Isolated pulmonary artery arising from a duct: A single-center review of diagnostic and therapeutic strategies

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Objective: Isolated pulmonary artery of ductal origin (IPADO) is a rare disease with diverse presentations. Diagnostic and therapeutic approaches vary widely given the low incidence. Reporting additional cases may help inform medical decision making.

Methods: We reviewed diagnostic data—including echocardiography, catheterization, and MRI—and outcomes for all patients with IPADO at our institution.

Results: Thirty-seven patients who met inclusion criteria were identified. The mean age at diagnosis was 3 months (range, 1 day-45 years). In 11 patients, the duct supplying the IPADO was patent (ie, patent ductus arteriosis [PDA]); the duct had closed (ligamentum) in the other 26 patients. When performed, catheterization delineated the anatomy in 90% (100% if PDA) versus 54% with magnetic resonance imaging. Patients with a PDA were more likely to undergo intervention (100% vs 58%, $P = .02$) and had earlier first intervention (1 vs 20 months; $P < .001$). Patients diagnosed at age ≤6 months were more likely to undergo intervention (86% vs 50%; $P = .03$) and unifocalization (81% vs 44%; $P = .04$), and had greater IPADO flow at follow-up (40% vs 14%; $P < .001$). Patients who underwent any intervention had greater IPADO flow than those without intervention (38% vs 0%).

Conclusions: Early IPADO diagnosis is important in long-term outcome. However, successful interventions can be performed on older patients. Diagnosis relies on angiography but magnetic resonance imaging may play an increasingly important role. Although initial intervention depends on individual factors, the ultimate goal should be early unifocalization. (J Thorac Cardiovasc Surg 2014;148:2245-52)

Ductal origin of a pulmonary artery (PA) is a rare condition. Fraentzel[1] published the first description of ''a case of abnormal communication of the aorta with the pulmonary artery'' in 1868. It is commonly associated with other congenital heart diseases—mostly tetralogy of Fallot and truncus arteriosus—with a prevalence up to 0.4%.[2] This lesion occurs in isolation as well, with an estimated incidence of ~1:200,000.[3]

When in isolation, this lesion is often described as a unilaterally ''absent'' PA, but we prefer isolated PA of ductal origin (IPADO), the descriptor used by Takatsuki and colleagues.[4] Although the extrapulmonary portion of the PA is missing, the intrapulmonary vessels are present and generally branch normally, suggesting the anomaly is related to a specific embryologic disruption involving involution of the proximal sixth aortic arch.[5-8] However, a connection persists between the intrapulmonary PA, which arises from the respective lung bud, and the distal sixth aortic arch, which develops into the ductus arteriosus.[8,9] The result is an intrapulmonary PA that is discontinuous from the central pulmonary circulation and fed directly by a ductus arteriosus (Figure 1).

IPADO presents in myriad ways, including recurrent infection, hemoptysis, pulmonary arterial hypertension (PAH), congestive heart failure, and scoliosis; it affects some patients severely at young ages, whereas others are asymptomatic with serendipitous diagnosis in adulthood.[10-12] Such heterogeneity has been associated with variable approaches to management. Treatment strategies include insertion of a surgical shunt to promote IPADO growth before unifocalization into the pulmonary circulation, unifocalizing the IPADO initially, or performing an interventional catheterization before a surgical intervention.[13-16]

Our aim was to provide data to help inform decision making given this variation in practice. Although the incidence of IPADO is insufficient to provide conclusive data, we attempted to identify potentially relevant diagnostic and therapeutic variables.

PATIENTS AND METHODS

Patients

We performed a retrospective, single-center cohort study of all patients with IPADO cared for at Boston Children's Hospital. We included patients with a patent foramen ovale, secundum atrial septal defect, or ventricular

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Abbreviations and Acronyms

IPADO = isolated pulmonary artery of ductal origin
MPA = main pulmonary artery
MRI = magnetic resonance imaging
PA = pulmonary artery
PAH = pulmonary arterial hypertension
PDA = patent ductus arteriosus

septal defect that was deemed hemodynamically insignificant (ie, pressure restrictive, no volume overload, and no intervention). Patients with a patent ductus arteriosus (PDA) were also included given the integral relationship between the ductus and IPADO. Exclusion criteria were cardiovascular lesions other than above, agenesis or severe hypoplasia of the ipsilateral lung, and iatrogenic discontinuity of a PA (eg, after PDA ligation). Our study was performed under a protocol approved by the Children's Hospital Committee for Clinical Investigation, which included a waiver for requiring patient consent.

Definitions and Outcomes

Nearly all patients underwent postoperative transthoracic echocardiography and many underwent nuclear scintigraphy lung perfusion and cardiac magnetic resonance imaging (MRI) scans. All studies were performed for clinical indications at the treating physician's discretion. All data were obtained from existing clinical records, with thorough attempts to obtain the most recent data for patients treated outside our institution.

No study end points were defined a priori and the most recent data were used for all patients. Study points of interest included age at first presentation and correct diagnosis, initial diagnosis or misdiagnosis, preoperative catheterization or cardiac MRI (if performed), age at first intervention, number and type of interventions (surgical: shunt or unifocalization; transcatheter: angioplasty or stenting), serious postprocedural complications, death, and duration of follow-up. Serious postprocedural complications included intra- or perioperative death and requirement for reintervention within 15 days.

A shunt was defined as a surgically created connection between the IPADO and systemic arterial system. Unifocalization was defined as a connection of the IPADO to the PA circulation and further classified as direct or with a tube graft. Direct unifocalization was anastomosis of the IPADO directly to the main pulmonary artery (MPA), including use of an MPA flap or noncircumferential piece of material to augment the anastomosis. A tube graft was a fully circumferential tube of any material interposed between the IPADO and MPA.

Study measures of interest included symptoms, history of PAH, treatment with anti-PAH medications, initial and most recent percentage of flow to the IPADO, and initial and most recent IPADO cross-sectional area (indexed to body surface area). Two orthogonal measurements were used to calculate cross-sectional area when available. PAH was defined as a directly measured mean pressure $\geq$25 mm Hg in the PA continuous with the right ventricle. If catheterization data were not available, PAH was defined as an estimated right ventricle pressure $\geq$50% systemic by echocardiographic measures. Catheterization and MRI adequately delineated anatomy if at least 1 IPADO diameter and the distance from IPADO to MPA were reliably measured.

Data Analysis

Data are presented as median (range) or frequency (% of appropriate cohort). Demographic, historical, diagnostic, and procedural variables were compared between the cohorts. To detect associations, the population was divided into 3 groups for comparisons: those who underwent any intervention or not, status of the IPADO-supplying duct at presentation (PDA or ligamentum arteriosum), and age at presentation ($\leq$6 vs >6 months). The χ^2 or Fisher exact test were used to compare nominal variables between 2 or more groups. The Mann-Whitney U and Kruskal-Wallis tests were used to compare medians between 2 or more groups. All data analyses and the standard scatter plots were created using Statistical Package for the Social Sciences software (version 19, IBM-SPSS Inc, Armonk, NY).

RESULTS

Patients

We identified 49 patients with an IPADO managed at Boston Children's Hospital from July 1951 through September 2011; 12 were excluded due to agenesis of the ipsilateral lung or an iatrogenic IPADO. Demographic and diagnostic variables of all patients are presented in Table 1. Most patients presented in infancy with respiratory symptoms, a murmur, or cyanosis. Two-thirds of the cohort was diagnosed after 2000. Five patients were initially misdiagnosed; 2 were correctly diagnosed within 6 months, whereas 3 were diagnosed 2, 3, and 20 years after presentation. All patients had an IPADO contralateral to aortic sidedness, in keeping with the postulated etiology.[8] Median duration of follow-up data was 7.3 years.

Of 37 patients included in our study, 26 had an IPADO connected to a ligamentum arteriosum, whereas 11 were supplied by a PDA. Patients with a PDA presented earlier (0.6 vs 4.3 months; $P = .04$) and with shorter duration from presentation to diagnosis (0.03 vs 1.6 months; $P < .001$) than those with a ligamentum. More than half of patients with a ligamentum presented with respiratory symptoms, whereas most with a PDA presented with a murmur or cyanosis. All patients presenting with hemoptysis had a ligamentum. Ductal status was not associated with era (ie, diagnosis before or after January 1, 2000).

Twenty-one patients were diagnosed before age 6 months. Younger patients presented roughly equally with respiratory symptoms, a murmur, or cyanosis, whereas most older patients presented with respiratory symptoms. No older patient presented with cyanosis. Both patients with hemoptysis presented at older age (20 and 70 months). Approximately 45% of younger patients had a PDA versus only 12% of older patients. Younger patients were also correctly diagnosed sooner after presentation (0.1 vs 4.8 months; $P < .001$) and had greater flow to the IPADO-supplied lung initially (33% vs 0%; $P < .001$) (Figure 2).

Diagnostic Studies

Nine PDA patients and 18 ligamentum patients underwent preoperative catheterization. Catheterization adequately delineated the anatomy in all cases with a PDA, but in only 12 with a ligamentum; pulmonary vein wedge angiograms were performed in all patients. Three PDA patients and 10 ligamentum patients had a preoperative MRI. MRI adequately delineated anatomy in 1 PDA patient