Cross Canada Round

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Case 1

HPI:

- 15 years old female was referred for Asthma difficult to control.
- Her main symptom is difficulty breathing during exercise
  - No improvement following short acting beta agonist administration

Examination:

- Normal

What is your differential diagnosis at this point?

Differential Diagnosis

Allergic
- Asthma
- Anaphylaxis

Psychogenic
- Anxiety disorder
- Depression, Munchausen syndrome & malingering
- Panic attack

Anatomical
- Tracheal stenosis
- Vocal cord paralysis
- Vocal cord polyps

Others
- Arrhythmia
- GERD
- Presence of foreign body

What investigations would you order at this point?

Investigations

- **Chest X ray.**
- **PFT:**
  - Spirometry.
  - Bronchodilator response.
  - Lung volumes.
  - MCT.
- **Exercises testing and flexible nasolaryngoscopy.**

Vocal Cord Dysfunction

Objectives case 1

- ✓ Case presentation.
- Define VCD and acknowledge the epidemiological part.
- Understand the pathophysiology.
- List the diagnostic test.
- Numerate the management strategies.

Definition

- Intermittent extrathoracic airway obstruction mainly during inspiration leading to dyspnea of varying intensity.

- Inappropriate adduction of the vocal cords during inhalation and sometimes exhalation.

Newman KB, Dubester SN. Semin Respir Crit Care Med 1994; 15: 161–167
Hoyte FC. Immunol Allergy Clin N Am. 2013;33(1):1–22. doi:10.1016/j.iac.2012.10.010

Historical Part

- 1842 as dysfunction of the laryngeal muscles sometimes seen in hysterical women.
- 1869 was first visualized during laryngoscopy by MacKenzie.
- 1974 VCD was described by Patterson in a 33 year old woman with 15 hospitalizations for what they termed “Munchausen’s stridor”

More than 70 terms have been used to describe
abnormal movement of the true vocal cords

Other names for VCD

- Laryngeal dyskinesia
- Vocal cord dysfunction (VCD)
- Inspiratory adduction
- Periodic occurrence of laryngeal obstruction (POLO)

- Munchausen’s stridor
- Episodic paroxysmal laryngospasm
  Psychogenic stridor
- Functional stridor
- Hysterical croup
- Emotional laryngeal wheezing

- Factitious asthma
- Pseudo-asthma
- Irritable larynx syndrome

Epidemiology

- Brugman and colleagues investigated 1530 patients with VCD and found that 65% were adults above 19 years of age.
- The median age was 36.5 years in adults and 14 years in pediatric patients.
- 2 – 3 : 1 Female predominance among patients with VCD.

Brugman S. The many faces of vocal cord dysfunction.. Am J Respir Crit Care Med 2003; 167: A58

Pathophysiology

- **Inhalation**:

The vocal cords abduct (open) widely , reaching a maximum width at midi nspiration.

- **Exhalation:**

The vocal cord movement varies significantly between individuals but generally adducts between 10 and 40 % from end inspiration.

Pathophysiology

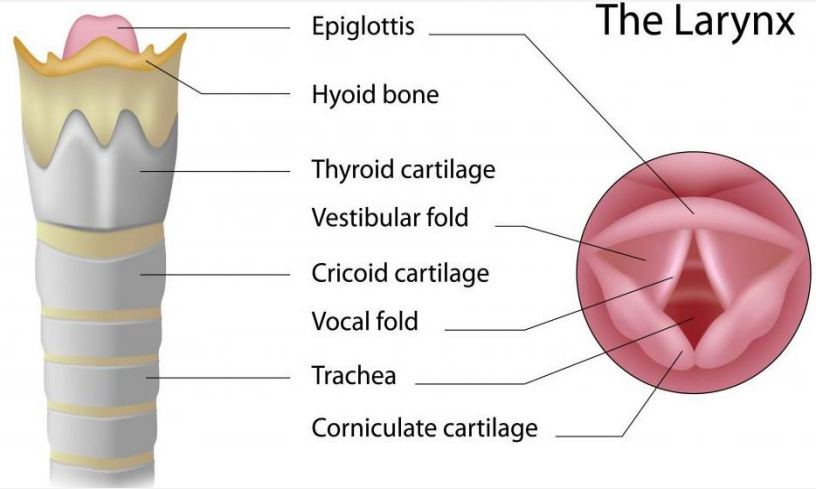
The 3 basic functions of the larynx are:

- Respiration
- Phonation

Can be initiated voluntarily

- Protection

Strictly reflexive and involuntary



Protection reflexes

- **Glottic closure reflex:**

- Mediated by the superior laryngeal nerve, the recurrent laryngeal nerve, and the vagal nerve.

- These nerves mediate closure of the 3 layers of the laryngeal structure.

- **Cough reflex:**

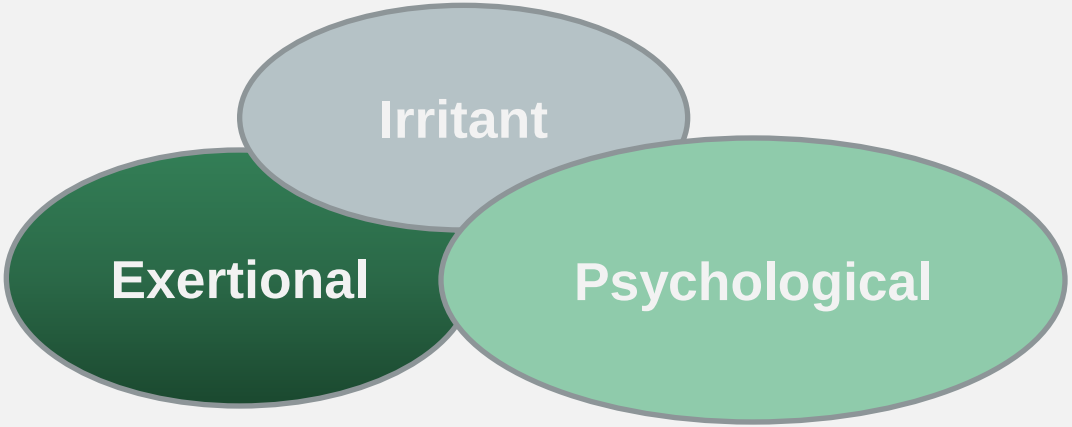
- Initiated by stimulating the sensory receptors.

- Cold, irritant, pressure and drive receptors.

Vocal cord dysfunction

Most likely due to laryngeal hyperresponsiveness:

- Increased sensitivity of the laryngeal receptors.
- Heightened response of the glottic and cough reflexes to a number of triggers.



Exercise

- Exertional VCD can be trigger by routine exercise or maximal activity.

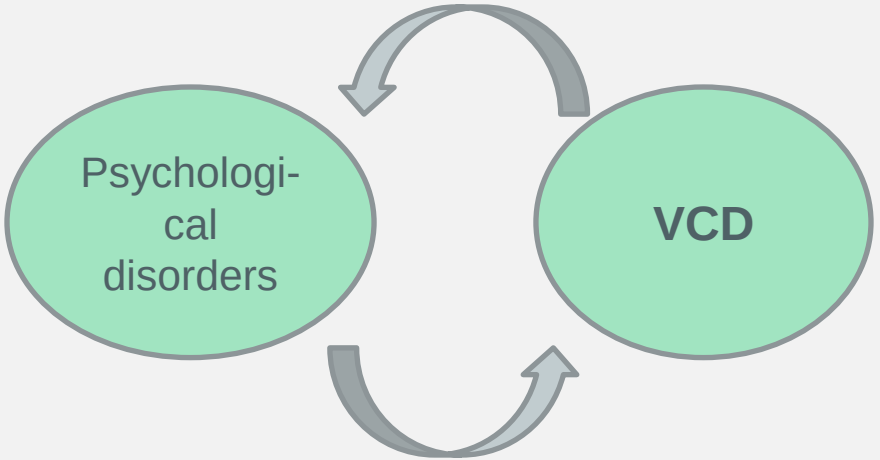
- A study by Morris et al., found that 12% of active duty military patients with exertional dyspnea had VCD triggered by exercise.

Morris MJ, Deal LE, Bean DR, Grbach VX, Morgan JA. Chest. 1999;116(6):1676–82.

Psychological triggers

It is still thought that psychological stimuli can trigger VCD.

- Anxiety disorder
- Stress
- Depression
- Somatoform disorder
- Conversion
- Disorder
- History of sexual abuse



Christopher KL, Morris MJ.Otolaryngol Clin N Am. 2010;43(1):43 66.doi:10.1016/j.otc.2009.12.002. viii.

Irritant triggers

- Laryngeal hyper-responsiveness could be triggered by different stimuli.

Intrinsic	**Other Diseases**
Gastroesophageal reflux disease Rhinitis	• Postnasal drip • Pharyngitis • Laryngitis • Sinusitis
Extrinsic	
Irritants Olfactory Visual stimuli	

Clinical Assessment

- **History:**

  - Throat or neck tightness
  - Shortness of breath or dyspnea
  - Sensation of choking or suffocation
  - More difficulty getting air in than out
  - Cough
  - Dizziness
  - Perioral or extremity numbness or tingling

- Questions should be directed to detect possible RFs including post-nasal drip and GER.

- Psychological assessment for any history of stress or abuse.

Clinical Assessment

- **History:**

TABLE 2 Differences and similarities of asthma and vocal cord dysfunction (VCD)

	Asthma	VCD
Time of onset	Quick (within minutes)	Sudden onset (within seconds)
Duration	Minutes to hours	Seconds to a few minutes
Dyspnoea during	Expiration	Inspiration
Area of limitation	Thorax, lower airways	Throat
Inhaled drug therapy	Highly effective	Often ineffective, aggravating
Induced by	Irritants, allergens, exercise	Irritants, stress, exercise

K. Kenn, R. Balkissoon Vocal cord dysfunction: what do we know? Eur Respir J. 2011 Jan; 37(1): 194–200.

Differential Diagnosis

Allergic
- Asthma
- Anaphylaxis
- Angioedema

Anatomical
- Tracheal stenosis
- Vocal cord paralysis
- Vocal cord tumors or polyps
- Laryngomalacia

Infectious
- Croup
- Epiglottitis

Psychogenic
- Somatoform disorder
- Conversion disorder
- Abuse
- Anxiety disorder, Depression, Munchausen syndrome & malingering

Others
- GERD
- Hypoparathyroidism
- Presence of foreign body1

Physical exam

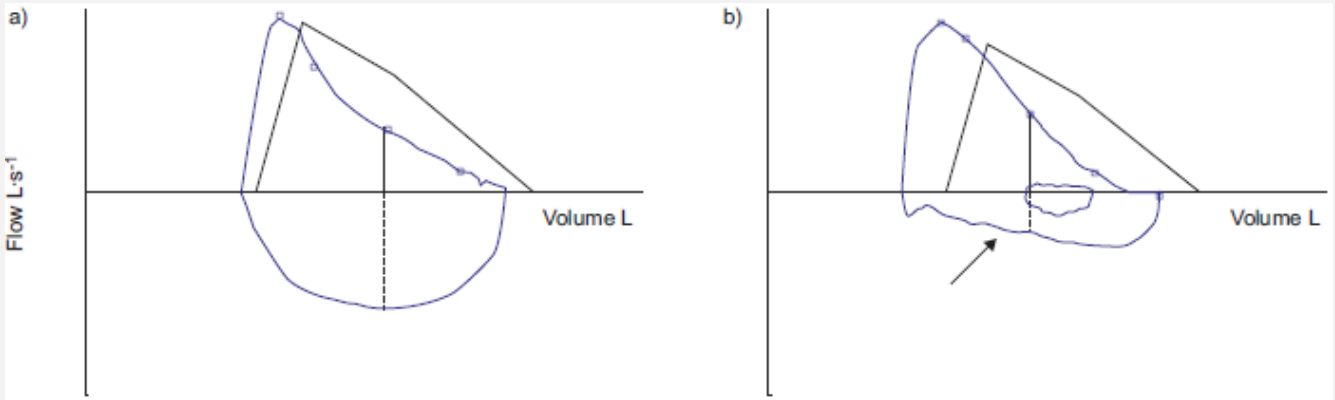
- At the time of an acute attack:

  Tachypnea or hyperventilation
  Stridor or wheezing
  Neck or chest retractions
  Pallor but no cyanosis
  Hoarseness or dysphonia

- One should also look for stigmata of posterior nasal drainage.
- GER (epigastric tenderness and halitosis).

Pulmonary function testing

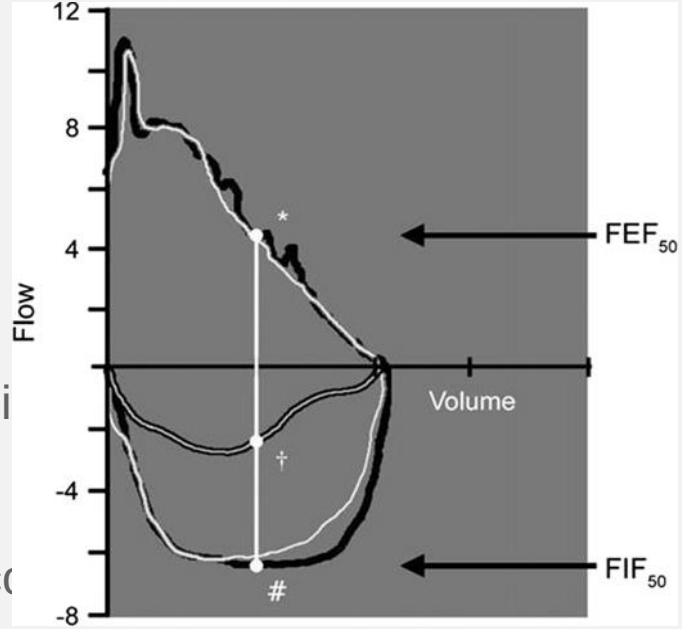
- Morris et al. reviewed 1500 cases of VCD in the published literature and found 28 % of reported VCD patients had flow-volume loop truncation on spirometry.



Morris MJ, Allan PF, Perkins PJ. Vocal cord dysfunction: etiologies and treatment. Clin Pulm Med. 2006;13(2):73–86.

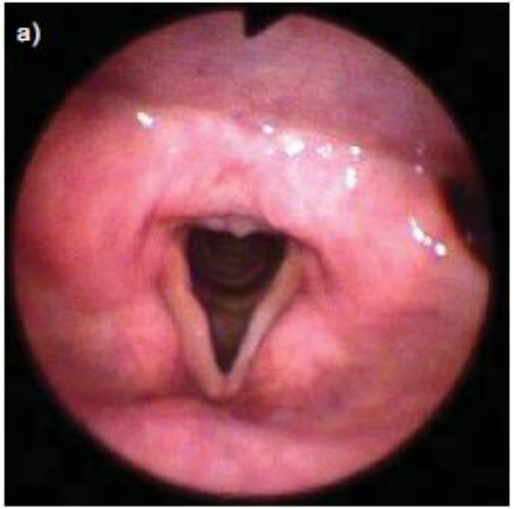
Pulmonary function testing

- FEF50/FIF50 ratio:
  - < 1 in normal individuals
  - > 1 in VCD

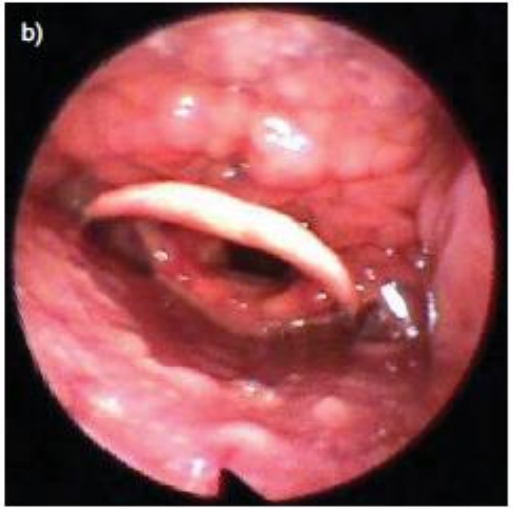


- The ratio is usually > 1 because truncation of the i
  FIF50.
- The ratio may be difficult to interpret in VCD and co
  obstruction or comorbid asthma.

Flexible laryngoscopy



Aryepiglottic fold swelling



Posterior phayngeal wall cobblestoning

Vocal fold nodules.

Broncho-provocation challenge

- Performing a laryngoscopy immediately after a bronchial challenge can help determine VCD.
- Gold standard.

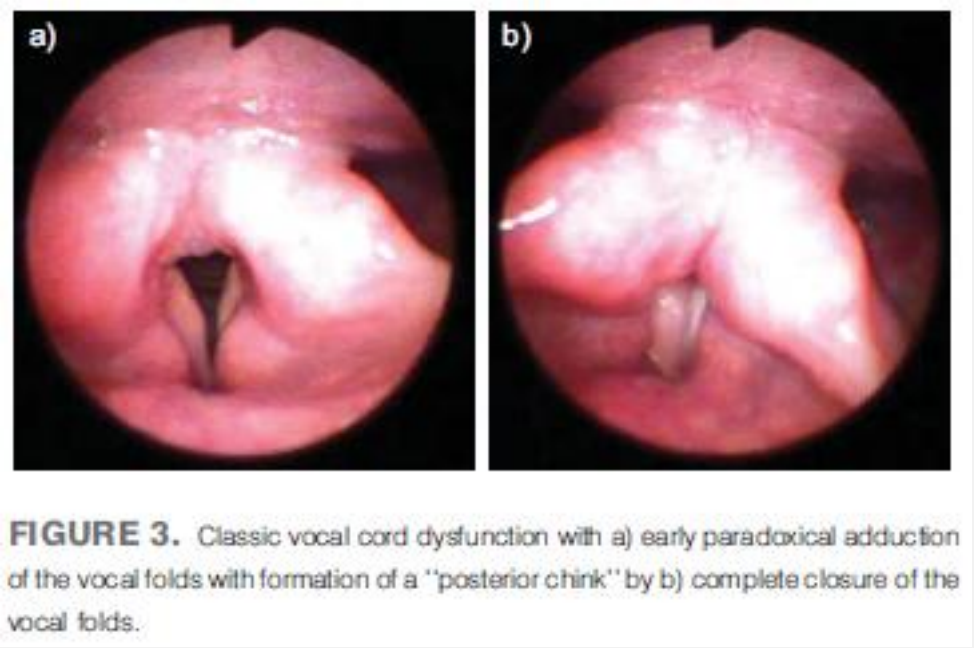


FIGURE 3. Classic vocal cord dysfunction with a) early paradoxical adduction of the vocal folds with formation of a ''posterior chink'' by b) complete closure of the vocal folds.

Management of VCD

- Team approach
- Patient education

Acute episode

- Reassure patient
- Instruct patient in breathing exercises.
  - Panting.
  - Diaphragmatic breathing.
  - Breathing through a straw.
  - Sniffing in during inhalation and exhaling through pursed lips.

Heliox

- Reduce air density.
- Decreases turbulent flow.
- Reduces work of breathing.
- Shown favorable responses in acute VCD episodes and has even demonstrated a sustained response after discontinuation.

Reisner C, Borish L. Heliox therapy for acute vocal cord dysfunction. Chest 1995; 108: 1477.

Other therapies

- Sedation and intubation.
- Continuous positive airway pressure ventilation.
- Topical lidocaine applied to the larynx.
- Superior laryngeal block with Clostridium botulinum toxin.
- Tracheotomy

Maillard I, Schweizer V, Broccard A, et al. Chest 2000; 118: 874–877.

Chronic Management

- Speech therapy and psychotherapy.
- Clinicians should discontinue unnecessary medications, such as bronchodilators and steroids, if coexistent asthma has been ruled out.
- Treatment of associated disease such as GERD or rhinosinusitis may reduce symptoms.

Therapy Monitoring

- Patient progress can be followed clinically or with more objective approaches, such as the VCDQ which has shown improvement in scores after speech therapy.

Table 2:The 12-item VCDQ questionnaire

Question	Disagree strongly	Disagree	Neither agree nor disagree	Agree	Agree strongly	Score
	1	2	3	4	5	
1. My symptoms are confined to my throat/upper chest						
2. I feel like I can't get breath past a certain point in my throat/upper chest because of restriction						
3. My breathlessness is usually worse when breathing in						
4. My attacks typically come on very suddenly						
5. I feel that there is something in my throat that I can't clear						
6. My attacks are associat with changes in my voice						
7. My breathing can be noisy during attacks						
8. I'm aware of other specific triggers that cause attacks						
9. My symptoms are associated with an ache or itch in my throat						
10. I am frustrated that my symptoms have not been understood correctly						
11. I am unable to tolerate any light pressure around the neck, e.g. tight clothes or bending the neck						
12. The attacks impact on my social life						
Total						(12–60)

Fowler SJ, Thurston A, Chesworth B, Cheng V, Constantinou P, Vyas A, et al. Clin Exp Allergy. 2015. doi:10.1111/cea.12550.

Learning points

- VCD is an important differential diagnosis of asthma, that is widely unrecognized.
- Can lead to high medical utilization, unnecessary medications use and other dangerous consequences.
- Suspect VCD in patients with asthma-like symptoms that do not respond to conventional asthma therapy or induced by exercise or stress.
- The gold-standard test for diagnosis is direct visualization of the vocal cords by laryngoscopy.

Refrences

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Any questions?

Case 2

Case 2

HPI:

- 5y old boy previously healthy presented with fever, vomiting, hypoxemia and consolidative process in right middle lobe
- Ongoing hypoxemia and right middle lobe opacity

What is your differential diagnosis?

Physiological causes of hypoxemia

Hypoventilation

Pulmonary hypoplasia.
Central causes of hypoventilation.
Neuromuscular disease.
Chest wall restriction.
Sedative agents.

Ventilation–perfusion mismatch

Airways obstruction (asthma)
Bronchiectasis.
Pulmonary oedema/haemorrhage.
Pulmonary parenchymal disease.
Pulmonary hypertension.

Right-to-left shunt

A- Intracardiac shunt:

CHD (5Ts).
Acyanotic lesions.

B. Intrapulmonary shunt:

Pulmonary (AVM)/ HHT
Hepatopulmonary syndrome.

Barrier to diffusion

Interstitial lung disease.
Interstitial oedema.
Pulmonary haemorrhage.
‘False’ hypoxaemia.
Haemoglobinopathies.

Bola SS, Narang I. 2014;2014:bcr2013201819. doi:10.1136/bcr-2013-201819.

What do you want to do next?

Investigations

- Chest x ray
- CT scan
- Blood gas
- Echocardiogram
- Bronchoscopy

Patient diagnosed with a left superior vena cava draining directly into the left atrium

- Referred to Cardiology.

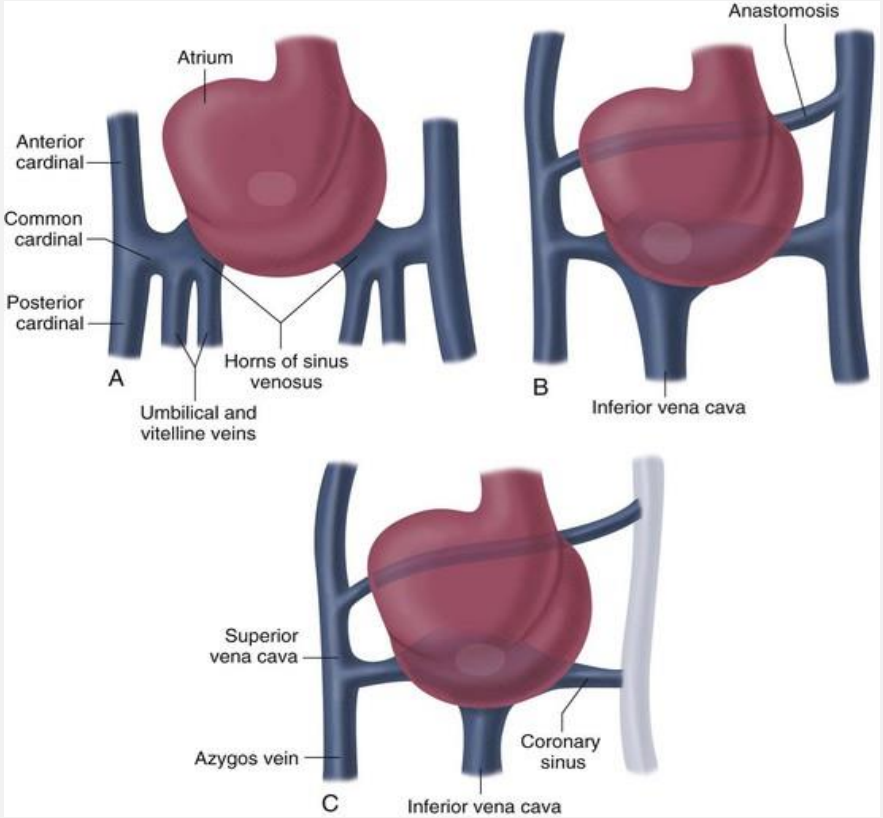
Objectives

- ✓ Case review.
- Review the epidemiology persistent left-sided superior vena cava (LSVC).
- Understand the basic embryologic origin of LSVC.
- Numerate the implications of having PLSVC.
- List the treatment options.

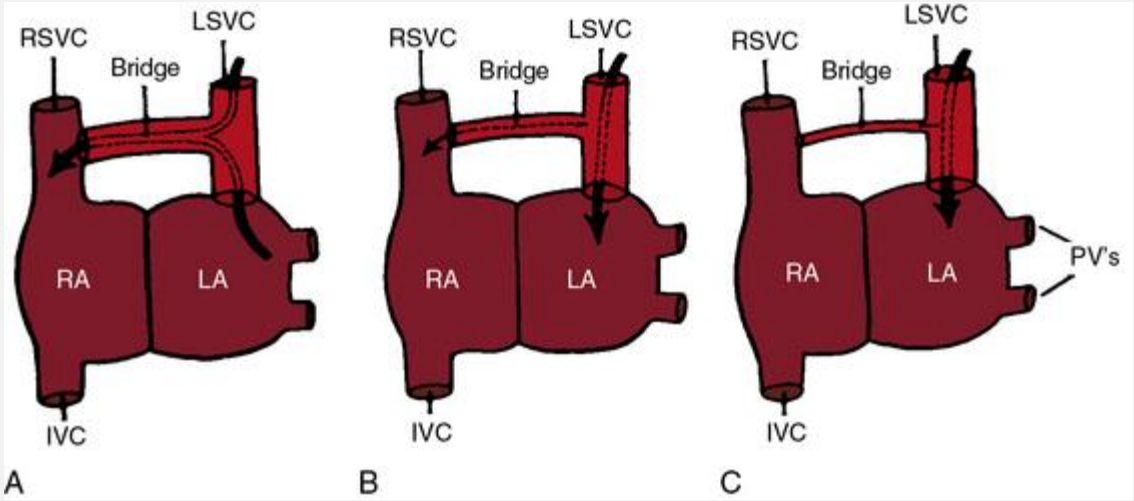
Persistent left-sided superior vena cava (LSVC)

- The common variant of systemic venous drainage.
- It is present in 0.5% of the general population and Up to 10% of those with established congenital heart disease.
- It results from failure of obliteration of the left common cardinal vein, and typically drains into the right atrium via the coronary sinus (CS) in 80-90%.
- LSVC drains into the left atrium (LA) in 10-20%.

Persistent left-sided superior vena cava (LSVC)



Persistent left-sided superior vena cava (LSVC)



Why should we be worried?

1- Right to left shunt would cause desaturation and exercise limitation.

2-Embolic complications:

- Stroke.
- Brain abscesses.

Other implications

- A PLSVC can cause problems during establishing central venous access (catheterization of the CS can cause hypotension, angina, perforation of the heart, tamponade and arrest).
- Pacemaker implantation (due to the tortuous course of the electrode, it can be difficult to fix the electrode).
- Cardiopulmonary bypass (isolated PLSVC impairs the use of retrograde cardioplegia).

Diagnosis

- Echocardiogram.
- Contrast Echo (agitated saline).
- Contrast CT.

Treatment

Treatment for a persistent L-SVC is most often performed by an open surgical procedure.

- Ligation of the L-SVC and over-sewing the atrial side.
- The L-SVC can be transposed to the right atrium or left pulmonary artery.
- The L-SVC can be connected to the right atrium via the coronary.

Learning points

- L-SVC can lead to hypoxemia and embolic phenomenon.
- Diagnosis of persistent L-SVC requires a high index of suspicion .
- Identification of this anomaly is important, so therapeutic measures can be taken to reduce/eliminate the risk of complications.

References

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