

**The natural history of patients with scimitar syndrome: an Italian multi-centric study of the Italian society of pediatric cardiology**

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**Objective:** Scimitar syndrome is a rare association of congenital cardiopulmonary anomalies. Surgical correction of this malformation is usually performed in symptomatic patients or in patients with an increased pulmonary blood flow. The aim of this study was to analyze the natural history of patients with scimitar syndrome who didn't require surgery.

**Materials:** Between January 1990 and December 2011, 36 patients from 7 Italian centers with diagnosis of scimitar syndrome who didn't require surgical correction were included. Excluded were patients with associated severe forms of congenital heart disease. Primary outcomes included the evaluation of their clinical status at follow-up.

**Results:** Median age of patients at diagnosis was 9 months (range 1 day-41 years). There were 20 females and 16 males. Twenty-one patients (58%) were symptomatic at the time of diagnosis (recurrent respiratory infections in 16 patients, congestive heart failure (CHF) in 10 patients, cyanosis in 1 patient, emophtoe in 1 patient. Seventeen patients (48%) had associated cardiac anomalies mainly including an atrial septal defect type in 13 patients, a ventricular septal defect in 5 patients and a patent ductus arteriosus in 5 patients.

Thirty patients underwent cardiac catheterization (84%). The mean pulmonary artery pressure was 26 mmHg (range 13-50 mmHg) and the mean Qp:Qs ratio was 1,6:1 (range from 1:1 to 3:1). Systemic arterial supply to the right lung was demonstrated in 20 patients (67%); 15 of which were treated by coil embolization (50%)(Table 1). Patients presenting with CHF have a higher mean pulmonary artery pressure and had a higher association to cardiac anomalies other than atrial septal defect ( $p<0.05$ ). Median age at last follow-up check was 7,5 years (1month-46years). One patient died at the age at 14 months for severe untreatable pulmonary hypertension. At the last clinical check, 18 patients are still symptomatic (51%) without reported worsening of symptoms and 15 patients (43%) are asymptomatic.

**Conclusions:** Scimitar syndrome is not a benign congenital heart malformation, especially when associated to other congenital heart disease and pulmonary artery hypertension. However it can present as an isolated lesion in almost half of the patients which in the majority of cases are asymptomatic.