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WA Coding Rule 0919/03 *Isolated pulmonary capillaritis* is superseded by ICD-10-AM/ACHI/ACS Coding Rule *Isolated pulmonary capillaritis* (Ref No: Q3670) effective 1 July 2021; (log in to view on the [IHPA Australian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](https://www.aihw.gov.au)).
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 Erythematosus etc.
- 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.]