



# Western Australian Coding Rule

## **0719/45 Costello syndrome**

WA Coding Rule 0612/02 *Costello Syndrome* is retired.

In ICD-10-AM/ACHI/ACS Eleventh Edition (effective 1 July 2019) ACS 0005 *Syndromes* was amended and new code U91 *Syndrome, NEC* created. Coders should be guided by the classification instructions in ACS 0005 *Syndromes*.

## **DECISION**

**WA Coding Rule 0612/02 *Costello Syndrome* is retired.**

[Effective 01 Jul 2019, ICD-10-AM/ACHI/ACS 11<sup>th</sup> Ed.]



# Western Australian Coding Rule

## 0612/02 Costello syndrome

### Q.

What code/s should be assigned for Costello syndrome? The Paediatrician describes the patient as having a history of Costello syndrome with associated seizure disorder. The patient is described as having “a background history of Costello syndrome, which comprises marked short stature, growth delay and developmental delay”.

Principal Diagnosis: Unprovoked seizure on background of Costello syndrome

The patient was admitted overnight for observations and additional anti-seizure medications were prescribed.

### A.

Referring to the ACS 0005 *Syndromes*, if no single code is available to describe all elements of a syndrome it can be difficult to code all elements separately. The guidelines for sequencing when coding syndromes for this case would dictate the coding of the seizure as the principal diagnosis.

If the syndrome is congenital assign Q87.- *Other specified congenital malformation syndromes affecting multiple systems* as an additional diagnosis to the specified manifestations already coded. When assigning multiple codes to represent the syndrome assign codes only for the manifestations that are relevant for the patient in question as all standard manifestations of a syndrome may not be present in every patient with the syndrome.

In this case the manifestations documented are short stature, growth delay and developmental delay.

For the case described, code assignment is:

Principal diagnosis: R56.8 *Other and unspecified convulsions*  
E34.3 *Short stature, not elsewhere classified*  
R62.9 *Lack of expected normal physiological development, unspecified*  
Q87.89 *Other specified congenital malformation syndromes, not elsewhere classified*

## DECISION

For the case described, code assignment is:

**Principal diagnosis: R56.8 *Other and unspecified convulsions***  
**E34.3 *Short stature, not elsewhere classified***  
**R62.9 *Lack of expected normal physiological development, unspecified***  
**Q87.89 *Other specified congenital malformation syndromes, not elsewhere classified***

[Effective 06 Jun 2012, ICD-10-AM/ACHI/ACS 7<sup>th</sup> Ed.]