Lymphangioma, any site
- 237.700 Neurofibromatosis
- 238.000 Teratoma, NOS
- 238.010 Teratoma, head and face
- 238.020 Teratoma, neck
- 238.030 Teratoma, abdomen
- 238.040 Teratoma, sacral, coccygeal

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238.080 Teratoma, other specified  
239.200 Neck cyst

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## Continued: Other Specified Codes Used in Metro Atlanta Congenital Defects Program

# 243.990	Hypothyroidism, congenital (Exclude even if other defects are present only if the record specifies hypothyroidism of prematurity <36 weeks. Include other types of hypothyroidism and hypothyroidism NOS only when another reportable defect is present regardless of gestational age)
# 251.200	Hypoglycemia, idiopathic
# 252.100	Hypoparathyroidism, congenital
253.280	Hypopituitarism, congenital
253.820	Diencephalic syndrome
255.200	Adrenogenital syndrome (adrenal hyperplasia)
257.800	Testicular feminization syndrome
# 270.100	Phenylketonuria (PKU)
# 270.200	Albinism
# 270.300	Maple syrup urine disease
# 270.600	Arginosuccinic aciduria
# 270.700	Hyperglycinemia
# 271.000	Glycogen storage diseases
# 275.330	Hypophosphatemic rickets
# 277.000	Cystic fibrosis <u>with no</u> mention of meconium ileus
# 277.010	Cystic fibrosis <u>with</u> mention of meconium ileus
277.400	Disorders of bilirubin excretion
277.510	Hurler syndrome Includes: lipochondrodystrophy
# 277.620	Alpha-1 antitrypsin deficiency
# 277.630	Pseudocholinesterase enzyme deficiency
279.110	DiGeorge syndrome
279.200	Combined immunodeficiency syndrome
# 282.000	Hereditary spherocytosis
# 282.100	Hereditary elliptocytosis
# 282.200	Glucose-6-phosphate dehydrogenase (G-6-PD) deficiency
# 282.600	Sickle cell anemia
# 284.000	Red cell aplasia
# 286.000	Hemophilia (all types)
# 286.400	von Willebrand disease
# 330.100	Cerebral lipidoses Includes: Tay-Sachs disease gangliosidosis
331.890	Familial degenerative CNS disease
335.000	Werdnig-Hoffman disease
345.600	Infantile spasms, congenital
# 351.000	Facial palsy
352.600	Moebius syndrome
362.600	Retinal degeneration, peripheral
362.700	Retinitis pigmentosa
363.200	Chorioretinitis
# 368.000	Esotropia
# 378.000	Exotropia
# 379.500	Nystagmus
425.300	Endocardial fibroelastosis
426.705	Congenital Wolfe-Parkinson-White syndrome
427.900	Cardiac arrhythmias, NEC. Never code premature atrial contractions, PACs.
453.000	Budd-Chiari, occlusion of hepatic vein
# 457.800	Other specified disorders of lymphatics (including chylothorax)

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## Continued: Other Specified Codes Used in Metro Atlanta Congenital Defects Program

# 520.600 Natal teeth  
 524.000 Abnormalities of jaw size  
           micrognathia  
           macrognathia

\* 524.080 Pierre Robin sequence

# 550.000- Inguinal hernia or patent processus vaginalis never  
 550.900 code in infants if <36 weeks gestation regardless of the  
 presence of a reportable defect.  
NOTE: for those ≥36 weeks:  
       Code in **males** only if another reportable defect is present;  
       in **females**, always code even if found in isolation

# 553.100 Umbilical hernia  
 553.200 Epigastric hernia

# 608.200 Torsion of testes or spermatic cord

T # 658.800 Amniotic bands (constricting bands, amniotic cyst)

# 685.100 Pilonidal sinus (sacrodermal), sacral sinus, sacral dimple  
 760.710 Fetal alcohol syndrome  
 760.718 Probable fetal alcohol syndrome  
       Includes: "facies"  
 760.750 Fetal hydantoin (Dilantin) syndrome

# 767.600 Erb's palsy

771 Congenital infections (in utero infections only)  
 Excludes: congenital syphilis (use 090.000)

771.000 Rubella, congenital  
 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  
 Excludes: human immunodeficiency virus (HIV) infection and  
           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  
 # 778.600 Hydrocele, congenital

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 Doc. 6digit88, Version 06/04

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**EXCLUSION LIST** for the MACDP  
Nonreportable birth defects

**Conditions Never to be Reported**

The following newborn and infant conditions include those descriptions considered to be excludable or nonreportable conditions in the MACDP. This includes certain biochemical disorders not considered part of the present MACDP case definition.

**Alphabetical** list of conditions that are never considered to be defects.

**Description**

Anal fissure  
Atrial contractions, premature  
Breast hypertrophy  
Bronchopulmonary dysplasia (Wilson-Mikity syndrome)  
Cephalohematoma  
Chalasia (gastroesophageal reflux)  
CNS hemorrhage  
Conjunctivitis  
Diastasis recti  
Epulis  
Gastroesophageal reflux  
Gum cysts - Includes epulis, ranula, mucocele  
Hydrocephalus secondary to intraventricular hemorrhage (IVH) or CNS bleed  
Hip click, with no follow-up or therapy  
T Heart murmur  
Hyaline membrane disease  
T Intestinal obstruction - requires chart review to determine if cause of obstruction is a reportable defect. If so, code only the cause.  
T Intussusception - requires chart review to determine if cause of intussusception is a reportable defect. If so, code only the cause.  
Inverted nipples  
Laryngotracheomalacia or tracheomalacia  
Meconium stained skin or nails  
Mucocele  
Neonatal acne  
Overriding (overlapping) sutures  
Petechiae  
Phimosis  
Pneumothorax  
Premature atrial contractions  
Protruding tongue  
Ranula  
Redundant foreskin  
Retractile testes  
Tracheomalacia  
T Volvulus - requires chart review to determine if cause of volvulus is a reportable defect. If so, code only the cause.  
Wilson-Mikity syndrome

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**EXCLUSION LIST** for the MACDP  
 Nonreportable birth defects

**Conditions Which may be Included Under Certain Conditions**

The following newborn and infant conditions include those descriptions considered to be excludable or nonreportable conditions in the MACDP, but which may be included under certain circumstances.

The following rules apply to coding these conditions:

- A. If a condition or defect listed appears in a chart, singly or in any combination with other defects listed only on the Exclusion List, do not fill out the case record form.
- B. If one of these conditions listed accompanies a reportable birth defect (from the 6-digit code manual and not on the exclusion list), then use the listed 6-digit code and record all defects (including those from this list) from the hospital record onto the case abstraction form.

**Alphabetical** list of conditions requiring no record abstraction to be performed unless associated with a reportable defect. The addition or revision dates of the changes in the list of conditions requiring no record abstraction are shown.

**Revised/  
 Changed  
 Date**

<u>Date</u>	<u>Code</u>	<u>Description</u>
	744.100	Accessory auricle
	757.650	Accessory nipple (supernumerary nipple, or skin tag)
	270.200	Albinism
	277.620	Alpha 1-antitrypsin deficiency
	T 658.800	Amniotic bands (constricting bands, amniotic cyst)
	757.310	Anal tags
10/1/92	T 746.400	Aortic valve insufficiency or regurgitation, congenital - Code cases designated as 'mild', 'minimal', 'trivial', or 'physiologic' only if another reportable defect is present. Code all other degrees of insufficiency or regurgitation, including those where the degree is not specified, regardless of whether another reportable defect is present.
	270.600	Argininosuccinic aciduria
	T 778.000	Ascites or anasarca, congenital. Includes: hydrops fetalis
	744.220	Bat ear
	T # 216.200	Benign neoplasm of skin, ear and external auditory canal Includes: auricle ear external meatus auricular canal external canal pinna Excludes: cartilage of ear
	T # 216.100	Benign neoplasm of skin, eyelid, including canthus Excludes: cartilage of eyelid
	T # 216.000	Benign neoplasm of skin, lip Excludes: vermilion border of lip

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**EXCLUSION LIST** for the MACDP  
 Nonreportable birth defects

**Alphabetical - Conditions Which may be Included Under Certain Conditions**

**Revised/  
 Changed  
 Date**

**Code**

**Description**

	T	#	216.700	Benign neoplasm of skin, lower limb, hip
	T	#	216.300	Benign neoplasm of skin, other and unspecified parts of face Includes: cheek, external nose, external eyebrow, temple
	T	#	216.800	Benign neoplasm of skin, other specified sites of skin Excludes: epibulbar dermoid cyst (use 743.810)
	T	#	216.400	Benign neoplasm of skin, scalp and skin of neck
	T	#	216.900	Benign neoplasm of skin, site unspecified
		#	216.500	Benign neoplasm of skin, trunk, except scrotum Includes: axillary fold perianal skin skin of: chest wall, abdominal wall, groin, buttock, anus, perineum, back, umbilicus, breast Excludes: anal canal, anus, NOS skin of scrotum
		#	216.600	Benign neoplasm of skin, upper limb, shoulder
			221.000	Benign skin neoplasm of female genital organs
			222.000	Benign skin neoplasm of male genital organs
			754.020	Bent nose, deviation of nasal septum
			744.820	Big lips
			757.385	Birth mark, NOS
			743.450	Blue sclera - if <36 weeks gestation, code only if another reportable defect is present. Always code if >36 weeks gestation.
			743.800	Brushfield spots
			757.390	Cafe au lait spots
			746.860	Cardiomegaly, congenital NOS
			744.230	Cauliflower ear
			330.100	Cerebral lipidoses (e.g., Tay-Sachs, gangliosidoses, etc.)
			756.200	Cervical rib
			755.500	Clinodactyly (incurving of fifth finger)
1/1/93			752.520	Cryptorchidism (see undescended testicle)
			277.010	Cystic fibrosis, with mention of meconium ileus
			277.000	Cystic fibrosis, with no mention of meconium ileus
			744.280	Darwin's tubercle
1/1/96	T		754.030	Dolichocephaly - if <36 weeks gestation, code only if another reportable defect is present. Always code if >36 weeks gestation.
1/1/93			743.800	Downward eye slant (antimongoloid)
			744.110	Ear tags, preauricular
			744.120	Ear tags, other
			744.230	Elfin ear, absent or decreased ear cartilage - if <36 weeks gestation, code only if another reportable defect is present.
			743.800	Epicanthal folds
			767.600	Erb's palsy

**EXCLUSION LIST** for the MACDP  
 Nonreportable birth defects

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Alphabetical - Conditions Which may be Included Under Certain Conditions

Revised/  
 Changed  
 Date

Code                    Description

		368.000	Esotropia
		378.000	Exotropia
		351.000	Facial palsy
		757.380	Flammeus nevus or port wine stain
		748.180	Flat bridge of nose
		754.040	Fontanelle (large or small)
	T	743.630	Fused eyelids - never code if <25 weeks gestation unless another reportable defect is present
		752.440	Fusion of vulva
		282.200	Glucose-6-phosphate dehydrogenase (G-6-PD) deficiency
		271.000	Glycogen storage disease
		746.990	Heart murmur - if chart review does not confirm a heart defect within 6 months, do not code as a defect even if other codable defects are present
		286.000	Hemophilia
		751.620	Hepatomegaly
		282.100	Hereditary elliptocytosis
		282.000	Hereditary spherocytosis
3/4/91		750.240	High arched palate
		778.600	Hydrocele, congenital
		752.480	Hymenal tags
		270.700	Hyperglycinemia
		251.200	Hypoglycemia, idiopathic
		252.100	Hypoparathyroidism, congenital
		275.330	Hypophosphatemic rickets
1/1/96	T	752.440	Hypoplastic labia majora - if <36 weeks gestation, code only if another reportable defect is present. Always code if ≥36 weeks gestation.
3/4/91	T	748.510	Hypoplasia of lung; pulmonary hypoplasia - exclude only if an isolated defect in infants <36 weeks gestation
		752.810	Hypoplastic scrotum - exclude if secondary to undescended testes
	T	243.990	Hypothyroidism, congenital (Exclude hypothyroidism of prematurity in infants <36 weeks gestation even if other reportable defects are present. Include other types of hypothyroidism and hypothyroidism NOS when another reportable defect is present regardless of gestational age)
		752.430	Imperforate hymen
		755.500	Incurving fingers (clinodactyly)
	T	550.000-	Inguinal hernia or patent processus vaginalis. Never code in infants <36 weeks gestation regardless of the presence of a reportable defect. For infants ≥36 weeks:
		550.900	code only if another reportable defect is present;
		550.901	In <b>females</b> , always code even if found in isolation
		550.902	
		757.450	Lanugo, excessive or persistent
		754.040	Large fontanelle

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**Alphabetical - Conditions Which may be Included Under Certain Conditions**

<u>Revised/ Changed Date</u>	<u>Code</u>	<u>Description</u>
	755.500	Long fingers and toes
	744.230	Lop ear
	744.245	Low set ears
	744.820	Macrocheilia (big lips)
	270.300	Maple syrup urine disease
	751.010	Meckel's diverticulum
	777.600	Meconium peritonitis
	777.100	Meconium plug
9/10/90	754.520	Metatarsus varus or adductus
	744.830	Microcheilia (small lips)
10/1/92	T 746.600	Mitral valve insufficiency or regurgitation, congenital - Code cases designated as 'mild', 'minimal', 'trivial', or 'physiologic' only if another reportable defect is present. Code all other degrees of insufficiency or regurgitation, including those where the degree is not specified, regardless of whether another reportable defect is present.
	757.386	Mongolian spots
	743.650	Nasal lacrimal duct obstruction
	520.600	Natal teeth
	745.500	Nonclosure of foramen ovale, NOS (see PFO)
	379.500	Nystagmus
9/10/90	756.080	Occiput, flat or prominent
3/5/90	457.800	Other specified disorder of lymphatics, including chylothorax
	755.600	Overlapping toes
10/14/92	T 747.000	Patent ductus arteriosus (PDA) 1) Always code if ≥36 weeks of gestation and defect last noted at ≥6 weeks of age. 2) If ≥36 weeks gestation and defect last noted <6 weeks of age, code only if the PDA was treated (e.g. by ligation or indomethacin) or if another reportable defect is present. 3) Never code if <36 weeks gestation or if treated with prostaglandins regardless of gestational age.
10/14/92	T # 745.500	Nonclosure of foramen ovale, NOS Patent foramen ovale (PFO) 1) Always code if ≥36 weeks of gestation and defect last noted at ≥6 weeks of age. 2) If ≥36 weeks gestation and defect last noted <6 weeks of age, code only if another reportable defect is present. 3) Never code if <36 weeks gestation regardless of presence of other defects.
	T 753.700	Patent urachus
	744.820	Patulous lips (wide lips)
8/1/93	747.325	Peripheral pulmonic stenosis (PPS) murmur - <u>do</u> collect if PPS documented by echocardiogram
	270.100	Phenylketonuria (PKU)

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Nonreportable birth defects

**Alphabetical - Conditions Which may be Included Under Certain Conditions**

<u>Revised/ Changed Date</u>		<u>Code</u>	<u>Description</u>
		685.100	Pilonidal or sacral dimple
		744.230	Pixie-like ear
		744.230	Pointed ear
		755.006	Polydactyly in blacks (postaxial, type B), includes only skin tags on hands or feet. All other types of postaxial polydactyly (i.e. extra finger with bone, nail, etc.) should always be coded.
		744.246	Posteriorly rotated ears
		744.410	Preauricular sinus, cyst or pit
		744.110	Preauricular tags
	T	747.680	Primary pulmonary artery hypertension
		752.450	Prominent clitoris
		277.630	Pseudocholinesterase enzyme deficiency
10/1/92	T	746.020	Pulmonary valve insufficiency or regurgitation, congenital Code cases designated as 'mild', minimal', 'trivial', or 'physiologic' only if another reportable defect is present. Code all other degrees of insufficiency or regurgitation, including those where the degree is not specified, regardless of whether another reportable defect is present.
		750.500	Pylorospasm (intermittent pyloric stenosis)
		751.580	Rectal fissures
		284.000	Red cell aplasia
		744.500	Redundant neck skin folds
		755.616	Rocker-bottom feet
		685.100	Sacral dimple
1/1/96	T	754.060	Scaphocephaly, no mention of craniosynostosis Always code if ≥36 weeks gestation.
	#		If <36 weeks gestation, code only if another reportable defect is present.
1/1/96	T	216.910	Sebaceous cysts
		744.900	Short neck
		282.600	Sickle cell anemia
		757.200	Sidney line
		757.200	Simian crease (transverse palmar crease)
		747.500	Single umbilical artery
		757.390	Skin cysts
		754.040	Small fontanelle
		744.830	Small lips
1/1/96	T	757.640	Small nipple (hypoplastic) Always code if ≥36 weeks gestation.
	#		If <36 weeks gestation, code only if another reportable defect is present
10/1/92	T	759.020	Splenomegaly
7/13/92		090.000	Syphilis, congenital
		759.240	Thymic hypertrophy
		755.630	Tibial torsion
		750.000	Tongue-tie

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Nonreportable birth defects

**Alphabetical - Conditions Which may be Included Under Certain Conditions**

<u>Revised/ Changed Date</u>		<u>Code</u>	<u>Description</u>
		608.200	Torsion of spermatic cord
		608.200	Torsion of testes
10/1/92	T	746.105	Tricuspid valve insufficiency or regurgitation, congenital - Code cases designated as 'mild', minimal', 'trivial', or 'physiologic' only if another reportable defect is present. Code all other degrees of insufficiency or regurgitation, including those where the degree is not specified, regardless of whether another reportable defect is present.
		759.900	Umbilical cord atrophy
		553.100	Umbilical hernias (completely covered by skin)
1/1/93	T	752.500-	Undescended testicle (cryptorchidism)
	T	752.520	1)If < 36 weeks gestation, code only if there is a medical/surgical intervention for this problem; 2)If ≥36 weeks gestation and defect last noted at <1 year of age, code only if there was a medical/surgical intervention for this problem or if another reportable defect is present 3)Always code if ≥36 weeks gestation and defect first noted at ≥1 of age.
		748.180	Upturned nose
		743.800	Upward eye slant (mongoloid)
		752.460	Vaginal cysts
		752.480	Vaginal tags
		286.400	von Willebrand's disease
3/14/91	T	755.130	Webbed toes 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
		744.500	Webbing of neck
		748.180	Wide nasal bridge
		755.600	Widely spaced first and second toes
		757.680	Widely spaced nipples

**EXCLUSION LIST** for the MACDP

Numerical list of conditions requiring no record abstraction unless associated with a reportable defect. The addition or revision dates of the changes in the list of conditions requiring no record abstraction are shown. .

**Revised/  
 Changed  
 Date**

**Code**

**Description**

7/13/92	090.000	Syphilis congenital
216		Benign neoplasm of skin (NOTE: All benign neoplasms should be coded ONLY if another reportable code is present)
		Includes:
		blue nevus
		pigmented nevus
		papilloma
		dermatofibroma
		syringoadenoma
		*dermoid cyst
		hydrocystoma
		syringoma
		Excludes: skin of female genital organs (use 221.000), skin of male genital organs (use 222.000)
#	216.000	Skin of lip Excludes: vermilion border of lip
#	216.100	Eyelid, including canthus Excludes: cartilage of eyelid
#	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 breast Excludes: anal canal anus, NOS skin of scrotum
#	216.600	Skin of upper limb, shoulder
#	216.700	Skin of lower limb, hip
#	216.800	Other specified sites of skin Excludes: epibulbar dermoid cyst (use 743.810)

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# 216.900 Site unspecified  
**EXCLUSION LIST** for the MACDP

Numerical list of conditions requiring no record abstraction unless associated with a reportable defect. The addition or revision dates of the changes in the list of conditions requiring no record abstraction are shown. .

Revised/  
 Changed  
 Date

Code                      Description

#	216.910	Sebaceous cyst
	221.000	Benign skin neoplasm of female genital organs
	222.000	Benign skin neoplasm of male genital organs
T	243.990	Hypothyroidism, congenital (Exclude even if other defects are present only if the record specifies hypothyroidism of prematurity <36 weeks. Include other types of hypothyroidism and hypothyroidism NOS only when another reportable defect is present regardless of gestational age.
	251.200	Hypoglycemia, idiopathic
	252.100	Hypoparathyroidism, congenital
	270.100	Phenylketonuria (PKU)
	270.200	Albinism
	270.300	Maple syrup urine disease
	270.600	Argininosuccinic aciduria
	270.700	Hyperglycinemia
	271.000	Glycogen storage diseases
	275.330	Hypophosphatemic rickets
	277.000	Cystic fibrosis, with no mention of meconium ileus
	277.010	Cystic fibrosis, with mention of meconium ileus
	277.620	Alpha 1-antitrypsin deficiency
	277.630	Pseudocholinesterase enzyme deficiency
	282.000	Hereditary spherocytosis
	282.100	Hereditary elliptocytosis
	282.200	Glucose-6-phosphate dehydrogenase (G-6-PD) deficiency
	282.600	Sickle cell anemia
	284.000	Red cell aplasia
	286.000	Hemophilia
	286.400	von Willebrand's disease
	330.100	Cerebral lipidoses (e.g., Tay-Sachs, gangliosidoses, etc.)
	351.000	Facial palsy
	368.000	Esotropia
	378.000	Exotropia
	379.500	Nystagmus
3/5/90	457.800	Other specified disorder of lymphatics, including chylothorax
	520.600	Natal teeth
T	550.000-	Inguinal hernia or patent processus vaginalis never code in infants if <36 weeks gestation regardless of the presence of a reportable defect.
	550.900	NOTE: for those ≥36 weeks: in <b>males</b> , code only if another reportable defect is present; in <b>females</b> , always code even if found in isolation
	553.100	Umbilical hernias (completely covered by skin)
	608.200	Torsion of spermatic cord

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608.200 Torsion of testes  
**EXCLUSION LIST** for the MACDP  
 Nonreportable birth defects

**Numerical - Conditions Which may be Included Under Certain Conditions**

**Revised/  
 Changed  
 Date**

**Code                      Description**

	T	658.800	Amniotic bands (constricting bands, amniotic cyst)
		685.100	Pilonidal or sacral dimple
	T	743.450	Blue sclera - if <36 weeks gestation, code only if another reportable defect is present. Always code if ≥36 weeks gestation.
	T	743.630	Fused eyelids - never code if <25 weeks gestation unless another reportable defect is present
		743.650	Nasal lacrimal duct obstruction
		743.800	Brushfield spots
		743.800	Downward eye slant (antimongoloid)
		743.800	Epicanthal folds
		743.800	Upward eye slant (mongoloid)
		744.100	Accessory auricle
		744.110	Ear tags, preauricular
		744.120	Ear tags, other
		744.220	Bat ear
		744.230	Cauliflower ear
		744.230	Elfin ear, absent or decreased ear cartilage
			If <36 weeks gestation, code only if another reportable defect is present.
		744.230	Lop ear
		744.230	Pixie-like ear
		744.230	Pointed ear
		744.245	Low set ears
		744.246	Posteriorly rotated ears
		744.280	Darwin's tubercle
		744.410	Preauricular sinus, cyst or pit
		744.500	Redundant neck skin folds
		744.500	Webbing of neck
		744.820	Macrocheilia (big lips)
		744.820	Patulous lips (wide lips)
		744.830	Microcheilia (small lips)
		744.900	Short neck
		745.500	Nonclosure of foramen ovale, NOS (see PFO)
10/14/92	T	745.500	Patent foramen ovale (PFO)
			1)Always code if ≥36 weeks of gestation and defect last noted at ≥6 weeks of age.
			2)If ≥36 weeks gestation and defect last noted <6 weeks of age, code only if another reportable defect is present.
			3)Never code if <36 weeks gestation regardless of presence of other defects.
10/1/92	T	746.020	Pulmonary valve insufficiency or regurgitation, congenital -
			Code cases designated as 'mild', 'minimal', 'trivial', or 'physiologic' only if another reportable defect is present. Code all other degrees of insufficiency or regurgitation, including those where the degree is not

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 T = Rev. 6/04  
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specified, regardless of whether another reportable defect is present.

**EXCLUSION LIST** for the MACDP  
 Nonreportable birth defects

**Numerical - Conditions Which may be Included Under Certain Conditions**

<u>Revised/ Changed Date</u>		<u>Code</u>	<u>Description</u>
10/1/92	T	746.105	Tricuspid valve insufficiency or regurgitation, congenital - Code cases designated as 'mild', minimal', 'trivial', or 'physiologic' only if another reportable defect is present. Code all other degrees of insufficiency or regurgitation, including those where the degree is not specified, regardless of whether another reportable defect is present.
10/1/92	T	746.400	Aortic valve insufficiency or regurgitation, congenital - Code cases designated as 'mild', minimal', 'trivial', or 'physiologic' only if another reportable defect is present. Code all other degrees of insufficiency or regurgitation, including those where the degree is not specified, regardless of whether another reportable defect is present.
10/1/92	T	746.600	Mitral valve insufficiency or regurgitation, congenital - Code cases designated as 'mild', minimal', 'trivial', or 'physiologic' only if another reportable defect is present. Code all other degrees of insufficiency or regurgitation, including those where the degree is not specified, regardless of whether another reportable defect is present.
		746.860	Cardiomegaly, congenital NOS
		746.990	Heart murmur - if chart review does not confirm a heart defect within 6 months, do not code as a defect even if other codable defects are present
10/14/92	T	747.000	Patent ductus arteriosus (PDA) 1) Always code if ≥36 weeks of gestation and defect last noted at ≥6 weeks of age. 2) If ≥36 weeks gestation and defect last noted <6 weeks of age, code only if the PDA was treated (e.g. by ligation or indomethacin) or if another reportable defect is present. 3) Never code if <36 weeks gestation or if treated with prostaglandins regardless of gestational age.
8/1/93		747.325	Peripheral pulmonic stenosis (PPS) murmur - <u>do</u> collect if PPS documented by echocardiogram
		747.500	Single umbilical artery
		747.680	Primary pulmonary artery hypertension
		778.000	Ascites or anasarca. Includes: hydrops fetalis
		748.180	Flat bridge of nose

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Revised 5/07 (EXCL1088)  
 Replaces 6/93 Exclusion List

748.180 Upturned nose  
 748.180 Wide nasal bridge

**EXCLUSION LIST** for the MACDP  
 Nonreportable birth defects

**Numerical - Conditions Which may be Included Under Certain Conditions**

**Revised/  
 Changed**

		<u>Date</u>	<u>Code</u>	<u>Description</u>
3/4/91	T	748.510		Hypoplasia of lung; pulmonary hypoplasia - exclude if isolated defect in infants <36 weeks gestation.
		750.000		Tongue-tie
3/4/91		750.240		High arched palate
		750.500		Pylorospasm (intermittent pyloric stenosis)
		751.010		Meckel's diverticulum
		751.580		Rectal fissures
		751.620		Hepatomegaly
		752.430		Imperforate hymen
		752.440		Fusion of vulva
1/1/96	T	752.440		Hypoplastic labia majora -if <36 weeks gestation, code only if another reportable defect is present. Always code if ≥36 weeks gestation.
3/4/91		752.450		Prominent clitoris
		752.460		Vaginal cysts
		752.480		Vaginal tags
		752.480		Hymenal tags
1/1/93	T	752.500-		Undescended testicle (cryptorchidism)
	T	752.520		1)If < 36 weeks gestation, code only if there is a medical/surgical intervention for this problem; 2)If ≥36 weeks gestation and defect last noted at <1 year of age, code only if there was a medical/surgical intervention for this problem or if another reportable defect is present 3)Always code if ≥36 weeks gestation and defect first noted at ≥1 of age.
1/1/93		752.520		Cryptorchidism (see undescended testicle)
		752.810		Hypoplastic scrotum - exclude if secondary to undescended testes
		753.700		Patent urachus
		754.020		Bent nose, deviation of nasal septum
1/1/96	T	754.030		Dolichocephaly - if <36 weeks gestation, code only if another reportable defect is present. Always code if ≥36 weeks gestation.
1/1/93		754.040		Fontanelle (large or small)
1/1/96		754.060		Scaphocephaly, no mention of craniosynostosis If <36 weeks gestation, code only if another reportable defect is present. Always code if ≥36 weeks gestation.
1/1/93		754.520		Metatarsus varus or adductus
		755.006		Polydactyly in blacks (postaxial, type B), includes only skin tags on hands or feet. All other types of postaxial polydactyly (i.e. extra finger with bone, nail, etc.) should always be coded.
3/14/91	T	755.130		Webbed toes Code webbing of the second and third toes only if

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Revised 5/07 (EXCL1088)  
 Replaces 6/93 Exclusion List

another reportable defect is present. Always code  
 webbing of other toes regardless of whether  
 another reportable defect is present

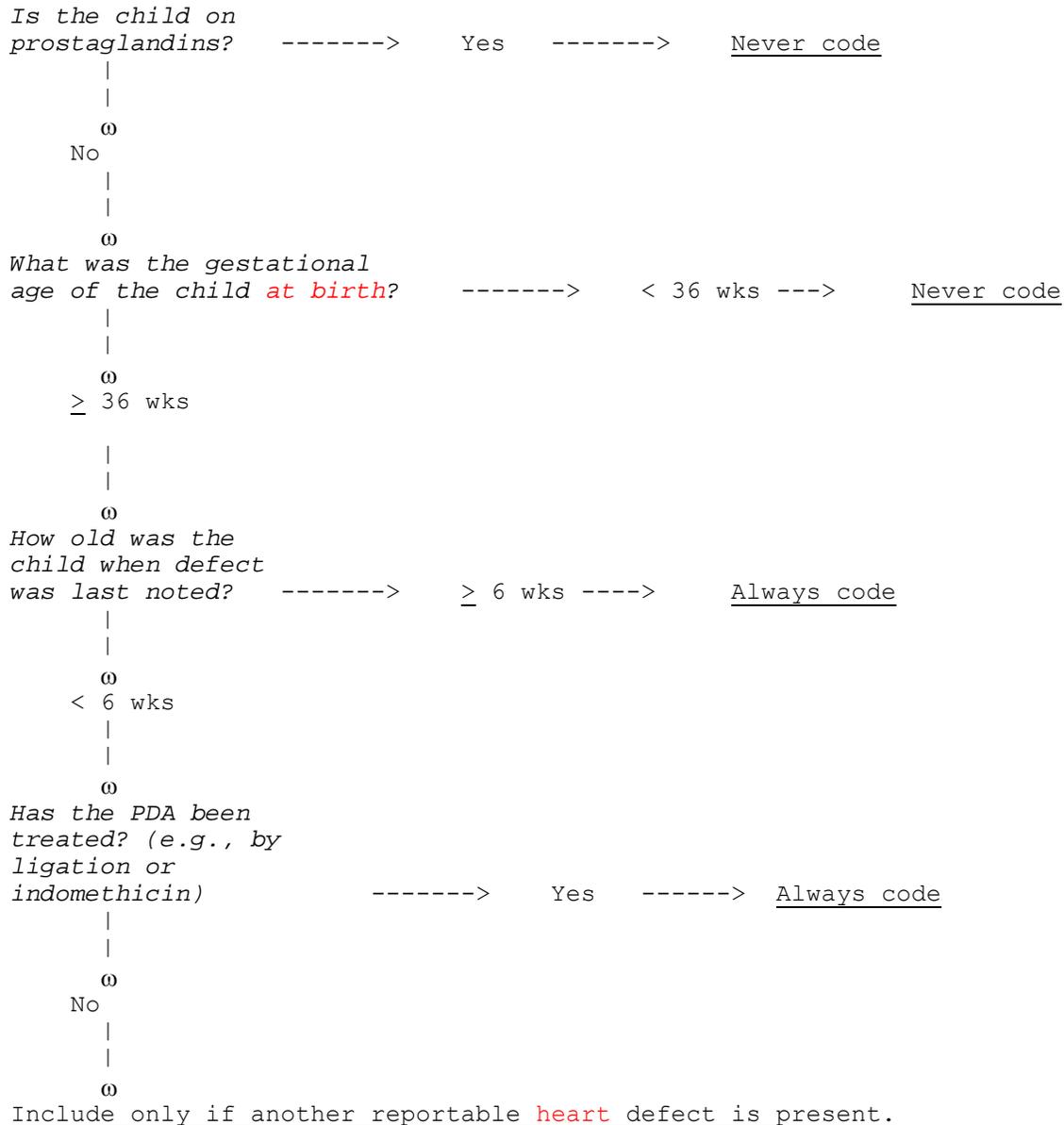
**EXCLUSION LIST** for the MACDP  
 Nonreportable birth defects

**Numerical - Conditions Which may be Included Under Certain Conditions**

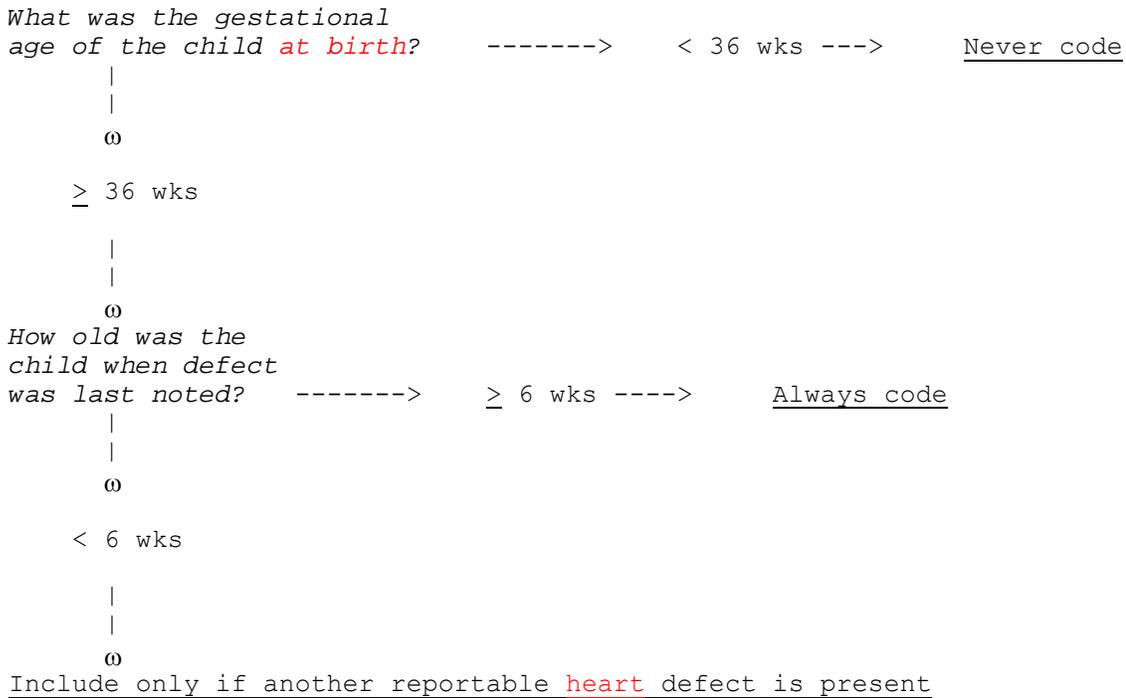
<u>Revised/ Changed</u>	<u>Date</u>	<u>Code</u>	<u>Description</u>
		755.500	Clinodactyly (incurving of fifth finger)
		755.500	Long fingers and toes
		755.600	Overlapping toes
		755.600	Widely spaced first and second toes
		755.616	Rocker-bottom feet
		755.630	Tibial torsion
		756.080	Occiput, flat or prominent
		756.200	Cervical rib
		757.200	Sidney line
		757.200	Simian crease (transverse palmar crease)
		757.310	Anal tags
		757.380	Flammeus nevus or port wine stain
		757.385	Birth mark, NOS
		757.386	Mongolian spots
		757.390	Cafe au lait spots
		757.390	Skin cysts
		757.450	Lanugo, excessive or persistent
	1/1/96	T 757.640	Small nipple (hypoplastic) If <36 weeks gestation, code only if another reportable defect is present. Always code if ≥36 weeks gestation.
tag)	9/10/90	757.650	Accessory nipple (supernumerary nipple, or skin
		757.680	Widely spaced nipples
		759.020	Splenomegaly
		759.240	Thymic hypertrophy
		759.900	Umbilical cord atrophy
		767.600	Erb's palsy
		777.100	Meconium plug
		777.600	Meconium peritonitis
		778.000	Ascites or anasarca, congenital
		778.600	Hydrocele, congenital

R = Rev. 6/07  
 N = Rev. 5/07  
 T = Rev. 6/04  
 \* = code created by CDC  
 # = on the MACDP Excl List

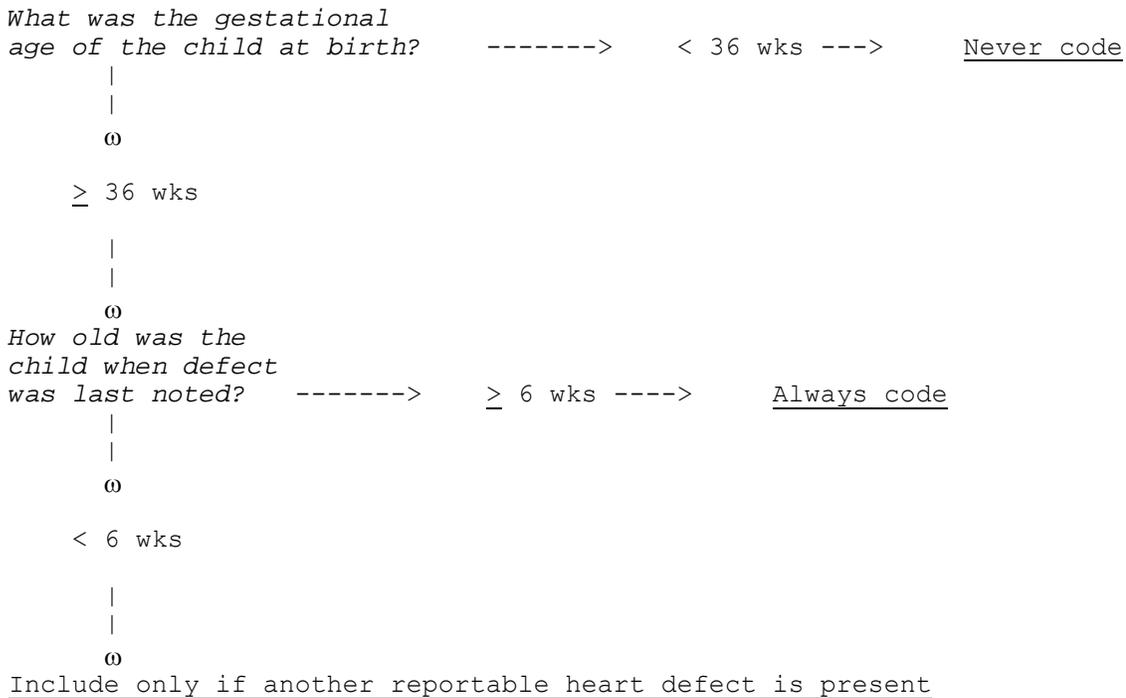
## MACDP Decision Tree for Determining Whether to Include Patent Ductus Arteriosus (PDA)



## MACDP Decision Tree for Determining Whether to Include Patent Foramen Ovale (PFO)



## MACDP Decision Tree for Determining Whether to Include Peripheral Pulmonary Stenosis (PPS)



May 22, 1996