Transition to Use of ICD-10-CM Coding for Birth Defects, Part 2

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Implementation of ICD-10-CM/PCS

- **Protecting Access to Medicare Act of 2014**

  "The Secretary of Health and Human Services may not, prior to October 1, 2015, adopt ICD–10 code sets as the standard for code sets under section 1173(c) of the Social Security Act (42 U.S.C. 1320d–2(c)) and section 162.1002 of title 45, Code of Federal Regulations."

- **The National Center on Health Statistics and the Centers for Medicare & Medicaid Services are working to identify a new implementation date.**

- **NBDPN strongly recommends that birth defects programs take advantage of the delay to continue development, revision, and implementation of their plans to transition to use of ICD-10-CM/PCS.**

ICD-10-CM Coding

- **Structure of codes**
- **Addition of new codes for increased specificity**
  - Ability to code and retrieve more individual diagnoses
- **Incorporation of new characteristics into the codes**
  - Laterality
  - Timing of examination
  - Birth order of infant in multiple gestations
- **A few areas with decreased specificity**
- **Reorganization of codes within organ systems**
- **Conditions moved to different areas of the code**
- **NBDPN code translation tools**

Code Structure in ICD-10-CM

- **Codes can be 3 to 7 characters long**
  - All have the potential to contain 7 characters
- **1st character – Always alphabetic**
  - A – T, V, Z
- **2nd character – Always numeric**
- **3rd-7th characters – Either alphabetic or numeric**
- **The last character can take on a variety of specified meanings for different codes**
- **Use of dummy placeholder "X" for characters without values**

Changes in ICD-10-CM Coding

- **Use of last character for specific meaning (code O09.0)**
  - O09.00 – Supervision of pregnancy with history of infertility, unspecified trimester
  - O09.01– Supervision of pregnancy with history of infertility, first trimester
  - O09.02– Supervision of pregnancy with history of infertility, second trimester
  - O09.03– Supervision of pregnancy with history of infertility, third trimester
- **Use of placeholder X (code O32.1)**
  - O32.1XX0 – Maternal care for breech presentation, single gestation
  - O32.1XX1 – Maternal care for breech presentation, fetus 1
  - O32.1XX2 – Maternal care for breech presentation, fetus 2

Chapter 17. Congenital Malformations, Deformations and Chromosomal Abnormalities

- Q00-Q07 Congenital malformations of the nervous system
- Q10-Q18 Congenital malformations of eye, ear, face, and neck
- Q20-Q28 Congenital malformations of the circulatory system
- Q30-Q34 Congenital malformations of the respiratory system
- Q35-Q37 Cleft lip and cleft palate
- Q38-Q45 Other congenital malformations of the digestive system
- Q50-Q56 Congenital malformations of genital organs
- Q60-Q64 Congenital malformations of the urinary system
- Q65-Q79 Congenital malformations and deformations of the musculoskeletal system
- Q80-Q89 Other congenital malformations
- Q90-Q99 Chromosomal abnormalities, not elsewhere classified

Malformations of the Nervous System

ICD-9-CM	ICD-10-CM
740 Anencephalus and similar anomalies	Q00 Anencephaly and similar malformations
741 Spina bifida 741.0 Spina bifida with hydrocephalus 741.9 Spina bifida without mention of hydrocephalus	Q01 Encephalocele Q02 Microcephaly Q03 Congenital hydrocephalus
742 Other congenital anomalies of nervous system 742.0 Encephalocele 740.1 Microcephalus 742.2 Reduction deformities of brain 742.3 Congenital hydrocephalus 742.4 Other specified anomalies of brain; macroencephaly 742.5 Other specified anomalies of spinal cord 742.8 Other specified anomalies of nervous system 742.9 Unspecified anomaly of brain, spinal cord, and nervous system	Q04 Other congenital malformations of brain Q05 Spina bifida Q05.0 Cervical spina bifida with hydrocephalus Q05.1 Thoracic spina bifida with hydrocephalus Q05.2 Lumbar spina bifida with hydrocephalus Q05.3 Sacral spina bifida with hydrocephalus Q05.4 Unspecified spina bifida with hydrocephalus Q06 Other congenital malformations of spinal cord Q07 Other congenital malformations of nervous system
	Q75 Other congenital malformations of skull and face bones Q75.3 Macrocephaly

Malformations of the Eye

ICD-9-CM	ICD-10-CM
743.0 Anophthalmos	Q10 Congenital malformations of eyelid, lacrimal apparatus, and orbit
743.1 Microphthalmos	
743.2 Buphthalmos; glaucoma	Q11 Anophthalmos, microphthalmos and macrophthalmos Q11.0 Cystic eyeball Q11.1 Other anophthalmos Q11.2 Microphthalmos Q11.3 Macrophthalmos
743.3 Congenital cataract and lens anomalies 743.30 Congenital cataract, unspecified 743.31 Capsular and subcapsular cataract 743.32 Cortical and zonular cataract 743.33 Nuclear cataract 743.34 Total and subtotal cataract, congenital	Q12 Congenital lens malformations Q12.0 Congenital cataract
743.4 Coloboma and other anomalies of anterior segment	Q13 Congenital malformations of anterior segment of eye
743.5 Congenital anomalies of posterior segment	Q14 Congenital malformations of posterior segment of eye
743.6 Congenital anomalies of eyelids, lacrimal system, and orbit	Q15 Other congenital malformations of eye Q15.0 Congenital glaucoma Q15.8 Other specified malformations of eye
743.8 Other specified anomalies of eye	
743.9 Unspecified anomaly of eye	

Malformations of the Ear, Face and Neck

ICD-9-CM	ICD-10-CM
744.0 Anomalies of ear causing hearing impairment	Q16 Congenital malformations of ear causing impairment of hearing
744.1 Accessory auricle	Q17 Other congenital malformations of ear Q17.0 Accessory auricle Q17.1 Macrotia Q17.2 Microtia Q17.9 Unspecified anomaly of ear
744.2 Other specified anomalies of ear 744.22 Macrotia 744.23 Microtia	
744.3 Unspecified anomaly of ear	
744.4 Branchial cleft cyst or fistula; preauricular sinus 744.41 Branchial cleft sinus or fistula 744.42 Branchial cleft cyst 744.46 Preauricular sinus or fistula 744.47 Preauricular cyst	Q18 Other congenital malformations of face and neck Q18.0 Sinus, fistula and cyst of branchial cleft Q18.1 Preauricular sinus and cyst Q18.2 Other branchial cleft malformations Q18.4 Webbing of neck
744.5 Webbing of neck	
744.8 Other specified anomalies of face and neck	
744.9 Unspecified anomalies of face and neck	

Malformations of the Circulatory System

ICD-9-CM	ICD-10-CM
745.0 Common truncus	Q20.0 Common arterial trunk
745.1 Transposition of great vessels 745.10 Complete transposition of great vessels 745.11 Double outlet right ventricle 745.12 Corrected transposition of great vessels 745.19 Other transposition of great vessels	Q20.1 Double outlet right ventricle
	Q20.2 Double outlet left ventricle
745.2 Tetralogy of Fallot	Q20.3 Discordant ventriculoarterial connection; transposition of great vessels
745.3 Common ventricle	Q20.4 Double inlet ventricle; common ventricle
745.4 Ventricular septal defect	Q20.5 Discordant atrioventricular connection; corrected transposition
745.5 Ostium secundum type atrial septal defect	Q20.6 Isomerism of atrial appendages
745.6 Endocardial cushion defects	Q21.0 Ventricular septal defect
745.7 Cor biloculare	Q21.1 Atrial septal defect; patent foramen ovale
745.8 Other	Q21.2 Atrioventricular septal defect
745.9 Unspecified defect of septal closure	Q21.3 Tetralogy of Fallot
	Q21.4 Aortopulmonary septal defect
	Q21.8 Other congenital malformations of cardiac septa

Malformations of the Circulatory System

ICD-9-CM	ICD-10-CM
746.00 Pulmonary valve anomaly, unspecified	Q22.0 Pulmonary valve atresia
746.01 Pulmonary valve atresia, congenital	Q22.1 Congenital pulmonary valve stenosis
746.02 Pulmonary valve stenosis, congenital	Q22.2 Congenital pulmonary valve insufficiency
746.09 Other anomalies of pulmonary valve	Q22.3 Other malformations of pulmonary valve
746.1 Tricuspid atresia and stenosis, congenital	Q22.4 Congenital tricuspid stenosis; tricuspid atresia
746.2 Ebstein's anomaly	Q22.5 Ebstein's anomaly
746.3 Congenital stenosis of aortic valve	Q22.6 Hypoplastic right heart syndrome
746.4 Congenital insufficiency of aortic valve	Q22.8 Other malformations of tricuspid valve
746.5 Congenital mitral stenosis	Q22.9 Unspecified malformation of tricuspid valve
746.6 Congenital mitral insufficiency	Q23.0 Congenital stenosis of aortic valve
746.7 Hypoplastic left heart syndrome	Q23.1 Congenital insufficiency of aortic valve
	Q23.2 Congenital mitral stenosis
	Q23.3 Congenital mitral insufficiency
	Q23.4 Hypoplastic left heart syndrome
	Q23.8 Other malformations of aortic and mitral valves
	Q23.9 Malformation of aortic and mitral valves, unspecified

Cleft Lip and Cleft Palate

ICD-9-CM	ICD-10-CM
749.0 Cleft palate 749.00 Cleft palate, unspecified 749.01 Unilateral, complete 749.02 Unilateral, incomplete 749.03 Bilateral, complete 749.04 Bilateral, incomplete	Q35 Cleft palate Q35.1 Cleft hard palate Q35.3 Cleft soft palate Q35.5 Cleft hard palate with cleft soft palate Q35.7 Cleft uvula Q35.9 Cleft palate, unspecified
749.1 Cleft lip 749.10 Cleft lip, unspecified 749.11 Unilateral, complete 749.12 Unilateral, incomplete 749.13 Bilateral, complete 749.14 Bilateral, incomplete	Q36 Cleft lip Q36.0 Cleft lip, bilateral Q36.1 Cleft lip, median Q36.9 Cleft lip, unilateral; cleft lip not otherwise specified
749.2 Cleft palate with cleft lip 749.20 Cleft palate with cleft lip, unspecified 749.21 Unilateral, complete 749.22 Unilateral, incomplete 749.23 Bilateral, complete 749.24 Bilateral, incomplete 749.25 Other combinations	Q37 Cleft palate with cleft lip Q37.0 Cleft hard palate with bilateral cleft lip Q37.1 Cleft hard palate with unilateral cleft lip Q37.2 Cleft soft palate with bilateral cleft lip Q37.3 Cleft soft palate with unilateral cleft lip Q37.4 Cleft hard and soft palate with bilateral cleft lip Q37.5 Cleft hard and soft palate with unilateral cleft lip Q37.8 Unspecified cleft palate with bilateral cleft lip Q37.9 Unspecified cleft palate with unilateral cleft lip

Malformations of Upper Gastrointestinal Tract

ICD-9-CM	ICD-10-CM
750.0 Tongue tie	Q38 Other congenital malformations of tongue, mouth, and pharynx Q38.0 Congenital malformations of lips, not elsewhere classified Q38.1 Ankyloglossia; tongue tie Q38.2 Macroglossia Q38.3 Other congenital malformations of tongue; microglossia Q38.4 Congenital malformations of salivary glands and ducts Q38.5 Congenital malformations of palate, not elsewhere classified Q38.6 Other congenital malformations of mouth Q38.7 Congenital pharyngeal pouch Q38.8 Other congenital malformations of pharynx
750.1 Other anomalies of tongue 750.10 Anomaly of tongue, unspecified 750.11 Aglossia 750.12 Congenital adhesions of tongue 750.13 Fissure of tongue 750.15 Macroglossia 750.16 Microglossia 750.19 Other anomalies of tongue	
750.2 Other specified anomalies of mouth and pharynx 750.21 Absence of salivary gland 750.22 Accessory salivary gland 750.23 Atresia, salivary gland 750.24 Congenital fistula of salivary gland 750.25 Congenital fistula of lip 750.27 Diverticulum of pharynx	Q39 Congenital malformations of esophagus Q39.0 Atresia of esophagus without fistula Q39.1 Atresia of esophagus with tracheo-esophageal fistula Q39.2 Congenital tracheo-esophageal fistula without atresia Q39.3 Congenital stenosis and stricture of esophagus Q39.4 Esophageal web Q39.5 Congenital dilatation of esophagus Q39.6 Congenital diverticulum of esophagus
750.3 Tracheoesophageal fistula, esophageal atresia and stenosis	
750.4 Other specified anomalies of esophagus	

Malformations of the Female Genital Organs

ICD-9-CM	ICD-10-CM
752.0 Anomalies of ovaries	Q50 Congenital malformations of ovaries, fallopian tubes, and broad ligaments Q50.0 Congenital absence of ovary Q50.3 Other congenital malformations of ovary Q50.6 Other congenital malformations of fallopian tube and broad ligament
752.1 Anomalies of fallopian tubes and broad ligaments	
752.2 Doubling of uterus	
752.3 Other anomalies of uterus	
752.4 Anomalies of cervix, vagina, and external female genitalia 752.41 Embryonic cyst of cervix, vagina, and external female genitalia 752.42 Imperforate hymen 752.43 Cervical agenesis 752.44 Cervical duplication 752.49 Other anomalies of cervix, vagina, and external female genitalia	Q51 Congenital malformations of uterus and cervix Q51.0 Agenesis and aplasia of uterus Q51.1 Doubling of uterus with doubling of cervix and vagina Q51.5 Agenesis and aplasia of cervix Q51.6 Embryonic cyst of cervix Q51.81 Other congenital malformations of uterus Q51.82 Other congenital malformations of cervix
	Q52 Congenital malformations of female genitalia Q52.0 Congenital absence of vagina Q52.1 Doubling of vagina Q52.3 Imperforate hymen Q52.4 Other congenital malformations of vagina Q52.6 Congenital malformation of clitoris Q52.7 Other and unspecified congenital malformations of vulva

Malformations of the Male Genital Organs

ICD-9-CM	ICD-10-CM
752.51 Undescended testes	Q53.0 Ectopic testis Q53.00 Ectopic testis, unspecified Q53.01 Ectopic testis, unilateral Q53.02 Ectopic testis, bilateral
752.52 Retractile testes	
752.61 Hypospadias	
752.62 Epispadias	Q53.1 Undescended testicle, unilateral Q53.10 Unspecified undescended testicle, unilateral Q53.11 Abdominal testis, unilateral Q53.12 Ectopic perineal testis, unilateral
752.63 Congenital chordee	
752.64 Micropenis	
752.65 Hidden penis	Q53.2 Undescended testicle, bilateral Q53.20 Unspecified undescended testicle, bilateral Q53.21 Abdominal testis, bilateral Q53.22 Ectopic perineal testis, bilateral
752.69 Other penile anomalies	
752.7 Indeterminate sex and pseudohermaphroditism	Q54 Hypospadias Q54.0 Hypospadias, balanic Q54.1 Hypospadias, penile Q54.2 Hypospadias, penoscrotal Q54.3 Hypospadias, perineal Q54.4 Congenital chordee
	Q64 Other congenital malformations of urinary system Q64.0 Epispadias

Malformations of the Urinary System

ICD-9-CM	ICD-10-CM
753.0 Renal agenesis and dysgenesis	Q60 Renal agenesis and other reduction defects of kidney Q60.0 Renal agenesis, unilateral Q60.1 Renal agenesis, bilateral Q60.2 Renal agenesis, unspecified Q60.3 Renal hypoplasia, unilateral Q60.4 Renal hypoplasia, bilateral Q60.5 Renal hypoplasia, unspecified Q60.6 Potter's syndrome
753.1 Cystic kidney disease 753.11 Congenital single renal cyst	
753.2 Obstructive defects of renal pelvis and ureter	
753.5 Exstrophy of urinary bladder	
753.6 Atresia and stenosis of urethra and bladder neck	Q61 Cystic kidney disease Q61.00 Congenital renal cyst, unspecified Q61.01 Congenital single renal cyst Q61.02 Congenital multiple renal cysts
753.7 Anomalies of urachus	
753.8 Other specified anomalies of bladder and urethra	Q62 Congenital obstructive defects of renal pelvis and congenital malformations of ureter Q62.0 Congenital hydronephrosis
	Q64 Other congenital malformations of urinary system Q64.0 Epispadias Q64.10 Exstrophy of urinary bladder, unspecified Q64.11 Supravesical fissure of urinary bladder Q64.12 Cloacal exstrophy of urinary bladder Q64.19 Other exstrophy of urinary bladder Q64.2 Congenital posterior urethral valves Q64.3 Other atresia and stenosis of urethra and bladder neck Q64.4 Malformations of urachus

Malformations of the Musculoskeletal System

ICD-9-CM	ICD-10-CM
754.0 Deformities of skull, face, and jaw	Q65 Congenital deformities of hip Q65.01 Congenital dislocation of right hip, unilateral Q65.02 Congenital dislocation of left hip, unilateral Q65.1 Congenital dislocation of hip, bilateral Q65.31 Congenital partial dislocation of right hip
754.1 Deformities of sternocleidomastoid muscle	
754.2 Deformities of spine	
754.3 Congenital dislocation of hip 754.30 Congenital dislocation of hip, unilateral 754.31 Congenital dislocation of hip, bilateral 754.35 Congenital dislocation of one hip with subluxation of other hip	Q66 Congenital deformities of feet Q66.0 Congenital talipes equinovarus Q66.4 Congenital talipes calcaneovalgus Q66.51 Congenital pes planus, right foot
754.4 Congenital genu recurvatum and bowing of long bones of leg 754.42 Congenital bowing of femur 754.43 Congenital bowing of tibia and fibula	Q67 Congenital musculoskeletal deformities of head, face, spine, and chest Q76.1 Congenital compression facies Q76.3 Plagiocephaly Q67.5 Congenital deformity of spine Q67.6 Pectus excavatum
754.5 Varus deformities of feet 754.51 Talipes equinovarus	
754.6 Valgus deformities of feet 754.61 Congenital pes planus 754.62 Talipes calcaneovalgus	Q68 Other congenital musculoskeletal deformities Q68.0 Congenital deformity of sternocleidomastoid muscle Q68.3 Congenital bowing of femur Q68.4 Congenital bowing of tibia and fibula Q68.6 Discoid meniscus
754.8 Other specified nonteratogenic anomalies 754.81 Pectus excavatum	
755.0 Polydactyly	Q69 Polydactyly
755.1 Syndactyly	Q70 Syndactyly Q70.4 Polysyndactyly
717.5 Derangement of meniscus, NEC	

Malformations of the Musculoskeletal System

ICD-9-CM	ICD-10-CM
755.2 Reduction deformities of upper limb 755.20 Unspecified reduction deformity of upper limb 755.21 Transverse deficiency of upper limb 755.22 Longitudinal deficiency of upper limb, not elsewhere classified 755.23 Longitudinal deficiency, combined 755.24 Longitudinal deficiency, humeral 755.25 Longitudinal deficiency, radioulnar 755.26 Longitudinal deficiency, radial 755.27 Longitudinal deficiency, ulnar 755.28 Longitudinal deficiency, carpals or metacarpals 755.29 Longitudinal deficiency, phalanges	Q71 Reduction defects of upper limb Q71.00 Congenital complete absence of unspecified upper limb Q71.01 Congenital complete absence of right upper limb Q71.02 Congenital complete absence of left upper limb Q71.03 Congenital complete absence of upper limb, bilateral Q71.1 Congenital absence of upper arm and forearm with hand present Q71.2 Congenital absence of both forearm and hand Q71.3 Congenital absence of hand and finger Q71.4 Longitudinal reduction defect of radius Q71.5 Longitudinal reduction defect of ulna Q71.6 Lobster-claw hand Q71.8 Other reduction defects of upper limb Q71.9 Unspecified reduction defect of upper limb
755.3 Reduction deformities of lower limb	Q72 Reduction defects of lower limb
755.4 Reduction deformities, unspecified limb	Q73 Reduction defects of unspecified limb
755.58 Cleft hand, congenital; lobster-claw hand	

Malformations of the Musculoskeletal System

ICD-9-CM	ICD-10-CM
755.5 Other anomalies of upper limb, including shoulder girdle 755.52 Sprengel's deformity 755.53 Radioulnar synostosis 755.54 Madelung's deformity 755.55 Acrocephalosyndactyly; Apert syndrome	Q74.0 Other congenital malformations of upper limb(s), including shoulder girdle
755.58 Cleft hand; lobster claw hand	Q74.1 Congenital malformation of knee Q74.2 Other congenital malformations of lower limb(s), including pelvic girdle Q74.3 Arthrogryposis multiplex congenita Q74.8 Other specified congenital malformations of limb(s) Q74.9 Unspecified congenital malformations of limb(s)
755.6 Other anomalies of lower limb, including pelvic girdle 755.61 Coxa valga, congenital 755.62 Coxa vara, congenital 755.63 Other congenital deformity of hip 755.64 Congenital deformity of knee (joint) 755.66 Other anomalies of toes	Q71.6 Lobster-claw hand
	Q65 Congenital deformities of hip Q65.81 Congenital coxa valga Q65.82 Congenital coxa vara Q65.89 Other specified congenital deformities of hip

Malformations of the Musculoskeletal System

ICD-9-CM	ICD-10-CM
756.0 Anomalies of skull and face bones	Q75 Other congenital malformations of skull and face bones Q75.0 Craniosynostosis Q75.3 Macrocephaly
756.1 Anomalies of spine	
756.2 Cervical rib	
756.3 Other anomalies of ribs and sternum	Q76 Congenital malformations of spine and bony thorax Q76.411 Congenital kyphosis, occipito-atlanto-axial region Q76.412 Congenital kyphosis, cervical region Q76.413 Congenital kyphosis, cervicothoracic region Q76.414 Congenital kyphosis, thoracic region Q76.415 Congenital kyphosis, thoracolumbar region Q76.42 Congenital lordosis
756.4 Chondrodystrophy	
756.5 Osteodystrophies 756.50 Osteodystrophy, unspecified 756.51 Osteogenesis imperfecta 756.52 Osteopetrosis 756.53 Osteopoikilosis 756.54 Polyostotic fibrous dysplasia of bone 756.55 Chondroectodermal dysplasia 756.56 Multiple epiphyseal dysplasia 756.59 Other osteodystrophies	Q77 Osteochondrodysplasia with defects of growth of tubular bones and spine Q77.1 Thanatophoric short stature Q77.2 Short rib syndrome Q77.3 Chondrodysplasia punctata Q77.4 Achondroplasia Q77.6 Chondroectodermal dysplasia
	Q78 Other osteochondrodysplasias Q78.0 Osteogenesis imperfecta Q78.1 Polyostotic fibrous dysplasia Q78.2 Osteopetrosis Q78.3 Progressive diaphyseal dysplasia Q78.8 Other specified osteochondrodysplasias; osteopoikilosis

Malformations of the Musculoskeletal System

ICD-9-CM	ICD-10-CM
756.6 Anomalies of diaphragm	Q79 Congenital malformations of musculoskeletal system, not elsewhere classified Q79.0 Congenital diaphragmatic hernia Q79.1 Other congenital malformations of diaphragm Q79.2 Exomphalos; omphalocele Q79.3 Gastroschisis Q79.4 Prune belly syndrome Q795. Other congenital malformations of abdominal wall Q79.6 Ehlers-Danlos syndrome Q79.8 Other congenital malformations of musculoskeletal system Q79.9 Congenital malformations of musculoskeletal system, unspecified
756.7 Anomalies of abdominal wall 756.71 Prune belly syndrome 756.72 Omphalocele 756.73 Gastroschisis 756.70 Other congenital anomalies of abdominal wall	
756.8 Other specified anomalies of muscle, tendon, fascia, and connective tissue 756.81 Absence of muscle and tendon 756.82 Accessory muscle 756.83 Ehlers-Danlos syndrome	
756.9 Other and unspecified anomalies of musculoskeletal system	

Other Congenital Malformations

ICD-9-CM	ICD-10-CM
757 Congenital anomalies of the integument	Q80 Congenital ichthyosis
758 Chromosomal anomalies	Q81 Epidermolysis bullosa
759 Other and unspecified congenital anomalies	Q82 Other congenital malformations of skin
	Q83 Congenital malformations of breast
	Q84 Other congenital malformations of integument
	Q85 Phakomatoses, not elsewhere classified
	Q86 Congenital malformation syndromes due to known exogenous causes, not elsewhere classified
	Q87 Other specified congenital malformation syndromes affecting multiple systems
	Q89 Other congenital malformations, not elsewhere classified
	Q90-99 Chromosomal abnormalities, not elsewhere classified

Congenital Malformations of Integument

ICD-9-CM	ICD-10-CM
757.0 Hereditary edema of legs	Q80 Congenital ichthyosis
757.1 Ichthyosis congenita	Q81 Epidermolysis bullosa
757.2 Dermatoglyphic anomalies	Q82 Other congenital malformations of skin Q82.0 Hereditary lymphedema Q82.4 Ectodermal dysplasia (anhydrotic) Q82.5 Congenital non-neoplastic nevus
757.3 Other specified anomalies of skin 757.31 Congenital ectodermal dysplasia 757.32 Vascular hamartomas 757.33 Congenital pigmentary anomalies of skin	Q83 Congenital malformations of breast
757.4 Specified anomalies of hair	Q84 Other congenital malformations of integument Q84.0 Congenital alopecia Q84.1 Congenital morphologic disturbances of hair, not elsewhere classified Q84.2 Other congenital malformations of hair Q84.3 Anonychia Q84.4 Congenital leukonychia Q84.5 Enlarged and hypertrophic nails 84.6 Other congenital malformations of nails
757.5 Specified anomalies of nails	
757.6 Specified congenital anomalies of breast	
757.8 Other specified anomalies of the integument	
759.5 Tuberous sclerosis	
759.6 Other hamartoses, not elsewhere classified	Q85 Phakomatoses, not elsewhere classified Q85.0 Neurofibromatosis Q85.1 Tuberous sclerosis Q85.8 Other phakomatoses, not elsewhere classified Q85.9 Phakomatosis, unspecified
237.7 Neurofibromatosis	

Other Congenital Malformations

ICD-9-CM	ICD-10-CM
759.0 Anomalies of spleen	Q86 Congenital malformation syndromes due to known exogenous causes, not elsewhere classified Q86.0 Fetal alcohol syndrome (dysmorphic) Q86.1 Fetal hydantoin syndrome
759.1 Anomalies of adrenal gland	
759.2 Anomalies of other endocrine glands	
759.3 Situs inversus	Q87 Other specified congenital malformation syndromes affecting multiple systems Q87.0 Congenital malformation syndromes predominantly affecting facial appearance Q87.1 Congenital malformation syndromes predominantly associated with short stature Q87.2 Congenital malformation syndromes predominantly involving limbs Q87.3 Congenital malformation syndromes involving early overgrowth Q87.4 Marfan syndrome Q87.5 Congenital malformation syndromes with other skeletal changes
759.4 Conjoined twins	
759.5 Tuberous sclerosis	
759.7 Multiple congenital anomalies, so described	
759.8 Other specified anomalies 759.81 Prader-Willi syndrome 759.82 Marfan syndrome 759.83 Fragile X syndrome	
760.7 Noxious influences affecting fetus or newborn via placenta or breast milk 760.71 Alcohol 760.77 Anticonvulsants	Q89 Other congenital malformations, not elsewhere classified Q89.0 Congenital absence and malformations of spleen Q89.1 Congenital malformations of adrenal gland Q89.2 Congenital malformations of other endocrine glands Q89.3 Situs inversus Q89.4 Conjoined twins Q89.7 Multiple congenital malformations, not elsewhere classified
	Q99.2 Fragile X chromosome

Chromosomal Abnormalities

ICD-9-CM	ICD-10-CM
758.0 Down syndrome	Q90 Down syndrome Q90.0 Trisomy 21, nonmosaicism Q90.1 Trisomy 21, mosaicism Q90.2 Trisomy 21, translocation
758.1 Patau'syndrome	
758.2 Edward syndrome	
758.3 Autosomal deletion syndromes 758.31 Cri-du-chat syndrome 758.32 Velo-cardio-facial syndrome; deletion 22q11.2 758.33 Other microdeletions 758.39 Other autosomal deletions	Q91 Trisomy 18 and 13
	Q92 Other trisomies and partial trisomies of the autosomes Q92.1 Whole chromosome trisomy, mosaicism Q92.2 Partial trisomy Q92.7 Triploidy and polyploidy
758.4 Balanced autosomal translocation in normal individual	Q93 Monosomies and deletions from the autosomes, not elsewhere classified Q93.2 Chromosome replaced with ring, dicentric or isochromosome Q93. Deletion of short arm of chromosome 4; Wolff-Hirschorn syndrome Q93.4 Deletion of short arm of chromosome 5; Crit-du-chat syndrome Q93.81 Velo-cardio-facial syndrome; deletion 22q11.2 Q93.88 Other microdeletions
758.5 Other conditions due to autosomal anomalies	
	Q95 Balanced rearrangements and structural markers, not elsewhere classified Q95.1 Chromosome inversion in normal individual Q95.2 Balanced autosomal rearrangement in abnormal individual

Chromosomal Abnormalities

ICD-9-CM	ICD-10-CM
758.6 Gonadal dysgenesis	Q96 Turner syndrome Q96.0 Karyotype 45,X Q96.1 Karyotype 46,X iso (Xq) Q96.3 Mosaicism, 45 X/46,XX or XY
758.7 Klinefelter syndrome	Q97 Other sex chromosome abnormalities, female phenotype, not elsewhere classified Q97. 0 Karyotype 47,XXX Q97.2 Mosaicism, lines with various numbers of X chromosomes Q97.3 Female with 46,XY karyotype
758.8 Other conditions due to chromosome anomalies 758.81 Other conditions due to sex chromosome anomalies 758.89 Other conditions due to chromosome anomalies	Q98 Other sex chromosome abnormalities, male phenotype, not elsewhere classified Q98.0 Klinefelter syndrome karyotype 47,XXY Q98.1 Klinefelter syndrome, male with more than two X chromosomes Q98.3 Other male with karyotype 46,XX Q98.5 Karyotype 47,XYY
758.9 Conditions due to anomaly of unspecified chromosome	Q99 Other chromosome abnormalities, not elsewhere classified Q99.0 Chimera 46,XX/46,XY Q99.1 46,XX true hermaphrodite Q99.2 Fragile X chromosome Q99.8 Other specified chromosome abnormalities

Additional Changes in ICD-10-CM

- **Persistent fetal circulation**
  - Included among "Other congenital anomalies of circulatory system" in ICD-9-CM (747.83)
  - Moved to "Cardiovascular disorders originating in the perinatal period "in ICD-10-CM (P29.3)
- **Other conditions not listed in ICD-9-CM but added to the congenital malformations codes in ICD-10-CM**
  - Congenital subglottic stenosis (Q31.1)
  - Congenital laryngomalacia (Q31.5)
  - Congenital tracheomalacia (Q32.0)
- **These conditions can be congenital, but often are related to prematurity or prolonged intubation. Many programs do not include them as congenital malformations.**

ICD-9-CM to ICD-10-CM Code Translation Tool

- **Developed specifically for birth defect programs that wish to translate data coded in ICD-9-CM to ICD-10-CM**
  - Differ in some instances from the General Equivalence Mappings (GEMs) available from NCHS
- **For each code in ICD-9CM, the tool provides all possible alternative codes in ICD-10-CM**
  - Many ICD-9-CM codes have only one corresponding code in ICD-10-CM
  - When there are more than one possible alternative ICD-10-CM codes, select the one that most closely matches the defect
  - If there is not enough information to select a single alternative, a default code is designated as the preferred code
  - The tool contains every code in ICD-9-CM, but not necessarily every code in ICD-10-CM
- **ICD-10-CM to ICD-9-CM code translation tool is also available**

ICD-9-CM to ICD-10-CM Code Translation Tool

ICD-9-CM	Default Code	ICD-10-CM
740.0 Anencephalus		Q00.0 Anencephaly
740.1 Craniorachischisis		Q00.1 Craniorachischisis
740.2 Iniencephaly		Q00.2 Iniencephaly
742.0 Encephalocele		Q01.0 Frontal encephalocele
		Q01.1 Nasofrontal encephalocele
		Q01.2 Occipital encephalocele
		Q01.8 Encephalocele of other sites
	D	Q01.9 Encephalocele, unspecified
749.10 Cleft lip, unspecified		Q36.9 Cleft lip, unilateral
749.11 Unilateral cleft lip, complete		Q36.9 Cleft lip, unilateral
749.12 Unilateral cleft lip, incomplete		Q36.9 Cleft lip, unilateral

Questions and Answers

- **Does the new delay in implementation of ICD-10-CM mean that birth defects surveillance programs cannot accept ICD-10-CM codes from data sources such as hospitals until October 1, 2015?**

  The new legislation states only that ICD-10-CM may not be adopted before October 1, 2015. We do not yet know whether it will be implemented on that date or at a later time. Implementation of ICD-10-CM affects all entities covered by the Health Insurance Portability and Accountability Act . Once it is implemented, ICD-10-CM coding will be required to process all claims for healthcare services; claims that utilize ICD-9-CM codes will not be accepted.

  There is nothing that prohibits a hospital or other source from submitting data coded in ICD-10-CM to birth defects programs, or that proibits a program from using data coded in ICD-10-CM, prior to October 1, 2015. But it seems unlikely that such sources will consistently utilize ICD-10-CM prior to the implementation date since they cannot use it for billing purposes. However, birth defects programs should keep in touch with data sources about when they will begin coding in ICD-10-CM and offer to help test procedures for data coding and submission prior to the implementation date.

Questions and Answers

- **Among chromosomal abnormalities, can ICD-10-CM coded data reflect the circumstance where a karyotype result is pending at the time a child with a birth defect is ascertained by a surveillance program?**

  ICD-10-CM coding does not include the ability to indicate that a diagnostic test such as a karyotype has been performed but the result is not yet available. It also does not include the ability to indicate the level of certainty of a defect diagnosis (e.g., possible, probable, definite). If a program wishes to capture this kind of detail, they will need to establish additional code or fields to reflect it. This will not be possible for programs that receive ICD-10-CM codes without any additional information about the defects or diagnostic tests that have been performed.

Questions and Answers

- **What are the anticipated impacts on birth defects surveillance and monitoring due to the transition from ICD-9-CM to ICD-10-CM? How will the training that physicians and coders receive impact reporting?**

  These are key questions. We do not know what all of the effects of the transition to ICD-10-CM coding will be. In general, ICD-10-CM codes are more specific than in ICD-9-CM. Many conditions represented by a single code in ICD-9-CM can be equally represented by combining two or more codes in ICD-10-CM. For monitoring, these equivalent groups of codes can be used for consistent defect definitions during the transition. And, specific ICD-10-CM codes that more closely define defects included in broader ICD-9-CM codes can then be monitored.

  The training in ICD-10-CM that physicians and coders receive will be critical to consistent and accurate reporting of defects. Unfortunately, this training and the resulting coding practices are likely to vary among states, data sources, and individual coders. Birth defects programs should keep in touch with data sources to become familiar with the training and coding practices they use.

Questions and Answers

- **At what point after the transition from ICD-9-CM to ICD-10-CM can we anticipate accurate defect coding and reporting from data sources and stability in the estimated prevalence of defects? How can we compensate for any anticipated distortions in reporting that may result from the transition?**

  At this point, it is difficult to anticipate how long it will take for defect coding and reporting, and the resulting estimates of defect prevalence, to stabilize. This will likely vary depending on the defect, the degree of equivalence between the relevant ICD-9-CM and ICD-10-CM codes, and the consistency of the coders. It may take a few years for any changes in some defect prevalences to be evident. For others, there may be no observable changes. It will be important for birth defects programs to review their data as it is received and compiled to watch for unexpected changes in defect frequency or distribution across data sources or geographic areas that might be related to ICD-10-CM coding. An early collaborative project for NBDPN programs could be to examine the prevalence of defects that are included in the annual report in the years before, during, and after the transition to ICD-10-CM.

Tools and Resources

- Code Translations from ICD-9-CM to ICD-10-CM, and from ICD-10-CM back to ICD-9-CM, for Birth Defects Surveillance (Excel file) – Developed for NBDPN: http://www.nbdpn.org/icd9_icd10_code_translation.php
- NBDPN Coding Tools Work Group Message Board - NBDPN members can post questions about ICD-10-CM coding, share experiences and tips, discuss common concerns, etc. Check it regularly!

  http://mms.nbdpn.org/members/forum/board_list.php
- ICD-10-CM Code, Guidelines, Addenda, and General Equivalence Mapping files: http://www.cdc.gov/nchs/icd/icd10cm.htm
- ICD-10-PCS Code, Guidelines, Addendum, and General Equivalence Mapping files: http://www.cms.gov/Medicare/Coding/ICD10/2014-ICD-10-PCS.html
- CDC Website on Public Health Transition to ICD-10-CM/PCS – Transition Planning, Trainings, Resources, FAQs: http://www.cdc.gov/nchs/icd/icd10cm_pcs.htm

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