P-90

Pulmonary hypertension in scimitar syndrome : a series of 90 consecutive patients from a single centre

Bonnet D. (1), Chemin A. (2), Delacourt C. (2), Boudjemline Y. (1), Ou P. (1), Lévy M. (1)
M3C-Necker, Université Paris Descartes, Sorbonne Paris Cité, Necker Enfants Malades, Paris, France (1)
Pneumologie Pédiatrique, Université Paris Descartes, Necker Enfants Malades, Paris, France (2)

Background : Scimitar syndrome (Scim.) combines 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.
Objective: To analyse the constellation of anatomic anomalies and their potential relation with PH and outcome in Scim.
Methods: We reviewed 90 consecutive cases of Scim for presenting symptoms, cardiac phenotype, extracardiac anomalies, surgical procedures and outcome. We also reviewed right heart catheterisation (RHC) for pulmonary hypertension (PH) when available.
Results: We identified 90 cases (53 females, 37 males) with Scim. Diagnosis was done in fetus in 10 pts, at birth in 25 pts, before 1 year of age in 34 pts, and after 1 year of age in 21 pts. 48/90 had an associated cardiac defect. The abnormal pulmonary venous return of the right lung was complete in 65 pts. Associated anomalies of systemic veins were present in 13 pts. Congenital stenosis of the scimitar vein was found in 10 pts and aberrant drainage in 2 pts. Systemic arterial supply to the right lung was present in 75 and considered significant in 60 pts. Extracardiac malformations were present in 21 pts. 73 RHC had been performed (the remaining 17 pts had normal estimation of pulmonary pressures on echo): 51 pts had PH at time of diagnosis. PH had different causes : PPHN in 17 neonates, PH due to massive overflow by the systemic supply in 10 pts; PH due to associated CHD in 7; postcapillary PH in 4; respiratory disease in 3; and finally, pulmonary arterial hypertension was observed in 26 pts with or without associated CHD. There were 24 deaths (18 neonates) that were directly related to refractory PH/cardiac failure in 9 pts, and to severe respiratory disease in 4.
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.