Government of Western Australia
Department of Health

Western Australian Coding Rule

0721/02 Isolated pulmonary capillaritis

WA Coding Rule 0919/03 *Isolated pulmonary capillaritis* is superseded by ICD-10-AM/ACHI/ACS Coding Rule ***Isolated pulmonary capillaritis*** (Ref No: Q3670) effective 1 July 2021; (log in to view on the IHPA Australian Classification Exchange).

Western Australian Coding Rule

0919/03 Isolated pulmonary capillaritis

Q.
What diagnosis code should be assigned for isolated pulmonary capillaritis?

A.
"Pulmonary capillaritis" may be:

- A manifestation of a systemic vasculitide e.g. Granulomatosis with polyangiitis (GPA), formerly known as Wegener granulomatosis; Systemic Lupus Erythematosis etc.

or

- Isolated pulmonary capillaritis: a small vessel vasculitis confined to the lungs and without concomitant systemic involvement.

Definition of isolated pulmonary capillaritis in the Orphanet rare disease nomenclature:
Isolated pauciimmune pulmonary capillaritis is a small vessel vasculitis restricted to the lungs that may induce diffuse alveolar hemorrhage with dyspnea, anemia, chest pain, hemoptysis, bilateral and diffuse alveolar infiltrates at chest X-rays, without any underlying systemic disease. ANCA are frequently positive but could be negative.

In Orphanet and ICD-11, this rare disease is classified as an interstitial lung disease.

Following the logic in national Coding Rule Q3249 *Neuroendocrine cell hyperplasia of infancy* (NEHI), assign J84.8 *Other specified interstitial pulmonary diseases* as a best fit, by following the Alphabetic Index pathway:

Disease, diseased
- lung
- - interstitial
- - - specified NEC J84.8

DECISION
A query will be submitted to IHPA. In the meantime, assign J84.8 *Other specified interstitial pulmonary diseases* for isolated pulmonary capillaritis.

[Effective 01 October 2019, ICD-10-AM/ACHI/ACS 11th Ed.]