Partial anomalous pulmonary venous connection (PAPVC)

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No disclosures
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Definition

• In partial anomalous pulmonary venous connection, one or more, but not all pulmonary veins drain directly either into a systemic vein or into the right atrium

• In 90% of cases the abnormal drainage is right-sided
• In 20% of pts. an entire lung (either right or left) is involved
1. Partial anomalous pulmonary venous drainage
The pulmonary veins connect normally and directly to the left atrium; PAPVC is caused by abnormal atrial septation draining the oxygenated blood from the anomalous vein(s) either exclusively into the right atrium or into both the left and right atrium.

- Sinus venosus defects: deficiency of the wall that normally separates the right upper [rarely the right lower] pulmonary vein from the superior vena cava (‘unroofing’)
1. Partial anomalous pulmonary venous drainage

- Malposition of the septum primum: leftward shift of the posterior and/or superior attachment of the septum primum resulting in right pulmonary veins draining into the right atrium rather than into the left atrium.

2. True anomalous pulmonary venous connections

- The anomalous pulmonary vein(s) directly connect(s) with a systemic vein (rarely the right atrium)
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~ 85%

~ 15%

Anomalous drainage with normal position of pulmonary veins
- Sinus venosus defects (unroofing) of pulmonary veins
  - Right upper pulmonary vein => proximal SVC
  - Right upper pulmonary vein => RA
  - Right lower/middle pulmonary vein => RA

Anomalous connection malposition of pulmonary vein(s)
- Right upper and middle/all right-sided pulmonary veins => proximal IVC: Scimitar-syndrome
  - Single right pulmonary vein => distal SVC/azygos vein
  - Upper left/all left pulmonary veins => vertical vein => innominate vein
  - Single/all left pulmonary veins => left-sided SVC
  - Single left pulmonary vein => right-sided SVC/azygos vein/subclavian vein
  - Single/all left pulmonary veins => proximal IVC

74%
12%
6%
9%

Scimitar syndrome

- complex malformation syndrome, in which part or even the entire right lung is drained by right pulmonary veins that connect anomalously to the inferior vena cava proximally to the liver veins.
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Scimitar syndrome – ‘infantile form’

• common concomitant malformations in *symptomatic infants* are
  - hypoplasia of the affected lung and its associated airways
  - unusual bronchial or vascular distribution patterns
  - hypoplastic right pulmonary artery
  - pulmonary sequestration
  - aortopulmonary collateral vessels
  - pulmonary vein stenosis
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Scimitar syndrome – ‘infantile form‘

'horse-shoe lung'

arterial supply for pulmonary sequester

aorto-pulmonary collaterals
Prevalence / Associated defects

- The overall prevalence of PAPVC is not known
  - often reported as an incidental finding in asymptomatic patients

- Coincidence with other cardiac abnormalities, most often an
  - atrial septal defect:
    - Sinus venosus defect in right-sided PAPVC (87%)
    - Secundum ASD in left-sided PAPVC
  - I-SVC

- malposition of the septum primum is associated with heterotaxia and polysplenia
there is return of oxygenated blood to the right side of the heart with subsequent recirculation through the pulmonary vasculature (pre-tricuspid left-to-right shunting)

significant shunting is associated with two or more anomalous connecting veins or an associated atrial septal defect, resulting in enlargement of the right atrium and ventricle, and dilation of the pulmonary artery

patients with single pulmonary vein involvement do not have significant haemodynamic and cardiac structural changes
the severity of clinical signs and symptoms is related to

- the degree of left-to-right shunting
- the presence of pulmonary hypertension
- the presence of other associated cardiac and pulmonary defects

- patients with a small degree of shunt are usually asymptomatic and are often identified incidentally by a cardiac murmur
- although patients with moderate to large shunts may be asymptomatic in childhood, left-to-right shunt increases with age, and symptoms appear before 40 years of age
presentation of scimitar syndrome varies widely depending upon the age at diagnosis:

• ‘infantile form’:
symptomatic infants tend to present with more severe symptoms related to heart failure and pulmonary hypertension:
- tachypnea
- poor feeding
- failure to thrive
- cyanosis

• ‘adult form’:
about one-half of patients who are diagnosed after the first year of life remain asymptomatic;
others may present with fatigue, dyspnea, and recurrent pneumonia,
or might be identified by an incidental finding on chest X-ray
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Imaging

Chest X-ray

- not diagnostic for PAPVC
- indicated in scimitar syndrome and pts. with concomitant pulmonary malformations
- should be performed before surgery
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Electrocardiogram

- not diagnostic for PAPVC and not helpful in excluding this diagnosis
- in hemodynamically relevant PAPVC, there may be **evidence of right heart enlargement:**
  - right axis deviation of the frontal plane QRS complex
  - evidence of right atrial hypertrophy and/or right ventricular volume load
- should be performed before surgery
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Electrocardiogram

3 y, hemodynamically relevant pre-tricuspid left-to-right shunt
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Diagnostic work-up

**Goal:**
- detailed demonstration of pulmonary venous return
- presence of concomitant cardiac / pulmonary malformations
- assessment of potential cardiac overload and thereby
  of the necessity of therapeutic intervention
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Imaging

Echocardiography

- most frequently used method for an initial diagnosis of PAPVC

- PAPVC should be considered
  • in pts. with unexpected right atrial or ventricular enlargement [especially if there are no other explanations for the finding(s) (eg, large ASD)]
  • in presence of a sinus venosus atrial septal defect
  • if there are fewer than four pulmonary veins connecting to the left atrium [although one must consider the possibility of a single pulmonary vein draining an entire lung]
  • in pts with unexplained dilation of central systemic veins

- a potential limitation of echocardiography is the availability of acoustic windows

- Transesophageal echocardiography (TEE) is more sensitive than transthoracic echocardiography (TTE) in detecting PAPVC
4 chamber view, demonstrating right heart enlargement secondary to a hemodynamically relevant pre-tricuspid left-to-right shunt without obvious large ASD in a 3-year-old girl
Right-sided PAPVC draining into the proximal SVC in combination with a sinus venosus-type ASD, representing the most common form of PAPVC.
Magnetic resonance imaging

- MR angiography (MRA) using gadolinium-based intravenous contrast agents provides enhanced visualization of the pulmonary vasculature including the anomalous pulmonary vein(s).
- MRI can provide additional information, including quantitation of heart chamber volumes, ventricular mass, the ratio of pulmonary to systemic blood flow (Qp/Qs), quantification of blood flow through left / right lung.
- Evaluation of pulmonary venous obstruction after surgical correction.
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Magnetic resonance imaging

Right PAPVC => SVC

Sinus venosus defect
Three-dimensional reconstructed magnetic resonance image of a right-sided partial anomalous pulmonary venous connection to the inferior right atrium (arrow)
Computed tomography

- provides more detailed anatomic information than echocardiography, and like CMR, is not limited by narrow acoustic windows
- enhanced visualization of the pulmonary vasculature
- is especially useful in pts. with concomitant pulmonary malformations
- first line diagnostic tool in scimitar syndrome
- evaluation of pulmonary venous obstruction after surgical correction
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Imaging

Computed tomography

Right-sided PAPVC => proximal SVC

Scimitar syndrome

3D CT in RAO and postero-anterior view.
although cardiac catheterization can be a definitive diagnostic procedure, it is not used as routine diagnostic tool any more

besides angiographic demonstration of PAPVC, cardiac catheterization also provides hemodynamic information, such as pulmonary vascular resistance, cardiac output, and ventricular pressures; shunt volume can be calculated by oximetry

may be indicated in pts. with concomitant cardiac malformations

Indicated as a therapeutic intervention to occlude aorto-pulmonary collaterals

may be indicated for (re)evaluation of pulmonary arterial hypertension

may be indicated for detailed evaluation of a suspected pulmonary venous obstruction after surgical correction
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Cardiac catheterization

Scimitar syndrome

Kinya Ashida et al. Circulation. 2001;103:e126-e127

Late phase of pulmonary arteriogram
Infantile form of scimitar syndrome with severe hypoplasia of right pulmonary artery, pulmonary sequestration, severe bronchomalacia of left main bronchus;
s/p embolization of abdominal a-p collaterals and endobronchial stenting of left main bronchus

Late phase of selective angiography of additional thoracic a-p collaterals displaying the draining vein

Selective angiography of the scimitar vein after embolization of these thoracic a-p collaterals
Unusual case: abnormal communication between RUPV and r-SVC, draining blood from the LA to SVC and RA

Carsten Beck et al. Circulation. 2006;113:e840-e841
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Differential Diagnoses

- haemodynamically relevant **atrial septal defects**
- ‘infantile form’ of scimitar syndrome: pulmonary malformations resulting in pulmonal arterial hypertension
Medical treatment
• rarely indicated in patients with partial anomalous pulmonary venous connection
• ‘infantile form’ of the scimitar syndrome: specific medication for pulmonary arterial hypertension and congestive heart failure may stabilize these patients

Catheter interventions
• occlusion of significant aorto-pulmonary collaterals (including the subdiaphragmatic arterial supply of sequestrations) may stabilize infants with the scimitar syndrome prior to surgery
• (stent-)angioplasty may be indicated to treat postoperative pulmonary venous obstruction

• in very rare cases selective embolisation in pts with PAPVC and additional connection to systemic veins or the left atrium
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Management

• **Surgery is the definitive treatment** in PAPVC;
  therapeutic goal is redirecting the pulmonary venous blood from the anomalously connected vein(s) to the left atrium

• elective surgery for correction of PAPVC should be performed in pre-school children; otherwise, once the diagnosis has been established

• however, asymptomatic patients with PAPVC with small left-to-right shunt do not require intervention, as the defect has no significant clinical impact, and they have a normal life expectancy without correction
The surgical procedure and technique vary and are dependent upon the anatomy and the presence of other associated cardiac anomalies.

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Surgery

Rerouting technique I

- A **double-patch technique** can be used by placing a second pericardial patch to enlarge the superior caval vein at the right lateral side.
- This can be necessary if an incision is made into the lateral side of the superior caval vein when the pulmonary venous connection is at a very high level towards the azygos vein; through this extra incision, the rerouting patch can be easily sutured.

The RA incision is oriented longitudinally and extended along the SVC/RA junction onto the SVC. The incision is carried to the upper limit of any anomalously connected pulmonary vein.

The anatomy is inspected to locate the pulmonary veins and any additional septal defects.

A pericardial patch is used to close the lower edge of the septal defect, and then continued up and around the superior edge of the highest pulmonary vein directing the pulmonary venous flow to the LA.

A second patch is used to augment the SVC/RA junction to ensure unobstructed drainage of the SVC to the RA.

The SVC is divided and the cardiac end is over sewn. A pericardial patch is used to close the stump of the SVC and the sinus venosus directing pulmonary venous flow to the LA.

The tip of the RA appendage is incised. It is important to divide all trabeculae to ensure an unobstructed pathway. An anastomosis is performed between the divided SVC and the RA appendage.

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Surgery

Replantation technique

- rerouting the venous return by replantation of the pulmonary veins into the left atrium
- an existing ASD could be used or enlarged for the exposure of the left atrial wall or an ASD could be created to obtain access to the left atrium
- pericardial patches could be used for the closure of the remaining defect(s) in the right atrial wall and ASD if there is one
Surgical view of the opened right atrium. The dotted line indicates the opening of the intra-atrial septum. Sometimes, an existing atrial septal defect could also be used or enlarged. The pulmonary veins are connected to the right atrium above the intra-atrial septum.

Detailed view of the opening of the intra-atrial septum. Mostly, the septum does not have to be removed but just incised to get a good exposure of the left atrial wall. The intra-atrial septum is partially removed and the back wall of the left atrium is visible.
After creation of an opening in the posterior wall of the left atrium, removal of the pulmonary veins from the right atrium is performed as an island (asterisk) from the right atrial lateral wall.

The pulmonary veins have been sutured into the opening of the left atrial wall and the gap in the right atrium is closed with a pericardial patch. Sometimes, this defect can be closed primarily.

Final result after closing the intra-atrial septum also with a pericardial patch. The dotted lines mark the position of the pulmonary veins.

doi:10.1093/mmcts/mmt001
left-sided anomalous pulmonary venous connection may be corrected by dissecting the vertical vein cranial to the anomalous vein with its subsequent reimplantation to the left atrial appendage
Surgical view of the opened right atrium in scimitar syndrome.
The right pulmonary veins are draining directly to the inferior caval vein.

Creation of a hole in the intra-atrial septum, sometimes an existing atrial septal defect can be used or enlarged.

doi:10.1093/mmcts/mmt001
A pericardial patch is used for rerouting the blood flow towards the left atrium.

Final situation after suturing the pericardial patch.
Correction of Scimitar syndrome with a long baffle from IVC to ASD.

Repair of Scimitar syndrome with reimplantation of SV in the right atrium and use of a short baffle.

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Surgery

Reimplantation technique in scimitar syndrome

Indiana University modification of SV repair via right thoracotomy and without the use of cardiopulmonary bypass.

Completion of repair of SV-Indiana University modification. The scimitar vein has been implanted into the left atrium and the clamp is about to be removed from the left atrium.

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Outcome

Pulmonary vein stenosis after repair of PAPVC

- Single centre experience, Toronto
  - 306 pts in 25 y;
  - 77% children, 236 pts, mean age 5.3 y

Pts with scimitar syndrome are at much higher risk

Single-centre (Boston), 80 pts in 35 years

36 pts. with scimitar vein surgery:
18 pts with postoperative pulmonary vein obstruction

19/80 pts died during f/u (mean 4.5 y)

Pts with severe pulmonary arterial hypertension are at much higher risk
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Outcome

Table IV. Univariate and multivariable analyses of risk factors for death

<table>
<thead>
<tr>
<th>Risk Factor</th>
<th>Univariate analysis</th>
<th>Multivariable analysis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age of presentation (≤1 y vs &gt;1 y)</td>
<td>n=80, log-rank test, P value</td>
<td>Hazard ratio (95% CI)</td>
</tr>
<tr>
<td>CHD excluding ASD</td>
<td>80, &lt;0.001</td>
<td>10.0 (2.9-34)</td>
</tr>
<tr>
<td>Noncardiac anomalies/syndromes</td>
<td>80, 0.04</td>
<td>3.8 (1.4-10)</td>
</tr>
<tr>
<td>PA pressure greater than half systemic</td>
<td>80, &lt;0.001</td>
<td>20.7 (2.8-155)</td>
</tr>
<tr>
<td>Left pulmonary vein stenosis</td>
<td>80, &lt;0.001</td>
<td>9.4 (3.1-28.5)</td>
</tr>
</tbody>
</table>

ASD: Atrial septal defect; PA: Pulmonary artery.

Conclusions

• Surgical intervention for scimitar syndrome is associated with a high rate of postoperative pulmonary vein obstruction that has a trend toward increased risk of occurrence when surgery is performed in infancy.
• Pulmonary hypertension and left pulmonary vein stenosis are independent risk factors for death.
• Normal pulmonary artery pressure and absence of other CHD excluding ASD are factors predictive of survival without surgical intervention.
• Aortopulmonary collaterals (APCs) are present in 70% of patients; closure of APCs does not cause pulmonary infarction, can reduce pulmonary artery pressure, and can potentially avoid the need for scimitar vein surgery.

Scimitar Syndrome
A European Congenital Heart Surgeons Association (ECHSA) Multicentric Study

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Background—Scimitar syndrome is a rare congenital heart disease. To evaluate the surgical results, we embarked on the European Congenital Heart Surgeons Association (ECHSA) multicentric study.

Methods and Results—From January 1997 to December 2007, we collected data on 68 patients who underwent surgery for scimitar syndrome. Primary outcomes included hospital mortality and the efficacy of repair at follow-up. Median age at surgery was 1.4 years (interquartile range, 0.46 to 7.92 years). Forty-four patients (64%) presented with symptoms. Surgical repair included intraatrial baffle in 38 patients (56%; group 1) and reimplantation of the scimitar vein onto the left atrium in 21 patients (31%; group 2). Eight patients underwent right pneumectomy, and 1 had a right lower lobe lobectomy (group 3). Four patients died in hospital (5.9%; 1 patient in group 1, 2.6%; 3 patients in group 3, 33%). Median follow-up time was 4.5 years. There were 2 late deaths (3.1%) resulting from severe pulmonary arterial hypertension. Freedom from scimitar drainage stenosis at 13 years was 83.8% in group 1 and 85.8% in group 2. Four patients in group 1 were reoperated, and 3 patients (2 in group 1 [6%] and 1 in group 2 [4.8%]) required balloon dilatation/stenotomy for scimitar drainage stenosis.

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.)
Scimitar Syndrome –
A European Congenital Heart Surgeons Association (ECHSA) Multicentric Study

• 68 pts underwent scimitar vein surgery in 10 y (’97-’07, 19 centres);
  - median 3 pts per centre (1-9)
  - 44 pts with symptoms
  - right lung hypoplasia in 35 pts
• Mean age at surgery: 1.4 y
  - 31 pts (45 %) < 1 y
  - 54 pts (79 %) < 10 y
• Surgical techniques:
  - intraatrial baffle: 38 pts (56 %)
  - reimplantation: 21 pts (31 %)
  - pneumectomy: 9 pts

• 30d mortality: 4 pts
• f/u (mean 4.5 y): 2 late deaths (PAH)

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Scimitar Syndrome –
A European Congenital Heart Surgeons Association (ECHSA) Multicentric Study

- Freedom from post-op scimitar vein obstruction (SVO) ~ 85 % after 13 y
- 4 reoperations and 3 cath interventions during f/u;
  - 3 pts with asymptomatic SVO without treatment

Circulation. 2010;122:1159-
Outcome and prognosis

- patients with partial anomalous pulmonary venous connection generally have an excellent outcome with low perioperative morbidity and mortality (0.4 %); the risk of postoperative pulmonary vein obstruction is considered to be below 2 % after 15 years.
- the exception are patients with the ‘infantile form’ of the scimitar syndrome who still have an increased risk (early mortality rate recently below 6%) despite improved perioperative management and postponing surgical repair beyond infancy; the risk of postoperative obstruction of the redirected scimitar vein is nowadays considered to be 15 % after 3 years.
- other potential postoperative complications after repair of right-sided partial anomalous pulmonary venous connection are:
  - stenosis of the superior vena cava (10-17 %)
  - sinus node dysfunction; the rate of subsequent pacemaker implantation varies considerably.
Follow-up recommendations

• postoperative findings after surgical repair requiring further diagnostics and potential treatment:
  - reobstruction of the redirected pulmonary vein(s)
  - obstruction of the superior vena cava
  - persisting pulmonal arterial hypertension
  - unequal distribution of pulmonary blood flow (obstruction of redirected veins?)
  - sinus node dysfunction with bradyarrhythmia

• besides clinical, echocardiographic, and electrocardiographic evaluation during outpatient visits, more extensive diagnostic is recommended in symptomatic patients:
  - Holter monitoring in case of suspected bradyarrhythmias
  - cardiopulmonary exercise test
  - MRI / CT imaging or catheterization for pulmonary venous obstruction.