

### Features and prognostic factors of Scimitar Syndrome in children

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**Objective :** the aim of this study was to assess the features of children with Scimitar syndrome and to determine factors for long term of outcome

**Methods :** this is a retrospective two-centers study of all patients less than 18 years diagnosed with isolated SS. Demographic data were collected, echocardiographic measurements at diagnosis and last follow up, hemodynamic data when available. Prognostic factors for survival and bad outcome were analyzed

**Results :** 52 patients (35 from Lyon center, France and 17 from Padova, Italy). Patients presented (median age 4.9 months) with respiratory symptoms in 48% or heart failure in 6% or were asymptomatic in 46%. Mean Z-scores for RV diameter, LV diameter, right PA and left PA branch were respectively +0.79, -0.6, -1.9 and +2.2. Mean pulmonary systolic pressure was 59mmHg: 52.5% of cases had no or mild PHT and 47.5% moderate or severe PHT. Fifteen patients were operated on, 26 received medical treatment and 9 percutaneous embolization of systemic artery were performed at a median age of 0.6years. Nine deaths occurred (17%) at median age of 0.4years : mortality was 35.7% in neonates, 16.6% in infants and 5.5% in children. Median FU was 13.4years. Survival rates were 87% at 6months, 85% at 1 and 5years and 78% at 12years of FU.

Stenosis of the scimitar vein, neonatal onset, symptoms at onset, systemic artery and moderate/severe PHT were associated with worse survival ( $p<0.0001$ ). RV systolic pressure, PA systolic, diastolic and mean pressure were higher (respectively : 70 vs 39.6, 61.9 vs 33.3, 22.7 vs 13.5 and 38.1 vs 20.5mmHg), and LV was lower (20.5 vs 29.9mm) in deceased cases than in survivors ( $p<0.0001$ ).

**Conclusion :** Overall outcomes of children with Scimitar syndrome is favourable except in cases with very early onset symptoms and/or moderate/severe PHT and/or stenosis of the scimitar vein.