Scimitar syndrome

a series of 90 consecutive patients from a single centre with a focus on associated pulmonary hypertension

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Background

Scimitar syndrome (Scim.) is a rare association of congenital cardiopulmonary anomalies with anomalous drainage of one or more of the right pulmonary veins to the inferior caval vein.

Pulmonary hypertension (PH) is a common finding but its causes are poorly understood.
Aims of the study

- To analyse in a large series of Scimitar
  - the constellation of anatomic anomalies
  - their potential relation with PH and outcome.
Methods & Population

- Retrospective review from 1985 to 2010
- 90 consecutive cases of Scim. (53 females, 37 males)
  - Presenting symptoms,
  - Cardiac phenotype,
  - Extracardiac anomalies,
  - Interventional & surgical procedures
  - Outcome.
- Right heart catheterisation (RHC) for pulmonary hypertension (PH) n=73
Age at diagnosis

- **foetal 10 pts**
  - 7 LV/RV asymmetry including 1 HLHS
  - 1 tetralogy of Fallot
  - 1 abnormal pulmonary venous connection
  - 1 hypoplastic right lung

- **neonatal 25 pts**

- **before 1 year of age in 34 pts**

- **after 1 year of age in 21 pts**
Associated defects

- 48/90 had an associated cardiac defect
  - ASD 12, VSD 10, Coarctation 5, Fallot 4, 17 other
- 13/90 had anomalies of systemic veins
- 65/90 had complete abnormal pulmonary venous return of the right lung
- 10/90 had congenital stenosis of the scimitar vein
  - with aberrant drainage in 2
- 75/90 had systemic arterial supply to the right lung
  - that was considered significant in 60 pts.
- 21/90 had extracardiac malformations
  - Including 7 diaphragmatic defects and 9 vertebral anomalies
Stenosis of the scimitar vein

Systemic supply

Primitive hepatic plexus

Systemic supply from renal artery
Pulmonary hypertension

- 73/90 had right heart catheterization
- the remaining 17 pts had normal estimation of pulmonary pressures on echo
  - Maximum TRV < 2.8 m/s and no other sign in favour of elevated pulmonary artery pressure

- Prevalence of PH at time of diagnosis
  - 55/90 soit 61%
Etiologies of PH in scimitar syndrome

- “Transient” PH (n=38)
  - Persistent pulmonary hypertension of the new-born in 17
  - PH due to massive overflow by the systemic supply in 10
  - PH due to associated CHD in 7
  - Postcapillary PH in 4 (including 2 Cor triatriatum and 2 pulmonary vein stenosis)

- PH due to respiratory disease (n=3)

- Pulmonary Arterial hypertension (n=14)
  - 8 patients with LR shunts (group A or B Dana Point)
  - 6 patients without LR shunts - idiopathic like PAH
Etiologies of PH in scimitar syndrome

- In 1/5 patients, different causes of PH were present either simultaneously or during follow-up
  - PPHN - systemic supply related - idiopathic like PAH

- Causes for idiopathic-like PAH remain unknown
Outcome

- There were 24/90 deaths (18 neonates) that were directly related
  - to refractory PH/cardiac failure in 9
  - to severe respiratory disease in 4

- Confirming that the infantile form of scimitar syndrome is the most worrying
Conclusion

- Surgical outcome of scimitar syndrome has been described favourable but this certainly due to the fact that only survivors and children without PAH undergo surgery.

- In our series, mortality was high and beside associated CHD, the management of PH of multifactorial origin is a remaining challenge.

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