Vocal Cord Dysfunction in Children and Adolescents

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Vocal cord dysfunction (VCD) is a nonorganic disorder of the larynx that involves unintentional paradoxical adduction of the vocal cords while breathing. The resultant symptoms can include dyspnea, chest tightness, cough, throat tightness, wheezing, or voice change. Most patients with VCD are female, and among adolescents and children, VCD tends to be triggered by exercise and is typically confused with exercise-induced asthma. Both gastroesophageal reflux disease (GERD) and psychiatric illness have been reported as having strong associations with VCD, although, to date, there is no evidence that either causes VCD. VCD often coexists with asthma, and should be suspected in any patient in whom asthma treatment fails. Confirming the diagnosis involves direct visualization of abnormal vocal cord motion, and this usually only occurs during symptoms. Adolescent athletes often require free running exercise challenge to reproduce their symptoms and confirm abnormal vocal cord motion laryngoscopically. The primary treatment for VCD involves a combination of patient education and speech therapy, and, in most cases, patients may resume their activities without significant limitation.

Introduction

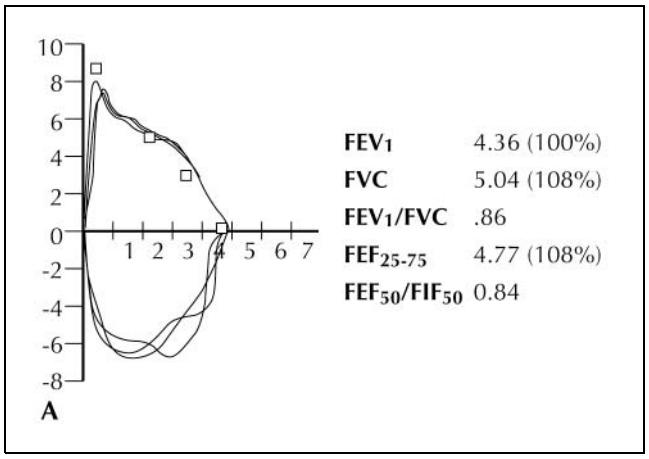
Vocal cord dysfunction (VCD) was first defined in 1983 by Christopher *et al.* [1•] in a report describing four adult patients referred with "refractory asthma" who were instead confirmed by larynogoscopy to have inspiratory vocal cord adduction. Previous reports used names such as "factitious asthma" [2] and "Munchausen's stridor" [3] to describe the same syndrome. Recently, the nomenclature has remained confusing, because some clinicians and authors refer to VCD as "paradoxical vocal-fold motion" [4] or "functional stridor" [5]. VCD is a nonorganic disorder, although symptoms are involuntary and often impair the patient's quality of life without providing any obvious secondary gain. VCD is also now recognized to involve adduction of the vocal cords during either inspiration, both inspiration and expiration, or, on rare occasion, during expiration only.

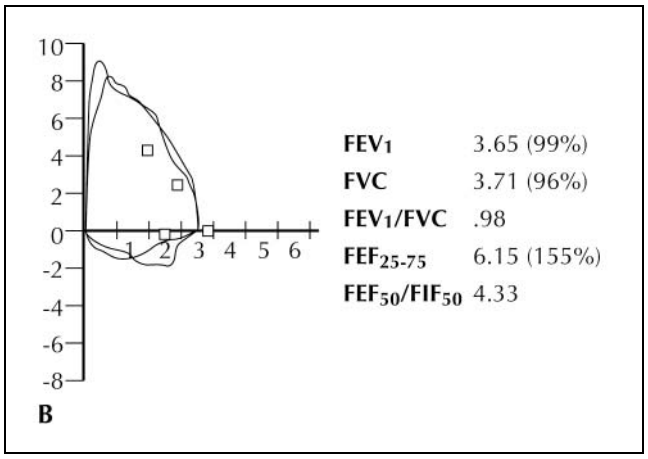
Vocal cord dysfunction commonly masquerades as asthma. The National Heart, Lung, and Blood Institute (NHLBI) Guidelines for the Diagnosis and Management of Asthma [6] specifically recommend considering the diagnosis of VCD in patients who fail to respond as expected to asthma therapy. VCD affects both children (primarily adolescents) and adults, although this review focuses on the pediatric-age group. Topics covered include the spectrum of the clinical presentation of VCD; the utility of diagnostic testing (such as spirometry, laryngoscopy, and provocative challenges); the importance of comorbidities, such as gastroesophageal reflux disease (GERD) and psychiatric disturbance; and therapeutic options for VCD.

Spectrum of Presentation

Vocal cord dysfunction symptoms are typically episodic, and include some combination of dyspnea, wheezing, cough, chest tightness, throat tightness, stridor, and voice change [7•]. These symptoms are often dramatic, resulting in escalating doses of asthma medications in an attempt to prevent recurrences. Both the onset and resolution of VCD symptoms are typically abrupt. There is a spectrum of severity of VCD, and although it is not a life-threatening disorder, many patients with VCD have required emergency department visits and/or hospitalizations. On rare occasion, there is a history of intubation or tracheostomy [4] for VCD symptoms. However, some VCD patients have not suffered a significant impairment in their quality of life or an excessive need for medical treatment.

Most VCD patients are female, and, within the pediatric age group, VCD tends to be associated with exercise and confused with exercise-induced asthma. The time course of symptoms can help discriminate VCD from exercise-induced asthma. A typical exercise-induced asthmatic response requires 5 to 8 minutes of intense exercise and persists for approximately 30 minutes untreated [8]. In contrast, exercise-induced VCD can occur within a minute or less of intense exercise, and symptoms often resolve within 2 to 5 minutes of stopping the activity. There is no sport-specificity with VCD. It has been reported in athletes participating in soccer, basketball, baseball, tae kwon do, figure skating, swimming, and boxing [9].





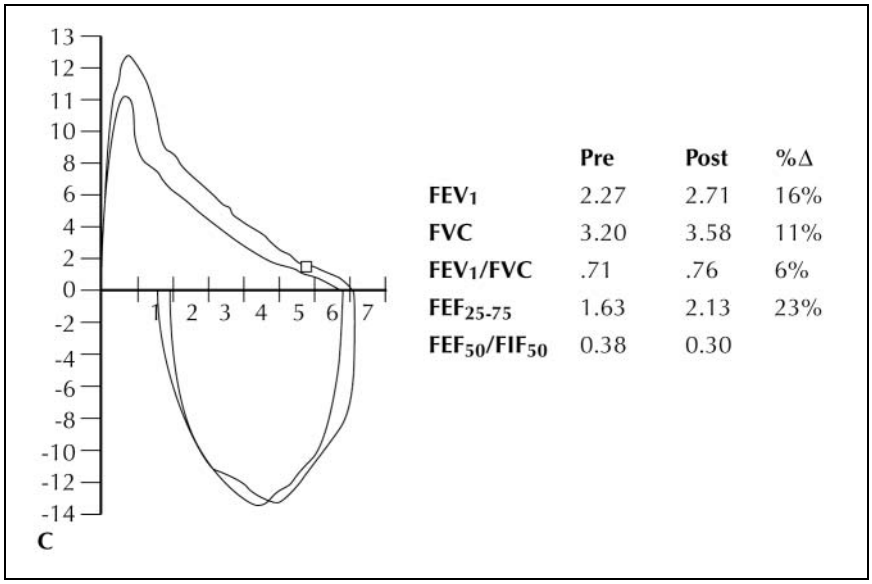


Figure 1. A, Normal spirometry; **B,** extrathoracic airflow obstruction in vocal cord dysfunction; **C,** reversible intrathoracic airflow obstruction in asthma.

Landwehr *et al.* [10] reported a series of seven adolescents (six female) with VCD mimicking exercise-induced bronchospasm. All subjects had been referred for evaluation of asthma aggravated by exercise, and had been treated unsuccessfully with bronchodilators and either inhaled or oral corticosteroids. Symptoms reported included throat tightness, dyspnea, wheezing, chest tightness, cough, and voice change. Each subject underwent exercise challenge and was confirmed laryngoscopically to have paradoxical vocal cord adduction during symptoms. One also had exercise-induced bronchospasm. All of the subjects evaluated were "straight-A" students.

McFadden and Zawadski [11] reported a series of seven elite athletes with VCD that masqueraded as exercise-induced asthma, five of whom were 15 to 18 years old at the time of their presentation. These five all had entered a higher level of competition just prior to the onset of symptoms. The symptoms reported included dyspnea, choking, cough, and wheezing. Episodes of symptoms occurred exclusively with exercise, were abrupt in onset, and either limited their performance or caused them to stop the exercise. Asthma medication neither prevented nor hastened resolution of symptoms. Provocations with exercise, inhaled methacholine, or hyperventilation of cold, dry air were carried out selectively. In two subjects, symptoms could only be provoked by active engagement in their sport.

Several other reports describe exercise-induced VCD in adolescents [12,13] and a recent report sponsored by the United States Olympic Committee [14] describes an observation of inspiratory stridor in 18 of 174 (10.3%) female elite athletes undergoing intense exercise testing. Athletes competing outdoors were found to have a higher prevalence of exercise-induced stridor than those competing indoors. It is not clear if these elite athletes were experiencing VCD, although the presence of stridor suggests that they were experiencing some degree of extrathoracic airflow obstruction.

Not all pediatric patients with VCD present with exercise-induced symptoms. Powell *et al.* [13] reported 22 subjects (18 female), ages 10 to 18, with VCD. Thirteen of these were nonathletes without exertion symptoms, and the authors propose "social stresses" as an associated factor. Pediatric VCD also has been reported in conjunction

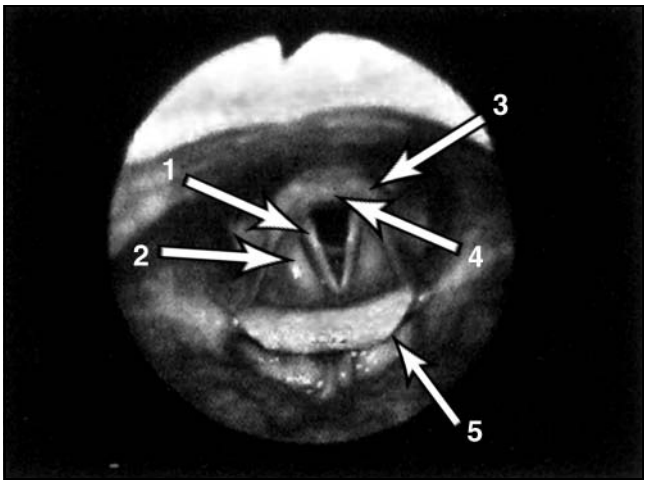


Figure 2. Relevant laryngoscopic findings in a normal larynx: *1*, vocal cords; *2*, false vocal cords; *3*, arytenoids; *4*, interarytenoid space; *5*, epiglottis.

with cystic fibrosis [14] and in infancy [15]. In clinical practice, asthma specialists and primary care physicians are likely to encounter a spectrum of clinical presentations of VCD. Although the prevalence of VCD in the general pediatric population is not known, it is very possible that the cases reported in the literature overrepresent the more severe end of the clinical spectrum, and that the diagnosis is still grossly underrecognized in clinical practice.

Diagnostic Testing

Spirometry

Spirometry is an essential component of the evaluation of any patient suspected of having either VCD or asthma. VCD patients typically have normal spirometry and flow volume loops (Fig. 1A) when asymptomatic, and findings of extrathoracic obstruction during VCD symptoms (Fig. 1B). Airflow obstruction due to VCD usually results in difficulty inspiring, resulting in an increase in the ratio of forced expiratory to inspiratory flow at 50% of vital capacity (FEF_{50}/FIF_{50}; normal value <1.0); truncation of the inspiratory portion of the flow volume loop; and no change in either the forced expiratory volume in 1 second (FEV_1) or forced vital capacity (FVC). When VCD involves vocal cord adduction during expiration, there can be a normal FEF_{50}/FIF_{50} ratio, truncation of both the inspiratory and expiratory portions of the flow volume loop, and a symmetric reduction in both the FEV_1 and FVC. The FEV_1/FVC ratio remains normal. Symptomatic asthma (Fig. 1C) typically results in a decrease in the FEV_1 without a change in FVC, resulting in a decrease in the FEV_1/FVC ratio. The FEF_{50}/FIF_{50} ratio in asthma is typically normal (<1.0), and the flow volume loop exhibits a normal inspiratory portion, with a reduction in the slope of the expiratory curve. These changes improve after treatment with a bronchodilator (see Fig. 1C).

Laryngoscopy

Once VCD is suspected, confirming the diagnosis requires direct laryngoscopic visualization of paradoxical vocal cord motion. This is sometimes possible during a baseline laryngoscopy in the absence of symptoms. More often, however, symptoms must be provoked to observe the abnormal cord motion. The endoscopist must be careful not to induce gagging [16]. Characteristic VCD abnormalities include adduction of the vocal cords and arytenoids while breathing, either at rest or during forced inspiratory or expiratory maneuvers. In severe asthma with chronic intrathoracic obstruction, there is often significant expiratory vocal cord adduction that is thought to serve as a compensatory reflex to maintain hyperinflated lungs. Late expiratory adduction of the vocal cords also occurs in normal and asthmatic subjects without airflow obstruction and, therefore, should not be used as the sole criterion for confirming VCD [17]. Figure 2 illustrates the relevant anatomic structures to examine during laryngoscopy.

Symptom provocation

Symptom provocation is usually necessary to confirm the diagnosis of VCD. Spirometry and laryngoscopy should be performed prior to provocation and should be repeated once symptoms are elicited. The provocative tests that are most often useful are methacholine challenge and exercise challenge. Methacholine challenge testing has an excellent negative predictive value for the diagnosis of clinical asthma. Therefore, a negative methacholine challenge in a patient with chronic dyspnea, chest tightness, wheezing, and/or cough essentially rules out the diagnosis of asthma and should raise suspicion for alternative diagnoses, including VCD. However, there are several issues that are important to consider when considering methacholine challenge in a child or adolescent with possible VCD. First, methacholine challenge is not nearly as sensitive for identifying patients with isolated exercise-induced bronchospasm (without other features of clinical asthma) [18]. Methacholine also often induces VCD symptoms that might result in a false-positive challenge result (a 20% fall in FEV_1 in the absence of bronchospasm). This can occur by either of two mechanisms. The first involves expiratory paradoxical cord adduction causing airflow obstruction during expiration. The second is more common and involves inspiratory vocal cord adduction causing a reduced volume of inspired air and a resultant restrictive spirometry abnormality in the absence of expiratory obstruction. This should be suspected when the FEV_1 and the FVC drop symmetrically during the provocation. Careful coaching by an astute pulmonary function technician can help minimize this phenomenon.

Exercise challenges are often more successful than methacholine in provoking VCD in patients reporting exercise as a trigger of their symptoms. Standardized treadmill or bicycle ergometer protocols exist, although either free-running or another unstandardized form of

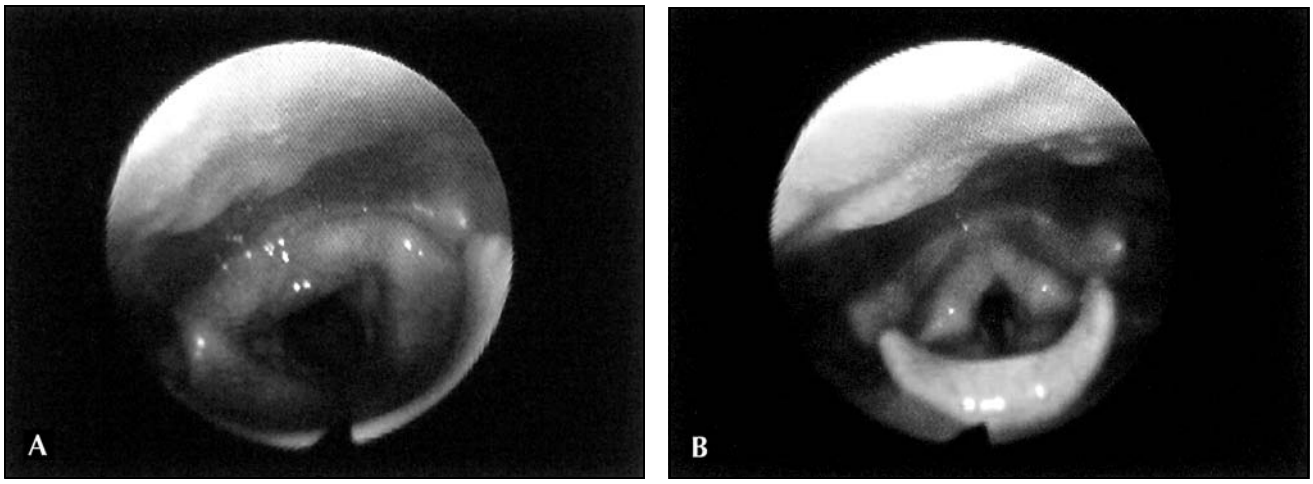


Figure 3. Paradoxical inspiratory adduction of the vocal cords and arytenoids induced by exercise in a 15-year-old female soccer player. After a brief, 5-minute warm-up (jogging), symptoms were provoked by a series of five 20-meter sprints on the sidewalk outside of the clinic. Symptoms and laryngoscopic findings resolved without treatment within 5 minutes. **A,** Resting breathing (inspiration); **B,** during exercise-induced dyspnea, throat tightness (inspiration).

exercise is more likely to reproduce VCD symptoms. As with methacholine challenges, misleading changes in spirometry can occur during VCD symptoms elicited by exercise challenge. Figure 3 illustrates laryngoscopic findings at rest and during exercise-induced VCD symptoms in a 15-year-old soccer player.

Gastroesophageal Reflux and Vocal Cord Dysfunction

Gastroesophageal reflux disease is a very common condition that has a strong association with asthma. GERD also might contribute to the failure of asthma symptoms to respond to treatment [6]. Overnight esophageal pH studies have demonstrated that GERD, in association with asthma, frequently occurs without symptoms of heartburn. GERD is also an increasingly recognized associated factor in both adults and children with VCD. Unfortunately, because nonrespiratory GERD symptoms are also frequently absent in VCD patients, and there have been no controlled studies documenting the presence of abnormal esophageal or laryngeal acid in VCD patients, the association between VCD and GERD is somewhat controversial. However, chronic laryngeal acid exposure causes predictable anatomic changes in the larynx, including arytenoid edema, interarytenoid edema, and interarytenoid pachyderma [19•]. Powell *et al.* [13] identified at least one of these changes in 21 of 22 adolescent VCD subjects. An uncontrolled retrospective case series [4] reported a GERD diagnosis in 8 of 10 VCD patients (one of two under age 18 had GERD). There is also a case report of an infant presenting with stridor, who was found to have GERD and VCD [15]. Additional controlled studies using overnight esophageal pH monitoring are needed to better define the relationship between VCD and GERD.

Psychiatric Issues

Many clinicians associate VCD with significant psychiatric impairment. VCD has been proposed to be a conversion disorder [1•]; however, the evidence for this is inconsistent [17]. Extensive psychiatric evaluations were carried out in four of the five adult subjects in the original VCD case series reported from the National Jewish Medical and Research Center in 1983 [1•]. Prior to their VCD diagnoses, each of these subjects had a history suggestive of severe, refractory asthma. Although various psychiatric disorders were diagnosed, psychological testing did not differentiate these subjects from patients with severe asthma. Newman and Dubester [17] reported the largest and most complete description of VCD cases in 1995 [7•]. A total of 95 adults with VCD were presented, including 53 with both VCD and asthma. Of the subjects with VCD alone, 21% had prior psychiatric hospitalizations, 73% had *Diagnostic and Statistical Manual* [*of Mental Disorders*], Third Edition, Revised (DSM-III-R) Axis I diagnoses (major psychiatric disorder), and 37% had an Axis II diagnosis (personality disorder). However, with the exception of the psychiatric hospitalizations, these striking findings were not significantly different from control subjects with severe asthma. Therefore, retrospective, controlled studies in adults suggest that VCD patients presenting with refractory asthma are psychiatrically impaired, although it is not clear whether that impairment preceded their symptoms, or whether this association also applies in VCD patients whose symptoms are less severe.

Vocal cord dysfunction in pediatric patients also has a reported association with psychiatric findings. In an uncontrolled series by Landwehr *et al.* [10], six of the seven subjects with VCD masquerading as exercise-induced asthma were found to have at least one DSM-III-R Axis II psychiatric diagnosis. "Perfectionism," depression, anxiety, "social stresses" [13,16], "intolerance of personal failure,"

and "the perception of family pressure to achieve a higher level of successs" [9] have all been noted as common among adolescent VCD patients. Although emotional stress is a common theme, the precise role of psychiatric factors in the pathophysiology of VCD is not clear.

Therapeutic Options

There are no well-controlled treatment trials validating various forms of VCD treatment. However, a treatment standard has emerged based on clinical practice that includes a combination of patient education and specialized speech therapy. Treating associated conditions, such as psychiatric disease, asthma, rhinosinusitis, and GERD, is also appropriate.

Patient education

Once the diagnosis of VCD is established, the next step is to explain to the patient what is known about VCD and its treatment. It is helpful to validate that the symptoms are distressing, unintentional, and important, despite the absence of a clear, organic explanation for them. Sharing a video or photographic image of the laryngoscopic findings simplifies the explanation and facilitates the patient's acceptance of the diagnosis. Acceptance of the diagnosis and motivation to proceed with treatment are important prognostic indicators.

Speech therapy

The most useful therapeutic intervention for VCD is speech therapy. The goal is to teach the VCD patient how to relax his or her larynx using breathing exercises and relaxation techniques that focus on the muscles of the larynx, neck, shoulders, and chest [9,12,16]. Some centers have found biofeedback to be a useful tool during speech therapy [20]. Unfortunately, relatively few speech pathologists are familiar with VCD treatment techniques, and, therefore, it is important to seek out appropriate local resources, including a speech pathologist who is enthusiastic about working with VCD patients.

Speech therapy treatment rarely requires more than three or four treatment sessions. After successful treatment, symptoms often do not disappear entirely, although they are less severe and less frequent. The adolescent athlete with VCD, with time, will typically resume competition without VCD symptoms limiting his or her performance. Unnecessary medications should be discontinued, although a gradual weaning is appropriate in most cases.

Asthma and vocal cord dysfunction

As mentioned earlier, many VCD patients also have asthma, and continuing appropriate asthma treatment is, therefore, important. In accordance with the NHLBI Guidelines, the goals for asthma treatment include minimizing asthma symptoms, maintaining normal or near-normal lung function, and minimizing any limitation in physical activity [6]. Optimizing the asthma medication regimen is best achieved by gradual weaning of medications once these goals have been met. It is very unusual for a pediatric patient with both asthma and VCD to require continuous systemic corticosteroid treatment chronically, and given the adverse effects of this treatment, every effort should be made to rely on inhaled rather than systemic corticosteroids.

Other vocal cord dysfunction treatments

Occasionally, acute VCD symptoms are severe and refractory enough to require emergent treatment. There are several acute interventions that might help avoid intubation, the most effective of which is administration of "heliox" [16]. A mixture of 70% helium and 30% oxygen, heliox is significantly less dense than ambient air and can maintain efficient laminar airflow during refractory upper airway obstruction, often aborting VCD attacks within minutes. Because heliox does not improve intrathoracic obstruction caused by asthma, a dramatic treatment response to it not only helps acutely, but can also be referred to during patient education and speech therapy efforts, as a demonstration of the exact location of the obstruction. Unfortunately, heliox is only available in selected emergency departments. Other helpful acute measures include the administration of oxygen and an anxiolytic agent, such as a benzodiazepine.

Iatrogenic vocal cord paralysis has been used effectively in selected VCD patients in whom vocal cord adduction is sustained chronically. This is achieved by injecting botulinum toxin ("Botox") into the thyroarytenoid muscle [4]. By blocking the release of acetylcholine at the motor endplate, these Botox injections relax the thyroaryntenoid muscle, which causes relative abducton of one or both vocal cords and restores glottic patency. Botox injections have prevented reliance on tracheostomy to maintain airway patency in some VCD patients, although it should be considered experimental and reserved for refractory cases.

Conclusions

Vocal cord dysfunction is a relatively common cause of respiratory symptoms in pediatric patients and should be considered in any patient in whom dyspnea, wheezing, chest tightness, and/or throat tightness fails to respond to asthma treatment. Pediatric VCD is most prevalent among adolescent girls, and symptoms are typically intermittent and triggered by exercise. The diagnostic workup for VCD should include spirometry, and suggestive findings include an FEF_{50}/FIF_{50} of more than 1.0 and truncation of the inspiratory portion of the flow volume loop. However, as with asthma, spirometry is often normal in the absence of symptoms. Confirming the diagnosis requires observing paradoxical vocal cord adduction larynogoscopically; this often requires provocation with either inhaled methacholine or exercise.

There appears to be a strong association between VCD and GERD, although the proof of this has so far been based on indirect findings of laryngeal inflammation during laryngoscopy. It is not known whether GERD causes VCD.

There is a continuum of VCD severity, and the most severe cases tend to associate strongly with psychiatric disturbance. However, controlled studies have so far not detected differences in psychopathology between VCD subjects and asthma controls. Emotional stress is a frequent association in VCD, regardless of its severity, although a causal link has not been established.

Patient education and specialized speech therapy are the most essential components of pediatric VCD treatment. Treating associated conditions, such as asthma, GERD, and psychiatric disease, is appropriate if any of these coexist with VCD. In the setting of acute VCD symptoms, administering heliox has been helpful. For chronic refractory VCD, some patients have benefited from botulinum toxin injection of the cricoarytenoid muscle.

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