

Paradoxical vocal cord dysfunction: when a wheeze is not asthma

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ABSTRACT

Vocal cord dysfunction (VCD) is an uncommon condition which often mimics asthma in presentation and severity. We present nine- and 11-year-old female siblings with vocal cord dysfunction, which is a dysfunction of the larynx involving unintentional paradoxical adduction of the vocal cords during inspiration. We evaluated the use of exercise testing in conjunction with pulmonary function testing in suspected vocal cord dysfunction. Although normal pulmonology function tests were elicited with the patient at rest, exercise testing revealed blunting of the expiratory loop with attenuation of the inspiratory loop unique to VCD. The child underwent video laryngoscopy in the specialised voice clinic, which confirmed vocal cord dysfunction. Exercise testing is a rapid and noninvasive means of diagnosing vocal cord dysfunction in a small subset of patients, but video laryngoscopy, with training manoeuvres to elicit paradoxical vocal cord movements in VCD, remains the gold standard of diagnosis of VCD.

Keywords: asthma, exercise testing, video-laryngoscopy, vocal cord dysfunction

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INTRODUCTION

Vocal cord dysfunction (VCD) is often misdiagnosed as “refractory asthma”. The clinical presentation includes dyspnoea, exertional dyspnoea, cough, throat tightness, wheezing and dysphonia. The majority of the patients presenting with VCD are female, and it is especially common among children and adolescents. The presentation is often dramatic, and has even led to patients being intubated, sometimes with tracheostomies performed; therefore, there needs to be a better awareness of this condition.

CASE REPORT

An 11-year-old girl was referred to the respiratory clinic with a history of exertional dyspnoea over several months. Her initial examination and pulmonary-function test were normal. She presented one month later with an “acute asthmatic attack” requiring frequent inhaled β -agonists and oral steroids in the Children’s Emergency Department. However, she was increasingly tachypnoeic in the ward, deteriorating with inspiratory and expiratory “wheezing”,

refractory to 48 hours of frequent nebulisations. She was transferred to the high-dependency unit and given adjunct intravenous magnesium sulphate for management of “status asthmaticus”. She gradually improved, and was discharged one week later from the hospital with minimal symptoms. Her parents reported that even after her discharge from the hospital, she continued to have periods of breathlessness.

She presented again six weeks later, at which time there was persistent dysphonia, hoarseness of voice and exercise limitation of 50 metres. She was commenced on 1.5 mg/kg of oral prednisolone along with a stepwise escalation of therapy, including oral theophylline, inhaled corticosteroids with long-acting β -agonists and sodium cromoglycate. However, there was no abatement of symptoms. Her degree of respiratory distress appeared to fluctuate from minimal to severe, with a persistent “wheeze” being present on physical examination. Her anteroposterior thoracic diameter was not increased. She had bilateral “wheezing” at rest, but this was loudest over the larynx. Her oxygen saturation was 100% in room air and she appeared comfortable at rest throughout these periods. The administration of β -agonists failed to relieve the symptoms. Interestingly, when she was asked to perform certain manoeuvres prior to auscultation, her wheezing diminished and disappeared. These manoeuvres included counting out loud, and closing her mouth and pinching her nose during auscultation.

She underwent a further pulmonary function test while she had bilateral wheeze, and this was found to be normal. She also underwent a high-resolution computed tomography of her thorax, which was reported as normal. She was then referred to the otolaryngology service for which flexible laryngoscopy was carried out on two separate occasions, one under sedation and the other by the bedside with hyperventilation manoeuvres. Both of which did not demonstrate any evidence of paradoxical movement of the vocal cords. She performed exercise testing after a period of six weeks of inhaled and oral corticosteroids. This demonstrated a plateau in the expiratory limb of the flow-volume curve at 3, 6, 10 and 15 minutes of exercise (Fig. 1). At the start of the test, the initial flow-volume loop was normal. The attenuation was more marked at the end of the exercise testing. She was later evaluated in the specialised voice clinic, where video laryngoscopy was carried out with training manoeuvres and vocal cord dysfunction was demonstrated (Fig. 2).

Interestingly, six months later, her nine-year-old sister presented to the emergency department with acute onset of

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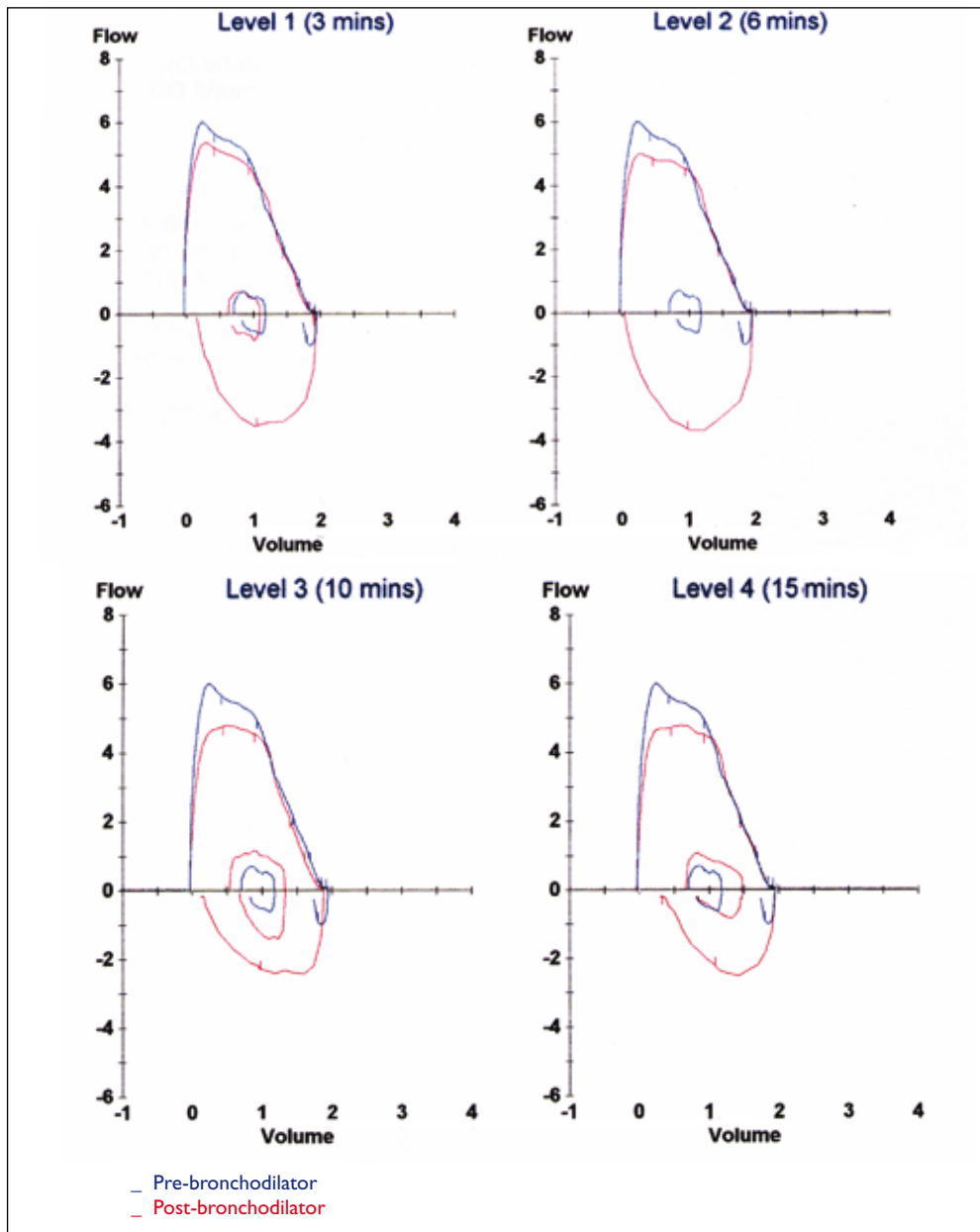


Fig. 1 Flow-volume loops done during exercise testing shows blunting of the expiratory limb.

shortness of breath with “wheeze”. The primary physician in charge of her sister diagnosed VCD. The clinical examination revealed a wheeze that was loudest over the larynx. Subsequent pulmonary function tests were normal. This child was sent for speech therapy. Both siblings are currently doing well on regular follow-up, with no further acute admissions for “exacerbations of asthma attacks”. They have been weaned off all medications.

DISCUSSION

VCD syndrome was first described by Duglison in a medical textbook in 1842 where he discovered disorders of the laryngeal muscles brought on by “hysteria”.⁽¹⁾ At the time, VCD was a condition that was confined mainly to the psychiatric literature. Psychogenic factors have been elucidated in a majority of VCD case reports and clinical studies. During the 1980s, VCD became more recognised as a clinical entity, with an emergence of

an increasing number of publications on this topic. In a prospective study of 52 Vancouver school children referred for poorly-controlled exercise-induced asthma, 14 (26.9%) of them had VCD.⁽²⁾ The largest series was published by the National Jewish Institute, where they reviewed their cumulative data over seven years. In their 95 adult patients with confirmed VCD on laryngoscopy, up to 10% of patients had VCD alone, while more than 30% had coexistent asthma.⁽³⁾

VCD tends to be triggered by exercise, and as a result, typically confused with exercise-induced asthma. The pathognomonic features of the adduction of the cords with posterior chinking of the vocal cord during inspiration or early expiration, or in either inspiration or expiration, has been frequently described,⁽⁴⁻⁶⁾ although its absence does not rule out VCD. In our patient, video laryngoscopy demonstrating vocal cord dysfunction was only elicited under training manoeuvres (including panting, phonation



Fig. 2 Photograph of the vocal cords during inspiration in a patient with VCD shows the paradoxical adduction with a posterior chink present.

and deep breathing). In the literature, most case studies of VCD report blunting of the inspiratory loop with only one case describing the attenuation of the expiratory limb.⁽⁷⁾ In our patient with VCD, we demonstrated blunting of the expiratory limb elicited as early as three minutes from the start of exercise testing and which reached a maximal attenuation at ten minutes.

The exact physiological mechanism of vocal cord dysfunction remains unknown. However, some postulate that possible pathways contributing to the spectrum of VCD include altered autonomic imbalance, arising from the central brain regions that are polysynaptically linked to the larynx that results in laryngeal hyper-responsiveness.⁽⁸⁾ Direct stimulation of the sensory nerve endings in the upper or the lower respiratory tract via noxious stimuli, such as environmental (cold air, smoke) or irritant exposure, is another theory.⁽⁹⁾

The clinical presentation includes dyspnoea, exertional dyspnoea, cough, throat tightness, wheezing, or dysphonia and can present in children as young as six years old. The primary treatment for VCD involves a combination of pharmacological, psychological, psychiatric and speech training. In most cases, patients may resume their activities without significant limitation. Panting manoeuvres are said to help to relieve the symptoms acutely.^(10,11) Speech therapy plays an important role in long-term management by providing respiratory retraining to de-emphasise laryngeal breathing. It has been suggested that during an acute attack, a mixture of helium: oxygen mixture (80:20) inhalation can help resolve the symptoms.⁽¹²⁾

VCD is an under-recognised disorder affecting more people than previously thought, with the majority of patients ranging from young girls to middle-aged women. In this case, the presentation of the sibling with VCD is interesting. The astute clinician must have a high index of suspicion and be knowledgeable about its presentation. Distinct history and physical examination findings should raise the suspicion for VCD in the treating physician. Prompt recognition of VCD, with appropriate referral

to the speech therapist may avert misdiagnosis of the patient and inappropriate treatment for refractory asthma. Inappropriate hospitalisation, high doses of unnecessary corticosteroids, intubation, and tracheostomy are among the iatrogenic causes of morbidity in this population of patients.

While the diagnosis is based on laryngoscopic evidence of inspiratory vocal cord adduction, this may have to be elicited under specialised and expert hands. Certain pulmonary function laboratory findings characteristic of the disorder may aid in the diagnosis especially if the expertise for video laryngoscopy is not available. Many of the children are asymptomatic at rest, and require exercise challenge or provocation challenge with methacholine to elicit symptoms and vocal cord abnormalities.⁽¹³⁻¹⁵⁾ It is worth giving them an adequate trial of inhaled corticosteroids or oral steroids for a period to ensure that the asthma is adequately controlled prior to the challenge. In conclusion, vocal cord dysfunction is not a common clinical entity. We present this case as an example of how exercise testing can be used as one of the diagnostic tools for vocal cord dysfunction, but referral to a specialised unit for video laryngoscopy remains the gold standard for diagnosis.

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