Birth Defects Coding and ICD-10-CM

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National Birth Defects Prevention Network (NBDPN)
18th Annual Meeting
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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.)
## NBDPN Annual Report
### Probable Change in Defect Prevalence

<table>
<thead>
<tr>
<th>Defect</th>
<th>ICD-9-CM Codes</th>
<th>ICD-10-CM Codes</th>
<th>Comments</th>
<th>Effect on Prevalence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Holoprosencephaly</td>
<td>742.2</td>
<td>Q04.2</td>
<td>ICD-9-CM includes other reduction defects</td>
<td>Decrease</td>
</tr>
<tr>
<td>Cloacal exstrophy</td>
<td>751.5</td>
<td>Q64.12</td>
<td>ICD-9-CM includes other anomalies of intestine</td>
<td>Decrease</td>
</tr>
<tr>
<td>Congenital posterior urethral valves</td>
<td>753.6</td>
<td>Q64.2</td>
<td>ICD-9-CM includes other atresia and stenosis of bladder neck</td>
<td>Decrease</td>
</tr>
<tr>
<td>Craniosynostosis</td>
<td>756.0</td>
<td>Q75.0</td>
<td>ICD-9-CM includes other anomalies of skull and face bones</td>
<td>Decrease</td>
</tr>
<tr>
<td>Interrupted aortic arch (IAA)</td>
<td>747.11</td>
<td>Q25.2, Q25.4</td>
<td>ICD-10-CM does not specify a code for IAA</td>
<td>Increase</td>
</tr>
<tr>
<td>Clubfoot</td>
<td>754.51, 754.70</td>
<td>Q66.0, Q66.89</td>
<td>ICD-10-CM includes other congenital deformities of feet</td>
<td>Increase</td>
</tr>
</tbody>
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No Change in Defect Prevalence

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<tr>
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<tbody>
<tr>
<td>Anophthalmia/Microphthalmia</td>
<td>743.0, 743.1</td>
<td>Q11.0-Q11.2</td>
<td></td>
<td>No change</td>
</tr>
<tr>
<td>Congenital cataract</td>
<td>743.30-743.34</td>
<td>Q12.0</td>
<td></td>
<td>No change</td>
</tr>
<tr>
<td>Anotia/Microtia</td>
<td>744.01, 744.23</td>
<td>Q16.0, Q17.2</td>
<td></td>
<td>No change</td>
</tr>
<tr>
<td>Atrioventricular septal defect/Endocardial cushion defect</td>
<td>745.60, 745.61, 745.69</td>
<td>Q21.2</td>
<td></td>
<td>No change</td>
</tr>
<tr>
<td>Ebstein anomaly</td>
<td>746.2</td>
<td>Q22.5</td>
<td></td>
<td>No change</td>
</tr>
<tr>
<td>Hypoplastic left heart syndrome</td>
<td>746.7</td>
<td>Q23.4</td>
<td></td>
<td>No change</td>
</tr>
<tr>
<td>Pulmonary valve atresia and stenosis</td>
<td>746.01, 746.02</td>
<td>Q22.0, Q22.1</td>
<td></td>
<td>No change</td>
</tr>
<tr>
<td>Tricuspid valve atresia and stenosis</td>
<td>746.1</td>
<td>Q22.4</td>
<td></td>
<td>No change</td>
</tr>
<tr>
<td>Choanal atresia</td>
<td>748.0</td>
<td>Q30.0</td>
<td></td>
<td>No change</td>
</tr>
<tr>
<td>Biliary atresia</td>
<td>751.61</td>
<td>Q44.2-Q44.3</td>
<td></td>
<td>No change</td>
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<tr>
<td>Cleft lip with cleft palate</td>
<td>749.2</td>
<td>Q37.0-Q37.9</td>
<td>-----------------</td>
<td>No change</td>
</tr>
<tr>
<td>Cleft lip alone</td>
<td>749.1</td>
<td>Q36.0-Q36.9</td>
<td>-----------------</td>
<td>No change</td>
</tr>
<tr>
<td>Cleft palate alone</td>
<td>749.0</td>
<td>Q35.1-Q35.9</td>
<td>-----------------</td>
<td>No change</td>
</tr>
<tr>
<td>Esophageal atresia/Tracheoesophageal fistula</td>
<td>750.3</td>
<td>Q39.0-Q39.4</td>
<td>-----------------</td>
<td>No change</td>
</tr>
<tr>
<td>Bladder extrophy</td>
<td>753.5</td>
<td>Q64.10, Q64.19</td>
<td>-----------------</td>
<td>No change</td>
</tr>
<tr>
<td>Hypospadias</td>
<td>752.61</td>
<td>Q54.0-Q54.9</td>
<td>-----------------</td>
<td>No change</td>
</tr>
<tr>
<td>Renal agenesis/hypoplasia</td>
<td>753.0</td>
<td>Q60.0-Q60.6</td>
<td>-----------------</td>
<td>No change</td>
</tr>
<tr>
<td>Diaphragmatic hernia</td>
<td>756.6</td>
<td>Q79.0, Q79.1</td>
<td>-----------------</td>
<td>No change</td>
</tr>
<tr>
<td>Limb deficiencies/Reduction defects of lower and unspecified limb</td>
<td>755.3-755.4</td>
<td>Q72.0-Q72.9, Q73.0-Q73.8</td>
<td>-----------------</td>
<td>No change</td>
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<tr>
<td>Deletion 22q11.2</td>
<td>758.32</td>
<td>Q93.81</td>
<td>-----------------</td>
<td>No change</td>
</tr>
<tr>
<td>Trisomy 13</td>
<td>758.1</td>
<td>Q91.4-Q91.7</td>
<td>-----------------</td>
<td>No change</td>
</tr>
<tr>
<td>Trisomy 18</td>
<td>758.2</td>
<td>Q91.0-Q91.3</td>
<td>-----------------</td>
<td>No change</td>
</tr>
<tr>
<td>Trisomy 21/Down syndrome</td>
<td>758.0</td>
<td>Q90.0-Q90.9</td>
<td>-----------------</td>
<td>No change</td>
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#### Possible Small Change in Defect Prevalence

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<tr>
<td>Anencephalus</td>
<td>740.0-740.1</td>
<td>Q00.0-Q00.1</td>
<td>ICD-10-CM includes acephaly</td>
<td>Small increase</td>
</tr>
<tr>
<td>Spina bifida without anencephalus</td>
<td>741.0, 741.9 without 740.0-740.1</td>
<td>Q05.0-Q05.9, Q07.01, Q07.03 without Q00.0-Q00.1</td>
<td>ICD-9-CM may include Arnold Chiari w/o spina bifida</td>
<td>Small decrease</td>
</tr>
<tr>
<td>Encephalocele</td>
<td>742.0</td>
<td>Q01.0-Q01.9</td>
<td>ICD-10-CM includes Arnold-Chiari type III</td>
<td>Small increase</td>
</tr>
<tr>
<td>Aortic valve stenosis</td>
<td>746.3</td>
<td>Q23.0</td>
<td>ICD-10-CM excludes aortic valve stenosis in HLHS</td>
<td>Small decrease</td>
</tr>
<tr>
<td>Atrial septal defect</td>
<td>745.5</td>
<td>Q21.1</td>
<td>ICD-10-CM specifies more individual types</td>
<td>Small increase</td>
</tr>
<tr>
<td>Coarctation of the aorta</td>
<td>747.1</td>
<td>Q25.1</td>
<td>ICD-9-CM includes hypoplasia of aorta and IAA; ICD-10-CM codes hypoplasia elsewhere, no code for IAA</td>
<td>Small decrease</td>
</tr>
<tr>
<td>Common truncus/Truncus arteriosus</td>
<td>745.0</td>
<td>Q20.0</td>
<td>ICD-10-CM excludes aortic septal defect</td>
<td>Small decrease</td>
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<tr>
<td>Double outlet right ventricle</td>
<td>745.11</td>
<td>Q20.1</td>
<td>ICD-9-CM includes dextrotransposition of the aorta</td>
<td>Small decrease</td>
</tr>
<tr>
<td>Single ventricle</td>
<td>745.3</td>
<td>Q20.4</td>
<td>ICD-10-CM specifies double inlet ventricle</td>
<td>Small increase</td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>745.2</td>
<td>Q21.3</td>
<td>ICD-9-CM includes Pentalogy of Fallot</td>
<td>Small decrease</td>
</tr>
<tr>
<td>Total anomalous pulmonary venous connection</td>
<td>747.41</td>
<td>Q26.2</td>
<td>ICD-10-CM has a separate code for unspecified anomalous pulmonary venous connection</td>
<td>Small decrease</td>
</tr>
<tr>
<td>Transposition of the great arteries</td>
<td>745.10, 745.12, 745.19</td>
<td>Q20.3, Q20.5</td>
<td>ICD-10-CM specifies ventricular inversion and levotransposition, includes dextrotransposition of the aorta</td>
<td>Small increase</td>
</tr>
<tr>
<td>Ventricular septal defect</td>
<td>745.4</td>
<td>Q21.0</td>
<td>ICD-9-CM includes Eisenmenger’s defect</td>
<td>Small decrease</td>
</tr>
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<tr>
<td>Limb deficiencies/Reduction defects of upper limb</td>
<td>755.2</td>
<td>Q71.0-Q71.9</td>
<td>ICD-10-CM includes lobster-claw hand</td>
<td>Small increase</td>
</tr>
<tr>
<td>Rectal and large intestinal atresia/stenosis</td>
<td>751.2</td>
<td>Q42.0-Q42.9</td>
<td>ICD-9-CM includes obstruction of large intestine</td>
<td>Small decrease</td>
</tr>
<tr>
<td>Small intestinal atresia/stenosis</td>
<td>751.1</td>
<td>Q41.0-Q41.9</td>
<td>ICD-9-CM includes obstruction of small intestine, NOS</td>
<td>Small decrease</td>
</tr>
<tr>
<td>Gastroschisis</td>
<td>756.73</td>
<td>Q79.3</td>
<td>Previous ICD-9-CM code 756.7 included omphaloele</td>
<td>Decrease or no change</td>
</tr>
<tr>
<td>Omphaloele</td>
<td>756.72</td>
<td>Q79.2</td>
<td>Previous ICD-9-CM code 756.7 included gastrochisis</td>
<td>Decrease or no change</td>
</tr>
<tr>
<td>Turner syndrome</td>
<td>758.6</td>
<td>Q96.0-Q96.9</td>
<td>ICD-10-CM codes pure gonadal dysgenesis elsewhere</td>
<td>Small decrease or no change</td>
</tr>
</tbody>
</table>
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

  - 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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For more information, contact CDC
1-800-CDC-INFO (232-4636)

The findings and conclusions in this presentation are those of the authors and do not necessarily represent the official position of the Centers for Disease Control and Prevention.