Case Presentation

Congenital and structural HD team
Presented by
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History & Physical examination

- a 7-years-old girl patient presented with dyspnea on exertion (NYHA FC III) and palpitation.
- O/E
  - General examination
    - underweight
  - Local cardiac examination
    - reveal apex shifted downwards & outwards, hyper dynamic, LV S3 gallop, a grade-III continuous murmur most loudly at the Rt sternal border.
CXR

What is the explanation?
The coronary sinus is defined as the blood conduit that is a continuation of the great cardiac vein from the valve of the great cardiac vein to the ostium of the coronary sinus.

The length varies from 3 to 5.5 cm. CS lies in the sulcus between the left atrium and ventricle.

The CS receives blood from the ventricular veins during ventricular systole and empties into the right atrium during atrial systole.

The Thebesian valve is a crescent shaped structure often found guarding the mouth of the CS as it opens to the right atrium.

**Coronary Sinus anomalies**

- An Absent coronary sinus is always associated with a persistent left superior vena cava (PLSVC) connecting to the left atrium.
- A Hypoplastic coronary sinus
- Atresia or stenosis of the coronary sinus ostium
- Enlargement of the coronary sinus
- Unroofed coronary sinus anomaly
Causes of dilated CS

With shunt:
- Low pressure shunt
  partial anomalous pulmonary venous drainage
- High pressure shunt
  coronary artery – CS fistula

Without shunt:
- Persistent LSVC
  Contrast with agitated saline

No evidence of Persistent LSVC
Partial anomalous pulmonary venous drainage – CS

Coronary artery – CS fistula

Low pressure shunt

High pressure shunt

Cardiac MSCT

Max PG = 60 mmHg

CW Doppler
MSCT

Large serpentine
Coronary sinus fistula

- Coronary artery fistula is a condition in which a communication exists between one or two coronary arteries and either:
  1. A cardiac chamber
  2. The coronary sinus
  3. The superior vena cava
     - The pulmonary artery
     - Coronary anomalies may be abnormalities of origin, distribution or termination.

CAVF are considered mainly to be termination abnormalities

<table>
<thead>
<tr>
<th>Origin</th>
<th>Frequency (%)</th>
<th>Drainage</th>
<th>Frequency (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>RCA</td>
<td>50-60</td>
<td>RV</td>
<td>14-40</td>
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<tr>
<td>LAD</td>
<td>25-42</td>
<td>RA</td>
<td>19-26</td>
</tr>
<tr>
<td>Both</td>
<td>5</td>
<td>LV</td>
<td>2-19</td>
</tr>
<tr>
<td>CX</td>
<td>18.3</td>
<td>PA</td>
<td>15-20.2</td>
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<tr>
<td>Diagonal</td>
<td>1.9</td>
<td>CS</td>
<td>7</td>
</tr>
<tr>
<td>Marginal</td>
<td>0.7</td>
<td>LA</td>
<td>5-6</td>
</tr>
<tr>
<td>Single coronary</td>
<td>3</td>
<td>SVC</td>
<td>1</td>
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</table>
The involved coronary artery is dilated because of increased blood flow and is often tortuous to an extent determined by the shunt volume.

Generally, the symptoms develop depending on the amount of the left-to-right shunt or the presence of coronary steal phenomenon of the fistulae, which usually present in young adults with angina (3–7%), exertional dyspnoea (60%), endocarditis in the fistula (20%), syncope, palpitations, myocardial ischemia and infarction, and manifest in older adults with congestive heart failure (19%), atherosclerosis, and cardiac arrhythmias.
What is the Plan?

Surgical closure

- Recommended
  - large and tortuous,
  - distal

- Difficult
  - the multiplicity of distal coronary artery-to-CS connections
  - the location of the connection on the posterior base of the heart

Transcatheter closure
Operative details

- Opening of the RA and closing the fistula by direct suture
- Closing PFO
- Closing of the RA

Follow-up

- ASA 75 mg/day life-long
- Prophylaxis against infective endocarditis
- Follow up Echo
Post operative Echo

6 months Post operative

THANK YOU