

Respiratory outcome in children with scimitar syndrome

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Background : Scimitar syndrome is a rare congenital association of an abnormal venous return of the right lung into the inferior vena cava, and of various heart and lung abnormalities, including a virtually constant right lung hypoplasia.

Objectives and methods : Our aim was to evaluate respiratory morbidities and lung function tests in the cohort of patients evaluated at our center since 1976.

Data related to the hospital course and to the follow-up controls of patients, as well as most recent lung function tests were collected.

Results : Eighty-one children were investigated. Twenty-six patients died, all with the infantile form. The median duration of follow-up of surviving children was 5.9 years. A high rate of respiratory morbidities was measured, with 38% and 43% of children reporting pulmonary infections or wheezing episodes during the last 12 months of follow-up, respectively. One third of children have been re-hospitalized for a respiratory cause. Lung function tests were obtained in 19 children. The median value of total lung capacity (TLC) was 76.0 % of the predicted value (IQR:67.5;89.0), and the median value of the ratio of the forced expiratory volume in one second to the forced vital capacity (FEV1/FVC) was -1.06 Z score (-2.13;0.14). Significantly lower TLC values were obtained in children with the infantile form ($p=0.008$), or with a history of thoracic surgery ($p=0.003$). FEV1/FVC Z score values were significantly lower in boys ($p=0.05$), and in children with a history of wheezing ($p<0.05$). Wheezing episodes were not associated to significant salbutamol-induced reversibility.

Conclusion: Respiratory complications are frequently observed in children with scimitar syndrome. Pulmonary hypoplasia appears as an independent marker of long term severity in these patients.