

Vocal Cord Dysfunction: Paradoxical Vocal Cord Motion

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Introduction

- Paradoxical vocal cord motion (PVCM)
 - Episodic laryngeal dyskinesia, VCM
 - Vocal cord adduction during inspiration/expiration causing a functional extrathoracic airway obstruction.
 - Symptoms include: wheeze, cough, dyspnea, SOB
 - More common than is appreciated, diagnosis frequently not considered.
 - Often confused with asthma and misdiagnosed.
 - Much morbidity caused from misdiagnosis.
 - Newman et al studied 95 patients with proven PVCM
 - Asthma was misdiagnosed an avg. 4.8 years, 28% intubated

Introduction cont'd

MEDICATIONS AT TIME OF ADMISSION, EXPRESSED AS NUMBER OF PATIENTS RECEIVING EACH MEDICATION DAILY

	VCD (<i>n</i> = 42) (<i>n</i>)	VCD + Asthma (<i>n</i> = 53) (<i>n</i>)	Asthma (<i>n</i> = 42) (<i>n</i>)
Beta-agonists	40	52	42
Theophylline	36	45	37
Prednisone	34	37	33
Inhaled steroids	25	29	30
H ₂ antagonist	22	22	15
Anxiolytic	13	12	7
Antidepressant	3	5	4
Neuroleptics	0	2	1
Methotrexate	4	2	2

History

- 1842: PVCM first recognized by Dunglison where he described disorders of laryngeal muscles brought on by “hysteria”.
- 1869: Mackenzie visualized cords close during inspiration in hysteric adults with stridor. First to attribute stridor to PVCM.

History cont'd

- 1902: William Osler defined the condition by describing patients with “laryngeal muscle spasms during inspiration and times of great distress.”
- Cause was considered purely psychogenic and confined to psychiatric literature.
- 1970's: otolaryngologic literature described a syndrome of episodic stridor, dyspnea, palpitations, diaphoresis that mimics or is mistaken for asthma, upper airway obstruction, or malingering.
- 1980's: First case reports and series began to emerge, consistency with diagnostic criteria.
- Today PVCMD is well-described disorder – caused by organic and non-organic disease - with various treatment regimens that depend upon the likely etiology.

Prevalence

- General population is unknown.
 - O'Connell et al, 164 patients
 - Up to 20% of females who underwent rhinolaryngoscopy for any reason had PVCMM.
- National Jewish Center, 1994, multiple patients diagnosed with refractory asthma:
 - 10% had PVCMM alone.
 - 30% had PVCMM with coexistent asthma.
- Patients diagnosed with PVCMM:
 - 56% had coexistent asthma.
- Because PVCMM is common among asthmatics and presents with symptoms similar to those seen in patients with only asthma, it is commonly overlooked and not included in the differential.

Demographics

- Juveniles – under age 18
 - 2 studies at different institutions found:
 - Average presenting age: 14.6 (range 9.0 – 18.0)
 - 82-86% of patients female.
 - Similarities among patients included: organized sports, social stressors, exercised-induced symptoms.
 - Powell et al found strong association with GERD.
 - Laryngospasm likely 2° reflux irritation but cause-effect relationship is yet established.

Demographics cont'd

- All age groups:
 - Avg. age at diagnosis 30 years (range 22-34)
 - 70-98% of patients were female, caucasian
- Documented psychological risk factors:
 - Medical profession, overweight, stress, anxiety, childhood abuse, psychiatric illness.
 - Increased incidence during wartime.
 - Psychopathology of pts with PVCMM varies but prevalence is same as that among asthmatics (excluding anxiety disorder).
- Documented physiological risk factors:
 - Asthma, brainstem abnormalities, CF, GERD

Etiology

- Causes of PVCMM with similar presentation:
 - Cortical injury – CVA, static encephalopathy, LMN injury from MG, ALS, medullary infarct.
 - Brainstem compression – reports more common in children, A-C malformation, aqueductal stenosis; 1 adult case report from giant posterior fossa arachnoid cyst.
 - Psychiatric – conversion and somatization d/o, unconsciously induced, no secondary gain; factitious d/o and malingering, consciously induced with 2° gain.

Etiology cont'd

- Voluntary wheezes are reproducible in all patients, not just the psychiatrically ill.
- Demonstrated by forced inspiration/expiration at low lung volumes - when glottic aperture is narrowest.
- Irritant-induced: newly documented phenomenon, temporal relationship shown between irritant exposure and PVCM, irritants included ammonia, cleaning chemicals, organic solvents, flux flames, smoke, chlorine from swimming pool.
 - Also includes GERD associated PVCM although no documented temporal relationship.

Proposed Classification

Organic Causes	Brainstem Compression
	Cortical or upper motor neuron injury
	Nuclear or lower motor neuron injury
	Movement disorders
	Gastroesophageal reflux
Nonorganic Causes	Factitious or malingering disorder
	Somatization/conversion disorder

Clinical Presentation

- Wide variety of symptoms including:
 - Cough
 - Inspiratory/expiratory wheeze
 - Dyspnea with/without exertion
 - Stridor
 - Hoarseness
 - Chest tightness
 - Reflux

Study evaluating 90 patients with documented PVCM:

-- Cough most common reported in up to 77%.

Clinical Presentation cont'd

- Hx sig. for frequent, episodic attacks leading to SOB and multiple ER visits.
- Previous diagnoses include asthma, refractory asthma, exercise-induced bronchospasm, COPD, anaphylaxis.
- Juveniles with PVCM have increased incidence of anxiety d/o when compared to asthmatics.
 - Anxiety precedes respiratory symptoms in PVCM
 - Anxiety follows respiratory symptoms in asthmatics.

20 patients with previously diagnosed PVCM

Diagnosis	
Asthma	12 patients (60% of total)
EIB* only	3 patients (15%)
COPD*	2 patients (10%)
Anaphylaxis	2 patients (10%)
Hoarseness	1 patient (5%)

* EIB = exercise-induced bronchospasm
and COPD = chronic obstructive pulmonary
disease.

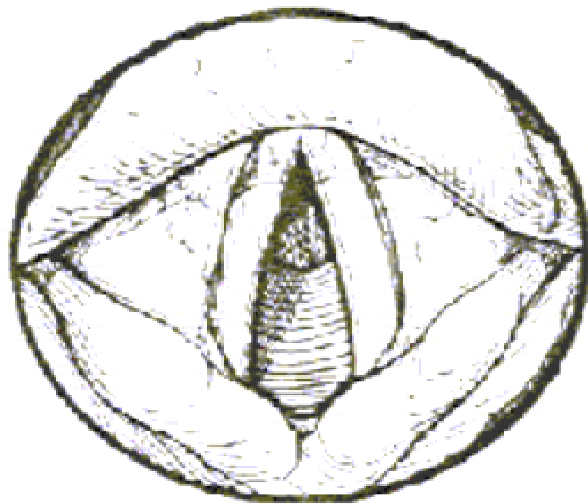
Physical Exam

- Classical finding on laryngoscopic exam is inspiratory anterior vocal cord closure with posterior chinking.
- This obstruction decreases laminar airflow through the glottis and produces an inspiratory wheeze or stridorous sound similar to that heard in asthmatics.
- Differs from paralysis:
 - PVCMM show normal cord movement during phonation.
- Differs from laryngospasm:
 - Laryngospasm shows adduction throughout the entire cord length without posterior chinking.

Physical Exam – posterior chinking

A

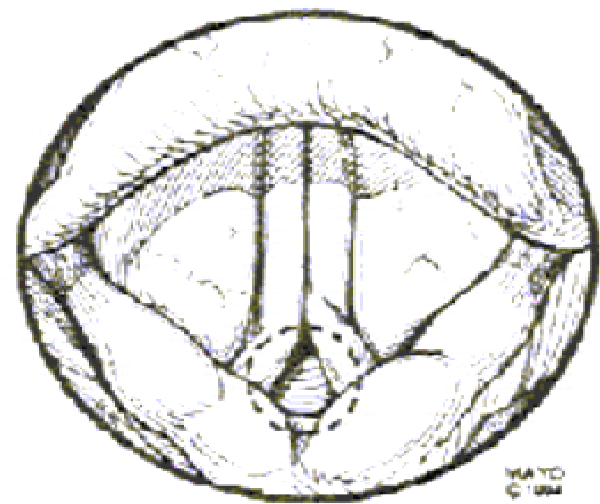
Anterior



Posterior

B

Anterior



Posterior

WATD
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Physical Exam cont'd

- Do asthmatics have physiologic PVCMM?
 - Patients with asthma alone can also exhibit adduction of the vocal cords during inspiration, although they show no posterior chinking.
- Higgenbottam et al induced broncho-constriction in patients and revealed a decrease in the glottic aperture during quiet respiration.
- This glottic narrowing allows a certain amount of positive end expiratory pressure (PEEP) to maintain airway patency preventing alveolar collapse.
- Such a physiologic response in asthmatics is suspected to be therapeutic not pathologic.
 - pursed-lip exhalation almost always relieves asthmatic wheezing.
- Where the transition from physiologic response to pathologic movement during expiration takes place is unclear.

Other Physical Exam Findings

- Wheezing originates over the larynx and is less evident over the rest of the lung fields.
- Tachypnea with neck extension and constant contraction of the anterior neck muscles.
- Patients with or without concurrent asthma are often hypoxic and therefore may appear cyanotic during exacerbations.

Differential Diagnosis

- Extensive, therefore separate by location and age group.
- Anatomic locations for extrathoracic airway obstruction include the trachea, larynx, glottis, and thyroid.
- Endobronchial obstruction must also be suspected as a foreign body, bronchial adenoma, bronchial carcinoid, or bronchogenic carcinoma can all present with dyspnea and/or wheezing.
- Because the site of obstruction is more specific to the presenting symptoms than the actual cause of the obstruction, it is helpful to develop a d/d according to age group and location of obstruction.

Adult		Pediatric	
Location	Cause	Location	Cause
Oral Cavity	Trauma	Oral Cavity	Macroglossia, angioedema
	Neoplasm		Dermoid cyst, Robin's Syndrome
	Angioedema		Ludwig's angina,
Pharynx	Neoplasm, Trauma	Pharynx	Lingual thyroid,
	Ludwig's Angina, Abscess		Peritonsillar abscess
	goiter angioedema		Tonsillar hypertrophy, Thyroglossal duct cyst
Larynx	Vocal cord dysfunction	Larynx	Vocal cord dysfunction
	Papillomatosis, Ludwig's Angina		Epiglottitis, vocal cord web
	Laryngospasm, angioedema		Bilateral vocal cord paralysis
	Bilateral vocal cord paralysis, myasthenia gravis, Bulbar Palsy		Laryngeal stenosis Laryngomalacia, cyst
	Prolonged intubation, anaphylaxis		Hemangioma, croup
Trachea	Cricoarytenoid arthritis, foreign body	Trachea	Prolonged intubation, Papillomatosis
	Blunt trauma, Neoplasm		Web, stenosis, foreign body
	Vascular anomalies, tracheitis		Cyst, thyroid tumor, Vascular anomalies, tracheomalacia
Endobronchial	Asthma, COPD, carcinoid syndrome	Endobronchial	Asthma
	Congestive heart failure		Tracheobronchitis

Lab Studies

- There is much controversy as to whether or not patients with only PVCM actually undergo respiratory compromise during acute exacerbations.
- The literature does show evidence of patients exhibiting no respiratory distress and presenting without hypoxemia, though such a finding is much less common.
- Different studies can be performed to distinguish between asthma and PVCM.

Lab studies cont'd

PVCM

Extrathoracic

Normal Aa gradient

No hypercapnea/acidosis

+ hypoxemia

Asthma

Intrathoracic

Elevated Aa gradient

+ hypercapnea/acidosis

+ hypoxemia

Arterial-Blood-Gas Analysis

Table 4. Arterial-Blood-Gas Analysis (Room Air) during an Episode of Wheezing and Dyspnea.*

PATIENT No.	pH	PO ₂ <i>torr</i>	PCO ₂ <i>torr</i>	P(A-a)O ₂ <i>torr</i>
1	7.47	75	35	3
2 †	7.40	89	39	11
3	7.48	64	34	15
4	7.43	75	37	1
5	7.40	68	36	9

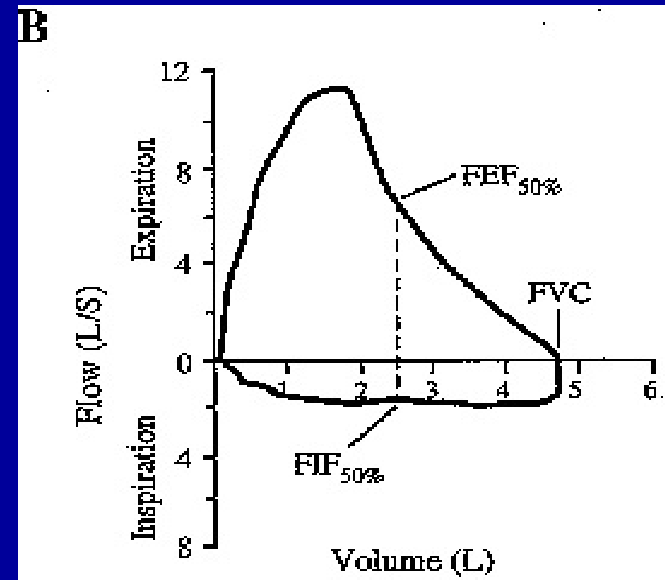
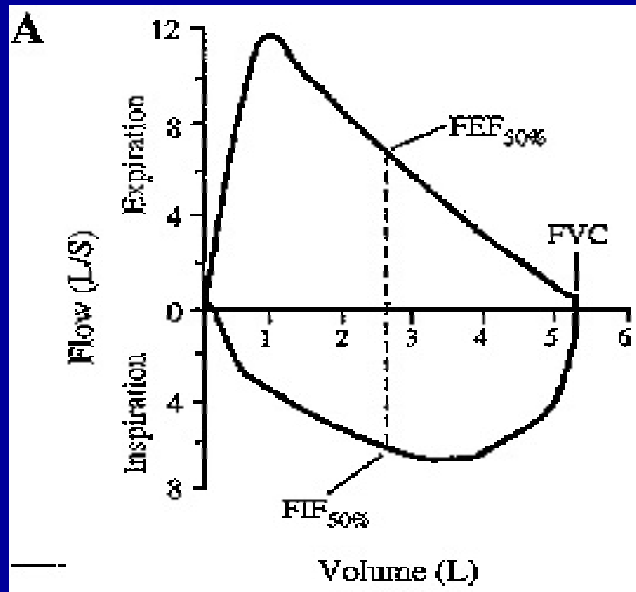
*PO₂ denotes partial oxygen tension, PCO₂ partial carbon dioxide tension, and P(A-a)O₂ alveolar-to-arterial oxygen-tension gradient.

†Measurements in Montgomery, Alabama (barometric pressure = 760 mm Hg). All other measurements were obtained in Denver, Colorado (barometric pressure = 630 mm Hg).

Lab studies cont'd

- While asymptomatic, patients have tendency to show normal laboratory/test values.
- Useful to stress patients during testing, exposing them to the same elements which typically initiate attacks.
 - commonly exercise induced or irritant induced
 - both methods are used for provocative testing.
- Methacholine challenges are used to detect bronchial hyperresponsiveness.
 - Useful to diagnose asthma when PFT's are inconclusive. Used for provocative testing to induce PVCN.

- PFT's with flow-volume loops have also been used to support the diagnosis of PVCN in symptomatic patients.
- Flow-volume loops of patients with PVCN often show flattening of the inspiratory curve, or a decrease in maximal inspiratory flow during acute attacks, and are normal while asymptomatic



PFT studies cont'd

- Inspiratory blunting is sensitive for symptomatic patients with PVCM but is not specific for VCD and may be produced by most types of extrathoracic airway obstruction.
- Parker et al evaluated 26 patients with PVCM
 - exercise flow-volume loops indicated the upper airway as a cause for symptoms in 74%
 - 62% showed inspiratory flow limitation
- Primary use of PFT's is to eliminate asthma from the differential diagnosis.

PFT studies cont'd

- Expiratory adduction and obstruction has been shown by laryngoscopy in these patients without evidence of expiratory flow-volume abnormalities.
 - Mechanism unknown, pursed-lip exhalation suspected
 - Elevates soft palate to posterior nasopharyngeal wall
 - Closes nasopharyngeal airway, increases resistance
 - Creates sufficient back pressure to open vocal cords and therefore shows no expiratory flow loop defect

Other lab studies

- Other PFT parameters have a high sensitivity and specificity for detecting extrathoracic airway obstruction but are not specific for VCD:
 - FEF50/FIF50
 - FEV1/FVC,
 - SRaw (specific airway resistance)
- Chest x-rays show no evidence of lung hyperinflation or peribronchial thickening.
- Low peripheral eosinophil count.

Diagnosis

- Difficult due to its episodic nature and presentation.
- Criteria for diagnosis:
 - Laryngoscopic confirmed adduction of vocal cords during inspiration, early expiration, or both inspiration and expiration with evidence of post. glottic chinking.
 - adduction occurring during only the last half of expiration is not pathologic
 - PVCMM cannot be ruled out when asymptomatic.
 - if the patient is asymptomatic, negative laryngoscopic findings do not exclude the diagnosis
 - Absence of gagging or coughing during laryngoscopy
 - must not confuse PVCMM with vocal cord motion produced by a laryngoscope induced gag reflex

Treatment

- The cause of the PVCMM must first be elicited.
- In PVCMM secondary to preexisting organic disease states the underlying disorder should be treated appropriately:
 - brainstem compression, encephalopathy, stroke, ALS, myasthenia gravis, GERD, etc.
- A history of previous exposure to irritants should be obtained.
- With no obvious source of causative organic disease - acute treatment is henceforth symptomatic.

Heliox therapy

- Gaseous mixture of oxygen and helium in ratios of 20/80 and 30/70 respectively.
 - mixture is less dense than air
 - inhalation reduces turbulence in the airway and eliminates respiratory noise
- Recommended for immediate relief of respiratory distress
 - reduces anxiety - the predisposing factor to many attacks
 - provides short-term relief of dyspnea
 - not effective for relief of symptoms due to asthma or other lower airway disease

LARYNGEAL DYSKINESIA

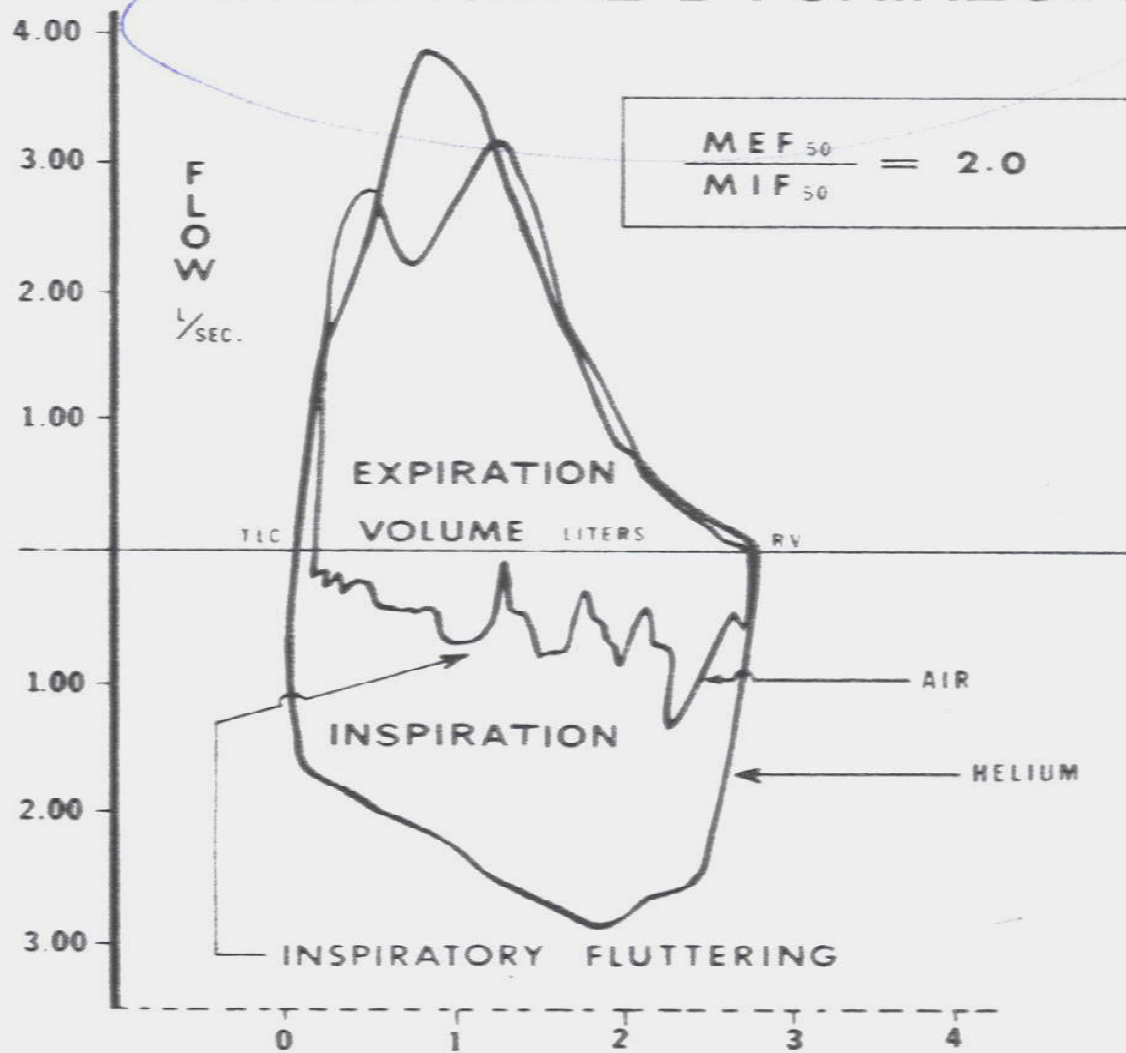


FIGURE 1. Tracing of a representative flow volume loop showing marked decrease in the inspiratory flows and inspiratory fluttering. Inspiratory flows improved while ventilating with an 80 percent helium/20 percent oxygen mixture.

Other Acute Therapy

- IPPV and CPAP
 - widen the rima glottidis and reduce turbulence
- Panting
 - physiologically increasing the glottic aperture
- Benzodiazepines / Reassurance
 - reduce anxiety and have been shown effective
- General anesthetic induction
 - small doses of propofol can relieve acute attacks
- Intralaryngeal injection of botulinum toxin type A
 - more invasive approach for severe exacerbation
- Conversely, therapy with bronchodilators / oxygen / corticosteroids
 - shown ineffective for relief in patients with PVCMM

Long-term Management

- requires a multidisciplinary approach involving speech therapy, psychiatric support and physician education regarding the syndrome
- Speech therapy
 - techniques aimed at focusing attention on expiration and abdominal breathing rather than on inspiration and laryngeal breathing
 - early recognition of symptoms allows relaxation of neck, shoulder and chest muscles promoting normal laryngeal breathing

Long-term management cont'd

- Psychotherapy
 - allows patient to explore for potential causes
 - trains the patient with relaxation techniques
- Psychotherapy should be initiated if:
 - insufficient improvement with speech therapy alone
 - evident psychological tumult in the patient's life
 - at the patient's request
- Education about the condition
 - useful for reducing stress.
 - Biofeedback training has been used as a long-term treatment approach -not considered primary agent

Management Summary

TABLE 2. Treatment options for patients with VCD

Acute treatment	Long-term treatment
Inhaled mixture of 70%-80% He/30%-20% O ₂	Biofeedback training
CPAP	Supportive psychotherapy
IPPV	Speech therapy
Benzodiazepines	Hypnotherapy
Panting exercises	Panting exercises
Reassurance	

CPAP, Continuous positive airway pressure; *IPPV*, intermittent positive pressure ventilation.

Prognosis

- long-term outcome unknown
 - most literature consists of case reports and retrospective studies.
 - One study followed three patients over a 10-year period - all showed continued symptomatic VCD at follow-up
- More trials needed before conclusions about management efficacy can be drawn.

Prognosis cont'd

- Initial response to standard management (speech, psychotherapy) is good:
 - interview with 15 patients all diagnosed with PVCMM who had received prior therapy.
 - took place an average of 20 months (range 11-62) after initial diagnosis of the disorder.
 - results showed most responded well with improved functioning and fewer symptoms after intervention

Conclusion

- PVCMM is an under recognized disorder that can result from many different etiologies
 - majority of patients are young to middle-aged females.
- Must have a high suspicion to make the diagnosis
- Many people every year are misdiagnosed and wrongly treated for refractory asthma and anaphylaxis
 - Inappropriate hospitalization, high doses of corticosteroids, intubation, and tracheostomy
- Strong association between people with VCD and those with asthma.

Conclusion cont'd

- The presentation of both patient groups can be identical
 - the finding of one in a patient does not rule out the presence of the other - it seems to make it more likely.
- Each disease carries its own unique treatment,
 - asthma therapy is ineffective against symptoms of VCD and vice-versa.
 - Success for both relies on correct diagnosis

Treatment of both must be maintained beyond resolution of the initial exacerbation.
- Little data is available about the long-term effects of therapy, but short-term studies have revealed promising results.
 - As more clinicians become aware about the spectrum of presentation seen with VCD, fewer misdiagnoses will be made.

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