Birth Defects Coding and ICD-10-CM

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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

Improved Defect Coding in ICD-10-CM

- **Increased specificity**
  - ICD-9-CM: 750.3 – Tracheoesophageal fistula, esophageal atresia and stenosis
  - ICD-10-CM:
    - 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
- **Laterality incorporated into defect codes**
  - Q65.00 – Congenital dislocation of unspecified hip, unilateral
  - Q65.01 – Congenital dislocation of right hip, unilateral
  - Q65.02 – Congenital dislocation of left hip, unilateral
  - Q65.1 – Congenital dislocation of hip, bilateral

Improved Defect Coding in ICD-10-CM

- **Gastroschisis and omphalocele**
  - ICD-9-CM: Both included in 756.79 – Anomalies of abdominal wall
  - ICD-10-CM:
    - Q79.2 – Exomphalos, Omphalocele
    - Q79.3 – Gastroschisis
- **Arnold-Chiari syndrome**
  - ICD-9-CM: Included in 741.0 - Spina bifida with hydrocephalus
  - ICD-10-CM:
    - Q07.00 – Arnold-Chiari syndrome without spina bifida or hydrocephalus
    - Q07.01 – Arnold-Chiari syndrome with spina bifida
    - Q07.02 – Arnold-Chiari syndrome with hydrocephalus
    - Q07.03 – Arnold-Chiari syndrome with spina bifida and hydrocephalus

Changes to Defect Coding in ICD-10-CM

- **Persistent fetal circulation**
  - ICD-9-CM: Included under 747.8 – Other specified anomalies of circulatory system
  - ICD-10-CM: Included under P29 – Cardiovascular disorders originating in the perinatal period
- **Neurofibromatosis**
  - ICD-9-CM: Included under 237 – Neoplasm of uncertain behavior of endocrine glands and nervous system
  - ICD-10-CM: Included under Q85 – Phakomatoses, not elsewhere classified
- **Fetal alcohol syndrome**
  - ICD-9-CM: Included under 760.7 – Noxious influences affecting fetus or newborn via placenta or breast milk
  - In ICD-10-CM: Included under Q86 – Congenital malformation syndromes due to known exogenous causes, not elsewhere classified
- **Epispadias**
  - ICD-9-CM: Included under 752 – Congenital anomalies of genital organs
  - ICD-10-CM: Included under Q64 – Other congenital malformations of urinary system

Changes to Defect Coding in ICD-10-CM

- **Megalencephaly vs. macrocephaly**
  - ICD-9-CM: Macroencephaly and megalencephaly included under 742.4 – Other specified anomalies of brain
    - There is no code for macrocephaly
  - ICD-10-CM: Megalencephaly included under Q04 – Other congenital malformations of brain
    - Macrocephaly included under Q75 – Other congenital malformations of skull and face bones
- **Conditions not listed in ICD-9-CM but added to 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.

Changes to Defect Coding in ICD-10-CM

- **Cleft lip and/or cleft palate**
  - ICD-9-CM
    - Unilateral vs. bilateral for both
    - Complete vs. incomplete for both
    - Includes cleft lip and/or cleft palate, unspecified
    - Does not distinguish hard vs. soft palate
  - ICD-10-CM
    - Unilateral, bilateral, median for cleft lip
    - Hard vs. soft cleft palate
    - Includes unspecified cleft palate (with or without cleft lip), but not unspecified cleft lip alone (included under unilateral cleft lip)
    - Does not distinguish complete vs. incomplete for either

Problematic Coding in ICD-10-CM Heart Valves

- **ICD-9-CM**
  - Pulmonary valve: atresia, stenosis, other, unspecified
  - Tricuspid valve: atresia and stenosis, Ebstein anomaly
  - Aortic valve: stenosis, insufficiency
  - Mitral valve: stenosis, insufficiency
- **ICD-10-CM**
  - Pulmonary valve: atresia, stenosis, insufficiency, other
  - Tricuspid valve: stenosis (includes atresia), Ebstein anomaly, other, unspecified
  - Aortic valve: stenosis (includes atresia), insufficiency
  - Mitral valve: stenosis (includes atresia), insufficiency
  - Aortic and mitral valves: other, unspecified

Problematic Coding in ICD-10-CM

- **Atrial septal defect (ASD) and patent foramen ovale (PFO)**
  - ICD-9-CM: 745.5 – Ostium secundum type ASD (includes ASD and PFO)
  - ICD-10-CM: Q21.1 – Atrial septal defect (includes ASD and PFO)
- **Interrupted aortic arch (IAA)**
  - ICD-9-CM: 747.11 – Interruption of aortic arch
  - In ICD-10-CM: IAA is not mentioned anywhere
    - Could be coded under Q25.2 – Atresia of aorta or Q25.4 – Other congenital malformations of aorta
- **Clinical genetic syndromes**
  - Many more specific syndromes mentioned
  - Some interspersed throughout defect codes
  - Conditions lumped together into single codes based on a single clinical feature (facial appearance, short stature, limb involvement, etc.)

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Probable Change in Defect Prevalence

Defect	ICD-9-CM Codes	ICD-10-CM Codes	Comments	Effect on Prevalence
Holoprosencephaly	742.2	Q04.2	ICD-9-CM includes other reduction defects	Decrease
Cloacal exstrophy	751.5	Q64.12	ICD-9-CM includes other anomalies of intestine	Decrease
Congenital posterior urethral valves	753.6	Q64.2	ICD-9-CM includes other atresia and stenosis of bladder neck	Decrease
Craniosynostosis	756.0	Q75.0	ICD-9-CM includes other anomalies of skull and face bones	Decrease
Interrupted aortic arch (IAA)	747.11	Q25.2, Q25.4	ICD-10-CM does not specify a code for IAA	Increase
Clubfoot	754.51, 754.70	Q66.0, Q66.89	ICD-10-CM includes other congenital deformities of feet	Increase

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No Change in Defect Prevalence

Defect	ICD-9-CM Codes	ICD-10-CM Codes	Comments	Effect on Prevalence
Anophthalmia/Microphthalmia	743.0, 743.1	Q11.0-Q11.2	----------	No change
Congenital cataract	743.30-743.34	Q12.0	----------	No change
Anotia/Microtia	744.01, 744.23	Q16.0, Q17.2	----------	No change
Atrioventricular septal defect/ Endocardial cushion defect	745.60, 745.61, 745.69	Q21.2	----------	No change
Ebstein anomaly	746.2	Q22.5	----------	No change
Hypoplastic left heart syndrome	746.7	Q23.4	----------	No change
Pulmonary valve atresia and stenosis	746.01, 746.02	Q22.0, Q22.1	----------	No change
Tricuspid valve atresia and stenosis	746.1	Q22.4	----------	No change
Choanal atresia	748.0	Q30.0	----------	No change
Biliary atresia	751.61	Q44.2-Q44.3	----------	No change

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No Change in Defect Prevalence

Defect	ICD-9-CM Codes	ICD-10-CM Codes	Comments	Effect on Prevalence
Cleft lip with cleft palate	749.2	Q37.0-Q37.9	----------	No change
Cleft lip alone	749.1	Q36.0-Q36.9	----------	No change
Cleft palate alone	749.0	Q35.1-Q35.9	----------	No change
Esophageal atresia/ Tracheoesophageal fistula	750.3	Q39.0-Q39.4	----------	No change
Bladder exstrophy	753.5	Q64.10, Q64.19	----------	No change
Hypospadias	752.61	Q54.0-Q54.9 excluding Q54.4	----------	No change
Renal agenesis/hypoplasia	753.0	Q60.0-Q60.6	----------	No change
Diaphragmatic hernia	756.6	Q79.0, Q79.1	----------	No change
Limb deficiencies/ Reduction defects of lower and unspecified limb	755.3-755.4	Q72.0-Q72.9, Q73.0-Q73.8	----------	No change

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No Change in Defect Prevalence

Defect	ICD-9-CM Codes	ICD-10-CM Codes	Comments	Effect on Prevalence
Deletion 22q11.2	758.32	Q93.81	----------	No change
Trisomy 13	758.1	Q91.4-Q91.7	----------	No change
Trisomy 18	758.2	Q91.0-Q91.3	----------	No change
Trisomy 21/Down syndrome	758.0	Q90.0-Q90.9	----------	No change

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Possible Small Change in Defect Prevalence

Defect	ICD-9-CM Codes	ICD-10-CM Codes	Comments	Effect on Prevalence
Anencephalus	740.0-740.1	Q00.0-Q00.1	ICD-10-CM includes acephaly	Small increase
Spina bifida without anencephalus	741.0, 741.9 without 740.0-740.1	Q05.0-Q05.9, Q07.01, Q07.03 without Q00.0-Q00.1	ICD-9-CM may include Arnold Chiari w/o spina bifida	Small decrease
Encephalocele	742.0	Q01.0-Q01.9	ICD-10-CM includes Arnold-Chiari type III	Small increase
Aortic valve stenosis	746.3	Q23.0	ICD-10-CM excludes aortic valve stenosis in HLHS	Small decrease
Atrial septal defect	745.5	Q21.1	ICD-10-CM specifies more individual types	Small increase
Coarctation of the aorta	747.1	Q25.1	ICD-9-CM includes hypoplasia of aorta and IAA; ICD-10-CM codes hypoplasia elsewhere, no code for IAA	Small decrease
Common truncus/ Truncus arteriosus	745.0	Q20.0	ICD-10-CM excludes aortic septal defect	Small decrease

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Possible Small Change in Defect Prevalence

Defect	ICD-9-CM Codes	ICD-10-CM Codes	Comments	Effect on Prevalence
Double outlet right ventricle	745.11	Q20.1	ICD-9-CM includes dextrotransposition of the aorta	Small decrease
Single ventricle	745.3	Q20.4	ICD-10-CM specifies double inlet ventricle	Small increase
Tetralogy of Fallot	745.2	Q21.3	ICD-9-CM includes Pentalogy of Fallot	Small decrease
Total anomalous pulmonary venous connection	747.41	Q26.2	ICD-10-CM has a separate code for unspecified anomalous pulmonary venous connection	Small decrease
Transposition of the great arteries	745.10, 745.12, 745.19	Q20.3, Q20.5	ICD-10-CM specifies ventricular inversion and levotransposition, includes dextrotransposition of the aorta	Small increase
Ventricular septal defect	745.4	Q21.0	ICD-9-CM includes Eisenmenger's defect	Small decrease

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Possible Small Change in Defect Prevalence

Defect	ICD-9-CM Codes	ICD-10-CM Codes	Comments	Effect on Prevalence
Limb deficiencies/ Reduction defects of upper limb	755.2	Q71.0-Q71.9	ICD-10-CM includes lobster-claw hand	Small increase
Rectal and large intestinal atresia/stenosis	751.2	Q42.0-Q42.9	ICD-9-CM includes obstruction of large intestine	Small decrease
Small intestinal atresia/stenosis	751.1	Q41.0-Q41.9	ICD-9-CM includes obstruction of small intestine, NOS	Small decrease
Gastroschisis	756.73	Q79.3	Previous ICD-9-CM code 756.7 included omphalocele	Decrease or no change
Omphalocele	756.72	Q79.2	Previous ICD-9-CM code 756.7 included gastroschisis	Decrease or no change
Turner syndrome	758.6	Q96.0-Q96.9	ICD-10-CM codes pure gonadal dysgenesis elsewhere	Small decrease or no change

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

- **Developed for birth defect programs that primarily use ICD-9-CM for defect coding**
- **Began by identifying all valid alternative ICD-10-CM codes in the general equivalence mappings (GEMs) developed by NCHS**
  - Made modifications appropriate for birth defects surveillance needs
- **For each code in ICD-9-CM, the tool provides possible alternative codes in ICD-10-CM**
  - Many ICD-9-CM codes have only one corresponding code in ICD-10-CM
  - When more than one possible alternative is available, programs can select the one that most closely matches the defect
  - If there is not enough information about the defect to select among multiple alternatives, 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
- **An ICD-10-CM to ICD-9-CM code translation tool is also available from the NBDPN website**

CDC Expanded Code for Birth Defects

- **Intended for use by programs that currently use the CDC/BPA code or a modification of it**
- **Analogous to ICD-10-CM as the CDC/BPA code is to ICD-9-CM**
  - Expand the range of numbers in each ICD-10-CM code
  - Maintain collapsibility to ICD-10-CM
  - Increase specificity, laterality, degree of severity, location of defect, etc.
  - Add specific conditions not coded in ICD-10-CM (e.g., syndromes)
  - Update terminology/medical knowledge
  - Resolve issues that were problematic in the CDC/BPA code
  - Ensure that all sections have codes for "Other" and "Unspecified" defects

CDC Expanded Code for Birth Defects

- **Development began in the early 1990's**
  - Identified defects frequently coded in the "Other" or "Other specified" categories in MACDP in order to give them individual codes
  - Identified defects recommended by MACDP staff and expert clinicians that are not coded in CDC/BPA
  - Identified codes and conditions that had been added or modified in the CDC/BPA code by other programs that use it
  - Reviewed the literature to identify conditions or variants not currently coded in CDC/BPA
- **Next will share with state programs that do their own coding**
  - Assess utility and obtain feedback, corrections, advice
  - Still need to harmonize the expansion with defect codes in BD-STEPS
- **MACDP plans to begin using the expanded code for births in January 2016**
  - Continue to use the CDC/BPA code for births through December 2015

ICD-10-CM Tools and Resources

- NBDPN Website Page on ICD-10-CM and Birth Defects: http://www.nbdpn.org/icd9_icd10_code_translation.php
  - Code Translations from ICD-9-CM to ICD-10-CM and from ICD-10-CM to ICD-9-CM for Birth Defects Surveillance (Excel files)
  - ICD-10-CM Implementation Plan Template
  - Slides from three webinars on "Transition to Use of ICD-10-CM Coding for Birth Defects"
- NBDPN Coding Tools Work Group Message Board (Members Only Section): http://www.viethconsulting.com/members/forum/board_list.php
  - NBDPN members can post questions about ICD-10-CM, share experiences and tips, discuss common concerns, etc.

ICD-10-CM Tools and Resources

- National Center for Health Statistics: http://www.cdc.gov/nchs/icd/icd10cm.htm
  - 2016 release of ICD-10-CM Code, Guidelines, and General Equivalence Mapping files
- Centers for Medicare & Medicaid Services: https://www.cms.gov/Medicare/Coding/ICD10/2016-ICD-10-PCS-and-GEMs.html
  - 2016 release of ICD-10-PCS Code, Guidelines, Addendum, and General Equivalence Mapping files
- CDC Website on Public Health Transition to ICD-10-CM/PCS: http://www.cdc.gov/nchs/icd/icd10cm_pcs.htm
  - Transition Planning, Trainings, Resources, FAQs
- American Health Information Management Association (AHIMA): http://www.ahima.org/topics/icd10
  - ICD-10- Implementation Toolkit, Preparation Checklist

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