ATTACHMENT 1

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

Doc. No. 6digit88
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.

S = Rev. 8/07
R = Rev. 6/07
N = Rev. 5/07
T = Rev. 6/04
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# = on the MACDP Excl List

Anencephalus and Similar Anomalies

740.0 Anencephalus

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

740.1 Craniorachischisis

740.100 Craniorachischisis

740.2 Iniencephaly

740.200 Closed iniencephaly
740.210 Open iniencephaly
740.290 Unspecified iniencephaly

741 Spina Bifida

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

741.0 Spina Bifida with Hydrocephalus

741.000 Spina bifida aperta, any site, with hydrocephalus

741.010 Spina bifida cystica, any site, with hydrocephalus and Arnold-Chiari malformation
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

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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, meningomyelocele)

742 Other Congenital Anomalies of Nervous System

742.0 Encephalocele

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

742.1 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

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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 oudated 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: cystalline
sub-cystalline
subarterial
conoventricular

N 745.485 Perimembranous VSD
Includes: membranous VSD

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 ≥36 weeks of gestation at birth and defect last noted at ≥6 weeks of age.
2)If ≥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 <= 4mm, 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 <= 4mm, assume it is a PFO and follow the coding instructions for 745.500.
2)If the defect size by echo is > 4mm, 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 crieria, 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 <= 4mm, 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

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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)

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 ≥36 weeks of gestation at birth and defect last noted at ≥6 weeks of age.

2)If ≥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 indomethicin) 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 ≥36 weeks of gestation at birth and defect last noted at ≥6 weeks of age.
2)If ≥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 ≥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

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# = on the MACDP Excl List

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 Megalogastria
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

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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

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 ≥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 Trigonocephaly, no mention of craniosynostosis
Always code if ≥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
turricephaly
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

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
macrodactylia
brachydactyly
triphalangeal 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

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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

755.900 Unspecified anomalies of unspecified limb

756 Other Congenital Musculoskeletal Anomalies

756.0 Anomalies of skull and face bones

Excludes: skull and face deformities in 754
Pierre Robin sequence (use 524.080)

756.000 Craniosynostosis, NOS
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
Hallermann-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

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: lipochondrodystrophy (use 277.510)

756.5 Osteodystrophies

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

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 anonychia
Absent nails
757.510 Enlarged or hypertrophic nails
757.515 Onychauxis
757.516 Pachyonychia
757.520 Congenital koilonychia
757.530 Congenital leukonychia
757.540 Club nail
757.580 Other specified anomalies of nails
757.585 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
T		Trisomy 18 or Edwards syndrome, but no cytogenetics result in record)
	758.295	Edwards phenotype - normal karyotype
T	758.298	Edwards syndrome suspected, cytogenetics pending

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

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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
Bonneville-Ullrich syndrome, NOS

758.7 Klinefelter syndrome

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

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)

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 Tuberous sclerosis

759.500 Tuberous 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

Other Specified Codes Used in Metro Atlanta Congenital Defects Program

List ordered alphabetically

524.000 Abnormalities of jaw size
micrognathia
macrognathia T

255.200 Adrenogenital syndrome

\# 270.200 Albinism

\# 277.620 Alpha-1 antitrypsin deficiency

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

\# 270.600 Arginosuccinic aciduria

\# 778.000 Ascites, congenital

216 Benign neoplasm of skin

T **(NOTE: All neoplasms should be coded ONLY if another reportable code is present)**

Includes: blue nevus pigmented nevus
papilloma dermatofibroma
syringoadenoma hydrocystoma

* * dermoid cyst syringoma

Excludes: skin of female genital organs (use 221.000),
skin of male genital organs (use 222.000)

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: vermillion border of lip

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

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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 ≥ 4cm in diameter or desribed 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 desribed 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: lipochondrodystrophy
#	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 -550.900	Inguinal hernia or patent processus vaginalis never 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

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 ≥ 4cm in diameter or desribed 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 ≥ 4cm in diameter or desribed 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

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 with no mention of meconium ileus
#	277.010	Cystic fibrosis with mention of meconium ileus
	277.400	Disorders of bilirubin excretion
	277.510	Hurler syndrome Includes: lipochondrodystrophy
#	277.620	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
	#	550.000-550.900	Inguinal hernia or patent processus vaginalis never 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

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			Code	Description
			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: vermillion 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- 550.900 550.901 550.902	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: In **males,** code only if another reportable defect is present; In **females**, always code even if found in isolation
		757.450	Lanugo, excessive or persistent
		754.040	Large fontanelle

Alphabetical - Conditions Which may be Included Under Certain Conditions

Revised/ Changed Date		Code	Description
		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 indomethicin) 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 - do collect if PPS documented by echocardiogram
		270.100	Phenylketonuria (PKU)

Alphabetical - Conditions Which may be Included Under Certain Conditions

Revised/ Changed Date		Code	Description
		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

Nonreportable birth defects

Alphabetical - Conditions Which may be Included Under Certain Conditions

Revised/ Changed Date		Code	Description
		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

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

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

Revised/ Changed Date		Code	Description
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 indomethicin) 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 - do 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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	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		Date	Code	Description
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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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

Revised/ Changed Date		Code	Description
		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.
9/10/90		757.650	Accessory nipple (supernumerary nipple, or skin tag)
		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

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

Is the child on prostaglandins? -------> Yes -------> Never code

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No

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What was the gestational age of the child at birth? -------> < 36 wks ---> Never code

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≥ 36 wks

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How old was the child when defect was last noted? -------> ≥ 6 wks ----> Always code

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< 6 wks

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Has the PDA been treated? (e.g., by ligation or indomethicin) -------> Yes ------> Always code

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No

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Include only if another reportable heart defect is present.

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

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

What was the gestational age of the child at birth? ------> < 36 wks ---> Never code

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≥ 36 wks

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How old was the child when defect was last noted? ------> ≥ 6 wks ----> Always code

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< 6 wks

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Include only if another reportable heart defect is present

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

What was the gestational age of the child at birth? -------> < 36 wks ---> Never code

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≥ 36 wks

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How old was the child when defect was last noted? -------> ≥ 6 wks ----> Always code

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< 6 wks

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Include only if another reportable heart defect is present

May 22, 1996