Scimitar Syndrome: 
The Curved Turkish Sabre

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Q20. Which of the following is the most unique component of Scimitar Syndrome?

a. Partial or complete pulmonary venous drainage abnormality
b. Curved anomalous right pulmonary vein draining into the inferior vena cava
c. Hypoplasia of the right lung
d. Hypoplasia of the right pulmonary artery
Q21. Which one of the following is the most commonly associated cardiovascular lesion to Scimitar Syndrome?

a. Atrial septal defect
b. Scimitar vein stenosis
c. Patent ductus arteriosus
d. Ventricular septal defect
A 27 years old female with history of asthma presented with fever and dyspnea

Patient gave history saying that her heart is on right side

So, a CXR was taken......
Layering right pleural effusion
Shift of the heart to the right
Subsegmental atelectasis
Dextocardia
Anomalous return of pulmonary veins
Right-sided aortic arch
Right lower lobe consolidation

Which finding(s) is(are) present?
A rare association of complex congenital cardiopulmonary anomalies

- 1-3 per 100,000 live births
  - Incidence might be higher

- Reported in 3 to 6% of patients with PAPVC

Scimitar Syndrome
Synonyms

- Pulmonary venolobar
- Hypogenetic lung syndrome
- Halasz’s syndrome
- Mirror image lung syndrome
- Epibronchial Rt pulm artery syndrome
- Vena cava bronchovascular syndrome

Scimitar syndrome
An anomalous vein connects between the pulmonary venous circulation and systemic venous circulation. This could be either partial (PAPVC) or total (TAPVC). The syndrome associated with PAPVC is more commonly known as "Scimitar syndrome"
The vein broadens as it curves downward, resulting in configuration resembling a TURKISH SWORD, SCIMITAR (KILIÇ)
“The scimitar vein is the sine qua non of this syndrome"
- Total or partial anomalous connection of the pulmonary veins to the inferior vena cava. This connection can be above or below the level of the diaphragm. Right lung is commonly involved.
- Variable degree of hypoplasia and malformation of the pulmonary arteries and lung in the same side.
- Often presence of aortic-pulmonary artery collateral arteries to the hypoplastic lung.

Components
- Abnormal bronchial supply to the right lung with sequestration
- Dextroposition
- Occasionally, there are 2 or more veins
- Anomalous venous drainage may involve entire / part of lung
- The anomalous vein can also drain into right atrium, portal vein

Components
19 to 75% of patients have associated congenital cardiovascular abnormalities

Most commonly **ASD, PDA, VSD, CoA, PA**

- Ipsilateral diaphragmatic anomalies
- Horse-shoe lung and localized bronchiectasis
- Vertebral anomalies
- Genitourinary tract abnormalities

**Associations**
- **Infantile form (n=25)**
  - Severely ill-High mortality (64% vs 3.2%)
  - Pulmonary hypertension
  - Large left to right shunt
  - Associated congenital heart defects
- **Childhood/Adult form (n=122)**
  - Recurrent pulmonary infections
  - Asymptomatic (incidental diagnosis)
  - Near normal PA pressures
  - ASD present in 20%
  - A-P artery absent in 50%

**Scimitar syndrome**

Dupuis C, et al. Am J Cardiol 1992;70:502-7*
- Increased PBF from the anomalous pulmonary vein
- Presence of systemic arterial supply to the right lung
- Reduction of the pulmonary vascular bed
- Stenosis of the anomalous PV
- Associated congenital cardiac malformations
- Persistent pulmonary hypertension of the newborn

Pulmonary Hypertension
Case 1

- **Infantile form**
  - 2 months old baby girl
  - 3,4 kg
  - Respiratory distress and feeding difficulty
  - Mild cyanosis (Sat 87%)

Scimitar syndrome
Case 1
Case 1

PAP 84/32/ 56 mmHg

Aorta 70/41/ 54 mmHg

Cardiac Cath
Case 1
Case 1  Obstruction

Left PVp mean 26 mmHg

Right PVp mean 16 mmHg
Case 1  Aorta-Pulmonary Collaterals
Case 1

What is your recommendation?

- Optimize medical treatment and follow
- Lobectomy
- Urgent corrective surgery
- Coil embolization of A-P collaterals and plan for a later surgical correction

Diagnosis: Bilateral Scimitar
Case 1
Case 1

- Improvement of symptoms
- Medical treatment optimized
- Pulmonary pressures still systemic
- Discharged home with plans for later surgery
- *Died at home 1 month after coil-embolization*

Post-procedure follow-up
Case 2

- **Infantile form**
  - 18 months old baby boy
  - 8.6 kg
  - Referred for Pulmonary atresia repair
  - Cyanosis (Sat 70%)

**Scimitar syndrome**
Case 2

Scimitar syndrome

- **Echo**
  - Pulmonary atresia
  - VSD
  - PDA/MAPCA
  - PAPVC
  - Hypoplastic RPA

- **CT**
  - Pulmonary atresia
  - Hypoplastic PAs
  - 2 A-P collaterals from abdominal aorta
  - R PVs to IVC
  - Hypoplastic R Lung
Case 2
Case 2
Case 2

Coil occlusion
Case 2

- Coil occlusion of the collateral during the time of cath
- Reimplantation of the Scimitar vein with a 5 mm PTFE tube
- Central shunt with a 4 mm PTFE tube
- Stormy postoperative course with an ICU stay of 39 days
- Discharged home @ POD 54
- Cardiac cath @ 3 months PA pressure systemic
Case 3

- **Adult form**
  - 27 years old female
  - Evaluated for bronchial asthma
  - Otherwise asymptomatic
  - **Echo**
    - Dextroposition
    - PAPVC
    - PFO

**Scimitar syndrome**
Case 3

- Normal pregnancy and delivery
- Asymptomatic currently
- Yearly follow-up by cardiology
  - *Echo Qp/Qs < 2.0*
  - *Normal RV size*
- **CXR**
  - Enlarged curved vascular structure coursing medially toward the R. diaphragm; this structure enlarges in diameter as it approaches diaphragm
  - Dextroposition of the heart
  - Hypoplasia of right lung

- **ECHO**
  - Scimitar vein, associated cardiac anomalies, PHT

- **CT**

- **MRI**

**Diagnostic Investigations**
CT and MRI

- Ideal - Wide field of view, excellent spatial orientation, unambiguous 3-D delineation of the course, connection & drainage
- Phase contrast cine MRI – quantification of PV blood flow, determination of Qp/ Qs, flow to the rt or left lungs

Diagnostic Investigations
- Cardiac Catheterization & Angiography
  - Definitive but not always necessary
  - PVR, Shunting, Scimitar stenosis
  - Opportunity
    - for embolization of S-P collaterals
    - Balloon dilatation of any pulmonary vein stenosis

Diagnostic Investigations
- Scimitar sign is simulated radiographically by hypoplasia of Rt. lung with aplasia of pulmonary artery
- Abnormal artery / arteries can simulate scimitar vein
- Abnormal meandering vein draining a hypoplastic Rt. lung to Lt atrium can simulate scimitar vein

Differential Diagnosis
- Disconnected bronchopulmonary mass with systemic arterial supply
- Scimitar a variant of sequestration?

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Scimitar Syndrome</th>
<th>Pulmonary Sequestration</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mediastinal Shift</td>
<td>Ipsilateral</td>
<td>Contralateral</td>
</tr>
<tr>
<td>Lung Involvement</td>
<td>Hypoplasia</td>
<td>Focal Consolidation</td>
</tr>
<tr>
<td>Systemic Arterial Supply</td>
<td>Less Common</td>
<td>More Common</td>
</tr>
<tr>
<td>Abnormal Venous Drainage</td>
<td>More common, typically IVC</td>
<td>Less Common</td>
</tr>
</tbody>
</table>

**Differential Diagnosis**
Surgical Indications

**Infantile Form**
- Failure to control symptoms
- Presence of pulmonary hypertension

**Adult Form**
- Recurrent lung infections
- $Qp/Qs > 2$ asymptomatic patients
  - Avoid sequela of volume overload
Surgical Indications

Infantile Form

- Failure to control symptoms
- Presence of pulmonary hypertension

PREOPERATIVE
Pulmonary Overcirculation → Occlusion of Systemic Arteries
- Redirect the anomalous pulmonary venous drainage to left atrium (REROUTING)
- Resection of the right lung
  - Pneumonectomy
  - Lobectomy
- Ligation of abnormal systemic arteries
- Repair of intra-cardiac lesions

Surgical Treatment
▪ Redirect the anomalous pulmonary venous drainage to left atrium (REROUTING)
  ▪ Baffling via a tunnel
  ▪ Direct re-implantation to left atrium

Surgical Treatment
Baffle Procedures

Ulf Gudjonsson, John W. Brown
Baffle Procedures

- **Zubiate**
  - Long tunnel predisposes to thrombosis
    - Inability to fix SV stenosis
    - Need for DHCA

- **Schumaker and Judd**
  - More direct flow to LA
    - SV stenosis can be addressed

- **Calhoun and Mee**
  - SV flow baffled through a relatively gentle angle
Re-implantation Procedures


- No need for CPB
- No baffles
- No tension or kinking

WARNING: Beware of the tendency of venous anastomoses to constrict with growth!

*Lam et al. 133:573-574*
Lung Resection

- **Lobectomy**
  - Difficult and seldom possible due to abnormal lobation of the hypoplastic lung

- **Pneumonectomy**
  - Anomalous vein traveling posterior to the hilum
  - Diffusely scarred bronchiectatic lung
  - Scoliosis, respiratory insufficiency, postpneumonectomy syndrome
Lung Resection

Indications

- Weight < 5kg
- Severe PHT
- Hypoplastic lung
- Sequestrated lung
- Entire drainage of lung through a single vein
- Recurrent RTI

Table 2. Operations and outcome of 6 patients with scimitar syndrome

<table>
<thead>
<tr>
<th>Patient</th>
<th>Procedure</th>
<th>Follow-Up</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Right pneumonectomy</td>
<td>Doing well, asymptomatic</td>
</tr>
<tr>
<td>2</td>
<td>Right lower lobectomy</td>
<td>Doing well, asymptomatic</td>
</tr>
<tr>
<td>3</td>
<td>Right pneumonectomy</td>
<td>Doing well, asymptomatic</td>
</tr>
<tr>
<td>4</td>
<td>Right pneumonectomy</td>
<td>Died after 2 days</td>
</tr>
<tr>
<td>5</td>
<td>Ligation of anomalous</td>
<td>PA supplying RLL on</td>
</tr>
<tr>
<td></td>
<td>vessels</td>
<td>CT angiogram</td>
</tr>
<tr>
<td>6</td>
<td>Died before surgery</td>
<td></td>
</tr>
</tbody>
</table>

CT = computed tomography, PA = pulmonary artery, RLL = right lower lobe.
SCIMITAR SYNDROME: TWENTY YEARS' EXPERIENCE AND RESULTS OF REPAIR

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Received for publication May 6, 1996 Revisions requested June 6, 1996; revisions received June 21, 1996 Accepted for publication June 24, 1996. Address for reprints: William G. Williams, MD, Chief, Cardiovascular Surgery, Division of Cardiovascular Surgery, The Hospital for Sick Children, 555 University Ave., Toronto, Ontario M5G 1X8, Canada.

Abstract

Background: Thirty-two patients with scimitar syndrome were seen in the period between 1975 and 1995. There were 11 male and 21 female patients. Median age at diagnosis was 7 months (mean 7.7 years, range 1 day to 70 years). Patients in whom the diagnosis was made during the first year of life (infantile group, n = 19) had more severe symptoms and had a higher incidence of heart failure (11/19 vs 0/13) and of pulmonary hypertension (11/19 vs 1/13) than did the patients in whom the diagnosis was made after age 1 year (adult group, n = 13). In 17 patients the anomalous pulmonary venous drainage was repaired by baffling the vein to the left atrium. The median age at this operation was 5.8 years (mean 14.8 years, range 6 months to 70 years).

Results: No deaths occurred in this surgical group during a mean follow-up period of 8.9 years (range 1.6 to 17 years). Eight patients (47%), however, had evidence of pulmonary venous stenosis after repair, and two required reoperation for pulmonary venous obstruction. All six children in the infantile group had postoperative pulmonary venous stenosis, compared with two of 11 older patients. Postoperative quantitative pulmonary perfusion scans performed in 15 patients demonstrated reduced flow to the right lung (24%, range 0% to 59%).

Conclusion: We conclude that age at detection of scimitar syndrome is important in predicting outcome. Surgical repair seldom results in normal blood flow to the right lung but abolishes left-to-right shunt. Postoperative pulmonary venous obstruction is prevalent, especially in the infants. (J THORAC CARDIOVASC SURG 1996;112:1161-69)
Age at detection is important predicting outcome (26 % Mortality)

- Postoperative pulmonary venous obstruction is prevalent, especially in infants (100% vs 18%)

- Surgical repair seldom results in normal blood flow to the right lung but abolishes left to right shunt

<table>
<thead>
<tr>
<th>Surgical Repair (n=17)</th>
<th>Infantile Group (n=6)</th>
<th>Adult Group (n=11)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Postrepair pulmonary vein stenosis</td>
<td>6</td>
<td>2</td>
</tr>
<tr>
<td>CNS complications</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>Perfusion scan of right lung (%)</td>
<td>19</td>
<td>27</td>
</tr>
</tbody>
</table>
Management of children with scimitar syndrome is complicated by an exceptionally high incidence of postoperative pulmonary venous obstruction and abnormally diminished perfusion of the right lung.
Freedom from pulmonary vein stenosis for patients with PAPVC stratified by type: scimitar versus non-scimitar types.

Bahaaldin Alsoufi et al.
Scimitar Syndrome
A European Congenital Heart Surgeons Association (ECHSA) Multicentric Study

Vladimo L. Vida, MD, PhD; Massimo A. Padalino, MD, PhD; Giovanna Boccuzzo, MPH; Erjon Tarja, MD; Hakan Berggren, MD; Thierry Carrel, MD; Sertac Cicik, MD; Giancarlo Crupi, MD;

Intraatrial baffle repair 38
Reimplantation 21
Pneumonectomy/Lobectomy 9
Preop Coil embolization 6

Conclusions—The surgical treatment of this rare syndrome is safe and effective. The majority of patients were asymptomatic at the follow-up control. There were a relatively high incidence of residual scimitar drainage stenosis that is similar between the 2 reported corrective surgical techniques used. (Circulation. 2010;122:1159-1166)

Key Words: congenital heart disease ■ multicenter study ■ scimitar syndrome
Indications

- Severe right lung hypoplasia
- Entire drainage of lung through a single vein
Figure 1. Kaplan-Meier estimate of overall survival according to the presence of pulmonary arterial hypertension (PAH).
Freedom from scimitar drainage stenosis after surgical repair

Median follow-up 4.5 years (1.6 to 8.3 years)

Vida VL et al. Circulation 2010;122:1159-1166
Scimitar syndrome is not a simple and benign congenital heart disease

Associated cardiac anomalies play an important role in the long-term results

Presence of pulmonary hypertension is associated with higher mortality rate

Conclusion
Both baffle repair and direct re-implantation of the scimitar vein are used according to surgeon preference and to the anatomic and pathological features of each case.

Still, there is no consensus as to which is the best surgical treatment option.

**Conclusion**
In memory
Hasan Solak, BSc
Chief Perfusionist

1966- 9 April 2013