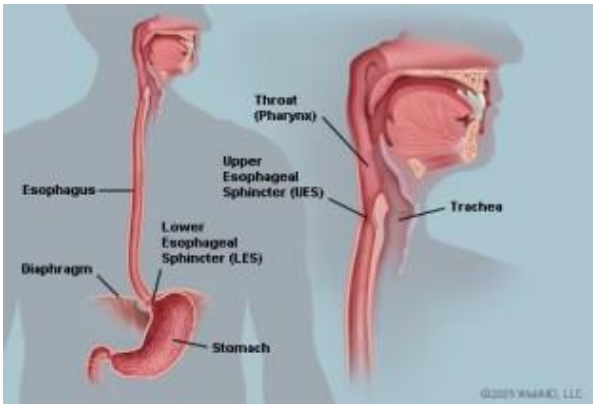
Cancer Association of South Africa (CANSA)

Research • Educate • Support

Fact Sheet
on
Sideropenic Dysphagia

Introduction

Sideropenic Dysphagia, also called Plummer–Vinson Syndrome (PVS) or Paterson–Brown–Kelly Syndrome, is a rare disease characterised by dysphagia (difficulty in swallowing), iron deficiency anaemia, and oesophageal webs. Treatment with iron supplementation and mechanical widening of the oesophagus generally provides an excellent outcome.



[Picture Credit: Oesophagus]

It generally occurs in postmenopausal women. Its identification and follow-up is considered relevant due to increased risk of post-cricoid carcinoma and squamous cell carcinomas of the oesophagus and pharynx.

Verma, S. & Mukherjee, S. 2020.

"Plummer-Vinson Syndrome (PVS) is a rare condition characterized by the classic triad of dysphagia, iron-deficiency anemia and esophageal web. Plummer Vinson Syndrome is more common in middle-aged women who appear to be at an increased risk of developing squamous cell carcinoma of the pharynx and proximal esophagus. This syndrome was named after two Mayo Clinic physicians, Henry Stanley Plummer (1874-1936) and Porter Paisley Vinson (1890-1959) who noted cases of iron deficiency and dysphagia in the presence of suspected spasm of the upper esophagus or abnormal angulation of the esophagus. In the United Kingdom, this condition is also known as Paterson-Brown-Kelly syndrome after two British laryngologists, Dr.Donald Ross Paterson (1863-1939) and Dr. Adam Brown-Kelly (1865-1941) who published their findings in 1919.Dr.Paterson was also the first physician to suggest an association between this syndrome and carcinoma while in charge of the department of Ear, Nose and Throat Medicine at the Cardiff Royal Infirmary."

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Incidence of Sideropenic Dysphagia in South Africa

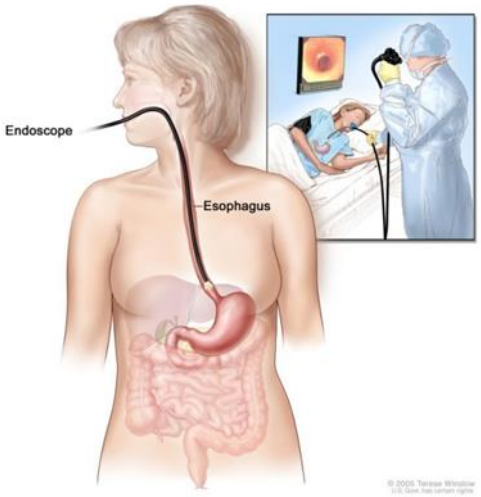
Because Sideropenic Dysphagia is not a cancerous condition in itself, the National Cancer Registry (2017) does not provide any information regarding the incidence of this condition.

Signs and Symptoms of Sideropenic Dysphagia

The list of signs and symptoms mentioned in various sources for Sideropenic Dysphagia include the following:

- Throat pain during swallowing
- Burning sensation during swallowing
- Sensation of food stuck in larynx
- Fatigue
- Pallor
- Pale inside of mouth
- Sideropenic anaemia
- Hypochromic anaemia
- Swallowing difficulty
- Mucosal webs in oesophagus
- Spoon-shaped fingernails
- Smooth tongue
- Red tongue
- Painful tongue

Diagnosis of Sideropenic Dysphagia



[Picture Credit: Oesophagoscopy]

The diagnosis is clinical with progressive and long-standing dysphagia associated with iron deficiency anaemia.

Oesophagoscopy (looking down the oesophagus with a special instrument) shows the oesophageal web as a thin diaphragm with pale, fragile, or normal appearing mucosa that partially obstructs the lumen of the oesophagus. The prognosis is good, despite the fact that the syndrome is associated with increased risk for post-cricoid carcinoma, pharyngeal and oesophageal cancers.

Steele, D.C., Owara, C. & McCarthy, D. 2020.

"We report a 39-year-old Native American female with an almost 20-year history of dysphagia that had increased in the 6 months prior to the initial evaluation. Investigation revealed a number of distinct esophageal disorders including Plummer-Vinson syndrome, gastroesophageal reflux disease with esophagitis, distal esophageal stricture, esophageal intramural pseudo-diverticulosis, and recurrent esophageal Candida infections. Although prolonged therapy with proton pump inhibitors,

fluconazole, nystatin, and repeated esophageal balloon dilations relieved her symptoms, her prognosis remains uncertain."

Causes of Sideropenic Dysphagia

The cause of this condition has been attributed to numerous factors that include alterations in oesophageal innervation. There is agreement that prolonged iron deficiency is necessary for the development of the syndrome; however, only a minority of patients with iron deficiency manifests the syndrome.

It is a rare disorder that can be linked to cancers of the oesophagus and throat. It is more common in women.

Paril, M., Malipatel, R. & Devarbhayi, J. 2020.

Background and aim: Plummer-Vinson syndrome (PVS) comprises triad of iron deficiency anemia, dysphagia, and post-cricoid esophageal web. PVS is rare nowadays due to improved nutritional status. However, we encountered patients with PVS regularly at our center. Data regarding PVS are limited; hence, we aimed to study the clinical features, treatment outcomes, and development of complications in patients with PVS.

Methods: The study was conducted over a 10-year period (January 2008 to January 2018) in a medical college setting. All adults with dysphagia, anemia, and post-cricoid web or those with iron deficiency anemia and post-cricoids web were included in the study. Patients were treated with iron supplementation and Savary-Gilliard bougie dilation of the web. Patients were followed-up for the recurrence of dysphagia and development of complications.

Results: Overall, 153 patients exhibited esophageal web, of which 132 (86.27%) patients had concomitant PVS and 21 (13.7%) patients did not. The mean age was 43.50 years (range 16-76) and 113 (85.6%) were women. Single session of Savary-Gilliard bougie dilation was successful in 90.7% of patients in relieving dysphagia and 9.3% developed recurrence, requiring repeated dilations. Four patients had concomitant squamous cell carcinoma of esophagus along with PVS and two developed upper gastrointestinal malignancy during follow-up.

Conclusion: Plummer-Vinson syndrome is predominantly seen in middle aged women and present with symptoms of iron deficiency anemia and early grade dysphagia. Single session of Savary-Gilliard bougie dilation was successful in majority of patients in relieving dysphagia. Overall risk of developing upper gastrointestinal malignancy was 4.5%.

Diagnosis of Sideropenic Dysphagia

The following tests may assist in making a diagnosis:

- Full Blood Count (FBC) will show a microcytic, hypochromic anaemia.
- Low Ferritin.
- Barium swallow may show the web. This may need to be enhanced with videofluoroscopy.
- A biopsy may be required if malignancy is suspected clinically.

Treatment of Sideropenic Dysphagia

Iron replacement can almost invariably be achieved by oral means. Adding vitamin C does not improve absorption significantly. 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.

Endoscopic dilatation or argon plasma coagulation therapy of the oesophageal web may occasionally be required in cases of persistent dysphagia.

Patil, M., Malipatel, R. & devarbhavi, H. 2021.

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