

TITLE: Paradoxical Vocal Fold Motion

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INTRODUCTION

Paradoxical vocal fold motion (PVFM) is an involuntary functional disorder of the larynx characterized by intermittent adduction of more than 50% of the vocal folds during inspiration. It is often misdiagnosed as asthma and resulted in unnecessary drug use and hospitalization. This paper serves to highlight the characteristics of PVFM and describes its diagnosis and management.

NOMENCLATURE

Since the description of PVFM as "hysteric croup" by Dunglison in 1842, there have been numerous terminologies used to describe this condition, including episodic laryngeal dyskinesia, Munchausen's stridor, pseudoasthma, functional upper airway obstruction, spasmodic croup, emotional laryngeal wheezing, irritable larynx syndrome, vocal cord dysfunction, etc. More recently, PVFM or paradoxical vocal cord movement has been used more frequently in the literature to give a more specific description of this condition.

EPIDEMIOLOGY

Various studies report the incidence of PVFM ranges from 3-10%. It affects mainly children and young adults with average age at diagnosis of 14.5 years in children and 33 years in adults. There is a reported 2:1 female predominance, with about 70-98% patients being female Caucasian. O'Connell et al reported that 20% of 164 female patients who underwent laryngoscopy for any reason were found to have PVFM.

PATHOPHYSIOLOGY

The exact etiology of PVFM remains unclear. Maschka et al proposed that PVFM might represent a spectrum of underlying diseases that manifest as a single clinical entity. In his classification system, Maschka divided the etiologies into organic and non-organic causes. Organic etiologies included neurologic diseases and GERD. Nonorganic causes were classified as either conversion disorders or malingering and were believed to represent the vast majority of cases.

Ayers and Gabbott suggested that PVFM may be caused by laryngeal hyper-responsiveness initiated by an initial inflammatory insult that resulted in altered autonomic balance. Subsequent stimuli from irritants and psychogenic factors induce local presynaptic reflexes leading to vocal fold adduction and glottic closure.

ASSOCIATED CONDITIONS

Several conditions have been closely linked to PVFM. The role of gastroesophageal reflux disease (GERD) has been elucidated in many studies. Powell et al found evidence of arytenoid edema or pachydermia on laryngoscopy in 21 of 22 pediatric PVFM patients. Loughlin et al reported abnormal 24-hour pH probe study in 10 of 12 PVFM adult patients. In a recent study by Cukier-Blaj et al, about 70 % of PVFM patients were found to have both abnormal reflux symptom index and laryngopharyngeal sensitivity scores consistent with laryngopharyngeal reflux disease.

Initially considered separate and distinct disorders, chronic cough and PVFM exhibit overlap in symptomatology, such as cough and dysphonia, and overlap in disease associations, namely asthma, GERD, and rhinosinusitis. Ryan and Gibson found evidence of PVFM in approximately 56% of individuals with chronic cough using hypertonic saline challenge and fiberoptic laryngoscopy.

Psychogenic factors have been considered to play a major role in PVFM since the original description of “hysteric croup”. Newman et al reported a previous psychiatric diagnosis in 73% of patient with PVCM. Powell et al noted that 55% of 20 adolescent females with PVCM had severe social stresses such as competitive sports. Prior history of sexual abuse has also been reported to play a part in the development of PVCM.

PVFM is often misdiagnosed as asthma although there might be close associations between the two conditions. Newman et al found that 56% of the 95 PVFM patients had coexistent asthma. A study by the National Jewish center in 1994 showed that 30% of patients diagnosed with refractory asthma had co-existing PVFM.

DIAGNOSIS

History and clinical features:

Patients with PVFM often present with chronic refractory cough and frequent episodic dyspnea attacks. Cough is commonly exacerbated by irritants such as perfume, smoke, and strong odor. These patients may show signs of asthma but are not responsive to the regimen of

bronchodilators and steroids. They may also present with hoarseness, dysphagia, and previous diagnosis of GERD.

Physical examination:

Physical examination may be normal when patients are not experiencing an acute attack. Wheezing or stridor may be heard loudest over the larynx. Direct visualization of vocal cords by laryngoscopy is the gold standard for diagnosis of PVFM. Classic finding on laryngoscopy is glottic closure of anterior vocal folds during inspiration with a small posterior chinking. If the patient is asymptomatic at the time of laryngoscopic examination, having the patient pant, breathe deeply and phonate may sometimes elicit symptoms. Laryngoscopy also helps identify other laryngeal pathologies that may co-exist with PVFM, including unilateral vocal cord paralysis, subglottic stenosis, nodules, laryngomalacia, and sulcus vocalis as described in Patel's study.

Pulmonary function test (PFT):

PFT with flow volume loops are often used as an adjunct to support the diagnosis of PVFM. In symptomatic patients, flow volume loops may show a flattening of the inspiratory limb secondary to decreased inspiratory flow rate. However, 70-80% of PVFM patients were found to have normal PFT when asymptomatic.

Methacholine challenge:

Methacholine is a cholinergic agent that can induce bronchoconstriction. It is often used after PFT as a diagnostic test in asthma patients. Although it has been shown to induce acute attacks in PVFM patients, its sensitivity is low. Perkins et al showed that only 2 of 8 PVFM patients previously diagnosed by laryngoscopy developed PVFM on methacholine challenge.

MANAGEMENT

Acute Management

The cause of the PVFM must first be elicited. Organic disease states such as brainstem compression, encephalopathy, stroke, myasthenia gravis, GERD should be treated appropriately. A history of previous exposure to irritants and sinusitis should also be obtained.

Heliox therapy may be used for immediate relief of respiratory distress in PVFM. Heliox is a gaseous mixture of oxygen and helium often found in ratios of 20/80 and 30/70 respectively. Because this mixture is less dense than air, inhalation reduces turbulence in the airway and eliminates respiratory noise. It provides short-term relief of dyspnea in patients with both PVFM and other forms of upper airway obstructive disease.

Other acute measures include IPPV (intermittent positive pressure ventilation) and CPAP (continuous positive airway pressure) which widen the rima glottidis and also reduce turbulence. Panting brings about acute relief by physiologically increasing the glottic aperture. Benzodiazepines and reassurance both reduce anxiety and therefore have been shown effective in terminating acute symptoms of PVFM.

Long-term Management

This requires a multidisciplinary approach involving speech therapy, psychiatric support, and physician education regarding the syndrome. Speech therapy is regarded as the cornerstone of treatment for PVFM. The primary role of speech therapy is to teach patients with PVFM how to control the laryngeal area and maintain an adequately open airway during respiration. Therapy with early recognition of symptoms allows relaxation of neck, shoulder and chest muscles and promotes normal laryngeal breathing. Breathing exercise with regular steady rhythm can help patients learn to avoid inspiratory bursts that may trigger PVFM.

Psychotherapy should be initiated in patients if there has been insufficient improvement with speech therapy alone or if significant psychological components are present in history. Psychotherapy allows the patient to explore for potential causes of the disorder and trains the patient with relaxation techniques. Educating the patient about the condition is another method useful for reducing stress. Biofeedback may be used in conjunction with psychotherapy for treating patients with PVFM.

Inspiratory muscle weakness has been linked with rapid, shallow breathing during heavy exercise and exercise-induced PVFM. Inspiratory muscle training aims at increasing strength and decreasing respiratory effort that may lead to a decreased sense of panic and struggle. The increased muscle strength may also enable patients to generate greater pressure differentials to overcome laryngeal obstruction.

The long-term outcome of patients with PVCM is unknown as most of the literature consists of case reports and retrospective studies. Short-term response to speech therapy and psychotherapy showed good response with improved functioning and fewer symptoms after intervention.

CONCLUSIONS

Symptoms of PVFM are non-specific and often misdiagnosed as asthma. PVFM is closely related to GERD, asthma, chronic cough, and psychiatric disorders. A detailed history and laryngoscopic examination are paramount in establishing the diagnosis of PVFM during symptomatic attacks. Management of PVFM is primarily medical with speech therapy and psychiatric evaluation as indicated. A prospective study is needed to evaluate the long-term outcomes of PVFM.

DISCUSSANT: Michael Underbrink, MD, MBA

Basically it's an unfortunate disease with an unclear etiology.

It's difficult to diagnose and typically we see this in the young adult and pediatric patients while they're experiencing some degree of a new stressor. It's typically exercise induced in teenagers, and in older adults it may be irritant induced. We try to get them to reproduce symptoms with that trigger, and in about one third of them it's nice when we can confirm those symptoms and signs with laryngoscopy.

I think it's worthwhile to examine these patients together with the speech pathologist. We teach relaxation breathing techniques and have the patient focus on expiratory breathing. If you have him inhale through the nose and exhale through the mouth you can break a laryngospasm very easily by remaining calm as you talk them through this.

I haven't had a case of this in which I had to use heliox therapy and I hope I never do. Medical treatment and followup is the cornerstone of treatment.

Pulmonologists are quite aware of this problem and they are pretty quick to send a consult your way if it is severe and the patients are non-responsive to treatment for asthma. Use of sedatives or muscle relaxants is not advisable because of their possible respiratory suppressive effect. A trial of heliox therapy should precede intubation if available.

Some people uniformly treat these patients with anti-reflux medications but I don't consider it necessary unless they have signs and symptoms of Laryngopharyngeal Reflux Disease (LPRD).

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