



Western Australian Coding Rule

0516/01 Tumour necrosis factor receptor-associated periodic syndrome

Q.

What is the correct code to assign for tumour necrosis factor receptor-associated periodic syndrome (TRAPS)?

A.

Tumour necrosis factor receptor-associated periodic syndrome (TRAPS) is a rare genetic auto-inflammatory syndrome. It presents as recurrent, prolonged episodes of fever typically associated with serosal, synovial and cutaneous inflammation.

It can be complicated by amyloidosis, resulting in kidney or liver failure. TRAPS is the most common autosomal dominant form of periodic fever syndrome and was originally known as familial Hibernian fever (FHF).

Hereditary periodic fever syndromes (HPFSs) are rare and distinct heritable disorders characterised by short and recurrent attacks of fever and severe localised inflammation that occur periodically or irregularly and that are not explained by usual childhood infections.

These attacks undergo spontaneous remission without antibiotic, anti-inflammatory, or immunosuppressive treatment. Between attacks, patients feel well and regain their normal daily functions until the next episode occurs. The episodes are usually associated with elevated serum levels of acute-phase reactants (e.g. fibrinogen, serum amyloid, an elevated erythrocyte sedimentation rate and leukocytosis).

The Index pathway for TRAPS is:

Fever

- periodic (Mediterranean) E85.0

E85.0 Non-neuropathic heredofamilial amyloidosis

DECISION

Tumour necrosis factor receptor-associated periodic syndrome (TRAPS) should be coded to E85.0 *Non-neuropathic heredofamilial amyloidosis* by following the pathway 'Fever/periodic' in the Alphabetic Index.

[Effective 25 May 2016, ICD-10-AM/ACHI/ACS 9th Ed.]