

Arkansas Reproductive Health Monitoring System
ARHMS web query: birth defect descriptions and codes

Birth Defects	Description	ARHMS 6-digit codes*
Any eligible birth defect	All eligible congenital malformations identified by ARHMS	740.000-759.999, plus a few others
Central Nervous System		
Any neural tube defect	Anencephaly, spina bifida, or encephalocele	740.000 – 740.100, 741.000 – 741.999, 742.000 – 742.090
Anencephalus	Absence of the skull, which results in damage to the fetal brain; this defect is incompatible with life either in the wound or shortly after birth	740.000 – 740.100
Spina bifida without anencephalus	Incomplete closure of the spinal neural tube; spinal cord or meninges may protrude through opening; surgical repair is necessary; permanent paralysis is common	741.000 – 741.999 w/o 740.000 – 740.100
Encephalocele	Defect in the skull resulting in parts of the brain protruding through an opening in the skull; surgery is usually necessary	742.000 – 742.090
Hydrocephalus without spina bifida	Abnormal fluid accumulation in the brain cavity; surgery for drainage system may be necessary	742.300 – 742.390 w/o 741.000 – 741.999
Holoprosencephaly	Abnormal development and division of the brain's hemispheres	742.260- 742.267
Eye/Ear		
Anophthalmia/microphthalmia	Absences or underdevelopment of the eyes;	743.000 – 743.104
Congenital cataract	Clouding of the lens of the eyes	743.320 – 743.326 743.350 - 743.364
Aniridia	Absences or a defect of the iris	743.420 – 743.424
Anotia/microtia	Absences or underdevelopment of the ears; surgery may help restore hearing in mild cases	744.010 – 744.014 744.210 – 744.214
Cardiovascular		
Any congenital heart defect (CHD)	Any congenital defect of the heart and directly related vessels	745.000-747.999
Any critical congenital heart defect	Any CHD likely identified by newborn screening pulse oximetry, includes common truncus, select transposition of great arteries, tetralogy of Fallot, pulmonary valve atresia, tricuspid atresia, hypoplastic left heart syndrome, total pulmonary venous return	745.000, 745.100, 745.110, 745.186, 742.200, 747.310, 746.000, 746.100, 746.700, or 747.420
Any septal heart defect	Ventricular septal defect or atrial septal defect; if defect does not close, surgery may be required later in childhood	745.400 – 745.486, 745.490, 745.599 745.510– 745.580,
Common truncus	A heart defect where the common arterial trunk fails to divide into the pulmonary artery and the aorta; immediate surgical repair is necessary	745.000

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Transposition of great arteries	A heart defect where the aorta arises from the right ventricle and pulmonary artery arises from the left ventricle (opposite of normal development); immediate surgical repair is necessary after birth	745.100, 745.110, 745.190
Tetralogy of Fallot	Four heart defects: ventricular septal defect, pulmonary valve atresia/stenosis, displacement of the aorta to the right, hypertrophy of the right ventricle; surgical repair is necessary	745.200, 747.310
Ventricular septal defect	Heart defect where there is one or more abnormal openings in the ventricular septum; may be called "a hole in the heart"	745.400 – 745.486, 745.490
Atrial septal defect	Heart defect where there is one or more abnormal openings in the atrial septum; may be called "a hole in the heart"	745.510– 745.580, 745.599
Atrioventricular septal defect (endocardial cushion defect)	A variety of septal heart defects in both the atrium and ventricular walls that results in imperfect fusion of endocardial cushions; surgical repair may occur	745.600– 745.690, 745.487
Pulmonary valve atresia and stenosis	Absences or constriction of the pulmonary valve; severe forms require surgical repair	746.000– 746.010
Tricuspid valve atresia	Absences of the tricuspid valve (opening between right atrium and right ventricle)	746.100
Ebstein's anomaly	A heart defect where the tricuspid valve is displaced downward into the right ventricle causing abnormal circulation in the heart	746.200
Aortic valve stenosis	Narrowing or stricture of the aortic valve	746.300
Hypoplastic left heart syndrome	A heart defect characterized by atresia of the aortic opening, hypoplasia of the ascending aorta, and abnormal development of the left ventricle; multiple surgical repairs or heart transplant is necessary	746.700
Coarctation of aorta	Localized narrowing of the aorta; surgical repair is usually necessary	747.100– 747.190
Total or partial anomalous pulmonary venous return	Veins carry oxygen-rich blood from the lungs to the heart are improperly attached to the heart; surgical repair is usually necessary	747.420, 747.430
Orofacial		
Any cleft lip or palate	Cleft lip or cleft palate, either isolated or in combination of both defects	749.001 – 749.290
Cleft palate without cleft lip	Failure of the fetal palate to fuse properly, forming a grooved depression in the roof of the mouth	749.001 – 749.090
Cleft lip with and without cleft palate	Failure of the fetal lip to fuse or join properly, forming a groove or fissure in the lip	749.101 – 749.290
Choanal atresia	A defect where membranous formations block the passage between the nose and the pharynx	748.010 – 748.014

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Gastrointestinal		
Esophageal atresia/ tracheoesophageal fistula	Narrowing or incomplete formation of the esophagus; abnormal passageway between the esophagus and trachea; surgical repair is necessary	750.300 – 750.330
Rectal and large intestinal atresia/stenosis	Absence, closure or constriction of the large intestine, rectum, or anus; surgical repair is usually necessary	751.200 – 751.240
Pyloric stenosis	A narrowing of the opening between the stomach and the large intestine (pyloric sphincter); surgical repair is usually necessary	750.510
Biliary atresia	Absence or underdevelopment of one or more of the ducts in the biliary tract, the passageway for bile between the liver and the small intestine	751.650
Genitourinary		
Renal agenesis/hypoplasia	Failure, or deviation, of the embryonic development of the kidneys; can be unilateral or bilateral; fatal if both kidneys are absent	753.000 – 753.010
Bladder exstrophy	Incomplete closure of the anterior wall of the bladder and abdominal cavity	753.500
Obstructive genitourinary defect	Atresia or stenosis of the urinary tract at any level	753.200 -753.290 753.600 -753.690
Hypospadias	A defect affecting males where the urinary meatus (urinary outlet) is on the underside of the penis	752.600 – 752.607, 752.620 752.625 – 752.627
Epispadias	A defect where the urinary meatus (urinary outlet) opens above the normal position	752.610
Musculoskeletal		
Any limb reduction defect	Any upper or lower limb reduction defect	755.200 – 755.394
Reduction deformity, upper limbs	Absent or partial development of the upper limbs (arm/hands)	755.200 – 755.294
Reduction deformity, lower limbs	Absent or partial development of the lower limbs (legs/feet)	755.300 – 755.394
Clubfoot	A defect where the foot is rotated internally at the ankle; may affect one or both feet	754.000- 754.504 754.600-754.604 754.730- 754.734
Gastroschisis	A defect where an abnormal opening of the abdominal wall results in protrusion of the intestines outside the body	756.710
Omphalocele	A defect where an abnormal opening at the naval results in protrusion of the intestines into the umbilicus	756.700
Congenital hip dislocation	A congenital birth defect where the ball of the femur (leg bone) is not aligned properly into the pelvis	754.300 – 754.304

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Diaphragmatic hernia	Failure of the diaphragm to form properly leaving a hole where abdominal organs may protrude into the chest cavity; Usually life-threatening and requires immediately surgery after birth	756.600 – 756.619
Craniosynostosis	Premature closing of the cranial sutures (grooves between the skull bones) before or after birth	756.000-756.024
Amniotic bands	Entrapment of fetal body parts (usually limbs or digits) by the fibrous bands in the amniotic sac during pregnancy; can result in major damage and amputation	658.800
Chromosomal		
Patau syndrome (Trisomy 13)	Extra (third) copy of chromosome 13; results in impaired facial development and severe mental disabilities; fetal and infant mortality is high	758.100 – 758.190
Down syndrome (Trisomy 21)	Extra (third) copy of chromosome 21; results in mild to severe mental disabilities, altered physical features in the face and digits, may have associated heart defects or intestinal atresia	758.000 – 758.090
Edward syndrome (Trisomy 18)	Extra (third) copy of chromosome 18; results in mental disabilities and skull malformations; heart and renal defects are common; survival beyond a few months is rare	758.200 – 758.290

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