

## Plummer–Vinson syndrome

*Synonyms: Paterson–Brown Kelly syndrome, Paterson–Brown–Kelly syndrome, Waldenström–Kjellberg syndrome, sideropenic dysphagia, sideropenic nasopharyngopathy.*

### What is Plummer–Vinson syndrome?

Plummer–Vinson syndrome (PVS) is a triad of microcytic hypochromic anaemia (iron deficiency), atrophic glossitis, and oesophageal webs or strictures.<sup>[1]</sup> A description of this syndrome was published by Henry Plummer in Chicago in 1911 and also by Porter Vinson in Philadelphia in 1919. Donald Ross Paterson and Adam Brown Kelly published independently of each other in London in 1919.<sup>[2]</sup>

### How common is Plummer–Vinson syndrome? (Epidemiology)

PVS most often affects middle-aged women (this may be related to a propensity for iron deficiency), although it has also been recognised presenting in a child or older adults.<sup>[3]</sup> <sup>[4]</sup> The web (containing mucosa and submucosa) occurs at the anterior post-cricoid area of the upper oesophagus.<sup>[5]</sup>

It is a rare condition and much of the literature is case reports of one or a few individuals rather than large series. Figures for incidence and prevalence are not available but it is becoming rarer in the developed world, probably because dietary inadequacy is becoming less common.<sup>[6]</sup>

A high prevalence of PVS was reported in Sweden in the early 20th century. This reduced after the introduction of iron supplementation in food.<sup>[7]</sup>

PVS appears to be more common in the Indian subcontinent than in Western countries. PVS is common in males as well as females in the Indian subcontinent.<sup>[8]</sup> <sup>[9]</sup>

## Risk factors

Iron deficiency – this is related to poor diet or blood loss. <sup>[10]</sup>

## Plummer-Vinson syndrome symptoms (presentation) <sup>[4] [11]</sup>

The presentation is usually with painless, intermittent dysphagia. It tends to be with solid foods but, if untreated, may progress to soft foods and even liquids. As with other causes of oesophageal strictures, webs and rings, the patient is able to give a good indication of the level of obstruction. In this condition the patient indicates the upper oesophagus. Dysphagia may lead to weight loss. This should be regarded as a sign of danger.

There may also be features of iron-deficiency anaemia like lethargy, tiredness and shortness of breath on exertion.

## Examination <sup>[10]</sup>

The features that appear on examination are those associated with iron deficiency. There may be pallor and even tachycardia if anaemia is marked. There may be koilonychia (spoon-shaped nails), angular cheilitis and glossitis.

## Investigation <sup>[11]</sup>

- FBC will show a microcytic, hypochromic anaemia. Ferritin is low.
- Barium swallow may show the web. This may need to be enhanced with videofluoroscopy.
- Biopsy may be required if malignancy is suspected clinically.

## Associated diseases

Various disorders have been known to associate PVS, such as [coeliac disease](#), [inflammatory bowel disease](#), [pernicious anaemia](#), thyroid disease, [Sjögren's syndrome](#) and [rheumatoid arthritis](#).

## Plummer-Vinson syndrome treatment<sup>[12]</sup> <sup>[13]</sup>

- Iron replacement can almost invariably be achieved by oral means. There is rarely any need for parenteral iron. Supplements may be needed long-term because after correction it is important to maintain a normal iron status. Causes of blood loss like menorrhagia may require attention.
- Regular endoscopic surveillance is required to detect transformation into squamous cell carcinoma.<sup>[14]</sup>
- Endoscopic dilatation or argon plasma coagulation therapy of the oesophageal web is occasionally required in cases of persistent dysphagia.<sup>[15]</sup>

## Complications

Post-cricoid carcinoma or carcinoma of the oesophagus may develop.<sup>[10]</sup>

## Prognosis and prevention

Correction of iron deficiency will correct the condition if malignancy has not yet occurred. Prevention of iron deficiency prevents the disease.

## Historical notes<sup>[2]</sup>

- Henry Plummer (1874–1937) was an American internist. He was a professor at the Medical School of Minnesota. He published: *Diffuse dilatation of the esophagus without anatomic stenosis (cardiospasm). A report of ninety-one cases*. Journal of the American Medical Association, Chicago, 1912, 58: 2013–2015.
- Porter Paisley Vinson (1890–1959) was an American surgeon at the Mayo Clinic. He published: *A case of cardiospasm with dilatation and angulation of the esophagus*. Medical Clinics of North America, Philadelphia, PA., 1919, 3: 623–627.
- Born in Inverness, Donald Ross Paterson (1863–1939) was an otolaryngologist. He worked in Cardiff and published: *A clinical type of dysphagia*. The Journal of Laryngology, Rhinology and Otology, London, 1919, 24: 289–291.

- Adam Brown Kelly (1865–1941) graduated from Glasgow but worked as an ENT surgeon in London, Berlin and Vienna. He published: *Spasm at the entrance of the esophagus*. The Journal of Laryngology, Rhinology and Otology, London, 1919, 34: 285–289.

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## Further reading

- [NDR \(Nutrition and Diet Resources\) UK](#)
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