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 arteriosus [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. Fraentzel¹ 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 $\sim 1:200,000$.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 ≥ 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 (≤ 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

FIGURE 1. Simultaneous injections into a left isolated pulmonary artery of ductal origin (IPADO) and right pulmonary artery. The right pulmonary artery catheter courses antegrade through the right ventricle, whereas the left IPADO catheter ascends retrograde from the right-sided aorta into the left patent ductus arteriosus.

and 6 ligamentum patients. No MRI defined the anatomy when catheterization failed to do so.

Therapeutic Strategies

Demographic, diagnostic, and therapeutic variables based on initial intervention are presented in [Table 2](#). Twenty-six patients underwent at least 1 IPADO intervention; all ultimately underwent surgery. Eight patients had initial shunt placement, whereas 18 had initial unifocalization. One of the shunt patients had initial transcatheter intervention (PDA stent) with subsequent shunt palliation. Another patient remains with a shunt with the plan to unifocalize; both patients currently with shunts have a left-sided IPADO. The other 6 initial shunt patients underwent subsequent unifocalization, all of which were with a tube graft. Patients who had an initial unifocalization were younger and underwent operation earlier than those with initial shunt ([Figure 3](#)). Those patients also had fewer surgical interventions and fewer procedures overall.

Among unifocalized patients, 13 underwent direct unifocalization and 11 had tube grafts. The direct unifocalization group was younger at diagnosis (0.7 vs 14 months; $P = .001$) and first intervention (1 vs 20 months; $P = .003$). The direct unifocalization group had fewer total (2 vs 4; $P = .01$) and surgical (1 vs 2; $P = .01$) interventions with more flow at most-recent follow-up (40% vs 32%).

Reinterventions

A total of 40 postoperative interventions were performed on 19 patients. Those procedures included 23 balloon

angioplasties, 6 angioplasties with stent placement, and 11 operations. All 8 shunt patients had a postoperative intervention; 5 of the 8 had a first reintervention related to issues with the shunt (ie, anastomotic stenosis or shunt thrombosis), whereas 3 had unifocalization at first reintervention. Of the 18 patients initially unifocalized, 7 had no reinterventions. Among the 11 with reinterventions, 1 had a reoperation for acute thrombosis of a unifocalized tube graft and 10 patients only had catheter-based reinterventions. The median duration to first reintervention was longer in the initially unifocalized group (5 vs 2.4 months) with fewer reinterventions performed per subject (1 vs 2).

Outcomes

All patients with a PDA underwent an intervention versus only 58% of patients with a ligamentum. Younger patients had greater IPADO flow at recent measurement (40% vs 14%; $P < .001$). Younger patients were also more likely to undergo intervention (86% vs 50%) and to undergo ultimate unifocalization (81% vs 44%) ([Figure 2](#)).

Among 16 patients who presented after age 10 months, 8 underwent intervention. Patients who underwent intervention were younger (median 31 vs 165 months; $P = .03$), the oldest of whom was diagnosed at 83 months. Of these 8, 2 had a PDA and 6 had a ligamentum. One of the patients with a PDA had direct unifocalization and had an increase in IPADO flow from 0% to 16%. The other patient with a PDA underwent initial catheter-based PDA stenting followed by shunt placement; the shunt thrombosed, no subsequent interventions were performed, and there was no flow to the IPADO at most recent follow-up. Two patients with a ligamentum were initially unifocalized with increases in flow from 0% to 28% and 30%. The other 4 patients had initial shunt placement followed by unifocalization; 1 had a minimal increase in flow but 2 had final indexed IPADO flows of 14% and 15% and the other had an increase in IPADO area from 12 to 40 mm²/m².

Complications

Four patients experienced serious postoperative complications: 3 had thrombosis of a shunt and 1 had thrombosis of a unifocalization tube graft. Two of the shunts were completely occluded and replaced; 1 was restored with thrombectomy. The shunt thrombectomy patient had partial thrombosis, significant lung disease with PAH, and died shortly after. Of the 2 patients with complete shunt occlusion, 1 had the smallest indexed cross-sectional area of the entire cohort and the other was well below the median. The patient with tube graft thrombosis underwent graft replacement on postoperative day 5 with no further complications.

Three known deaths occurred that were related, at least in part, to the IPADO. Two had been correctly diagnosed initially, but both patients were born prematurely with significant lung disease. One died from sepsis in infancy and

TABLE 1. Demographic, diagnostic, and follow-up data for the entire population and by cohort

	All patients (N = 37)	Any intervention (n = 26)	No intervention (n = 11)	P value*
Discontinuous PA				
Left	17 (46%)	12 (46%)	5 (45%)	
Right	20 (54%)	14 (54%)	6 (55%)	
Age at diagnosis, mo	3.2 (0-539)	2.4 (0-83.5)	66.2 (0.1-539)	.01
Era of diagnosis				.008
Before January 1, 2000	12 (32%)	5 (19%)	7 (64%)	
On or after January 1, 2000	25 (68%)	21 (81%)	4 (36%)	
Sex				
Male	15 (41%)	11 (42%)	4 (36%)	
Female	22 (49%)	15 (58%)	7 (64%)	
Status of duct at diagnosis				.01
Patent	11 (30%)	11 (42%)	0 (0%)	
Ligamentum	26 (70%)	15 (58%)	11 (100%)	
Age of presenting complaint, mo	2.3 (0-516)	1.5 (0-69)	5.2 (0-516)	
Presenting symptom/sign				
Respiratory symptoms	15 (41%)	10 (38%)	5 (45%)	
Murmur	10 (27%)	7 (27%)	3 (27%)	
Cyanosis	6 (16%)	4 (15%)	2 (18%)	
Hemoptysis	2 (5%)	2 (8%)	0 (0%)	
Other†	4 (11%)	3 (12%)	1 (9%)	
Initial misdiagnoses	5 (14%)	2 (8%)	3 (27%)	
Time from presentation to diagnosis, mo	1 (0-244)	0.4 (0-34)	3.4 (0.1-244)	.01
Right ventricular hypertension				
At time of diagnosis	19 (51%)	11 (42%)	8 (73%)	
Ever	22 (59%)	14 (52%)	8 (73%)	
At most recent follow-up	8 (22%)	1 (4%)	7 (64%)	<.001
Pulmonary antihypertensive medications				
Ever	4 (11%)	2 (8%)	2 (18%)	
At most recent follow-up	2 (5%)	1 (4%)	1 (9%)	
Symptoms at most recent follow-up	6 (16%)	2 (8%)	4 (36%)	.03
Age at first intervention, mo		4.5 (0.1-91.6)	NA	
First intervention type				
Surgical		25 (96%)	NA	
Transcatheter		1 (4%)	NA	
First operation				
Shunt		8 (31%)	NA	
Unifocalization		18 (69%)	NA	
Total interventions		2 (1-6)	NA	
Total surgical interventions		1 (1-3)	NA	
Ultimately unifocalized		24 (92%)	NA	
Age at unifocalization, mo		4.9 (0.1-97)	NA	
Initial IPADO cross-sectional area				
Indexed, mm ² /m ²	17.5 (0-69.1)	24.6 (6.6-69.1)	0 (0-5)	<.001
Most recent IPADO cross-sectional area				
Indexed, mm ² /m ²	50.6 (0-189)	53.3 (23.7-189)	0 (0-0)	.1
First flow distribution to IPADO, %	11 (0-59)	15 (0-59)	0 (0-1)	.04
Most recent flow distribution to IPADO, %	34 (0-59)	38 (0-59)	0 (0-1)	.008
Duration of follow-up, y	7.3 (0.1-59)	7.4 (0-20)	6.5 (2-59)	

Data are presented as median (range) or frequency (%). PA, Pulmonary artery; NA, not applicable; IPADO, isolated pulmonary artery of ductal origin. *Comparing any intervention and no intervention cohorts. †Other diagnoses include abnormal chest radiograph, electrocardiogram, and screening obstetric ultrasound.

the other was the patient who experienced partial postoperative shunt occlusion. The third patient was diagnosed with IPADO at age 44 years after an MRI, and ultimately died from heart failure related to PAH at age 60 years.

DISCUSSION

Importance of Age at Intervention and Outcome

Many studies have suggested that early diagnosis and management of patients with IPADO is important.¹⁷⁻²⁰

FIGURE 2. Scatter plots comparing first measured indexed isolated pulmonary artery of ductal origin (IPADO) cross-sectional area (CSA). A, Most recent indexed IPADO CSA. B, Most recent flow to the IPADO based on age and presence of right ventricle (RV) hypertension at most recent follow-up. Circles indicate diagnosed at age <6 mo, triangles indicate diagnosed at age ≥6 mo. No fill indicates normal right ventricle pressure at most recent follow-up, solid fill indicates right ventricle hypertension at most recent follow-up. *Percent flow to IPADO is expressed as percent of total pulmonary blood flow.

We believe that unifocalization and achievement of pulmonary flow distribution as close to normal as possible should be the primary treatment goals. In our cohort, patients diagnosed earlier were more likely to undergo unifocalization, suggesting that age at diagnosis is important. Interestingly, we did not observe an association between age and the total number of interventions performed. This may be due to the heterogeneity in presentation and therapeutic strategies. Also, older patients may have vascular disease in the affected lung, precluding aggressive attempts to rehabilitate the IPADO. We also did not observe differences in complication rates

between cohorts based on age, suggesting the risk of intervention is not necessarily greater in younger patients. Most patients, even those diagnosed after age 10 months, had reasonable improvement in IPADO flow and area, especially when unifocalized. Overall, we believe that diagnosis and intervention before age 6 months should be a primary goal, but that patients of any age can yield benefit from unifocalization.

Associations With Initial Ductal Status

Our data demonstrate that patients with a PDA have potential advantages compared with those in whom the ductus has closed. Patients with a PDA presented at an earlier age and were diagnosed more rapidly; 9 of 11 were diagnosed before age 4 months and 5 were diagnosed as neonates (the oldest patient was aged 49 months and an outlier). This association of age and ductal status may be related to the fact that many PDA patients presented with murmurs in early infancy, likely leading to evaluation by a pediatric cardiologist. Also, fewer PDA patients presented with recurrent respiratory symptoms or infections compared with patients with a ligamentum. This nonspecific presentation may have led to more extensive evaluation in ligamentum patients before referral to a cardiologist, prolonging time to diagnosis. In fact, half the ligamentum patients were diagnosed after 1 year, with the oldest diagnosed at nearly 45 years of age; again, ductal status was not associated with era of diagnosis. We further speculate that this later diagnosis may be related to the development of hemoptysis. Although hemoptysis is incompletely understood in patients with congenital cardiac disease, prolonged diagnosis may lead to the development of increased pulmonary vascular resistance and systemic-to-pulmonary collaterals that may play a role in this phenomenon.^{10,21,22} Several studies have described the deleterious consequences of a delay in diagnosis.^{17,23} Our data corroborate this, because nearly one-third of patients with a ligamentum had PAH at most recent follow-up.

Therapeutic Strategies

Therapeutic strategies in this series fell into 2 basic approaches: shunt placement followed by direct unifocalization and direct unifocalization initially.²⁴ Krammoh and colleagues¹⁶ espoused a variant of the first strategy, recommending initial stenting of the PDA supplying the IPADO with subsequent unifocalization. They hypothesized that increased flow would improve the technical feasibility and success of unifocalization. They believed this was especially useful in patients with hypoplastic branch PAs, because small PAs lead to greater pulmonary vascular resistance with a higher propensity for thrombosis of a surgical shunt. Conversely, other authors have postulated that the optimal strategy is direct unifocalization as early as

TABLE 2. Demographic, diagnostic, and follow-up data based on initial surgical intervention

Variable	Shunt (n = 8)	Unifocalization (n = 18)	P value
Discontinuous PA			
Left	5 (62.5%)	11 (61%)	
Right	3 (37.5%)	7 (39%)	
Age at diagnosis, mo	22.9 (1.5-83.5)	0.8 (0-49.9)	.001
Era of diagnosis			.13
Before January 1, 2000	0 (0%)	5 (28%)	
On or after January 1, 2000	8 (100%)	13 (72%)	
Status of duct at diagnosis			.23
Patent	2 (25%)	9 (50%)	
Ligamentum	6 (75%)	9 (50%)	
Right ventricular hypertension			
At time of diagnosis	1 (17%)	10 (56%)	
Ever	3 (50%)	11 (61%)	
At most recent follow-up	0 (0%)	1 (6%)	
First intervention			
Surgery	7 (87.5%)	18 (100%)	
Catheterization	1 (12.5%)	0 (0%)	
Age at first operation, mo	35.5 (2.8-91.6)	2.4 (0.1-58.1)	.003
Subsequent unifocalization	6 (75%)	NA	
Time from shunt to unifocalization	5.6 (2.6-28.3)	NA	
Any intervention(s) after first operation	8 (100%)	11 (61%)	.048
Total interventional catheterizations per patient			
Balloon dilation	1.5 (0-2)	1 (0-3)	
Stent placement	0 (0-0)	0 (0-2)	
Duration from surgery to first reintervention, mo	2.4 (0.1-5.5)	5 (0.2-46.7)	.07
Total surgical interventions per patient	2 (1-3)	1 (1-3)	<.001
Total interventions per patient	3.5 (2-5)	2 (1-4)	.02
Age at unifocalization, mo	39 (10.4-97)	2.4 (0.1-58.1)	.001
Initial IPADO cross-sectional area			
Indexed, mm ² /m ²	13.2 (6.6-26)	32.9 (9.5-69.1)	.14
Most recent IPADO cross-sectional area			<.001
Indexed, mm ² /m ²	40.1 (25.2-189)	71.7 (23.7-171.4)	
First flow distribution to IPADO, %	7 (0-41)	22 (0-59)	.25
Most recent flow distribution to IPADO, %	14 (0-41)	40 (16-59)	<.001
Duration of follow-up, y	4.2 (0.9-10.4)	8.5 (0.1-20)	

Data are presented as median (range) or frequency (%). PA, Pulmonary artery; NA, not applicable; IPADO, isolated pulmonary artery of ductal origin.

possible, followed by transcatheter intervention(s) to augment IPADO growth.¹⁸

Patients who underwent initial unifocalization in our cohort had superior outcomes (Figure 4). Multiple factors may explain this observation, some of which may be related to intrinsic pathophysiology and others to the specific therapy. One issue is that patients who underwent shunt placement were older and more likely to have a ligamentum. It is plausible that those patients had more hypoplastic or dysplastic pulmonary vasculature, impeding flow and growth of the IPADO. Earlier unifocalization should best restore normal PA physiology and may allow the IPADO and distal vasculature to match somatic growth.

It has been hypothesized that unifocalization technique is relevant.^{25,26} Our data suggest that patients with direct unifocalization have superior outcomes to those with an interposition graft. Other authors have described this association.^{18,19} One explanation is that tube grafts do

not grow, thereby impeding flow, IPADO growth, and normal vascular arborization in the long term. They may also require more PA interventions to maintain optimal IPADO growth. However, 9 of 11 tube graft patients had a ligamentum and all had initial shunt placement, possibly confounding this association. Overall, we believe that direct unifocalization is the preferred therapeutic goal.

Direct Angiography Versus MRI

The diagnostic standard for defining IPADO anatomy has been angiography. As its role increases, MRI's ability to delineate anatomy has been questioned, especially if the duct is atretic or closing.²⁷ Other authors have argued that evolving imaging techniques address this issue.²⁸ Our data confirm that angiography can delineate the anatomy, especially if the duct is patent. Although not 100% sensitive, pulmonary venous wedge angiography is

FIGURE 3. Scatter plot comparing age at presentation and most recent flow to the isolated pulmonary artery of ductal origin (IPADO) based on initial operation and status of the IPADO at diagnosis. Circles indicate patent ductus arteriosus (PDA) at diagnosis, triangles indicate ligamentum at diagnosis. No fill indicates initial shunt, solid fill indicates initial unifocalization. *Percent flow to IPADO is expressed as percent of total pulmonary blood flow.

necessary to delineate anatomy in cases with a ligamentum. MRI failed to delineate the anatomy when catheterization was unsuccessful in our experience. However, MRI delineated the anatomy in 60% of patients with a ligamentum. Although our sample size is small, the utility of MRI in patients with ligamentum should not be overlooked.

Limitations

Our study has numerous limitations. It was a retrospective, single center study with a small sample size. In addition, there was significant heterogeneity in diagnostic and therapeutic strategies. Furthermore, no strict criteria were developed. Lastly, we had limited statistical capability so

FIGURE 4. Bar graphs comparing patients who underwent unifocalization versus those who did not. *Significant P value. IPADO, Isolated pulmonary artery of ductal origin; RV, right ventricle.

age, PDA, era, and other variables had potentially confounding effects.

CONCLUSIONS

IPADO is a rare condition that presents in a variety of ways, with significant consequences, including impaired lung growth, PAH, and hemoptysis. Early diagnosis and intervention appear to be important to optimize long-term outcomes. We recommend a therapeutic strategy with initial catheterization or MRI to adequately delineate the anatomy and the ultimate goal of IPADO unifocalization as early as possible. A preceding procedure to increase flow to the IPADO may be warranted, but should be undertaken cautiously in patients with a small IPADO or PAH. Close follow-up with transcatheter interventions to augment IPADO growth as indicated may lead to optimal outcomes.

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