

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 06/04
Replaces Version 06/93

Explanation of 6-Digit Code

6th Digit Code - Master

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

Notes:

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

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

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

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

CONGENITAL ANOMALIES

Anencephalus and Similar Anomalies

740.0 Anencephalus

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

740.1 Craniorachischisis

- 740.100 Craniorachischisis

740.2 Iniencephaly

- 740.200 Closed iniencephaly
- 740.210 Open iniencephaly
- 740.290 Unspecified iniencephaly

741 Spina Bifida

Includes: Spina bifida aperta (open lesions)
myelocele
rachischisis

Spina bifida cystica (closed lesions)
meningocele
meningomyelocele
myelomeningocele

Excludes: Spina bifida occulta (see 756.100)
craniorachischisis (see 740.100)

741.0 Spina Bifida with Hydrocephalus

- 741.000 Spina bifida aperta, any site, with hydrocephalus
- 741.010 Spina bifida cystica, any site, with hydrocephalus and Arnold-Chiari malformation
Arnold-Chiari malformation, NOS
- 741.020 Spina bifida cystica, any site, with stenosed aqueduct of Sylvius
- 741.030 Spina bifida cystica, cervical, with unspecified hydrocephalus
Spina bifida cystica, cervical, with hydrocephalus but without mention of Arnold-Chiari malformation or aqueduct stenosis
- 741.040 Spina bifida cystica, thoracic, with unspecified hydrocephalus, no mention of Arnold-Chiari
- 741.050 Spina bifida cystica, lumbar, with unspecified hydrocephalus, no mention of Arnold-Chiari
- 741.060 Spina bifida cystica, sacral, with unspecified hydrocephalus, no mention of Arnold-Chiari
- 741.070 Spina bifida of any site with hydrocephalus of late onset
- 741.080 Other spina bifida, meningocele of specified site with hydrocephalus
- 741.085 Spina bifida, meningocele, cervicothoracic, with

- hydrocephalus
- 741.086 Spina bifida, meningocele thoracolumbar, with hydrocephalus
- 741.087 Spina bifida, meningocele, lumbosacral with hydrocephalus
- 741.090 Spina bifida of any unspecified type with hydrocephalus

741.9 Spina bifida without mention of hydrocephalus

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

742 Other Congenital Anomalies of Nervous System

742.0 Encephalocele

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

742.1 Microcephalus

- 742.100 Microcephalus

742.2 Reduction deformities of brain

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

742.3 Congenital hydrocephalus

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

- 742.300 Anomalies of aqueduct of Sylvius
- 742.310 Atresia of foramina of Magendie and Luschka
Dandy-Walker syndrome
- 742.320 Hydranencephaly
- 742.380 Other specified hydrocephaly
Includes: communicating hydrocephaly
- # 742.385 Hydrocephalus secondary to intraventricular hemorrhage (IVH) or CNS bleed

742.390 Unspecified hydrocephaly, NOS

742.4 Other specified anomalies of brain

742.400 Enlarged brain and/or head
 megalocephaly
 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

743.5 Congenital anomalies of posterior segment

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

743.6 Congenital anomalies of eyelids, lacrimal system, and orbit

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

743.8 Other specified anomalies of eye

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

743.9 Unspecified anomalies of eye

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

744 Congenital Anomalies of Ear, Face, and Neck

744.0 Anomalies of ear causing impairment of hearing

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

744.1 Accessory auricle

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

744.2 Other specified anomalies of ear

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

744.3 Unspecified anomalies of ear

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

744.4 Branchial cleft, cyst, or fistula; preauricular sinus

- 744.400 Branchial cleft, sinus, fistula cyst, or pit
- # 744.410 Preauricular sinus, cyst, or pit
- 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)
- 745.180 Other specified transposition of great vessels
Includes: double outlet right ventricle
- 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

- 745.400 Roger's disease
- 745.410 Eisenmenger's syndrome
- 745.420 Gerbode defect
- T** 745.480 Other specified ventricular septal defect (includes membranous,
perimembranous, muscular, mid-muscular, cystalline, sub-
cystalline, subarterial, inlet, conoreentricular)
- 745.490 VSD (ventricular septal defect), NOS
Excludes: common atrioventricular canal type (use 745.620)
- 745.498 Probable VSD

745.5 Ostium secundum type atrial septal defect

- 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.
- 745.510 Ostium (septum) secundum defect

745.520 Lutembacher's syndrome
745.580 Other specified atrial septal defect
745.590 ASD (atrial septal defect), NOS
Auricular septal defect, NOS
Partial foramen ovale
PFO vs. ASD

745.6 Endocardial cushion defects

745.600 Ostium primum defects
745.610 Single common atrium, cor triloculare biventriculare
745.620 Common atrioventricular canal
with ventricular septal defect (VSD)
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

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

746.010 Stenosis of pulmonary valve
See 746.995 if valve not specified
(e.g., "pulmonary stenosis");

Excludes: pulmonary infundibular
stenosis (use 746.830)

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.

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

746.100 Tricuspid atresia, stenosis, hypoplasia

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.
Exclude: Ebstein's anomaly (code as 746.200)

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

- T** # 746.400 Aortic valve insufficiency or regurgitation, congenital bicuspid aortic valve.
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.
- * 746.480 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

- 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.

746.7 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)

746.810 Levocardia
746.820 Cor triatriatum
746.830 Pulmonary infundibular (subvalvular) stenosis

- 746.840 Trilogy of Fallot
- 746.850 Anomalies of pericardium
- # 746.860 Anomalies of myocardium
 - cardiomegaly, congenital, NOS
 - cardiomyopathy, congenital
 - cardiomyopathy, hypertrophic
- 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
- 746.886 Ventricular hypertrophy (right or left)
- 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)
- 746.995 "Pulmonic" or "pulmonary" atresia, stenosis, or hypoplasia, NOS
 - (no mention of valve or artery)

747 Other Congenital Anomalies of Circulatory System

- T** # 747.000 Patent ductus arteriosus (PDA)
 - 1) Always code if ≥ 36 weeks of gestation and defect last noted at ≥ 6 weeks of age.
 - 2) If ≥ 36 weeks gestation and defect last noted < 6 weeks of age, code only if the PDA was treated (e.g. by ligation or indomethacin) or if another reportable defect is present.
 - 3) Never code if < 36 weeks gestation or if treated with prostaglandins regardless of gestational age.
 - (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
 747.215 Interrupted aortic arch

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
 747.290 Unspecified anomalies of aorta

747.3 Anomalies of pulmonary artery

747.300 Pulmonary artery atresia, absence or agenesis
 Use 746.995 if artery or valve is not specified

747.310 Pulmonary artery atresia with septal defect

747.320 Pulmonary artery stenosis
 Use 746.995 if artery or valve is not specified

747.325 Peripheral pulmonary artery stenosis
 Includes: peripheral pulmonic stenosis (PPS)

peripheral pulmonic stenosis (PPS) murmur only if
 documented by echocardiogram)

747.330 Aneurysm of pulmonary artery
 dilatation of pulmonary artery

747.340 Pulmonary arteriovenous malformation or aneurysm

747.380 Other specified anomaly of pulmonary artery
 Includes: pulmonary artery hypoplasia

747.390 Unspecified anomaly of pulmonary artery

747.4 Anomalies of 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)
- T** 747.680 Other anomalies of peripheral vascular system
Includes: primary pulmonary artery hypertension ONLY if it
occurs with another reportable defect
- 747.690 Unspecified anomalies of peripheral vascular system

747.8 Other specified anomalies of circulatory system

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

747.9 Unspecified anomalies of circulatory system

- 747.900 Unspecified anomalies of circulatory system

748 Congenital Anomalies of Respiratory System

748.0 Choanal atresia

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

748.1 Other anomalies of nose

748.100 Agenesis or underdevelopment of nose
748.110 Accessory nose
748.120 Fissured, notched, or cleft nose
748.130 Sinus wall anomalies
748.140 Perforated nasal septum
748.180 Other specified anomalies of nose
flat bridge of nose
wide nasal bridge
small nose and nostril
absent nasal septum
748.185 Tubular nose, single nostril, proboscis
748.190 Unspecified anomalies of nose
Excludes: congenital deviation of the nasal
septum (use 754.020)

748.2 Web of larynx

748.205 Web of larynx-glottic
748.206 Web of larynx-subglottic
748.209 Web of larynx-NOS

748.3 Other anomalies of larynx, trachea, and bronchus

748.300 Anomalies of larynx and supporting cartilage
T 748.310 Congenital subglottic stenosis - Never code if chart states
the condition was acquired or secondary to endotracheal (ET)
intubation or ventilation
748.330 Other anomalies of trachea
Excludes: vascular ring compression of the
trachea (use 747.250)
748.340 Stenosis of bronchus
748.350 Other anomalies of bronchus
748.360 Congenital laryngeal stridor, NOS
748.380 Other specified anomalies of larynx and bronchus
748.385 Cleft larynx, laryngotracheoesophageal cleft
748.390 Unspecified anomalies of larynx, trachea, and bronchus

748.4 Congenital cystic lung

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

748.5 Agenesis or aplasia of lung

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

748.6 Other anomalies of lung

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

748.8 Other specified anomalies of respiratory system

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

748.9 Unspecified anomalies of respiratory system

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

749 Cleft Palate and Cleft Lip

749.0 Cleft palate alone

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

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

749.1 Cleft lip alone

Includes: alveolar ridge cleft
cleft gum
harelip

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

749.2 Cleft lip with cleft palate

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

750 Other Congenital Anomalies of Upper Alimentary Tract

750.000 Tongue tie
Ankyloglossia

750.1 Other anomalies of tongue

Excludes: protruding tongue (never a defect)

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

750.2 Other specified anomalies of mouth and pharynx

750.200 Pharyngeal pouch
750.210 Other pharyngeal anomalies
750.230 Other anomalies of salivary glands or ducts
750.240 High arched palate
750.250 Other anomalies of palate
750.260 Lip fistulae or pits
750.270 Other lip anomalies
Includes: notched lip, prominent philtrum,
long philtrum
Excludes: cleft lip (see 749)
750.280 Other specified anomalies of mouth and pharynx
Excludes: receding jaw (see 524.0)
large and small mouth (see 744.8)

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

750.300 Esophageal atresia without mention of T-E fistula
750.310 Esophageal atresia with mention of T-E fistula
750.320 Tracheoesophageal fistula without mention of esophageal atresia
750.325 Tracheoesophageal fistula - "H" type
750.330 Bronchoesophageal fistula with or without mention of esophageal atresia
750.340 Stenosis or stricture of esophagus
750.350 Esophageal web
750.380 Other tracheoesophageal anomalies

750.4 Other specified anomalies of esophagus

- 750.400 Congenital dilatation of esophagus
giant esophagus
- 750.410 Displacement of esophagus
- 750.420 Diverticulum of esophagus
esophageal pouch
- 750.430 Duplication of esophagus
- 750.480 Other specified anomalies of esophagus

750.5 Congenital hypertrophic pyloric stenosis

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

750.6 Congenital hiatus hernia

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

750.7 Other specified anomalies of stomach

- 750.700 Microgastria
- 750.710 Megalogastrica
- 750.720 Cardiospasm
achalasia of cardia, congenital
- 750.730 Displacement or transposition of stomach
- 750.740 Diverticulum of stomach
- 750.750 Duplication of stomach
- 750.780 Other specified anomalies of stomach

750.8 Other specified anomalies of upper alimentary tract

- 750.800 Other specified anomalies of upper alimentary tract

750.9 Unspecified anomalies of upper alimentary tract

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

751 Other Congenital Anomalies of Digestive System

751.0 Meckel's diverticulum

- 751.000 Persistent omphalomesenteric duct
persistent vitelline duct
- # 751.010 Meckel's diverticulum

751.1 Atresia and stenosis of small intestine

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

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

- 751.200 Stenosis, atresia or absence of large intestine
Stenosis, atresia or absence of appendix
- 751.210 Stenosis, atresia or absence of rectum with fistula
- 751.220 Stenosis, atresia or absence of rectum without mention of
fistula
- 751.230 Stenosis, atresia or absence of anus with fistula
Includes: imperforate anus with fistula
- 751.240 Stenosis, atresia or absence of anus without mention of fistula
Includes: imperforate anus without fistula

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

- 751.300 Total intestinal aganglionosis
- 751.310 Long-segment Hirschsprung's disease; aganglionosis beyond the
rectum
- 751.320 Short-segment Hirschsprung's disease; aganglionosis involving
no more than the anal sphincter and the rectum
- 751.330 Hirschsprung's disease, NOS
- 751.340 Congenital megacolon
congenital macrocolon, not aganglionic

751.4 Anomalies of intestinal fixation

- 751.400 Malrotation of cecum and/or colon
- 751.410 Anomalies of mesentery
- 751.420 Congenital adhesions or bands of omentum and peritoneum; Ladd's
bands
- 751.490 Other specified and unspecified malrotation
- 751.495 Malrotation of small intestine alone

751.5 Other anomalies of intestine

- 751.500 Duplication of anus, appendix, cecum, or intestine
enterogenous cyst
- 751.510 Transposition of appendix, colon, or intestine
- 751.520 Microcolon
- 751.530 Ectopic (displaced) anus
- 751.540 Congenital anal fistula
- 751.550 Persistent cloaca

- * 751.560 Duodenal web
- # 751.580 Other specified anomalies of intestine
Includes: rectal fissures
- 751.590 Unspecified anomalies of intestine

751.6 Anomalies of gallbladder, bile ducts, and liver

- 751.600 Absence or agenesis of liver, total or partial
- 751.610 Cystic or fibrocystic disease of liver
- # 751.620 Other anomalies of liver
hepatomegaly
hepatosplenomegaly (also use code 759.020)
Excludes: Budd-Chiari (use 453.000)
- 751.630 Agenesis or hypoplasia of gallbladder
- 751.640 Other anomalies of gallbladder
duplication of gallbladder
- 751.650 Agenesis or atresia of hepatic or bile ducts
Includes: biliary atresia
Excludes: congenital or neonatal hepatitis
(use 774.480 or 774.490)
- 751.660 Choledochal cysts
- 751.670 Other anomalies of hepatic or bile ducts
- 751.680 Anomalies of biliary tract, NEC

751.7 Anomalies of pancreas

Excludes: fibrocystic disease of pancreas (277.000)
diabetes mellitus,
congenital
neonatal

- 751.700 Absence, agenesis or hypoplasia of pancreas
- 751.710 Accessory pancreas
- 751.720 Annular pancreas
- 751.730 Ectopic pancreas
- 751.740 Pancreatic cyst
- 751.780 Other specified anomalies of pancreas
- 751.790 Unspecified anomalies of pancreas

751.8 Other specified anomalies of digestive system

- 751.800 Absence of alimentary tract, NOS
(complete or partial)
- 751.810 Duplication of alimentary tract
- 751.820 Ectopic digestive organs, NOS
- 751.880 Other specified anomalies of digestive system

751.9 Unspecified anomalies of digestive system

- 751.900 Unspecified anomalies of digestive system
congenital of digestive system, NOS
anomaly, NOS
deformity, NOS
obstruction, NOS

- 752 Congenital Anomalies of Genital Organs**
 Excludes: congenital hydrocele (778.600)
 testicular feminization syndrome (257.800)
 syndromes associated with anomalies in
 number and form of chromosomes (758)
- 752.0 Anomalies of ovaries**
- 752.000 Absence or agenesis of ovaries
 - 752.010 Streak ovary
 - 752.020 Accessory ovary
 - 752.080 Other specified anomalies of ovaries
 - 752.085 Multiple ovarian cysts
 - 752.090 Unspecified anomalies of ovaries
- 752.1 Anomalies of fallopian tubes and broad ligaments**
- 752.100 Absence of fallopian tube or broad ligament
 - 752.110 Cyst of mesenteric remnant
 epoophoron cyst
 cyst of Gartner's duct
 - 752.120 Fimbrial cyst
 parovarian cyst
 - 752.190 Other and unspecified anomalies of fallopian tube
 and broad ligaments
- 752.2 Doubling of uterus**
- 752.200 Doubling of uterus
 doubling of uterus (any degree) or
 associated with doubling of cervix and
 vagina
- 752.3 Other anomalies of uterus**
- 752.300 Absence or agenesis of uterus
 - 752.310 Displaced uterus
 - 752.320 Fistulae involving uterus with digestive or
 urinary tract
 Includes: uterointestinal fistula
 uterovesical fistula
 - 752.380 Other anomalies of uterus
 bicornuate uterus
 unicornis uterus
 - 752.390 Unspecified anomalies of uterus
- 752.4 Anomalies of cervix, vagina, and external female genitalia**
- 752.400 Absence, atresia or agenesis of cervix
 - 752.410 Absence or atresia of vagina, complete or partial
 - 752.420 Congenital rectovaginal fistula
 - # 752.430 Imperforate hymen
 - # 752.440 Absence or other anomaly of vulva
 fusion of vulva
 hypoplastic labia majora - Always code if ≥36weeks gestation. If
 <36 weeks gestation, code only if another reportable defect is
 present.

- # 752.450 Absence or other anomaly of clitoris
Includes: clitoromegaly
enlarged clitoris
clitoral hypertrophy
prominent clitoris
- # 752.460 Embryonal cyst of vagina
- 752.470 Other cyst of vagina, vulva, or canal of Nuck
- # 752.480 Other specified anomalies of cervix, vagina, or external female genitalia
Includes: vaginal tags
hymenal tags
- 752.490 Unspecified anomalies of cervix, vagina, or external female genitalia

752.5 Undescended testicle

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

752.6 Hypospadias and epispadias

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

752.7 Indeterminate sex and pseudohermaphroditism

- Excludes: pseudohermaphroditism:
female, with adrenocortical disorder (see 255.200)
male, with gonadal disorder with specified chromosomal anomaly (see 758)
- 752.700 True hermaphroditism
ovotestis
- 752.710 Pseudohermaphroditism, male
- 752.720 Pseudohermaphroditism, female
pure gonadal dysgenesis
Excludes: gonadal agenesis (758.690)
- 752.730 Pseudohermaphrodite, NOS
- 752.790 Indeterminate sex, NOS
ambiguous genitalia

752.8 Other specified anomalies of male genital organs

- 752.800 Absence of testis
monorchidism, NOS
- # 752.810 Aplasia or hypoplasia of testis and scrotum
- 752.820 Other anomalies of testis and scrotum
polyorchidism
bifid scrotum
Excludes: torsion of the testes or spermatic
cord (use #608.200)
- 752.830 Atresia of vas deferens
- 752.840 Other anomalies of vas deferens and prostate
- 752.850 Absence or aplasia of penis
- # 752.860 Other anomalies of penis
absent or hooded foreskin
redundant foreskin (never a defect)
- 752.865 Small penis, hypoplastic penis, or micropenis
- 752.870 Cysts of embryonic remnants
cyst: hydatid of Morgagni
Wolffian duct
appendix testis
- 752.880 Other specified anomalies of genital organs
microgenitalia
macrogenitalia

752.9 Unspecified anomalies of genital organs

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

753 Congenital Anomalies of Urinary System

753.0 Renal agenesis and dysgenesis

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

753.1 Cystic kidney disease

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

753.2 Obstructive defects of renal pelvis and ureter

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

753.3 Other specified anomalies of kidney

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

753.4 Other specified anomalies of ureter

- 753.400 Absence of ureter
- 753.410 Accessory ureter
double ureter, duplex collecting system
- 753.420 Ectopic ureter
- 753.480 Other specified anomalies of ureter
Includes: ureterocele
- 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
turriccephaly
oxycephaly
- * 754.090 Deformity of skull, NOS

754.1 Anomalies of sternocleidomastoid muscle

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

754.2 Certain congenital musculoskeletal deformities of spine

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

754.3 Congenital dislocation of hip

- 754.300 Congenital dislocation of hip
- 754.310 Unstable hip
prelaxation 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
 macrodactyly
 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

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 Arthrogyrosis multiplex congenita
 - Includes: distal arthrogyrosis syndrome
 - Temporarily includes: flexion contractures of individual joints
- 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
- 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: agenesi s 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
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

T

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 Trisomy 18 or Edwards syndrome, but no cytogenetics result in record)
- T** 758.295 Edwards phenotype - normal karyotype
- T** 758.298 Edwards syndrome suspected, cytogenetics pending

758.3 Autosomal deletion syndromes

- 758.300 Antimongolism syndrome
Clinical antimongolism syndrome:
karyotype - partial or total deletion of:
21
G, NOS
NOS
- 758.310 Cri du chat syndrome
Clinical Cri du chat syndrome:
karyotype - deletion of:
5
B, NOS
NOS
- 758.320 Wolff-Hirschorn syndrome
Clinical Wolff-Hirschorn syndrome:
karyotype - deletion of:
4
B, NOS
NOS
- 758.330 Deletion of long arm of 13
deletion of long arm of D, NOS
- 758.340 Deletion of long arm of E
deletion of long arm of 17 or 18
- 758.350 Deletion of short arm of E
deletion of short arm of 17 or 18
- 758.360 Monosomy G mosaicism
- 758.370 Deletion in band 11 of long arm of 22 (22q11 deletions)
Note: Code added for use with births on or after 4/1/2001
- 758.380 Other loss of autosomal material
- 758.390 Unspecified autosomal deletion syndromes

758.4 Balanced autosomal translocation in normal individual

- 758.400 Balanced autosomal translocation in normal individual

758.5 Other conditions due to autosomal anomalies

- 758.500 Trisomy 8
- 758.510 Other trisomy C syndromes
Trisomy: 6, 7, 9, 10, 11, 12, or C, NOS
- 758.520 Other total trisomy syndromes
Trisomy 22
Trisomy, NOS
- 758.530 Partial trisomy syndromes
- 758.540 Other translocations
Excludes: balanced translocation in normal individual (use 758.400)
- 758.580 Other specified anomalies of autosomes, NOS
Includes: marker autosome
- 758.585 Polyploidy
- 758.586 Triploidy
- 758.590 Unspecified anomalies of autosomes

758.6 Gonadal Dysgenesis

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

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

758.7 Klinefelter syndrome

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

758.8 Other conditions due to sex chromosome anomalies

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

758.9 Conditions due to anomaly of unspecified chromosomes

- 758.900 Mosaicism, NOS
- 758.910 Additional chromosome(s), NOS
- 758.920 Deletion of chromosome(s), NOS
- 758.930 Duplication of chromosome(s), NOS
- 758.990 Unspecified anomaly of chromosome(s)

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

759.5 Tuberos sclerosi

- 759.500 Tuberos sclerosi
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: vermilion 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

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

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

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 ≥ 4 cm in diameter or described as large, huge, or of medical significance is present.

286.000 Hemophilia (all types)
 774.490 Hepatitis, neonatal, NOS
 774.480 Hepatitis, neonatal, other specified

282.100 Hereditary elliptocytosis
 # 282.000 Hereditary spherocytosis

771.220 Herpes simplex (in utero infections only)
 Includes: encephalitis
 meningoencephalitis

202.300 Histiocytosis, malignant
 277.510 Hurler syndrome
 Includes: lipocondrodystrophy

778.600 Hydrocele, congenital
 # 270.700 Hyperglycinemia
 # 251.200 Hypoglycemia, idiopathic
 # 252.100 Hypoparathyroidism, congenital
 # 275.330 Hypophosphatemic rickets
 253.280 Hypopituitarism, congenital
 # 243.990 Hypothyroidism, congenital (Exclude even if other defects are present only if the record specifies hypothyroidism of prematurity. Other types of hypothyroidism or hypothyroidism NOS should continue to be on the routine exclusion list.)

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

208.000 Leukemia, congenital, NOS
 214 Lipoma

214.300 Lipoma, intra-abdominal organs
 214.200 Lipoma, intrathoracic organs
 214.810 Lipoma, lumbar or sacral lipoma
 paraspinal lipoma

214.100 Lipoma, other skin and subcutaneous tissue
 214.800 Lipoma, other specified sites
 214.000 Lipoma, skin and subcutaneous tissue of face
 214.400 Lipoma, spermatic cord
 214.900 Lipoma, unspecified site

457.800 Lymphatics - other specified disorders of (including chylothorax)
 524.000 Macrognathia
 # 270.300 Maple syrup urine disease
 # 777.600 Meconium peritonitis
 # 777.100 Meconium plug syndrome
 524.000 Micrognathia
 352.600 Moebius syndrome

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

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
- # 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 \geq 4cm in diameter or described as large, huge, or of medical significance is present.
 - # 228.010 Hemangioma, skin & subcutaneous, NOS
 Always code if multiple hemangiomas of any size are present, if one or more cavernous hemangiomas of any size are present, or if a single hemangioma measuring \geq 4cm in diameter or described as large, huge, or of medical significance is present.
 - 228.020 Hemangioma, intracranial (Always code regardless of size, type or number)
 - 228.030 Hemangioma, retinal (Always code regardless of size, type or number)
 - 228.040 Hemangioma, intra-abdominal (Always code regardless of size, type or number)
 - 228.090 Hemangioma, of other sites (Always code regardless of size, type or number)
 - 228.100 Cystic hygroma
 - Lymphangioma, any site
 - 237.700 Neurofibromatosis
 - 238.000 Teratoma, NOS
 - 238.010 Teratoma, head and face
 - 238.020 Teratoma, neck
 - 238.030 Teratoma, abdomen
 - 238.040 Teratoma, sacral, coccygeal
 - 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 <u>with no</u> mention of meconium ileus
# 277.010	Cystic fibrosis <u>with</u> mention of meconium ileus
277.400	Disorders of bilirubin excretion
277.510	Hurler syndrome Includes: lipochondrodystrophy
# 277.620	Alpha-1 antitrypsin deficiency
# 277.630	Pseudocholinesterase enzyme deficiency
279.110	DiGeorge syndrome
279.200	Combined immunodeficiency syndrome
# 282.000	Hereditary spherocytosis
# 282.100	Hereditary elliptocytosis
# 282.200	Glucose-6-phosphate dehydrogenase (G-6-PD) deficiency
# 282.600	Sickle cell anemia
# 284.000	Red cell aplasia
# 286.000	Hemophilia (all types)
# 286.400	von Willebrand disease
# 330.100	Cerebral lipidoses Includes: Tay-Sachs disease gangliosidosis
331.890	Familial degenerative CNS disease
335.000	Werdnig-Hoffman disease
345.600	Infantile spasms, congenital
# 351.000	Facial palsy
352.600	Moebius syndrome
362.600	Retinal degeneration, peripheral
362.700	Retinitis pigmentosa
363.200	Chorioretinitis
# 368.000	Esotropia
# 378.000	Exotropia
# 379.500	Nystagmus
425.300	Endocardial fibroelastosis
426.705	Congenital Wolfe-Parkinson-White syndrome
427.900	Cardiac arrhythmias, NEC. Never code premature atrial contractions, PACs.
453.000	Budd-Chiari, occlusion of hepatic vein
# 457.800	Other specified disorders of lymphatics (including chylothorax)

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

520.600 Natal teeth
 524.000 Abnormalities of jaw size
 micrognathia
 macrognathia
 * 524.080 Pierre Robin sequence
 # 550.000- Inguinal hernia or patent processus vaginalis never
 550.900 code in infants if <36 weeks gestation regardless of the
 presence of a reportable defect.
 NOTE: for those ≥36 weeks:
 Code in **males** only if another reportable defect is present;
 in **females**, always code even if found in isolation
 # 553.100 Umbilical hernia
 553.200 Epigastric hernia
 # 608.200 Torsion of testes or spermatic cord
T # 658.800 Amniotic bands (constricting bands, amniotic cyst)
 # 685.100 Pilonidal sinus (sacrodermal), sacral sinus, sacral dimple
 760.710 Fetal alcohol syndrome
 760.718 Probable fetal alcohol syndrome
 Includes: "facies"
 760.750 Fetal hydantoin (Dilantin) syndrome
 # 767.600 Erb's palsy

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

 771.000 Rubella, congenital
 771.090 TORCH infection, unspecified
 771.100 Cytomegalovirus (CMV)
 771.210 Toxoplasmosis
 771.220 Herpes simplex
 Includes: encephalitis
 meningoencephalitis
 771.280 Congenital infection, other specified
 Excludes: human immunodeficiency virus (HIV) infection and
 acquired immunodeficiency syndrome (AIDS)
 774.480 Hepatitis, neonatal, other specified
 774.490 Hepatitis, neonatal, NOS
 # 777.100 Meconium plug syndrome
 # 777.600 Meconium peritonitis
 # 778.000 Ascites, congenital
 # 778.600 Hydrocele, congenital

HHS:PHS: CDC:NCBDDD:DBDDD:06/16/04
 Doc. 6digit88, Version 06/04

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

EXCLUSION LIST for the MACDP
 Nonreportable birth defects

Conditions Which may be Included Under Certain Conditions

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

The following rules apply to coding these conditions:

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

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

**Revised/
 Changed
 Date**

<u>Date</u>	<u>Code</u>	<u>Description</u>
	744.100	Accessory auricle
	757.650	Accessory nipple (supernumerary nipple, or skin tag)
	270.200	Albinism
	277.620	Alpha 1-antitrypsin deficiency
	T 658.800	Amniotic bands (constricting bands, amniotic cyst)
	757.310	Anal tags
10/1/92	T 746.400	Aortic valve insufficiency or regurgitation, congenital - Code cases designated as 'mild', 'minimal', 'trivial', or 'physiologic' only if another reportable defect is present. Code all other degrees of insufficiency or regurgitation, including those where the degree is not specified, regardless of whether another reportable defect is present.
	270.600	Argininosuccinic aciduria
	T 778.000	Ascites or anasarca, congenital. Includes: hydrops fetalis
	744.220	Bat ear
	T # 216.200	Benign neoplasm of skin, ear and external auditory canal Includes: auricle ear external meatus auricular canal external canal pinna
	T # 216.100	Excludes: cartilage of ear Benign neoplasm of skin, eyelid, including canthus
	T # 216.000	Excludes: cartilage of eyelid Benign neoplasm of skin, lip
	T # 216.700	Excludes: vermilion border of lip Benign neoplasm of skin, lower limb, hip
	T # 216.300	Benign neoplasm of skin, other and unspecified parts of face

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Includes: cheek, external nose, external eyebrow, temple

EXCLUSION LIST for the MACDP
 Nonreportable birth defects

Alphabetical - Conditions Which may be Included Under Certain Conditions

**Revised/
 Changed
 Date**

Code

Description

	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
			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

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unless another reportable defect is present
EXCLUSION LIST for the MACDP
 Nonreportable birth defects

Alphabetical - Conditions Which may be Included Under Certain Conditions

**Revised/
 Changed
 Date**

Code Description

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

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9/10/90 754.520 Metatarsus varus or adductus
EXCLUSION LIST for the MACDP
 Nonreportable birth defects

Alphabetical - Conditions Which may be Included Under Certain Conditions

**Revised/
 Changed
 Date**

Code

Description

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

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744.110 Preauricular tags
EXCLUSION LIST for the MACDP
 Nonreportable birth defects

Alphabetical - Conditions Which may be Included Under Certain Conditions

**Revised/
 Changed
 Date**

		<u>Code</u>	<u>Description</u>
	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
		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

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553.100 Umbilical hernias (completely covered by skin)

EXCLUSION LIST for the MACDP
 Nonreportable birth defects

Alphabetical - Conditions Which may be Included Under Certain Conditions

**Revised/
 Changed**

<u>Date</u>		<u>Code</u>	<u>Description</u>
1/1/93	T	752.500-	Undescended testicle (cryptorchidism)
	T	752.520	1)If < 36 weeks gestation, code only if there is a medical/surgical intervention for this problem; 2)If ≥36 weeks gestation and defect last noted at <1 year of age, code only if there was a medical/surgical intervention for this problem or if another reportable defect is present 3)Always code if ≥36 weeks gestation and defect first noted at ≥1 of age.
		748.180	Upturned nose
		743.800	Upward eye slant (mongoloid)
		752.460	Vaginal cysts
		752.480	Vaginal tags
		286.400	von Willebrand's disease
3/14/91	T	755.130	Webbed toes Code webbing of the second and third toes only if another reportable defect is present. Always code webbing of other toes regardless of whether another reportable defect is present
		744.500	Webbing of neck
		748.180	Wide nasal bridge
		755.600	Widely spaced first and second toes
		757.680	Widely spaced nipples

EXCLUSION LIST for the MACDP

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

**Revised/
 Changed
 Date**

Code

Description

7/13/92	090.000	Syphilis congenital
	216	Benign neoplasm of skin (NOTE: All benign neoplasms should be coded ONLY if another reportable code is present) Includes: blue nevus pigmented nevus papilloma dermatofibroma syringoadenoma *dermoid cyst hydrocystoma syringoma Excludes: skin of female genital organs (use 221.000), skin of male genital organs (use 222.000)
#	216.000	Skin of lip Excludes: vermilion border of lip
#	216.100	Eyelid, including canthus Excludes: cartilage of eyelid
#	216.200	Ear and external auditory canal Includes: auricle ear external meatus auricular canal external canal pinna Excludes: cartilage of ear
#	216.300	Skin of other and unspecified parts of face Includes: cheek, external nose, external eyebrow, temple
#	216.400	Scalp and skin of neck
#	216.500	Skin of trunk, except scrotum Includes: axillary fold perianal skin skin of: chest wall abdominal wall groin buttock anus perineum back umbilicus breast Excludes: anal canal anus, NOS skin of scrotum
#	216.600	Skin of upper limb, shoulder
#	216.700	Skin of lower limb, hip
#	216.800	Other specified sites of skin Excludes: epibulbar dermoid cyst (use 743.810)
#	216.900	Site unspecified
#	216.910	Sebaceous cyst

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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

	221.000	Benign skin neoplasm of female genital organs
	222.000	Benign skin neoplasm of male genital organs
T	243.990	Hypothyroidism, congenital (Exclude even if other defects are present only if the record specifies hypothyroidism of prematurity <36 weeks. Include other types of hypothyroidism and hypothyroidism NOS only when another reportable defect is present regardless of gestational age.
	251.200	Hypoglycemia, idiopathic
	252.100	Hypoparathyroidism, congenital
	270.100	Phenylketonuria (PKU)
	270.200	Albinism
	270.300	Maple syrup urine disease
	270.600	Argininosuccinic aciduria
	270.700	Hyperglycinemia
	271.000	Glycogen storage diseases
	275.330	Hypophosphatemic rickets
	277.000	Cystic fibrosis, with no mention of meconium ileus
	277.010	Cystic fibrosis, with mention of meconium ileus
	277.620	Alpha 1-antitrypsin deficiency
	277.630	Pseudocholinesterase enzyme deficiency
	282.000	Hereditary spherocytosis
	282.100	Hereditary elliptocytosis
	282.200	Glucose-6-phosphate dehydrogenase (G-6-PD) deficiency
	282.600	Sickle cell anemia
	284.000	Red cell aplasia
	286.000	Hemophilia
	286.400	von Willebrand's disease
	330.100	Cerebral lipidoses (e.g., Tay-Sachs, gangliosidoses, etc.)
	351.000	Facial palsy
	368.000	Esotropia
	378.000	Exotropia
	379.500	Nystagmus
3/5/90	457.800	Other specified disorder of lymphatics, including chylothorax
	520.600	Natal teeth
T	550.000-	Inguinal hernia or patent processus vaginalis never code in infants if <36 weeks gestation regardless of the presence of a reportable defect.
	550.900	
		<u>NOTE:</u> 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
	608.200	Torsion of testes
T	658.800	Amniotic bands (constricting bands, amniotic cyst)
	685.100	Pilonidal or sacral dimple

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EXCLUSION LIST for the MACDP
 Nonreportable birth defects

Numerical - Conditions Which may be Included Under Certain Conditions

<u>Revised/ Changed Date</u>	<u>Code</u>	<u>Description</u>
	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.

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EXCLUSION LIST for the MACDP
 Nonreportable birth defects

Numerical - Conditions Which may be Included Under Certain Conditions

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

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EXCLUSION LIST for the MACDP
 Nonreportable birth defects

Numerical - Conditions Which may be Included Under Certain Conditions

Revised/
 Changed

	<u>Date</u>	<u>Code</u>	<u>Description</u>
		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 another reportable defect is present. Always code webbing of other toes regardless of whether another reportable defect is present
		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

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Revised 6/93 (EXCL1088)
 Replaces 6/89 Exclusion List

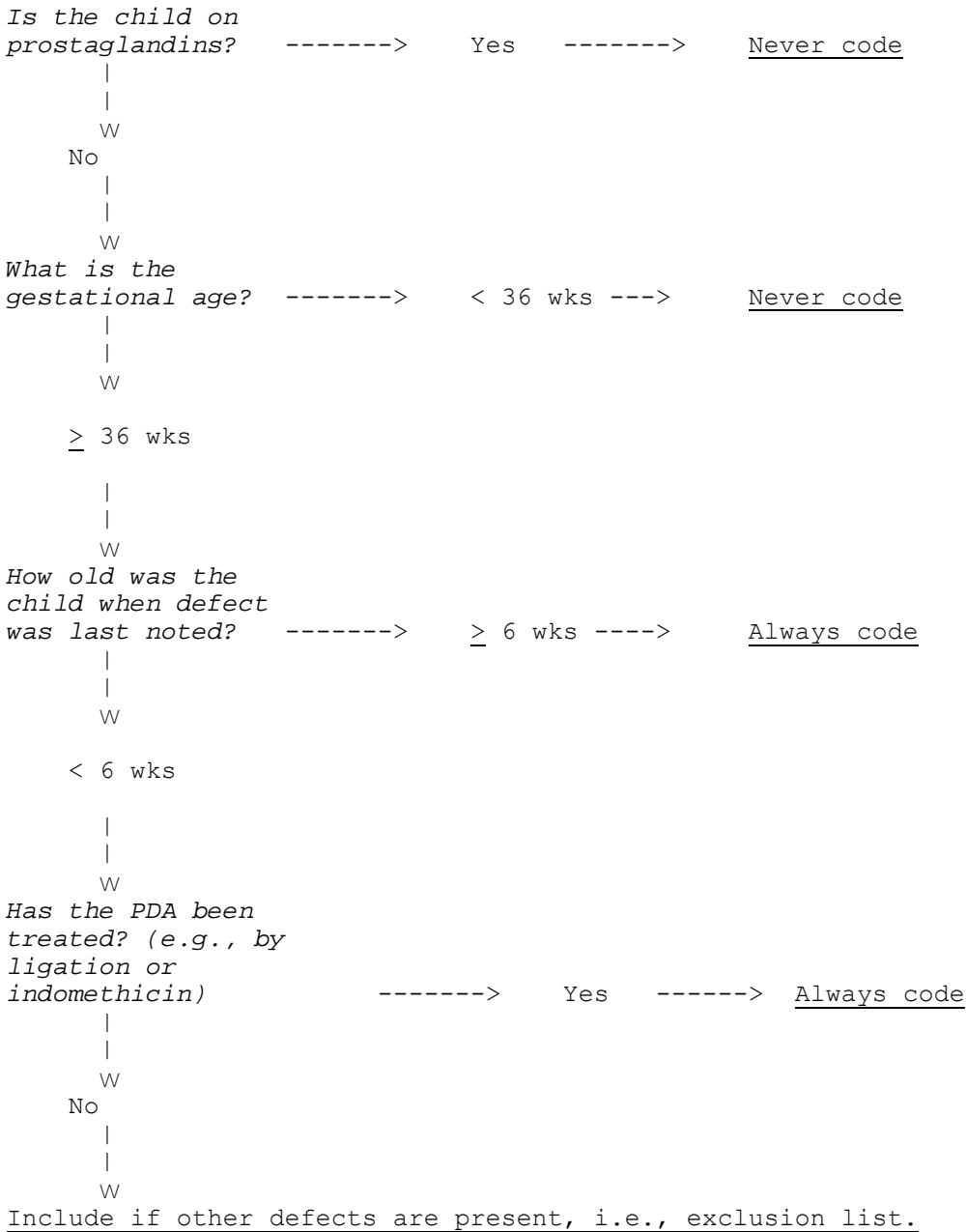
EXCLUSION LIST for the MACDP
 Nonreportable birth defects

Numerical - Conditions Which may be Included Under Certain Conditions

<u>Revised/ Changed Date</u>	<u>Code</u>	<u>Description</u>
	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

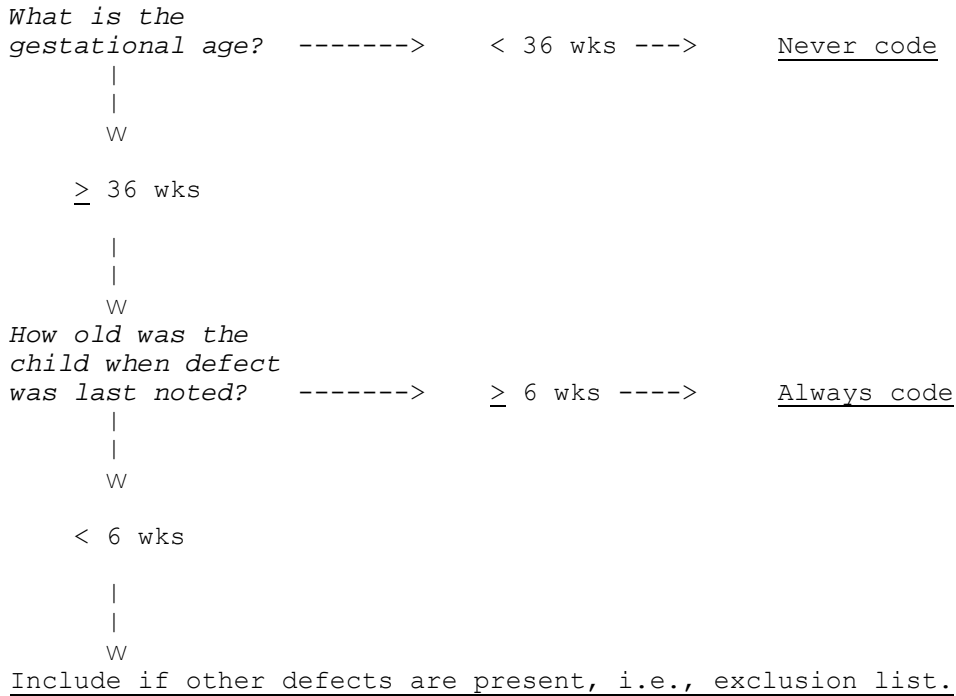
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MACDP Decision Tree for Determining Whether to Include Patent Ductus Arteriosus (PDA)



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



May 22, 1996