

## Rare Diseases that cause Dysphagia: Plummer-Vinson Syndrome

Violet O Cox\*

Department of Communication Science and Disorders, Cleveland State University, USA

\*Corresponding author: Violet O Cox, Department of Communication Science and Disorders, Cleveland State University, USA, Tel: +216-687-6909; E-mail: v.cox@csuohio.edu

Received date: October 26, 2018; Accepted date: November 8, 2018; Published date: November 14, 2018

Copyright: © 2018 Cox VO. This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

### Abstract

The general field of rare diseases is particularly challenging because of the delays in reaching a diagnosis as well as the many trials and errors before effective treatment can be reached. To date there are approximately 7,000 rare diseases. Most of these are derived from a genetic basis thus affecting the individual from birth or early childhood. Furthermore, many of these diseases affect multiple systems creating devastating effects on patient and family alike. For example, many of these rare diseases impact the patient's motor, cognitive and respiratory skills. Frequently, patients are unable to swallow safely. This may lead to aspiration pneumonia and even death. The slow turn around rate in precisely identifying and treating these diseases can have a significant impact on mortality. This chapter presents a summary of a rare disease, Plummer-Vinson syndrome (PVS). One of the main characteristics of PVS is the significant dysphagia (swallowing disorder) that occurs due to the presence of esophageal webs. PVS is discussed in terms of its history, etiology, epidemiology, clinical presentation, diagnosis, treatment and the significant impact on swallowing that results.

**Keywords:** Plummer-Vinson syndrome; Dysphagia; Esophageal web; Iron deficiency anemia

### Introduction

Rare diseases whether in adults or children have received increased attention over the last several years. This rise in attention is most likely due to the global sharing of information via the internet improvement in the understanding and treatment of more common diseases and the development of laws related to the treatment of rare medical conditions [1,2]. To date, there is no consensual definition of a rare disease but there are at least three broadly accepted methods of defining rare diseases. Some of these definitions rely mainly on [1] the number of individuals surviving with the disease, [2] the availability of treatments for the disease and [3] the severity of the disease.

In the United States of America, the Orphan Drug Act of 1983 (PL-97-414) [4] (United States Department of Health and Human Services, Public Health Service, Office of the Assistant Secretary for Health. Report of the National Commission on Orphan Diseases. Rockville, MD: Office of the Assistant Secretary for Health, 1989) [5] Created by Congress, defines a rare disease as any disease or condition which occurs infrequently. Later, the Rare Disease Act of 2002, which was based on the 1983 Orphan Drug Act, specifically defined a rare disease as "those which affect small patient populations, typically populations smaller than 200,000 individuals in the United States," or roughly 1 in 1,500 individuals. Initially, the term "Rare Diseases" was synonymous with "Orphan Diseases" largely because these diseases did not offer a lucrative market for drug companies. However, the attitude of drug companies later changed with the enactment of the Orphan Drug Act that provided financial incentives whereby these companies could develop new drugs to treat rare diseases.

In the country of Japan, a rare disease, according to their laws, is one that affects less than 50,000 individuals or 4/10,000. Whereas in Italy, there is no stated definition of a rare disease, but different

measures has been used by various health institutions to categorize a disease as rare. For example, the Italian National Health Plan varies in its definition from 1 in 20,000 to 1 in 200,000. Countries that comprise the European Union define a disease as rare if it affects less than 1 individual in 2,000.

The National Center for Advancing Translational Sciences (NCATS) estimates that there are about 7,000 rare diseases with a further estimate of 25 million people living in the USA with one of these rare diseases and an estimated 30 million in Europe. This suggests that a disproportionately large number of individuals continue to survive with a rare disease. One of the most difficult conundrums is that well over 6,000 rare diseases present with a wide range of underlying symptoms and disorders that vary from disease to disease and from patient to patient. This is further compounded by the fact that many ubiquitous symptoms can camouflage rare diseases thus leading to a misdiagnosis of the disease, and hence a delay of appropriate treatment, and not surprisingly, the emergence of a further sequelae of psychosocial ills and even morbidity. Of the many challenges diagnosticians face, the paucity of medical and scientific information about these diseases appears to be the most impactful, because it leads to a reduction in timely diagnosis and intervention.

### Challenges

Rare diseases offer extreme challenges to physicians and the general medical community as evidenced by the long delay and missteps associated with diagnoses [4]. Recognizing and treating rare diseases create a further challenge to the medical profession in that 80% of these rare diseases are "genetic in nature, with the inherent problem of limited resources, lack of research, scarce expertise and patients that are relatively few in number and are geographically spread out" (United States Department of Health and Human Services, Public Health Service, Office of the Assistant Secretary for Health. Many rare diseases have accompanying common disorders such as dysphagia and cognitive deficits that require diagnosis and treatment provided by a

speech-language pathologist. Unfortunately, physicians do not encounter rare diseases on a regular basis, and not surprisingly, other health care specialists such as speech-language pathologists, physical therapists and occupational therapists. The “trickle-down” effect here is that in the realm of common diseases, health care specialists receive the referral from the physician to treat the patient, thus intervention occurs timely, but in the case of a rare disease, this timely dissemination of information does not occur. Most speech-language pathologists are familiar with common neurological diseases that cause swallowing and a plethora of speech and language disorders and can readily diagnose and treat most of these deficits.

In the case of a rare disease in which dysphagia is part of the symptomatology, lack of familiarity with the disease may fail to trigger a timely referral for speech intervention. A delay in referral will most likely lead to a much later intervention process. In attempting to unravel these challenges, it is probably a good idea to discuss briefly dysphagia, which will be the center of the discussion of a forthcoming text “Rare disorders that Cause Dysphagia” [6].

## Dysphagia

Swallowing under normal circumstances occurs with relatively little effort, yet it is a complex function. It is a process whereby masticated food, liquid or saliva is forwarded through the mouth and finally to the stomach. Dysphagia is a common medical term used to describe a disorder of or difficulty with this transference of the food to the stomach. Based on the Taber’s Cyclopedic Medical Dictionary, dysphagia is classified into five subcategories [7];

Constricta dysphagia: disorder of swallowing due to narrowing of the pharynx or the esophagus.

Lusoria dysphagia: swallowing disorder that arises from the compression of the esophagus by the right subclavian artery.

Oropharyngeal dysphagia: difficulty transferring food from the mouth to the pharynx and esophagus.

Paralytica dysphagia: swallowing disorders due to paralysis of the muscles mouth, pharynx or esophagus

Spastica dysphagia: swallow disorder that arises from spasms of the pharynx or esophagus.

While dysphagia for the most part may not be a primary medical diagnosis, it is usually a strong symptom of an underlying medical condition whether acquired or congenital. Its clinical signs can range from coughing and or choking during, after or before swallowing, food sticking anywhere along the swallowing tract, regurgitation of food material, painful swallowing, drooling, unexplained weight loss or nutritional deficiencies [7]. There are wide sequelae of physical, emotional and psychosocial consequences that accompany dysphagia. Aspiration pneumonia, malnutrition, increased mortality, prolonged hospitalization, advanced disability, declined quality of life and social isolation are but a few of the consequences of dysphagia [8].

## Prevalence of dysphagia

Dysphagia can result from a myriad of specific disorders and categories of disorders most of which are well known to speech-language pathologists who routinely treat the swallowing problem. There are wide varieties of common neurological diseases associated with dysphagia. It is difficult to be precise in terms of the exact number of cases of dysphagia by disease or condition; but there are many

reports that provide acceptable estimates. It is well documented that dysphagia commonly results from strokes. Some disease states that are commonly associated with dysphagia are neurological, congenital and developmental, obstructions, muscular disorders, rheumatological disorders as well as other nonspecific causes (Table 1).

Dysphagia is a relatively common and increasingly prevalent clinical problem. Estimated reports suggest that close to 10 million individuals are evaluated for some aspect of dysphagia annually in the United States of America. According to Bhattacharyya, approximately 1 in 25 adults in the United States will experience some type of swallowing problem. Furthermore, about 22% of adults in primary care settings and 13.5% of the general population have some form of dysphagia. In addition, of those in the primary care setting who suffer with dysphagia, 80% are most likely to be females and 20% male [9]. In spite of these numerical approximations, actual estimates of the prevalence of dysphagia is problematic for two basic reasons [1] dysphagia can result from a multiplicity of diseases common and rare [2] and it can affect both the young and elderly. Thus, it is not surprising that there are many under-reported incidences of dysphagia. Nevertheless, a plethora of published reports substantiates the prevalence of dysphagia in older individuals. Schindler and Kelly in 2002 reported that persons 65 yrs and over account for two-thirds of individuals with dysphagia [10]. According to data released by the US Census Bureau, in April 2010 [11], the total US population increased by 9.7%, and the number of persons age 65 yrs and older increased by 14.9%. The census projected that for the next 18 yrs, 10,000 more Americans will become seniors each day, a natural phenomenon because of the aging Baby Boomer population. It is expected that by 2030, 1 in 5 US residents will be 65 yrs of age or older.

Many studies have reported the presence of dysphagia in 50% of nursing home patients, with an increased risk of aspiration pneumonia and other complications more so in the elderly than in younger patients [12,13]. However, it must be pointed out that aging does not cause dysphagia, but because the aging process is associated with measurable changes in neuromuscular activities, the risk for dysphagia is maximized in this population [14]. It has long been established that physiological deterioration is a hallmark of aging; however, it is not known how much of this deterioration is due to age and how much too age-related diseases and life-style. For example in normal aging, there is cerebral atrophy, nerve function deterioration and region-dependent decline in muscle mass that may impact swallowing [15]. Humbert and Robbins made an interesting distinction between “normal healthy aging swallow” presbyphagia and an otherwise disordered swallow-dysphagia [16]. Presbyphagia has to do with characteristic changes that occur in the swallow mechanism in older individuals who are healthy [17]. Clinicians are becoming much more aware of these distinctions within the scheme of swallowing and are now able to make appropriate diagnoses. Added to this increased awareness, is the fact that common conditions that precipitate dysphagia are well known to speech-language pathologists. These etiologies are routinely addressed in various textbooks on dysphagia (Table 1).

In recent decades, there has been a burgeoning of medical information that has led to the identification of hitherto unknown diseases that carry sequelae of medical conditions including dysphagia. Given the rarity of these diseases, it is not surprising that many practicing speech-language pathologists are unaware of their existence. There is no question that knowledge of the etiology of dysphagia can inform intervention. Therefore, the purpose of this book is to identify and unpack rare diseases that are contributory to dysphagia in an easy

and readily accessible form for the medical speech-language pathologist.

Neurologic	Congenital /Developmental	Muscular	Rheumatologic	Obstruction	Other
Stroke	Cerebral palsy	Scleroderma		Oral cancer	Chronic obstructive Pulmonary disease
Parkinson's	Learning disabilities	Achalasia	Sjogren's syndrome	Laryngeal cancer	Head and neck cancer
Multiple sclerosis	Cleft lip/palate		Dermatomyositis	Esophageal cancer	Muscle weakness due to aging
Dementia	Polymyotiss			Thrush	
Motor neuron diseases			Systemic lupus erythematosus		
Myasthenia gravis					
Brain tumors					
Huntington's Chorea					
Guillain-Barre syndrome					
Poliomyelitis					
Peripheral neuropathy					
HIV/AIDS					

**Table 1:** Conditions that cause Dysphagia.

## Plummer-Vinson Syndrome

### Definition

Plummer-Vinson syndrome (PVS), also known as Patterson-Brown-Kelly syndrome as well as Sideropenic dysphagia, is listed as a rare disease by the Office of Rare Diseases (ORD) of the National Institutes of Health (NIH). Based on the definition of a rare disease, this means that PVS affects less than 200,000 persons in the United States. PVS falls within the categories of blood and digestive diseases. It is generally defined by its classic presentation of triad disorders. These are dysphagia, iron-deficiency anemia and laryngeal webbing. The main cause of the dysphagia is believed to be associated with the presence of a web in the cervical esophagus. Abnormal motility of the pharynx and the esophagus is also implicated. The swallowing difficulties prominent in PVS are highly correlated with the small thin growths of tissue-webbing-that tend to block the upper esophagus. However according to early researchers it has been found that, while the pathogenesis of PVS is still largely unknown, it is felt that the most probable cause is the iron deficiency [18]. This may be because in iron deficiency the loss of iron-dependent enzymes causes degeneration of tissue, atrophy and the eventual formation of webs that are frequently seen in the upper esophageal region.

### History

Plummer-Vinson syndrome is eponymically linked to two prominent physicians of the Mayo Clinic, Henry Stanley Plummer an internist/endocrinologist and founder member of the Clinic, and Porter Paisley Vinson a surgeon [1]. Plummer identified a number of

patients with a history of iron deficiency anemia, dysphagia and spasm of the upper esophagus in the absence of anatomic stenosis. Later Vinson corroborated Plummer's findings when he reported similar esophageal aberrations in patients with dysphagia.

Another eponym for the disorder is Paterson-Kelly syndrome, named after two British laryngologists, Donald Ross Patterson and Adam Brown-Kelly. These physicians not only independently reported, but were actually the first to describe the characteristic features of the syndrome [19]. The nomenclature for the disorder changes depending on location. In the United States, it is referred to as Plummer-Vinson, whereas in the United Kingdom, it is commonly known as Patterson-Kelly Syndrome [20]. The term "sideropenic dysphagia" is sometimes used for the disorder simply because iron-deficiency or sideropenia is a key factor in the syndrome.

### Etiology

While the cause of PVS remains unclear, many researchers have linked iron and nutritional deficiencies, as well as genetic factors to the root cause. This rare disorder, in many cases of the condition, is associated with cancers of the throat and the esophagus. The pervasive iron-deficiency theory present in PVS has been met with some measure of speculation even though most of the earlier reports of the disorder cited iron deficiency in the pathogenesis of esophageal webs and dysphagia in a majority of predisposed patients. Nevertheless, it is note-worthy, that the improvement in dysphagia in many cases following iron therapy does provide evidence for some association between the iron-deficiency and the dysphagia [21]. Other etiologic factors such as malnutrition, genetic predisposition and autoimmune processes have been proposed. The latter is based on the association

between Plummer-Vinson syndrome and certain autoimmune disorders such as celiac disease-which was the most frequently mentioned associated disease in the case reports published in recent years - thyroid disease and rheumatoid arthritis [19].

### Epidemiology

Currently, there are no reliable data about the incidence and prevalence of PVS as the disorder is now considered to be very rare. However, in the early part of the 20<sup>th</sup> century, the disease was noted to be prevalent in most middle-aged Caucasian women specifically in Northern European countries. Since the syndrome is so rare, only case reports as opposed to “series” of patient reports have been published in recent literature. The rarity of PVS appears to correlate with improvement in nutritional status, availability of health care and the widespread addition of iron and iron-supplemented diets. Although PVS has been more frequently observed in females between 40-70 yrs of age, there have been a few reported incidences of the disorder in children as well as in males [22]. Bakshi in 2015 reported a high occurrence of PVS in India in both male and female [1]. A review of the English language case reports published between 1999 and 2005 revealed that 25 out of 28 cases (89%) were females with a mean age of 47 ranging from 28 yrs of age to 80 [19].

### General clinical presentation

Head and Neck		
Term Identifier	Name	Definition
HP: 0100825	Cheilitis	Inflammation of the lip
HP: 0000160	Narrow mouth	Decreased width of oral aperture
HP: 0000206	Glossitis	Inflammation of the tongue
HP: 0012473	Tongue atrophy	Wasting of the tongue
HP: 0010284	Intra-oral hyperpigmentation	Increased pigmentation focal or generalized of the oral mucosa
Blood and blood- forming tissues		
Term identifier	Name	Definition
HP: 0004840	Hypochromic microcytic anemia	A type of anemia characterized by an abnormally low concentration of hemoglobin in the erythrocytes and lower than normal size of the erythrocytes.
HP: 0001891	Iron deficiency anemia	
Digestive System		
Term identifier	Name	Definition
HP: 0100594	Esophageal web	Thin (2-3 mm) membranes of normal esophageal tissue consisting of mucosa and submucosa that can be congenital or acquired. Congenital webs commonly appear in the middle and inferior third of the esophagus, and

		they are more likely to be circumferential with a central or eccentric orifice. Acquired webs are much more common than congenital webs and typically appear in the cervical area (post-cricoid). Clinical symptoms of this condition are selective (solid more than liquids) dysphagia, thoracic pain, nasopharyngeal reflux, aspiration, perforation and food impaction (the last two are very rare).
HP: 0002015	Dysphagia	Difficulty swallowing

**Table 2:** Plummer-Vinson Human Phenotype Ontology Associations.

Most disease symptoms vary from person to person; consequently, it is not surprising that people with the same disease may not manifest all of the known symptoms of the particular disease. The Human Phenotype Ontology foundry (HPO) provides a standardized vocabulary of phenotypic abnormalities identified in human diseases. Currently, six categories of HPO associations are linked to PVS (Table 2). While most patients will not exhibit all of the symptoms established for PVS, all of them will manifest dysphagia, iron deficiency and an esophageal web. Table 3 provides a distribution of symptoms seen across patient groups.

Clinical Symptoms
<b>Present in all cases of PVS</b>
Decreased serum ferritin
Dysphagia
Esophageal web
Hypochromic microcytic anemia
Iron deficiency anemia
<b>Present in 80-99% cases of PVS</b>
Easy fatigability
Glossitis
Pallor
<b>Present in 5%-29% cases of PVS</b>
Abdominal pain
Cheilitis
Concave nail
Intra-oral hyperpigmentation
Narrow mouth
Tongue atrophy

**Table 3:** Clinical Symptoms in Plummer-Vinson.

Apart from the classic symptoms described above, there are a number of other presenting disorders such as glossitis, angular cheilitis and koilonychia commonly seen in PVS.

**Glossitis:** The National Institutes of Health (NIH) defines Glossitis as a general term for inflammation of the tongue. Of the many causes

of Glossitis, iron deficiency is recognized as a major contributor. Bayraktar suggests that atrophy of the upper GI mucosa caused by iron deficiency leads to the inflammatory nature of the tongue [2]. This condition is often characterized by depapillation of the dorsal aspects of the tongue. In some cases of Glossitis, the tongue can become so swollen that chewing, swallowing and speaking may be difficult.

**Angular cheilitis:** This inflammatory condition affects one or both corners (angles) of the mouth. Typically, erythema (redness), cracked lips, bleeding and ulcerated corners of the lips characterizes the condition. These all tend to cause pain. Angular cheilitis can result from many conditions such as fungus, bacteria, malnutrition. In the case of malnutrition, angular cheilitis is most often caused by the iron deficiency anemia that is linked to PVS.

**Koilonychias:** A condition in which the fingernails present as thin with lifted outer edges, resembling the shape of a spoon, hence also known as “spoon nails.” This condition is present in cases of iron deficiency or poor absorption of iron as in PVS.

## Diagnosis

As has been previously discussed, the diagnostic criteria for PVS feature dysphagia, esophageal webs and iron deficiency anemia as the three key players [23].

Of great interest is the interrelatedness of these three different symptoms that apparently must exist together in order to make the case for PVS. As it turns out, these three different symptoms may be more connected than originally thought. However, the disorder is not limited to these three conditions. It seems as though other shadowy symptoms manifested in PVS are all linked to the big triad (Figure 1).

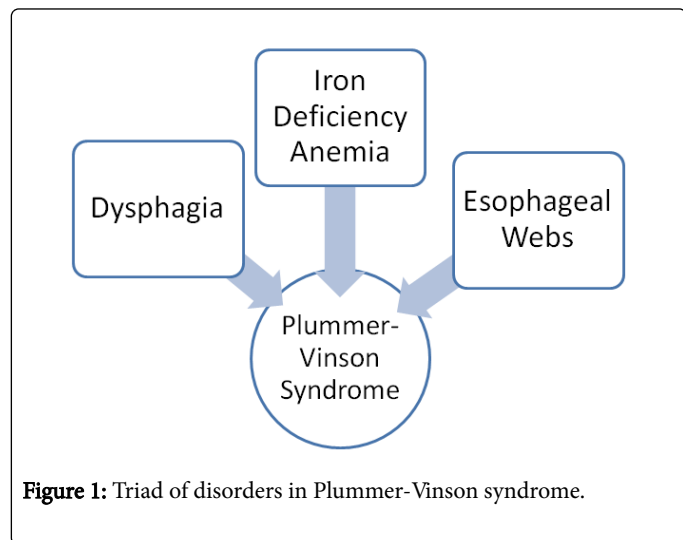


Figure 1: Triad of disorders in Plummer-Vinson syndrome.

For example, it is postulated that the esophageal webs prominent in PVS may be due to long term iron deficiency anemia consequently, the fatigue, weakness and pallor that are present in individuals with PVS are all related to the classic anemia that can be routed back to iron deficiency which eventually leads to the formation of these esophageal webs. In turn, the esophageal webs, which are thin mucosal folds that protrude into the lumen of the proximal esophagus, contribute to the classic dysphagia.

## Dysphagia

Dysphagia is one of the main clinical features of Plummer-Vinson syndrome, but because there are many other causes of dysphagia, a differential diagnosis has to be made. The post-cricoid dysphagia seen in PVS is also present in motility disorders such as achalasia, as well as other disorders such as scleroderma, diabetes mellitus, gastroesophageal reflux disease (GERD) and most commonly in neurological and skeletal muscular disorders, therefore these diseases must first be ruled out.

The nature of the dysphagia seen in individuals with PVS varies. In some patients, it has been described as painless occurrence ranging from intermittent to progressive. Classically, most patients experience choking or fear of choking on specific food textures. Usually these dysphagic symptoms tend to occur more so with solid textures. Typical video fluoroscopic evaluation of the swallow may reveal delayed emptying of material in the hypo pharyngeal region. Some patients may also experience a sudden onset of the dysphagia, but usually it tends to be insidious. It is not uncommon for patients to experience weight loss. This weight loss may be attributed to either avoidance of eating for fear of choking or compensatory changes in diet. The presence of the esophageal webs has reportedly caused choking spells and aspiration on solid foods in some patients, although others have experienced regurgitation on both solids and liquids [23]. Patients usually identify the neck area or above the suprasternal notch as the site of the obstruction, when trying to explain the choking they experience.

Endoscopic studies have linked the manifestation of dysphagia to the size of the luminal diameter in the region of the esophageal web. Two independent studies suggest that in most patients with PVS, dysphagia should be graded in terms of the size of the luminal diameter, specifically, if the lumen is less than 12 mm. These studies described four levels of severity. Grade I in which the patient exhibited difficulty swallowing solids. This was found in 52% of the cases. Grade II in which the patients were only able to swallow semi-solid foods. This was present in 36% of the cases. On the other hand, in grade III dysphagia, 8% of the patients could only swallow liquids, and grade IV, 4% had an inability to tolerate liquids. These two latter grading's appeared to be less common [24]. Most cases of PVS confirm difficulty swallowing solid foods and in all cases, the dysphagia was associated with the presence of esophageal webs.

## Esophageal webs

The barium swallow X-ray is used to detect esophageal webs but the best way for demonstration is via video fluoroscopy [25]. Esophageal webs are also detectable by upper gastrointestinal endoscopy. Visually, webs appear smooth, thin, and gray. They typically occur in the proximal part of the esophagus and may be visually undetected or accidentally ruptured unless the endoscope is introduced under direct visualization [26]. Esophageal webs can cause some patients to experience a feeling of chest pain. Typically, these patients may not associate the pain with dysphagia initially, as they may be continuing to consume food in their usual manner and mistaking the source of the pain to be unrelated to swallowing. Webs are often located in the post cricoid region of the esophagus, which is a muscular tube that connects the pharynx with the stomach. It courses down posterior to the trachea and heart and just anterior to the spinal column. Because of its lengthy course, pain in the esophagus can often be mistaken for heart-burn or indigestion.

Besides dysphagia and the presence of esophageal webs, another equally important element in the diagnosis of PVS, is the presence of iron deficiency anemia. In fact, this is frequently the major discovery before dysphagia is identified.

### Iron-deficiency anemia

The presence of Iron deficiency anemia (IDA) is part of PVS diagnosis. It is believed that IDA leads to rapid loss of iron-dependent enzymes due to its high cell turnover. Loss of these enzymes causes mucosal degenerations, atrophic changes and web formation which have been shown to lead to dysphagia [18]. This notion is supported by reports that the dysphagia in PVS tends to be relieved when the patient is treated with iron supplements [27,28].

IDA is associated with other nutritional deficiencies in riboflavin, thiamine and pyridoxine. Other complications link to dysphagia that result from IDA are mucosal changes in the oropharynx such as stomatitis (inflammation of the mouth and lips), atrophy of the lingual epithelium, depapillation and angular cheilosis. Although frequent occurrences of dysphagia, hypopharyngeal and oral cancers have been reported in some cases of IDA, these reports are still inconclusive [24].

As mentioned earlier in this chapter, in diagnosing PVS from other causes of dysphagia, it is important to determine the cause of the dysphagia. The clinician has to bear in mind that causes of dysphagia other than PVS are common, therefore, these sources must be ruled out. Goel et al. identified a number of benign causes that result in esophageal dysphagia [24]. Some of these are zenker's diverticulum, esophageal strictures secondary to corrosive injury, surgical anastomosis of the esophagus specifically after surgery to repair a tracheoesophageal fistula. Other esophageal motility disorders such as scleroderma or achalasia cardia are some causes of dysphagia in PVS.

### Management of dysphagia

In general, dysphagia in PVS has an excellent recovery outcome even though it is a precancerous condition. Patients with PVS are at risk for squamous cell carcinoma of the hypopharynx or upper esophageal region. The good news in treatment for many patients with PVS, is that with treatment for IDA, the dysphagia as well as the esophageal webs tend to resolve over time [27,28].

The usual symptoms of dysphagia secondary to PVS described by most patients are, difficulty swallowing solids, sensation of discomfort, tightness or fullness typically described either in the neck or chest region. Management of this type dysphagia is never the sole responsibility of the speech-language pathologist (SLP) since the primary concern is to improve the IDA. The SLP however, still plays a vital role in terms of recommending strategies for safe swallowing such as posturing techniques if indicated, adjusting bolus viscosity and size, having the patient alternate solid foods with liquids and other compensatory strategies.

Management of the IDA in PVS has been closely linked to the successful improvement of dysphagia in the patient. Different modes of management have been described in the literature. One approach is to treat with iron supplements. Iron supplements have proven to improve the swallowing deficits experienced by most patients. IDA is caused by the lack of iron deficiency anemia; consequently, it develops when there is not enough iron in the body for hemoglobin synthesis. The lack of iron causes myasthenic changes to occur in muscles that are involved in the swallowing mechanism. This muscle weakness causes

atrophy of the esophageal mucosa and leads to the formation of webs. Thus, it makes sense to treat the iron deficiency in an effort to reverse the process that caused the web to occur in the first place and simultaneously eliminate or reduce the dysphagia. Not all webs respond to the iron therapy denser webs may result in more severe esophageal dysphagia and may require a more aggressive management such as the use of endoscopic dilatation.

### Summary

PVS is a rare iron-deficiency anemia that is associated with esophageal webs and dysphagia usually in women. Typically, patients complain of choking and or fear of swallowing. Even though the dysphagia is usually painless, some patients have complained ofodynophagia. Symptoms secondary to the iron deficiency such as muscle weakness tend to dominate the clinical presentation of PVS. While the etiopathogenesis of PVS is still unknown, iron deficiency anemia is paramount in the discovery of the disorder. PVS in most cases can be effectively managed with iron supplements and endoscopic dilatation. However, since patients with PVS are at an increased risk for squamous cell esophageal carcinoma the medical team follows them closely.

### References

1. Bakshi SS (2015) Plummer-Vinson Syndrome. Mayo Clinic Proceedings 91: 404.
2. Bayraktar UD, Bayraktar S (2010) Treatment of iron deficiency anemia associated with gastrointestinal tract diseases. World J Gastroenterol 16: 2720-2725.
3. Bhattacharyya N (2014) The prevalence of dysphagia among adults in the United States. Otolaryngol Head Neck Surg 151: 765-769.
4. Svenstrup J, Jorgensen, Winther (2015) Rare disease diagnosis: A review of research, social media and large-scale data mining. Rare Dis 3: e1083145.
5. United State Department of Health and Human Services Public Health Service (1989) Report of the National Commission on Orphan Diseases.
6. Cox VO (in Press). Rare diseases that cause Dysphagia. Plural Publishing.
7. Groher M , Crary M (2016) Dysphagia: Clinical Management in Adults and Children (2nd ed.). St. Louis Missouri.
8. Eyigor S (2013) Dysphagia in Rheumatological Disorders. World J of Rheumatol 3: 45-50.
9. Wilkins T, Gilles RA, Thomas AM, Wagner PJ (2007) The prevalence of Dysphagia in primary care patients: a HamesNet Research Network study. J Am Board Fam Med 20: 144-150.
10. Schindler JS, Kelly JH (2002) Swallowing disorders in the elderly. Laryngoscope 112: 589-602.
11. United States Census Bureau (2010) Census.
12. Marik PE, Kaplan D (2003) Aspiration pneumonia and dysphagia in the elderly. Chest 124: 328-336.
13. Rofes L, Arreola V, Romea M, Palomere E, Almirall J, et al. (2010) Pathophysiology of oropharyngeal dysphagia in the frail elderly. Neurogastroenterol Motil 22: 851-858.
14. Carucci LR , Turner MA (2015) Dysphagia revisited: Common and unusual causes. Radiographics. 35: 105-122.
15. Masoro EJ (1987) Biology of aging. Current state of knowledge. Arch Intern Med 117: 166-169.
16. Humbert IA, Robbins J (2008) Dysphagia in the elderly. Phys Med Rehabil Clin N Am 19: 853-866.
17. Robbins J, Hamilton JW, Lof GL, Kempster GB (1992) Oropharyngeal swallowing in normal adults of different ages. Gastroenterology 103: 823-829.

18. Okamura H, Tsutsumi S, Inaki S, Mori T (1988) Esophageal web in Plummer-Vinson syndrome. *Laryngoscope* 98: 994-998.
19. Gottfried N (2006) Plummer-Vinson syndrome. *Orphanet J of rare Dis* 1: 36.
20. Slater S (1991) The Brown Kelly-Patterson or Plummer-Vinson Syndrome: an old score finally settled. *Journal of the Royal College of Physicians London* 3: 257-258.
21. Chisholm M (1974) The association between webs, iron and post-cricoid carcinoma. *Postgrad Med J* 50: 215-219.
22. Mansell NJ, Jani P, Bailey CM (1999) Plummer-Vinson syndrome – a rare presentation in a child. *J Laryngol Otol* 113: 475-476.
23. Skolka M, Pauly-Hubbard H (2017) Rapidly Progressive Plummer Vinson Syndrome. *J Rare Disord Diagn Ther* 3: 6.
24. Goel A, Lakshmi CP, Bakshi SS, Sony N, Koshy S (2016) Single-center prospective study of Plummer-Vinson syndrome. *Diseases of the Esophagus* 29: 837-841.
25. Chung S, Roberts-Thomson IC (1999) Gastrointestinal: upper Oesophageal web. *J Gastroenterol Hepatol* 14: 611.
26. Hoffmann RM, Jaffe PE (1995) Plummer-Vinson syndrome A case report and literature review. *Arch Intern Med* 155: 2008-2011.
27. Sugiura Y, Nakagawa M, Hashizuma T, Nemoto E, Kaseda S, et al. (2015) Iron Supplementation Improved Dysphagia Related to Plummer-Vinson syndrome. *Keio J Med* 64: 48-50.
28. Tahara T, Shibata T, Okubo M, Yoshioka D, Ishizuka T, et al. (2014) A case of Plummer-Vinson syndrome showing rapid improvement of Dysphagia and esophageal web after two weeks of iron therapy. *Case Rep Gastroenterol* 8: 211-215.