Scimitar Syndrome in Children and Adults: Natural History, Outcomes, and Risk Analysis

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Background. **Scimitar syndrome involves congenital anomalous pulmonary venous return to the inferior vena cava. Optimal management remains controversial. We describe the natural history of disease, nonsurgical and surgical outcomes, and risk factors for poor outcomes at our institution.**

Methods. **Patients with anomalous pulmonary venous return to the inferior vena cava documented on echocardiography at our institution between January 1994 and January 2015 were reviewed retrospectively. The study protocol IRB-AAAO1805 was approved.**

Results. **Forty-seven patients were identified, including 20 infants with significant associated congenital heart defects (42.6%, including 7 with single ventricle physiology), and 10 infants (21.3%) and 16 noninfants (34.0%) with isolated scimitar syndrome. Median follow-up was 3.55 years. Noninfants exhibited lower incidences of right pulmonary artery hypoplasia ($p < 0.001$), aortopulmonary collaterals ($p = 0.004$), and scimitar vein obstruction at the caval confluence ($p = 0.032$). Eighteen patients (38.3%) underwent surgical repair for scimitar syndrome. Overall mortality after baffle repair or scimitar vein reimplantation was 37.5% (3 of 8) for infants and 0% (0 of 6) for noninfants ($p = 0.209$). Overall mortality for medically managed infants was 46.7% (7 of 15) compared with 0% (0 of 8) for noninfants ($p = 0.052$). Multivariable analyses identified infantile onset as an independent risk factor for stenosis or obstruction after repair (hazard ratio 9.34, $p = 0.048$), and single ventricle physiology as an independent risk factor for mortality among unrepaired patients (hazard ratio 29.8, $p = 0.004$).**

Conclusions. **The severity of scimitar syndrome depends on presenting age and associated congenital heart disease. Nonsurgical and surgical outcomes are suboptimal for infantile disease, which is a risk factor for stenosis after repair. Single ventricle physiology is associated with poor prognosis.**

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Scimitar syndrome, a rare association of congenital heart and lung abnormalities, features anomalous pulmonary venous return to the inferior vena cava, typically from the right lung. Associated findings classically include atrial septal defects [1], aortopulmonary collaterals (APC) [2], and hypoplasia of both the right pulmonary artery (RPA) and right lung [3].

Scimitar syndrome patients may be classified into three groups [4–6]. Those diagnosed with other clinically significant congenital heart defects (CHDs), excluding atrial septal defect and patent ductus arteriosus, represent the "CHD infantile" group. These patients present early in infancy but also have other CHDs that influence the course of illness. Among patients without significant associated CHDs, those in the "isolated infantile" group often have significant pulmonary hypertension and present with heart failure before 1 year of age [4], whereas those in the milder "isolated adult" group have symptoms, if any, years to decades after birth [5].

Owing to the rarity of scimitar syndrome, there are insufficient data from which to draw statistically informed conclusions regarding indications for medical versus surgical management. Here, we present one of the largest single-center experiences of scimitar syndrome, including both pediatric and adult cases, and both surgical and nonsurgical cases.

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Patients and Methods

Patients with scimitar syndrome who were evaluated at our institution between January 1994 and January 2015 were identified by searching our congenital echocardiography, cardiac surgery, and cardiology databases. Anomalous pulmonary venous return to the inferior vena cava documented on echocardiography served as the inclusion criteria. The study protocol was approved by the Columbia University Medical Center Institutional Review Board (protocol IRB-AAAO1805). Individual consent was waived owing to the retrospective nature and minimal risk of this study.

A retrospective chart review was performed. The primary outcome variables included mortality, postrepair stenosis or obstruction of scimitar drainage, and reoperation.

Continuous variables were nonnormally distributed and expressed as median with interquartile range, with comparisons performed using the Wilcoxon rank sum test or Kruskal-Wallis one-way analysis of variance. Categoric variables were expressed as counts and percentages, with comparisons performed using Fisher's exact test. Time-related event survival estimates were achieved using the Kaplan-Meier method, and survival distributions were compared for univariate risk analysis using the log rank test. Survival analysis for continuous variables required assigning data to groups defined by value greater or less than the median. Multivariable risk analysis was achieved as sample size permitted using Cox regression models with multiple predictors. A forward selection approach was used to add prospective univariate risk associates ($p < 0.10$) to the Cox model in order of ascending log rank statistic, and a threshold $p < 0.10$ justified inclusion in the model. All data analyses were performed using STATA 12 software (StataCorp, College Station, TX). For all tests, a $p < 0.05$ was considered statistically significant.

Results

Patient Characteristics and Diagnostic Findings

Patient characteristics and diagnostic findings are summarized in Table 1. Forty-seven patients with scimitar syndrome were identified, including 10 (21.7%) isolated infantile, 20 (43.5%) CHD infantile, and 16 (34.8%) isolated adult patients. One patient, who was less than 1 month old at last follow-up, without significant associated CHDs, and never symptomatic, was unable to be classified and was excluded from analyses dependent on scimitar classification. All isolated infantile and CHD infantile patients were symptomatic with heart failure at the time of diagnosis, compared with only 53.8% of isolated adult patients ($p = 0.001$). The median age of heart failure onset was 37.7 years for isolated adult patients.

An atrial septal defect was observed in 29 patients (61.7%), including 80% of isolated infantile patients, 85% of CHD infantile patients, and 18.8% of isolated adult patients ($p < 0.001$). Ventricular septal defect represented the most common significant associated CHD. Concurrent left anomalous pulmonary venous return was diagnosed in 4 patients, including 1 patient with bilateral scimitar syndrome, in which all left and right pulmonary veins except the right upper pulmonary vein drained into the inferior vena cava. Notably, we also identified 7 patients with single ventricle physiology. Six had hypoplastic left heart syndrome (HLHS), including 5 with mitral atresia and 1 patient with Shone's complex. The remaining patient had hypoplastic right ventricle with tricuspid atresia.

Scimitar vein obstruction at the inferior vena cava was noted at the time of diagnosis in 19 patients (43.2%), and was more common among CHD infantile patients than isolated adult patients on pairwise comparison ($p = 0.017$). Right pulmonary artery (RPA) hypoplasia was observed in isolated adult patients (21.4%) less often than in both isolated infantile (80%) and CHD infantile (90%) patients ($p < 0.001$). Similarly, the incidence of APCs was lower for isolated adult patients (18.8%) than the other two groups (70% each, $p = 0.004$). Pulmonary sequestration ($p = 0.065$) and right lung hypoplasia ($p = 0.084$) also trended toward lower frequency among isolated adult patients.

Surgical Outcomes

Eighteen patients (38.3%) underwent surgical repair for scimitar syndrome. Twelve (66.7%) received an intraatrial baffle, including all 6 noninfants who received surgery. Scimitar vein reimplantation was performed in 5 infant cases (27.8%), 3 of which involved single ventricle physiology whereby the scimitar vein was reimplanted to the right atrium. Finally, 1 isolated infantile patient underwent a right pneumonectomy as definitive management. All baffle and reimplantation repairs were performed through median sternotomy. Among infants who underwent surgery, no statistically significant differences were observed when comparing the use of scimitar vein reimplantation versus intraatrial baffle repair in terms of postrepair stenosis, heart failure recurrence, mortality, or need for reoperation (Supplemental Table 1). Cardiac surgery procedures performed concurrently with scimitar repair are summarized in Supplemental Table 2.

Outcomes of surgical management for scimitar syndrome are shown in Table 2. A summary of all isolated infantile and CHD infantile patients who underwent surgical repair is provided in Supplemental Table 3. Of the 11 patients with infant-onset scimitar syndrome who underwent baffle or reimplantation repair, 3 (27.3%) had early postoperative mortality in hospital. Of the remaining 8 patients who survived to discharge, follow-up data are available for 5 patients. Although late mortality was not observed for any infantile patients who underwent surgery during a median follow-up period of 9.1 months, heart failure symptoms redeveloped in all 5 patients at a median 6.8 months after repair, with 4 (80%) receiving a reoperation for scimitar repair stenosis or obstruction. In contrast, no mortality was observed among the 6 isolated adult patients who underwent surgery during a median follow-up period of 3.92 years, although 2 (33%) showed signs of heart failure