

APPENDIX C - BIRTH DEFECTS CODES FOR ITEM 49**Code Other Lethal or Life Threatening Birth Defect**

100 Other Lethal or Life-Threatening Birth Defect which is not listed below

Code Central Nervous System Defects

100 Other lethal or life threatening CNS Defects (DESCRIBE)
101 Anencephaly
102 Meningomyelocele
103 Hydranencephaly
104 Congenital Hydrocephalus
105 Holoprosencephaly
106 Microcephaly
107 Hypopituitary
108 Septic Optic Dysplasia
109 Encephalocele

Code Congenital Heart Defects

200 Other lethal or life threatening congenital heart defects
(DESCRIBE)
201 Truncus Arteriosus
202 Transposition of the Great Vessels
203 Tetralogy of Fallot
204 Single Ventricle
205 Double Outlet Right Ventricle
206 Complete Atrio-Ventricular Canal
207 Pulmonary Atresia
208 Tricuspid Atresia
209 Hypoplastic Left Heart Syndrome
210 Interrupted Aortic Arch
211 Total Anomalous Pulmonary Venous Return
212 Coarctation of the Aorta
213 Atrial septal defect (ASD)
214 Ventricular septal defect (VSD)
215 Arrhythmias
216 Ebsteins Anomaly
217 Pericardial Effusion
218 Pulmonary Stenosis
219 Hypertrophic Cardiomyopathy

Code Gastro-Intestinal Defects

300 Other lethal or life threatening GI Defects (DESCRIBE)
301 Cleft Palate
302 Tracheo-Esophageal Fistula
303 Esophageal Atresia
304 Duodenal Atresia
305 Jejunal Atresia
306 Ileal Atresia
307 Atresia of Large Bowel or Rectum
308 Imperforate Anus
309 Omphalocele

310	Gastroschisis
311	Pyloric Stenosis
312	Annular Pancreas
313	Biliary Atresia
314	Meconium Ilius
315	Malrotation Volvulus
316	Hirschsprung's Disease

Code **Genito-Urinary Defects**

401	Bilateral Renal Agenesis
402	Bilateral Polycystic, Multicystic, or Dysplastic Kidneys
403	Obstructive Uropathy with Congenital Hydronephrosis
404	Exstrophy of the Urinary Bladder
400	Other Lethal or Life Threatening Genito-Urinary defects not listed above (DESCRIBE)

Code **Chromosomal Abnormalities**

501	Trisomy 13
502	Trisomy 18
503	Trisomy 21
504	Other Chromosomal Abnormality (DESCRIBE)
505	Triploidy

Code **Other Birth Defects**

601	Skeletal Dysplasia (DESCRIBE)
602	Congenital Diaphragmatic Hernia
603	Hydrops Fetalis with anasarca and one or more of the following: ascites, pleural effusion pericardial effusion
604	Oligohydramnios sequence including all 3 of the following: <ol style="list-style-type: none"> 1. Oligohydramnios documented by antenatal ultrasound 5 or more days prior to delivery. 2. Evidence of fetal constraint on postnatal physical exam (such as Potter's facies, contractures, or positional deformities of limbs), and 3. Postnatal respiratory failure requiring endotracheal intubation and assisted ventilation.
605	Inborn Error of Metabolism (DESCRIBE)
606	Myotonic Dystrophy requiring endotracheal intubation and assisted ventilation.
607	Conjoined Twins
608	Tracheal Agenesis or Atresia
609	Thanatophoric Dysplasia Types 1 and 2
610	Hemoglobin Barts

Code **Pulmonary Defects**

800	Other Lethal or Life Threatening Pulmonary Malformation (DESCRIBE)
801	Congenital Lobar Emphysema
802	Congenital Cystic Adenomatoid Malformation of the Lung
803	Sequestered Lung
804	Aveolar Capillary Dysplasia

Code **Vascular and Lymphatic Defects**

900	Other Vascular or Lymphatic (DESCRIBE)
901	Cystic Hygroma
902	Hemangioma
903	Sacrococcygeaal Teratoma
904	Cerebral AV Malformation

Code **Other Diagnoses**

121	Hematologic
122	Hemolytic Disease of the Newborn (Not ABO)

The following conditions should *NOT* be coded as Major Birth Defects

Extreme Prematurity, Limb Abnormalities, Syndactyly, Polydactyly, Hypospadias, Patent Ductus Arteriosus, Pulmonary Hypoplasia (use code 401 for bilateral renal agenesis, or 604 for Oligohydramnios Sequence, if applicable), Intrauterine Growth Retardation, Small Size for Gestational Age, Fetal Alcohol Syndrome, Hypothyroidism, Intrauterine Infection, Persistent Pulmonary Hypertension (PPHN)