Vocal Cord Dysfunction:
Paradoxical Vocal Cord Motion

Todd M Weiss, Senior Medical Student
Faculty Advisor: Francis B. Quinn, M.D.
The University of Texas Medical Branch
Department of Otolaryngology
Summer Grand Rounds
July 2001
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
### Medications at Time of Admission, Expressed as Number of Patients Receiving Each Medication Daily

<table>
<thead>
<tr>
<th>Medication</th>
<th>VCD  (n=42) (n)</th>
<th>VCD + Asthma  (n=53) (n)</th>
<th>Asthma  (n=42) (n)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Beta-agonists</td>
<td>40</td>
<td>52</td>
<td>42</td>
</tr>
<tr>
<td>Theophylline</td>
<td>36</td>
<td>45</td>
<td>37</td>
</tr>
<tr>
<td>Prednisone</td>
<td>34</td>
<td>37</td>
<td>33</td>
</tr>
<tr>
<td>Inhaled steroids</td>
<td>25</td>
<td>29</td>
<td>30</td>
</tr>
<tr>
<td>H$_2$ antagonist</td>
<td>22</td>
<td>22</td>
<td>15</td>
</tr>
<tr>
<td>Anxiolytic</td>
<td>13</td>
<td>12</td>
<td>7</td>
</tr>
<tr>
<td>Antidepressant</td>
<td>3</td>
<td>5</td>
<td>4</td>
</tr>
<tr>
<td>Neuroleptics</td>
<td>0</td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>Methotrexate</td>
<td>4</td>
<td>2</td>
<td>2</td>
</tr>
</tbody>
</table>
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.
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 PVCM 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’Connel et al, 164 patients
  – Up to 20% of females who underwent rhinolaryngoscopy for any reason had PVCM.

• National Jewish Center, 1994, multiple patients diagnosed with refractory asthma:
  – 10% had PVCM alone.
  – 30% had PVCM with coexistent asthma.

• Patients diagnosed with PVCM:
  – 56% had coexistent asthma.

• Because PVCM 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 PVCM varies but prevalence is same as that among asthmatics (excluding anxiety disorder).

• Documented physiological risk factors:
  – Asthma, brainstem abnormalities, CF, GERD
Etiology

• Causes of PVCM 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; factious d/o and malingering, consciously induced with 2º 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

<table>
<thead>
<tr>
<th><strong>Organic Causes</strong></th>
<th>Brainstem Compression</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Cortical or upper motor neuron injury</td>
</tr>
<tr>
<td></td>
<td>Nuclear or lower motor neuron injury</td>
</tr>
<tr>
<td></td>
<td>Movement disorders</td>
</tr>
<tr>
<td></td>
<td>Gastroesophageal reflux</td>
</tr>
<tr>
<td><strong>Nonorganic Causes</strong></td>
<td>Factitious or malingering disorder</td>
</tr>
<tr>
<td></td>
<td>Somatization/conversion disorder</td>
</tr>
</tbody>
</table>
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

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Number of Patients</th>
<th>Percentage of Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Asthma</td>
<td>12 patients</td>
<td>60%</td>
</tr>
<tr>
<td>EIB* only</td>
<td>3 patients</td>
<td>15%</td>
</tr>
<tr>
<td>COPD*</td>
<td>2 patients</td>
<td>10%</td>
</tr>
<tr>
<td>Anaphylaxis</td>
<td>2 patients</td>
<td>10%</td>
</tr>
<tr>
<td>Hoarseness</td>
<td>1 patient</td>
<td>5%</td>
</tr>
</tbody>
</table>

* 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:
  – PVCM show normal cord movement during phonation.

• Differs from laryngospasm:
  – Laryngospasm shows adduction throughout the entire cord length without posterior chinking.
Physical Exam – posterior chinking
Physical Exam cont’d

• Do asthmatics have physiologic PVCM?
  – Patients with asthma alone can also exhibit adduction of the vocal cords during inspiration, although they show no posterior chinking.

• Higenbottam 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.
<table>
<thead>
<tr>
<th>Adult</th>
<th>Location</th>
<th>Cause</th>
<th>Pediatric</th>
<th>Location</th>
<th>Cause</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Oral Cavity</td>
<td>Trauma</td>
<td>Oral Cavity</td>
<td>Macroglossia, angioedema</td>
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<td></td>
<td>Neoplasm</td>
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<td>Dermoid cyst, Robin’s Syndrome</td>
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<tr>
<td></td>
<td>Angioedema</td>
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<td>Ludwig’s angina,</td>
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<tr>
<td>Pharynx</td>
<td>Neoplasm, Trauma</td>
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<td>Pharynx</td>
<td>Lingual thyroid,</td>
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<td>Larynx</td>
<td>Vocal cord dysfunction</td>
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<td>Larynx</td>
<td>Vocal cord dysfunction</td>
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</tr>
<tr>
<td>Larynx</td>
<td>Papillomatosis, Ludwig’s Angina</td>
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<td>Larynx</td>
<td>Epiglotitis, vocal cord web</td>
<td></td>
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<tr>
<td>Larynx</td>
<td>Laryngospasm, angioedema</td>
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<td></td>
<td>Bilateral vocal cord paralysis</td>
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<tr>
<td>Trachea</td>
<td>Blunt trauma, Neoplasm</td>
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<td>Trachea</td>
<td>Web, stenosis, foreign body</td>
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<td>Trachea</td>
<td>Vascular anomalies, tracheitis</td>
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<td>Cyst, thyroid tumor, Vascular</td>
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<td>Endobronchial</td>
<td>Asthma, COPD, carcinoid syndrome</td>
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<td>Endobronchial</td>
<td>anomalies, tracheomalacia</td>
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<tr>
<td>Endobronchial</td>
<td>Congestive heart failure</td>
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<td></td>
<td>Asthma</td>
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<tr>
<td>Endobronchial</td>
<td></td>
<td></td>
<td></td>
<td>Tracheobronchitis</td>
<td></td>
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</tbody>
</table>
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

**PVCN**

Extrathoracic
Normal Aa gradient
No hypercapnea/acidosis
+ hypoxemia

**Asthma**

Intrathoracic
Elevated Aa gradient
+ hypercapnea/acidosis
+ hypoxemia
# Arterial-Blood-Gas Analysis

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>pH</th>
<th>PO₂</th>
<th>PCO₂</th>
<th>P(A-a)O₂</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>7.47</td>
<td>75</td>
<td>35</td>
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<td>2 +</td>
<td>7.40</td>
<td>89</td>
<td>39</td>
<td>11</td>
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<td>3</td>
<td>7.48</td>
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<tr>
<td>4</td>
<td>7.43</td>
<td>75</td>
<td>37</td>
<td>1</td>
</tr>
<tr>
<td>5</td>
<td>7.40</td>
<td>68</td>
<td>36</td>
<td>9</td>
</tr>
</tbody>
</table>

*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 PVCM.
• PFT’s with flow-volume loops have also been used to support the diagnosis of PVCM in symptomatic patients.
• Flow-volume loops of patients with PVCM 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
  – PVCM cannot be ruled out when asymptomatic.
    • if the patient is asymptomatic, negative laryngoscopic findings due not exclude the diagnosis
  – Absence of gagging or coughing during laryngoscopy
    • must not confuse PVCM with vocal cord motion produced by a laryngoscope induced gag reflex
Treatment

• The cause of the PVCM must first be elicited.
• In PVCM 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
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 PVCM
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>
<thead>
<tr>
<th>Acute treatment</th>
<th>Long-term treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Inhaled mixture of 70%-80% He/30%-20% (O_2)</td>
<td>Biofeedback training</td>
</tr>
<tr>
<td>CPAP</td>
<td>Supportive psychotherapy</td>
</tr>
<tr>
<td>IPPV</td>
<td>Speech therapy</td>
</tr>
<tr>
<td>Benzodiazepines</td>
<td>Hypnotherapy</td>
</tr>
<tr>
<td>Panting exercises</td>
<td>Panting exercises</td>
</tr>
<tr>
<td>Reassurance</td>
<td></td>
</tr>
</tbody>
</table>

*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 PVCM 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

• PVCM 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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