Government of Western Australia
Department of Health

Western Australian Coding Rule

0725/84 Hereditary periodic fever syndrome

Q.
What code is assigned for a hereditary periodic fever syndrome (such as Tumour necrosis factor receptor-associated periodic syndrome (TRAPS))?

A.
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 hereditary periodic fever syndrome and was originally known as familial Hibernian fever (FHF).

Hereditary periodic fever syndromes 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).

For hereditary periodic fever syndrome, assign E85.01 *Familial Mediterranean fever* via Alphabetic Index:

Fever
- periodic (Mediterranean) E85.01

DECISION

For hereditary periodic fever syndrome, such as tumour necrosis factor receptor-associated periodic syndrome (TRAPS), assign E85.01 *Familial Mediterranean fever*.

This WA Coding Rule 0725/84 *Hereditary periodic fever syndrome* supersedes WA Coding Rule 0516/01 *Tumour necrosis factor receptor-associated periodic syndrome (TRAPS)*.

This Rule has a minor modification to correspond with an update in ICD-10-AM/ACHI/ACS Twelfth Edition.

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Effective 1 July 2025, ICD-10-AM/ACHI/ACS 13th Ed.

As per the Patient Activity Data Policy (MP 0164/21) Western Australian Coding Rules must be followed.

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Western Australian Coding Rule

0516/01 Tumour necrosis factor receptor-associated periodic syndrome (TRAPS)

Q.
How is tumour necrosis factor receptor-associated periodic syndrome (TRAPS) coded?

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.]