Paradoxical Vocal Fold Motion

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Grand Rounds Presentation
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Historical perspectives

• **1842**: Dunglison described a disorder of laryngeal muscles brought on by “hysteria” and named it “hysteric croup”.

• **1869**: Mackenzie first to visualize vocal cord adduction during inspiration in adults with stridor.
Historical perspectives

- 1902: Sir William Osler defined the condition by describing patients with “laryngeal muscle spasms during inspiration and times of great distress.”
- 1970’s: Patterson described a 33 year old woman with “Munchausen’s stridor”
  - During that time the disorder was considered purely psychogenic and confined to psychiatric literature.
- 1980’s: Christopher reported 5 patients with uncontrollable asthma and described the condition as “vocal cord dysfunction”.
Terminologies in the literature

- Vocal cord dysfunction
- Laryngeal asthma
- Functional upper airway obstruction
- Emotional laryngeal wheezing
- Irritable laryngeal syndrome
- Episodic paroxysmal laryngospasm
Introduction

- Paradoxical vocal fold motion (PVFM) is an involuntary functional disorder
- Episodic adduction of vocal folds during inspiration resulting in greater than 50% glottic closure
- Often misdiagnosed as asthma
Misdiagnosis

Morbidity resulting from misdiagnosis.

- Newman et. al. studied 95 patients with proven PVCM
- Asthma was misdiagnosed an avg. 4.8 years
- Unnecessary steroid treatments and hospitalizations.
• Failure to diagnose PVFM leads to unnecessary treatment and hospitalization
Epidemiology

- Overall incidence ranges from 3-10%
- Affects mostly children and young adults
  - Children: avg age – 14.5 years
  - Adult: 33 years
  - 2:1 Female to male ratio
  - 70-98% of patients were female Caucasian

- O’Connel et al, 164 patients
  - 20% of females who underwent laryngoscopy had PVFM.
The underlying etiology and origin of PVFM remain unclear
Theories

- **Inflammatory:**
  - Gastroesophageal reflux disease
  - Initial inflammatory response
  - Laryngeal hyperresponsiveness

- **Neuropathic:**
  - Superior laryngeal neuralgia (afferent)
  - Recurrent laryngeal neuralgia (efferent)
  - Alteration of autonomic balance
    - Medulla, midbrain, pre-frontal cortex
    - Reduction of threshold for glottic closure
Vocal cord function

- Posterior cricoarytenoid muscle (PCA) is the principal abductor of vocal cords
- During inspiration, glottic opening is controlled through medullary respiratory center which sends efferent signals to PCA via RLN.
- During expiration, there is a decrease in the tonic activity of the PCA muscle, resulting in a 10–40% narrowing of the rim glottides. This narrowing starts just before expiration and continues for almost 95% of the expiratory phase
Possible pathophysiology

- Ayers and Gabbott suggested that PVFM may be caused by laryngeal hyperresponsiveness initiated by an initial inflammatory insult and resulting in altered autonomic balance, which may be short lived or become persistent.

- If persistent, subsequent stimuli will induce local presynaptic reflexes causing airway narrowing, either at the glottic level or in patients with asthma in the lower airways.
glottis narrowing

inspiratory effort

reflexive adductor activation

powerful negative pressure in the trachea
Precipitating factors

- Airway irritants
- GERD
- Infection
- Extreme temperatures
- Physical exercise
- Emotional stressors
- Psychological factors
## Maschka Classification

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

- **Powell et al. (2000)**
  - 22 patients (aged ≤ 18) with PVFM
  - 21/22 patients with arytenoid edema or pachydermia on fiberoptic laryngoscopy

- **Loughlin et al. (1996)**
  - 12 adult patients with PVFM
  - 11/12 patients with evidence of LPR on fiberoptic exam
  - 10/12 with abnormal 24-hour pH probe study
  - All patients responded to anti-reflux treatment with complete cessation of PVFM
GERD and PVFM

TABLE III.
Number of Patients With Paradoxical Vocal Fold Motion With Normal and Abnormal Reflux Symptom Index Scores and Laryngopharyngeal Sensitivity Thresholds.

<table>
<thead>
<tr>
<th>Number of Patients</th>
<th>Number</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal RSI + normal LS</td>
<td>1</td>
<td>1.3</td>
</tr>
<tr>
<td>Normal RSI + abnormal LS</td>
<td>11</td>
<td>14.7</td>
</tr>
<tr>
<td>Normal LS + abnormal RSI</td>
<td>10</td>
<td>13.3</td>
</tr>
<tr>
<td>Abnormal RSI + abnormal LS</td>
<td>53</td>
<td>70.7</td>
</tr>
</tbody>
</table>

RSI = reflux symptom index; LS = laryngopharyngeal sensitivity.
Irritant-induced PVFM

- Airborne pollutants
- Chemicals
- Perfume
- GERD
- Sinusitis
- Post-nasal drip
Chronic cough and PVFM

Chronic Cough

PVFM

GERD
Airway Hyper-responsiveness
Rhinosinusitis
Causes of Chronic cough

- GERD/ LPR
- Rhinosinusitis
- Asthma
- ACE inhibitor
- COPD
- CHF
Psychiatric factors

- Psychiatric co-morbidities in 50-70 % patients
  - Mood disorders
  - Anxiety disorders
  - Somatoform disorders
  - Conversion disorders
  - Personality disorders
- Competitive sports
- History of sexual abuse
- Wartime
PVFM and Asthma

- Newman et al. (1995)
  - 95 PVFM patients
  - 56% with coexisting asthma

- National Jewish Center, 1994, multiple patients diagnosed with refractory asthma:
  - 10% had PVFM alone.
  - 30% had PVFM with coexistent asthma.
<table>
<thead>
<tr>
<th></th>
<th>PVFM</th>
<th>Asthma</th>
</tr>
</thead>
<tbody>
<tr>
<td>Incidence</td>
<td>Less common</td>
<td>More common</td>
</tr>
<tr>
<td>Age and sex</td>
<td>Young, female</td>
<td>Any</td>
</tr>
<tr>
<td>Triggers</td>
<td>Usually exercise or emotional stress</td>
<td>Many triggers</td>
</tr>
<tr>
<td>Psychological factors</td>
<td>Important role</td>
<td>Less important role</td>
</tr>
<tr>
<td>History of allergy</td>
<td>Usually absent</td>
<td>May be present</td>
</tr>
<tr>
<td>Family history</td>
<td>Usually absent</td>
<td>May be present</td>
</tr>
<tr>
<td>Sensation of tightness</td>
<td>Throat</td>
<td>Chest</td>
</tr>
<tr>
<td>Inspiratory stridor</td>
<td>More common, heard loudly over larynx</td>
<td>Rare</td>
</tr>
<tr>
<td>Sputum production</td>
<td>Rare</td>
<td>Common</td>
</tr>
<tr>
<td>Nocturnal awakening</td>
<td>Rare</td>
<td>Common</td>
</tr>
<tr>
<td>with symptoms</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Response to</td>
<td>No response</td>
<td>Good response</td>
</tr>
<tr>
<td>bronchodilators</td>
<td></td>
<td>Common</td>
</tr>
<tr>
<td>Hypoxemia</td>
<td>Rare</td>
<td>Common in allergic asthma</td>
</tr>
<tr>
<td>Eosinophilia</td>
<td>Rare</td>
<td>May show hyperinflation and peribronchial thickening</td>
</tr>
<tr>
<td>Chest radiograph</td>
<td>Usually normal</td>
<td></td>
</tr>
<tr>
<td>Residual volume</td>
<td>Normal</td>
<td>May be increased</td>
</tr>
<tr>
<td>Total lung capacity</td>
<td></td>
<td>Obstructive pattern</td>
</tr>
<tr>
<td>Flow volume loop</td>
<td>Flattening of inspiratory loop</td>
<td>Usually positive</td>
</tr>
<tr>
<td>Bronchial</td>
<td>May be positive</td>
<td>Usually positive</td>
</tr>
<tr>
<td>provocation test</td>
<td>Inspiratory adduction of the anterior 2/3 of vocal folds with posterior chink</td>
<td>Usually normal</td>
</tr>
<tr>
<td>Laryngoscopy</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
### Differential diagnosis of paradoxical vocal cord motion disorder

- Asthma
- Anaphylaxis
- Aspiration of foreign body
- Angioedema
- Chronic obstructive pulmonary disease
- Croup
- Epiglottitis
- Extrinsic airway compression
- Larygomalacia
- Laryngospasm
- Laryngeal tumour
- Spastic dysphonia/laryngeal dystonia
Diagnosis

- Clinical history
- Fiberoptic laryngoscopy
- Speech evaluation
- Pulmonary function test
- Methacholine challenge test
Symptoms

- Cough
- Dyspnea
- Dysphonia
- Stridor
- Wheezing
- Dysphagia
- Anxiety
Typical history

- Chronic refractory cough for months
- Frequent episodic attacks causing dyspnea
- Hoarse voice and fatigue
- Asthma-like symptoms refractory to steroid or bronchodilator
- Cough worsened with perfume, smoke, strong odor
- Diagnosis of GERD/ LPR and on PPI
- Athletes with choking sensation during exercise
Physical Exam

- Direct visualization via laryngoscopy while patient is having symptoms is gold standard for diagnosis.
- 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.
Laryngoscopy findings
Laryngoscopy

Normal

PVFM

Ibrahim et al. 2007 Postgrad Med J.
## Associated laryngeal pathology

<table>
<thead>
<tr>
<th>Laryngeal pathology</th>
<th>n</th>
</tr>
</thead>
<tbody>
<tr>
<td>Unilateral vocal fold motion impairment</td>
<td>4</td>
</tr>
<tr>
<td>Subglottic stenosis</td>
<td>3</td>
</tr>
<tr>
<td>Vocal nodules</td>
<td>1</td>
</tr>
<tr>
<td>Laryngomalacia</td>
<td>1</td>
</tr>
<tr>
<td>Sulcus vocalis</td>
<td>1</td>
</tr>
</tbody>
</table>
Pulmonary function test

- Decreased maximum inspiratory flow rate
- Truncated inspiratory limb of flow volume loop
- 70-80% of asymptomatic PVFM patients have normal PFT
Flow volume loop

Normal

PVFM
Methacholine challenge

- Cholinergic agonist causing bronchoconstriction
- Followed by pulmonary function test
  - Positive challenge when decrease of FEV1 > 20 % of baseline
- Asthma patients more sensitive to methacholine challenge than non-asthmatics.
- Although Methacholine may trigger PVFM, it is NOT a sensitive test for PVFM:
  - Perkins et al. shows only 2 out of 8 previously diagnosed PVFM developed PVFM after methacholine challenge.
Exercise-induced PVFM

- Often misdiagnosed as exercise-induced asthma
- Morris (1999): 15% of patients with exertional dyspnea have PVFM
- Occurs suddenly during increased physical activities
- Tense, shallow breathing with excessive tension at laryngeal level
- Rapid resolution once activities cease
- Patients learn to limit activities to avoid attacks
Diagnosis of exercise-induced PVFM

Tervonen et al. Laryngoscope July 2009
Management

- Medical
  - Acute vs. long-term
- Speech therapy
- Psychotherapy
  - Surgery – tracheostomy
Acute treatment

- **CPAP**
  - widen the rima glottidis and reduce turbulence
- **Panting**
  - physiologically increasing the glottic aperture
- **Benzodiazepines / Reassurance**
  - reduce anxiety and have been shown effective
- **Heliox**
  - Reduce turbulent flow since it is less dense than air
- **Intralaryngeal injection of botulinum toxin type A**
  - more invasive approach for severe exacerbation
Medical therapy

- Acid suppressive therapy
- Allergy treatments
- Sinusitis treatments
- Avoidance of irritants
- Stop unnecessary medication (eg. asthma Rx)
Inhaled anticholinergic agent

- Doshi et al. (2006)
  - Retrospective study
  - Ipratropium bromide (anticholinergic)
  - Vagus nerve autonomic hyperresponsiveness
  - Reduced exercise-induced PVFM in 49 patients

- No prospective study yet
Speech therapy

- Cornerstone of treatment
- 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
- Sullivan et al found that 95% of patients with PVFM treated with speech therapy were able to control their symptoms.
Psychotherapy

- Recognition of stressors
- Development of coping techniques
- Relaxation techniques for fear of toxic exposure
- Biofeedback training exercises
- Increase social support network
- Decrease dependency on family or the sick role
Breathing exercise

- Breathing with regular steady rhythm
- Exhaling against increased resistance by saying “sh” or “s”
- Patients learn to avoid inspiratory bursts that interrupt the normal respiratory pattern
Inspiratory muscle training

- Useful in treating exercise-induced PVFM
- One-way adjustable pressure valve
- Inhalation against increased resistance
- Increase muscle strength
- Decrease respiratory effort and struggle
- Generate sufficient pressure gradient
Improvement of cough after breathing exercise

A

SEVERITY OF COUGH

Pre - Therapy
Post - Therapy

SUBJECT

Female 1  Female 2  Female 3  Female 4  Male 1

1  2  3  4

0  1  2  3  4  5  6  7  8  9  10
Prognosis

• Long-term outcome unknown
  • most literature consists of case reports and retrospective studies.
• 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
Conclusions

- PVFM is often misdiagnosed as asthma
- Exact pathophysiology of PVFM is unclear
- Gold standard for diagnosis of PVFM is direct visualization via laryngoscopy
- Management is primarily medical with acid suppression therapy, speech and psychological evaluation.
Thank You