

# CHAPTER 17

CONGENITAL MALFORMATIONS, DEFORMATIONS AND CHROMOSOMAL  
ABNORMALITIES (Q00-Q99)

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## CHAPTER SPECIFIC CATEGORY CODE BLOCKS

- 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

## CHAPTER NOTES

- Codes from this chapter are not for use on maternal or fetal records.
- These codes are assigned when a malformation, deformation or chromosomal abnormality is documented, and the code may be the principal or first-listed diagnosis on a record or a secondary diagnosis.
- Codes from Chapter 17 may be used throughout the life of the patient.
- Block, category, subcategory and code title changes have been made in Chapter 17; for example in ICD-9-CM, code 758.1 is titled “Patau’s Syndrome,” whereas the counterpart codes in ICD-10-CM are titled “Trisomy 13.”
- Many codes for congenital conditions and chromosomal abnormalities have been expanded in ICD-10-CM; for example, chromosomal anomalies are classified to category 758 in ICD-9-CM; however in ICD-10-CM, there are nine categories for chromosomal abnormalities, not elsewhere classified.

## CONGENITAL MALFORMATIONS, DEFORMATIONS AND CHROMOSOMAL ABNORMALITIES

- When a malformation, deformation or chromosomal abnormality does not have a unique code assignment, assign additional code(s) for any manifestations that may be present.
- When the code assignment specifically identifies the malformation, deformation or chromosomal abnormality, manifestations that are an inherent component of the anomaly should not be coded separately.
- Additional codes should be assigned for manifestations that are not an inherent component.
- If a congenital malformation or deformity has been corrected, a personal history code should be used to identify the history of the malformation or deformity.
- Although present at birth, malformation, deformation or chromosomal abnormality may not be identified until later in life.
- Whenever the condition is diagnosed by the physician, it is appropriate to assign a code from codes Q00-Q99.

## CONGENITAL MALFORMATIONS, DEFORMATIONS AND CHROMOSOMAL ABNORMALITIES (cont.)

- For the birth admission, the appropriate code from category Z38, Liveborn infants, according to place of birth and type of delivery, should be sequenced as the principal diagnosis, followed by any congenital anomaly codes, Q00-Q99.
- Congenital anomalies or syndromes may occur as a set of symptoms or multiple malformations.
- For syndromes with specific codes, additional codes may be assigned to identify manifestations not included in the specific code.

## CODING EXAMPLES

1. Assign the code(s) for the following diagnosis: Cleft palate involving both the soft and hard palate, with bilateral cleft lip.

**Answer:** Q37.4 Cleft, (congenital) lip (unilateral), bilateral, with cleft palate, hard with soft

**Rationale:** Careful review of the documentation is indicated to select the one code that combines these conditions. Cleft lip and palate are congenital defects caused when the bones and tissues don't fuse together in utero. The palate is the roof of the mouth, and consists of the soft (back part near the throat) and the hard (front part behind the teeth) palates. Frequently cleft lip and palate are both present. A cleft lip can be either unilateral or bilateral. The unilateral cleft lip has a gap on one side of the lip under either the left or right nostril, but in a bilateral cleft lip, the gap is on both side of the lip. ICD-9-CM classified cleft palate as unilateral versus bilateral and complete versus incomplete, while ICD-10-CM classifies it by hard, soft, hard with soft, uvular and unspecified. ICD-9-CM classifies cleft lip by unilateral versus bilateral and complete versus incomplete while ICD-10-CM uses the terms bilateral, median, or unilateral.

## CODING EXAMPLES (cont.)

**Rationale to example one cont.:** Cleft lip and palate in ICD-10-CM is classified according to hard versus soft palate with unilateral versus bilateral cleft lip. The terms complete versus incomplete were used in ICD-9-CM to classify this condition, and are no longer present in ICD-10-CM.

2. Assign the code for the following diagnosis: Frontal encephalocele with hydroencephalocele.

**Answer:** Q01.0 Encephalocele, frontal

**Rationale:** Encephalocele has been expanded in ICD-10-CM from one code to five codes. An encephalocele is defined as a congenital malformation in which brain tissue protruding through a skull defect. Hydroencephalocele is included in code Q01.0.

## CODING EXAMPLES (cont.)

3. Assign the code(s) for the following diagnosis: Penoscrotal hypospadias.

**Answer:** Q54.2 Hypospadias, penoscrotal

**Rationale:** In ICD-9-CM, there was one code to identify this condition, whereas in ICD-10-CM codes are available for hypospadias balanic, penile, penoscrotal, perineal, congenital chordee, other hypospadias, and unspecified. Hypospadias refers to a congenital condition in which the urethral meatus lies on the ventral position of the penile shaft and may be located as far down as in the scrotum or perineum.



## CODING EXAMPLES (cont.)

4. This newborn was delivered by cesarean section and transferred immediately to NICU because of previous anomalies identified via sonogram and fetal echocardiogram. The newborn male has complete transposition of the great vessels with cyanosis. The baby received IV prostaglandin and underwent uneventful corrective surgery at four days, and was discharged 10 days post surgery. What diagnosis codes are assigned?

**Answer:** Z38.01 Newborn (infant) (liveborn) (singleton), born in hospital, by cesarean  
Q20.3 Transposition (congenital) vessels, great (complete) (partial)

**Rationale:** In this case, the newborn code is listed first. Transposition of the great vessels (TGV) is a congenital heart defect in which the aorta and the pulmonary artery are transposed. Because this is a cyanotic heart defect (too little oxygen) the cyanosis is inherent and not separately coded.

## TRAINING SOURCES

American Health Information Management Association  
[www.ahima.org](http://www.ahima.org)