Multimodality hemodynamic evaluation for optimizing management of anomalous origin of the right pulmonary artery from the aorta in an adolescent

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ABSTRACT

The hemodynamics of anomalous origin of the pulmonary artery (PA) from the aorta is challenging. Different sources of blood supply to the lungs lead to a unique state of differential flow, pressure, and pulmonary vascular resistance in each lung. The decision for surgical reimplantation of the anomalous PA during infancy is easy. The assessment of operability beyond infancy, however, is perplexing. In this report, we describe stepwise multimodal hemodynamic evaluation and successful surgical management in a 15-year-old boy with an isolated anomalous origin of the right PA from the aorta. We also report 5-year hemodynamic data confirming sustained benefit over the long term, thus providing much-needed clinical validation of often cited Poiseuille's and Ohm's laws.

Keywords: Anomalous origin of pulmonary artery, cardiac catheterization, cardiac hemodynamics, lung perfusion, magnetic resonance imaging

INTRODUCTION

Anomalous origin of the pulmonary artery (PA) from the aorta is a rare cardiac malformation, with most (>95%) patients presenting with heart failure during infancy.[1-4] The management during infancy is straightforward, with surgical reimplantation of anomalous PA yielding good outcomes.[3,4] In contrast, operability assessment in an older child, even in the absence of coexisting heart disease, may be challenging.[5-8] Differential blood flow and vascular resistance in the left and the right lung makes the hemodynamic assessment difficult.[6,7] We demonstrate stepwise multimodality hemodynamic evaluation that helped in successful surgical repair in an adolescent boy. We also provide a 5-year follow-up data confirming sustained clinical and hemodynamic benefits thus achieved.

CASE REPORT

A 15-year-old boy presented to the outpatient department with exertional dyspnea. At presentation, he had a heart rate of 80 beats/min, blood pressure of 120/76 mmHg, and respiratory rate of 16 breaths/min with oxygen saturation of 98% in room air. The physical exam was unremarkable except for a loud P2 and a grade 2/6 pansystolic murmur at the left lower sternal border. He did not have heart

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failure. The child had a normal perinatal and childhood history, including growth and development.

Transthoracic echocardiogram showed the usual arrangement of atriums, concordant atrioventricular, and ventriculoarterial connections. The atrial and ventricular septum were intact. The aortic arch was left-sided with no patent ductus arteriosus (PDA). He had normal biventricular function but with a dilated left ventricle. The right ventricular systolic pressure (RVSP), estimated by mild tricuspid regurgitation (TR), was 50 mmHg. Saline contrast echocardiography and imaging in the suprasternal view revealed an anomalous origin of the right PA (RPA) from the ascending aorta [Figure 1 and Videos 1 and 2], the details of which are published elsewhere.[9] Cardiac catheterization confirmed the origin of RPA from the left posterior aspect of the ascending aorta adjacent to the main PA (MPA) [Figure 2 and Video 3]. MPA continued as the left PA (LPA). The pressure in RPA was systemic while pressure in LPA measured 47/12/27 mmHg. Since, the entire venous return was reaching the left lung, the blood flow and pulmonary vascular resistance (PVR) of the left lung were estimated to be 4.6 L/min and 3.7 WU, respectively. The estimation, however, is imperfect owing to the differential flow to the left and the right lung. Being connected to the aorta, a similar estimation for the right lung was not possible using the Fick's principle [Table 1]. Hence, a phase-contrast magnetic resonance imaging (MRI) study was performed on the same day as cardiac catheterization. The blood flow in the right lung and the left lung was measured as 1.8 L/min and 4.6 L/min, respectively. The blood flow in the ascending aorta proximal and distal to anomalous RPA measured 6.5 L/min and 4.6 L/min, respectively [Table 1]. The similar blood flow in the left PA and distal ascending aorta provided much-needed internal validation of flow measurements on MRI. Combining pressure data from cardiac catheterization and flow data from MRI, the calculated PVR of the right and the left lung was 45.5 WU and 3.7 WU, respectively [Table 1]. Of note, the blood flow and PVR of the left lung, estimated by Fick's method and MRI, were the same thus providing further validation of the multimodal hemodynamic assessment. Thereafter, assuming restoration of parallel pulmonary circulation postoperatively, using Poiseuille's and Ohm's laws[10] for the prospective parallel circuits, we predicted the PVR of the postoperative pulmonary circuit to be 3.5 WU [Table 1].

The patient underwent successful surgical implantation of RPA to MPA using aortic interposition homograft under cardiopulmonary bypass. Echo showed normal RV function with TR predicted RVSP of 40 mm Hg. He was started on sildenafil (20 mg twice daily). He had an uneventful postoperative recovery and was discharged on the postoperative day 5. At the 2-month follow-up, the patient was in NYHA functional Class I. The echocardiogram showed normal biventricular function, normal flow in branch pulmonary arteries, and TR predicted RVSP of 30 mmHg. Catheter angiography demonstrated unobstructed flow in RPA. PA pressure was measured at 32/8/17 mmHg while PVR was calculated as 2.3 WU [Table 2]. On Tc99 macroaggregated albumin (MAA) lung perfusion scintigraphy, the right lung received 21% of the lung perfusion, with the remaining 79% going to the left lung [Figure 3]. The flow pattern was the same following 9:50 min of exercise on Bruce protocol achieving 11.30 metabolic equivalents METS. Sildenafil was discontinued and regular clinical and echocardiographic follow-up was planned.

At a 5-year follow-up, he is in NYHA Class I. The echocardiogram showed normal biventricular function with TR-predicted RVSP of 30 mmHg. Cardiac catheterization confirmed unobstructed PA flow with PA pressure

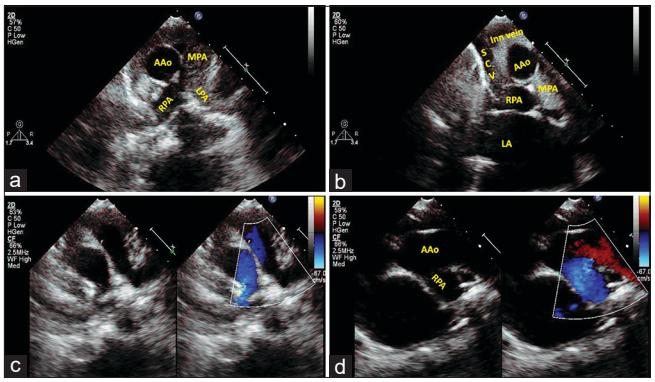

Figure 1: Saline contrast echocardiography in parasternal short-axis view (a) and suprasternal view (b) shows no opacification of the RPA indicating a lack of connection of the right PA with the main PA. Transthoracic echocardiography in modified high parasternal short axis (c) and suprasternal view (d) clarified the anomalous origin of the RPA from the left posterolateral aspect of the distal ascending aorta. AAo: Ascending aorta, Inn vein: Innominate vein, LA: Left atrium, LPA: Left pulmonary artery, MPA: Main pulmonary artery, RPA: Right pulmonary artery, SCV: Superior caval vein. Modified with permission from Gupta and Gupta[8]

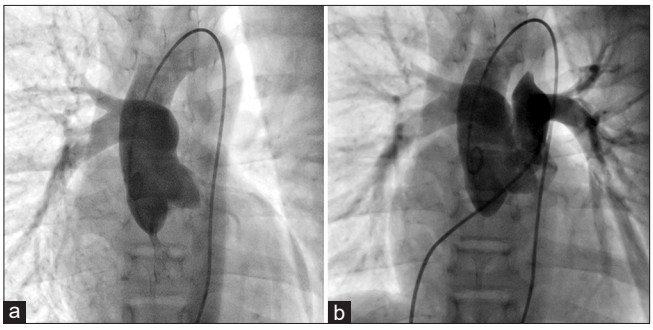

Figure 2: Aortic root angiogram in anteroposterior projection alone (a) and simultaneously with MPA angiogram using 2 catheters and 2 injectors (b) shows an anomalous origin of the RPA from the posteromedial aspect of the ascending aorta close to the MPA. MPA: Main pulmonary artery, RPA: Right pulmonary artery

Table 1: Hemodynamic parameters obtained and calculated from cardiac catheterization and magnetic resonance imaging data at presentation

	Saturations (%)	Pressures (mm Hg)
SCV	66	-
RA	67	a 8 v 10 m 8
RV	68	47 ed 10
LPA	68	47/15/27
RPA	99	120/75/92
PCW	100[a]	Mean 10[a]
LV	99	122 ed 10
Ao	99	122/75/92
FA	99	138/77/96
Calculated variables[#]		
Qp right (L/min)	Cannot be calculated by oximetry	
Qp left (L/min)	4.6	
Qp total (L/min)	Cannot be calculated by oximetry	
Qs (L/min)	4.3	
PVR left (WU)	3.7	
PVR right (WU)	Cannot be calculated by oximetry	
PVR total* (WU)	Cannot be calculated by oximetry	
Cardiac MRI		
Qp right=Flow at RPA (L/min)	1.8	
Qp left=Flow at LPA (L/min)	4.6	
Flow at Asc Ao proximal to RPA (L/min)	6.5	
Qs=Flow at Asc Ao distal to RPA (L/min)	4.6	
Calculated variables using MRI based flow and catheterization based pressure data		
Qp right (L/min)	1.8	
Qp left (L/min)	4.6	
Qp (L/min)	6.5	
Qs (L/min)	4.6	
PVR left (WU)	3.7	
PVR right (WU)	45.5	
Estimated PVR total* (WU)	3.5	

[a]Assumed, [#]Height 156 cm; Weight 43 kg; Hb 10 g/dL; BSA 1.37 m^2; HR 100/min; VO_2 142 L/min/m^2, *Assuming post-RPA re-implantation parallel circulation $PVR_{total}=1/(1/PVR_{left} + 1/PVR_{right})$. PA: Pulmonary artery, Ao: Aorta, FA: Femoral artery, LPA: Left PA, LV: Left ventricle, PCW: Pulmonary capillary wedge, PVR: Pulmonary vascular resistance, RA: Right atrium, RPA: Right PA, RV: Right ventricle, SCV: Superior caval vein, Qp: Pulmonary blood flow, Qs: Systemic blood flow, MRI: Magnetic resonance imaging, Hb: Hemoglobin, BSA: Body surface area, HR: Heart rate

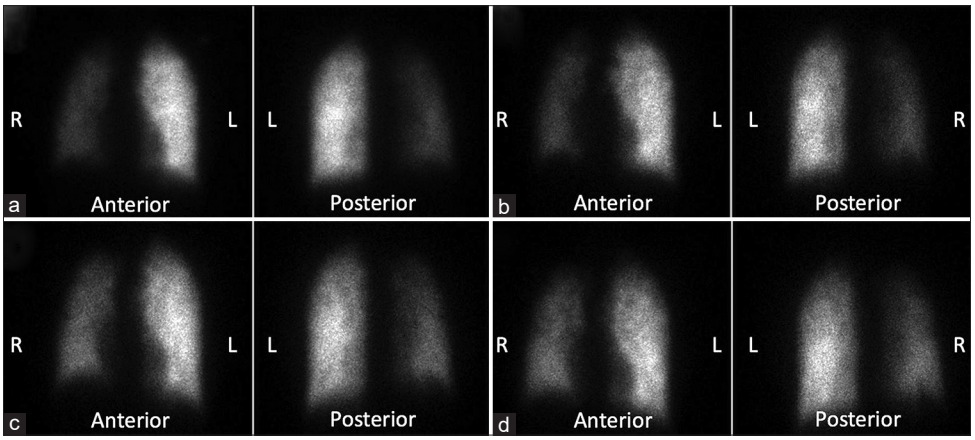

Figure 3: Rest and exercise Tc99 MAA lung scintigraphy scan 2 months (a and b) and 5 years (c and d) postsurgical repair showing significantly reduced perfusion in the right lung at 2 months (a and b) with improvement at 5-year follow-up study (c and d). On both occasions, compared to the rest scan, the perfusion pattern did not change with exercise

measured at 31/7/15 mmHg [Figure 4 and Video 4]. The calculated PVR was 1.3 WU. Tc99 MAA lung perfusion scintigraphy showed 31% of the lung perfusion reaching the right lung with the remaining 69% reaching the left lung. The flow pattern was the same following exercise [Figure 3]. The patient could exercise for 9:24 min on Bruce protocol achieving 10.7 METS, thus confirming good effort tolerance.

Table 2: Hemodynamic parameters obtained by cardiac catheterization at 2 months and 5 years after surgical reimplantation of right pulmonary artery

	2 months postoperative^		5 years postoperative^^	
	Saturation	Pressure	Saturation	Pressure
SCV	64	-	68	-
RA	64	a 8 v 5 m 6	-	a 8 v 5 m 6
RV	65	32 ed 6	-	30 ed 6
PA	64	32/8/17	70	31/7/15
PCW	100[a]	Mean 10[a]	100[a]	Mean 10[a]
LV	99	120 ed10	-	130 ed 10
Ao	99	120/61/87	-	130/67/90
FA	99	130/62/90	99	146/67/93
Calculated variables				
Qp	3.0		3.8	
Qs	3.1		3.7	
PVR	2.3		1.3	
SVR	25.9		22.7	
Calculated variables (individual lungs)^^^				
Qp left lung	2.4		2.6	
Qp right lung	0.6		1.2	
PVR left lung	2.9		1.9	
PVR right lung	10.9		4.2	

[a]Assumed, ^Height 162 cm; Weight 45 kg; Hb 13 g/dL; BSA 1.42 m^2; HR 80/min; VO_2 136 L/min/m^2, ^^Height 178 cm; Weight 54 kg; Hb 14 g/dL; BSA 1.63 m^2; HR 80/min; VO_2 134 L/min/m^2, ^^^Taking lung perfusion scan estimated Qp left: Right=79:21 and 69:31 at 2 months and 5 years following surgery, respectively. Ao: Aorta, FA: Femoral artery, LV: Left ventricle, PA: Pulmonary artery, PCW: Pulmonary capillary wedge, PVR: Pulmonary vascular resistance, RA: Right atrium, RV: Right ventricle, SCV: Superior caval vein, SVR: Systemic vascular resistance, Qp: Pulmonary blood flow, Qs: Systemic blood flow, Hb: Hemoglobin, BSA: Body surface area, HR: Heart rate

DISCUSSION

Anomalous origin of the PA from the aorta is rare congenital heart disease, more common on the right than on the left. In the majority, anomalous origin of PA coexists with other cardiac malformations, the most common being PDA.[1,2] In a few, however, it exists as an isolated anomaly.[1,2,5-8] Irrespective of associated lesions, anomalous origin of the PA is a serious disease, with most patients presenting with heart failure during infancy and up to 70% dying within the 1st year if left unoperated.[1,3,4] Presentation beyond the first decade is limited to case reports.[3,5-8,11-13]

Different sources of blood supply to the left and the right lung result in a unique state of differential flow, pressure, and PVR in each lung, thus making the hemodynamics uniquely complex.[10] The lung with an anomalous PA from the aorta does not contribute to oxygenation while it continues to be exposed to systemic pressures. Besides the lung with anomalous PA, the contralateral lung also may develop vascular changes consequent to excessive flow and other mechanisms such as reflex vasoconstriction or neurogenic crossover from the lung with anomalous

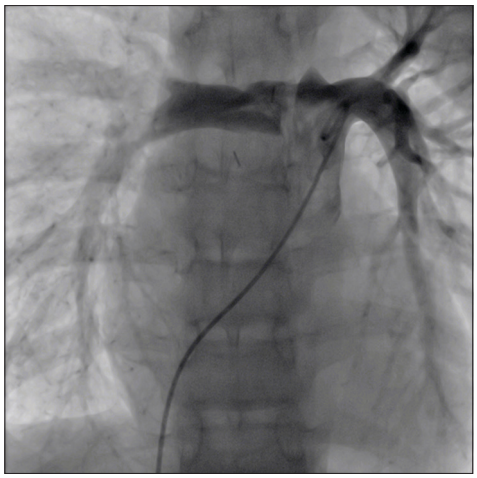

Figure 4: PA angiogram in anteroposterior projection shows unobstructed flow in the RPA [as seen in Video 4]. PA: Pulmonary artery, RPA: Right pulmonary artery

PA.[1,14] The contralateral lung's PVR may sometimes be too high to preclude surgical repair. However, as in our case, certain patients continue to have normal or only a mild elevation of pressure in the lung with normally connected PA.[8] In the absence of associated cardiac lesions, the lung with a normally connected PA receives the entire systemic venous return and PVR in that lung may be calculated using oximetry. Nonetheless, use of flow assessment on phase contrast MRI and pressure data obtained during cardiac catheterization is necessary to measure PVR in the lung supplied by the anomalous PA.

Unlike normal hearts with pulmonary circulation in parallel, the pulmonary blood flow in the isolated anomalous origin of a PA from the aorta is a circulation in series. Surgical reimplantation of anomalous PA would convert pulmonary circulation to circulation in parallel wherein the inverse of the total resistance is the sum of the inverse of its component, and the total resistance of a parallel circuit is lower than the individual components [Figure 5].[10] This is in contrast to circulation in series, wherein the total resistance is the sum of individual components and, therefore, is higher than either lung [Figure 5].[10]

Intuitively, only mild elevation of pressure in the LPA indicates the operable status of our patient. Nonetheless, the age of the patient and limited guidance from the literature about the long-term outcome prompted a detailed hemodynamic assessment. Some authors have used hemodynamic assessment to decide on management but most reports lack information about PVR[13] with none providing information about postoperative hemodynamics.[7,12]

As we have shown, even though PVR in the right lung was prohibitively high, the total PVR of the prospective parallel circuit, predicted to be 3.5 WU, was well within operability limits. This near-perfect prediction

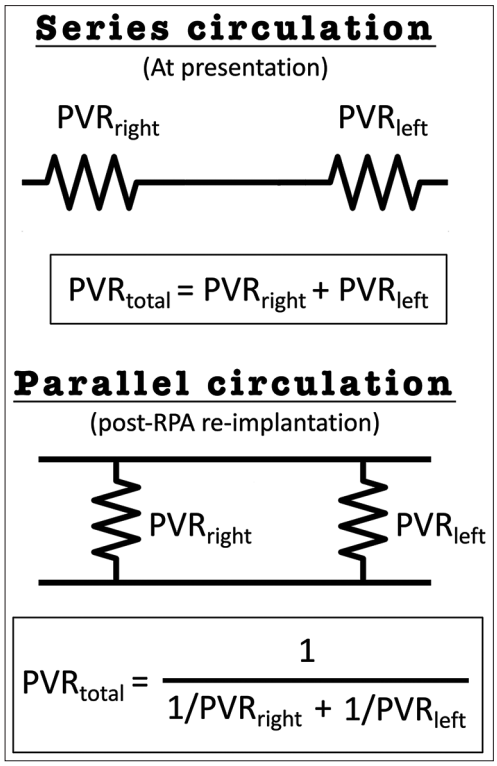

Figure 5: Line diagram showing estimated PVR in post-RPA reimplantation parallel circulation compared to series circulation at presentation. PVR: Pulmonary vascular resistance, RPA: Right pulmonary artery

of postoperative PVR substantiates the accuracy of fundamental principles while also emphasizing the importance of multimodal hemodynamic assessment. Besides other findings, we believe that the prediction of postoperative PVR lends further support to the operability in patients presenting late. With the establishment of parallel circulation postoperatively, the fall in PVR was as per expectation [Figure 5].[10] The reduction in LPA pressure is not only due to reduced flow but also secondary to the escape from harmful neurohormonal influences from the right lung.[7] The same, however, cannot be extrapolated to cases with high PVR in the lung with a normally connected PA although a modest reduction in PVR has been observed even in such cases operated in adulthood.[1,7,12] In an encouraging report, Sha *et al.* described a successful surgical repair of the anomalous origin of RPA despite a coexisting large PDA feeding the left lung at the age of 33 years with the mean LPA pressure of 70 mmHg and ascending aortic pressure measured at 130/98 mmHg. Although a bit unexpected, reported normalization of PA pressure at a 5-year follow-up is reassuring.[7]

Despite a fall in PVR, one may argue against surgical reimplantation as the affected lung with vascular changes is unlikely to contribute well to lung perfusion. The reduction in PVR also may be a reflection of redistribution of pulmonary blood flow with the blood preferentially reaching the less affected left lung. In our patient, nevertheless, the right lung perfusion improved from 21% at the 2-month follow-up to 39% at the 5-year follow-up. This, along with a further reduction in PVR from 2.3 WU at 2-month follow-up to 1.3 WU at the last follow-up, possibly signifies reverse remodeling. In the only other case with long-term follow-up data of a patient operated at 5 months of age, Fontana *et al.* reported that despite normalized PA pressure, the right lung perfusion was 39% at 7 years following surgery.[1] The right lung perfusion, however, reduced to 14% at 10-year postsurgery, thus mandating long-term follow-up to understand the extent of benefit from surgical repair. Much lesser perfusion in the right lung despite normal pressure and total PVR also highlights the problems in the conventional flow-based estimation of PVR. In our case, the calculations of PVR in the individual lungs, based on differential lung perfusion, further highlight favorable reduction in the PVR of the right as well as the left lung [Table 2].

The extent of improvement depends on the age at surgery and baseline PVR of the lung with normal PA connection. Early surgery provides maximum hemodynamic benefits, although normalization may be incomplete if operated after 4 months of age.[1,15] The immediate outcome is largely dependent on the PVR of the lung with a normally connected PA, whereas the long-term benefits will depend on the progression or regression of pulmonary vascular disease in the lung with an anomalously connected PA. Besides other findings, prediction of postoperative PVR may aid in decision-making and favor surgical repair in some patients who present late. Nonetheless, irrespective of the short-term outcomes of surgical repair, considering the risk of residual or recurrent pulmonary hypertension and perfusion deficit in the affected lung a careful long-term follow-up is mandatory.

CONCLUSION

The isolated anomalous origin of the PA has unique hemodynamics. A detailed multimodal hemodynamic evaluation and the use of hemodynamic principles for parallel circulation help in the optimal management of patients presenting late. Despite late presentation, surgical reimplantation in carefully selected patients with normal or mildly elevated contralateral PVR may have favorable outcomes following surgical reimplantation.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient (s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initial s will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed

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Nil.

Conflicts of interest

There are no conflicts of interest.

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