Hemodynamic rounds: Dilemma in the management of disconnected pulmonary arteries with double arterial ducts

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ABSTRACT

Segmental pulmonary arterial hypertension occurs when the right and left pulmonary arteries are nonconfluent and receive blood supply from different sources. High blood pressure confined to limited lung segments may accelerate progression of pulmonary vascular resistance. Calculation of segmental vascular resistance and assessment of operability in such situations are done after integrating catheter hemodynamics, magnetic resonance imaging techniques, or perfusion scintigraphy. When an isolated pulmonary artery perfused by a restrictive ipsilateral arterial duct is associated with a large nonrestrictive contralateral arterial duct connected to the other pulmonary artery, leading to unilateral pulmonary arterial hypertension and features of high vascular resistance, it offers unique challenges to decision-making.

Keywords: Differential lung perfusion, double patent arterial ducts, isolation of pulmonary artery, pulmonary vascular resistance, segmental pulmonary hypertension

INTRODUCTION

In patients with isolation of pulmonary artery, a patent arterial duct may maintain the ipsilateral lung perfusion. The size of this duct determines the adequacy of lung perfusion and its differential pulmonary vascular resistance. As the contralateral lung receives the entire systemic venous return, it may demonstrate a higher than normal pulmonary arterial pressure.[1] A young asymptomatic child was incidentally diagnosed to have isolation of left pulmonary artery supplied by an extremely restrictive left-sided arterial duct from the base of the left innominate artery. A second large nonrestrictive right-sided arterial duct from the right aortic arch led to severe unilateral right pulmonary arterial hypertension. This differential lung perfusion with an obligatory left-to-right ductal shunt to the left lung and a dependent bidirectional ductal shunt to the right lung created difficulties in deciding operability. This report discusses the issues involved in managing patients with different sources of lung perfusion causing segmental elevation of pulmonary vascular resistance.

CASE REPORT

A 3-year-old girl child with adequate growth (13 kg) was treated with occasional use of inhalers for bronchial asthma until a recent echocardiographic evaluation suspected an arterial duct. Absence of abnormal cardiovascular findings delayed a cardiac referral. Clinical examination showed no features of dysmorphic syndromes, normal volume pulses, and normal blood pressure including pulse pressure. Pulse oximetry recorded saturations of 95% in room air in all four limbs. Jugular venous pressure was not elevated. There was no cardiomegaly or abnormal precordial pulsations. The second heart sound showed normal split with loud

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pulmonary component. There were no murmurs or additional sounds. Chest X-ray showed cardiomegaly with a left ventricular contour and a cardiothoracic ratio of 60%, right aortic arch, dilated main pulmonary artery segment, multiple end-on vessels in the medial two-thirds of the right lung without any plethora in the lateral one-third of the lung field along with relative oligemia of the left lung [Figure 1]. There was right-axis deviation to 105° with right ventricular hypertrophy on the electrocardiogram without any evidence of left ventricular volume overload on the lateral chest leads.

Echocardiogram showed intact atrial and ventricular septum, normal-sized ventricles without any volume overload, and normal biventricular systolic function. The dilated pulmonary trunk continued as a large right pulmonary artery. The mean right pulmonary artery pressure predicted from the peak of a trivial Grade I pulmonary regurgitant jet on Doppler was 56 mmHg [Figure 2]. The aortic arch was right sided with mirror image branching pattern. There was a large 9-mm tubular arterial duct from the undersurface of the right arch to the right pulmonary artery, with an early systolic right-to-left shunt and diastolic left-to-right flow. A small restrictive second arterial duct originating from the base of left innominate artery connected to an isolated left pulmonary artery with continuous flows and showed a peak gradient of 64 mmHg [Figure 3]. The pulmonary venous return from both lungs demonstrated normal and equal flow patterns to the left atrium. Abdominal aortic Doppler did not show features of run-off such as diastolic flow reversal.

Absence of clinical signs of aortic run-off to suggest a significant left-to-right shunt at arterial level, absence of murmurs, lack of vascularity in the peripheral one-third of right lung, absence of left ventricular forces on the electrocardiogram, and lack of left ventricular enlargement on echocardiogram coupled with bidirectional flow in the right-sided duct posed a doubt about the operability with concerns of raised pulmonary vascular resistance in the right lung. Despite favorable Z-scores of mitral valve and left ventricular internal diastolic dimension at +1.8 and +1.2, respectively, the left heart dilatation could have a contribution from the obligatory shunt through the left-sided duct. This retained the doubts about the vascular resistance of the right lung. A cardiac catheterization was planned to assess the hemodynamics after informed consent under conscious sedation.

The right pulmonary artery pressures were nearly equal to aortic pressures [Figure 4]. The left pulmonary arterial trace was nonpulsatile trace with a very low mean pressure of 10 mmHg. There was a 6% step-up in oxygen saturations in the right pulmonary artery without any step-down in the descending aorta, indicating absence of basal right-to-left shunt through the right duct [Table 1]. Even though aortic samples showed mild desaturation possibly due to sedation, it was not proved as pulmonary veins were not sampled in the absence of atrial communication. Different sources of pulmonary blood flow posed difficulties to calculate shunt ratio and vascular resistances by Fick principle. To predict the relative differences between systemic and pulmonary vascular resistance, a 6F Berman angiographic catheter was advanced through the pulmonary artery and right arterial duct was occluded with the balloon. There was a significant drop in the right pulmonary artery pressure with a corresponding increase of the aortic pressures accompanied by a relative bradycardia. Left arterial duct angiogram demonstrated a good-sized left hilar pulmonary artery [Figure 5].

Surgical unifocalization of the left pulmonary artery to the pulmonary trunk and division of the right arterial duct on cardiopulmonary bypass was followed by an uneventful recovery. She was asymptomatic at 3-month follow-up with no evidence of residual pulmonary hypertension.

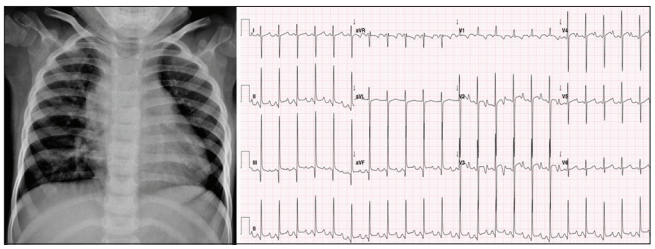

Figure 1: Chest X-ray showing mild cardiomegaly of left ventricular contour, right aortic arch, multiple end-on vessels in the medial two-thirds of the right lung, and lack of vascularity in the lateral one-third of the right lung along with oligemia of the left lung. Electrocardiogram showing right-axis deviation, right ventricular hypertrophy, and fails to demonstrate large left ventricular forces

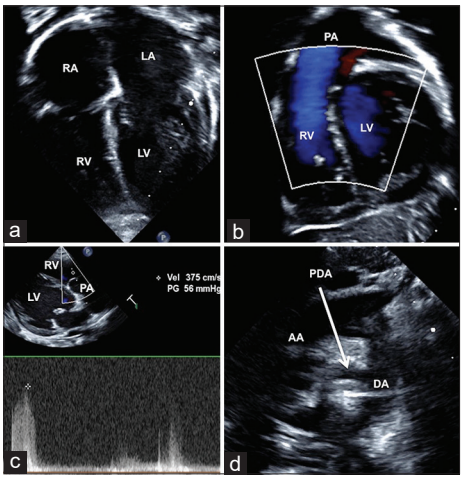

Figure 2: There is no evidence of left ventricular volume overload in apical four-chamber view (a) and subxiphoid short-axis view (b). The mean pulmonary artery pressure predicted from the peak of the pulmonary regurgitation jet on Doppler (c) was 56 mmHg. Long axial view of the aortic arch (d) showing a large right-sided arterial duct

Table 1: Hemodynamic data

Site	Oximetry			Pressures (mmHg)	Postballoon (mmHg)
	PO_2	SO_2	PCO_2		
SVC	43.8	76.2	38		
IVC	35.3	64.5	37		
RA	41.7	73.2	38	1	
RV				SYS-84 EDP-6	
RPA	46.5	79.2	39	80/38/60	72/27/48
LPA	72	92.1	38	10	
LV				SYS-85 EDP-8	
Ascending aorta	65.3	91.7	38	86/42/67	
Descending aorta	72.4	92.9	37	84/43/65	124/70/97

mmHg: Millimeters of mercury, PO_2: Partial pressure of oxygen, SO_2: Oxygen saturation on co-oximetry, PCO_2: Partial pressure of carbon dioxide, SVC: Superior vena cava, IVC: Inferior vena cava, RA: Right atrium, RV: Right ventricle, RPA: Right pulmonary artery, LPA: Left pulmonary artery, LV: Left ventricle, SYS: Systolic, EDP: End diastolic pressure

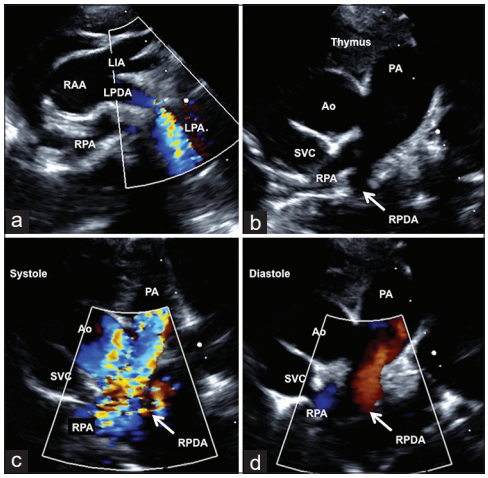

Figure 3: Suprasternal view (a) demonstrates a LPDA from RAA supplying the isolated LPA). A high parasternal short-axis view (b) with color flow imaging in systole (c) and diastole (d) showing pulmonary trunk (PA) continuing as RPA and a RPDA inserting into the RPA with systolic right-to-left flows and diastolic left-to-right flows. LPDA: Left-sided arterial duct, RAA: Right aortic arch, LPA: Left pulmonary artery, RPA: Right pulmonary artery, RPDA: Right-sided arterial duct

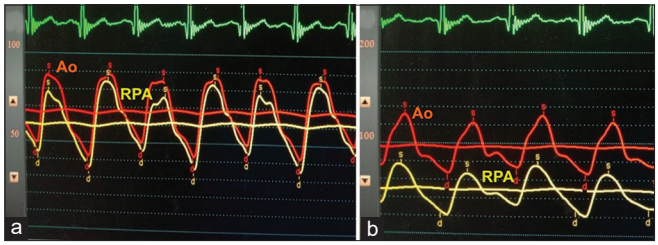

Figure 4: Simultaneous recording of both aortic and right pulmonary artery pressures (a) are almost similar with a 6 mmHg difference between the two vessels. After balloon occlusion of the right-sided arterial duct (b), an increase of aortic pressure and fall of right pulmonary artery pressure was accompanied by a relative bradycardia due to Branham effect

DISCUSSION

Isolation of left pulmonary artery results from involution of the proximal embryonic left sixth (pulmonary) arch, leading to loss of its connection to the pulmonary trunk. A left-sided arterial duct derived from the distal left sixth arch maintains left lung perfusion.[2] The pulmonary artery pressures in the contralateral side may show varying degrees of elevation as it receives the entire systemic venous return.[1] Bilateral arterial duct patency is very rare only observed in patients with pulmonary atresia and nonconfluent pulmonary arteries.[3] Presence of a very large arterial duct on the right side in our patient led to transmission of systemic pressures to the right lung that was already receiving the entire systemic venous return. This resulted in severe right pulmonary arterial hypertension.

Operability in the arterial level shunt is decided based on clinical signs of aortic run-off, cardiomegaly, flow murmurs, plethoric lung fields, and left ventricular forces on electrocardiogram. A left ventricular volume overload coupled with a clear left-to-right shunt on echocardiogram will confirm operability.[4] Our patient lacked all these findings and hence warranted an additional invasive hemodynamic study. It could be argued that the postoperative pulmonary artery pressures after left lung unifocalization would never increase significantly as the left lung vascular bed protected by a very restrictive left-sided arterial duct would accommodate the right ventricular output. However, this strategy carried a risk of a left lung hyperperfusion injury.

Calculation of differential and total pulmonary vascular resistance is challenging when the lungs are perfused from different sources at different pressures. When the oxygen saturations in the right and left pulmonary artery vary, the pulmonary flow cannot be calculated by Fick principle. Cardiac magnetic resonance imaging uses two different methods for quantifying shunts.[5] The volumetric method based on gradient echo cine imaging is not applicable in this patient as the right and left lungs receive blood supply from the right and left ventricle, respectively. The second approach utilizes velocity-encoded phase-contrast imaging of the hilar pulmonary arteries and aorta to quantify the shunt. This method fails when significant turbulence in the pulmonary artery causes aliasing that can be anticipated in the left pulmonary artery in our patient due to

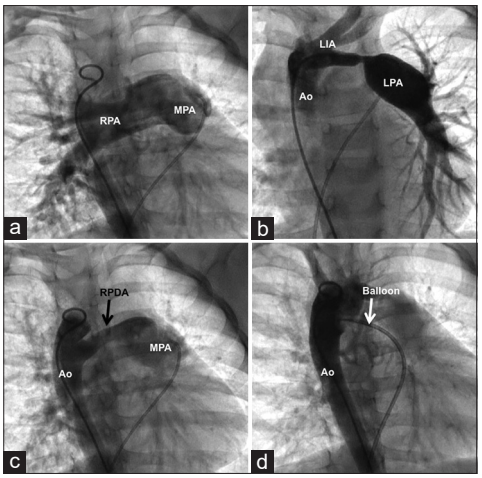

Figure 5: Pulmonary angiogram (a) showing pulmonary trunk (MPA) continuing as a dilated RPA which is dilated in the medial zones and pruned in the lateral zones of right lung. The isolated LPA filled from a left-sided arterial duct (b) arising from the base of LIA. Aortogram (c) showing right aortic arch, a large right-sided tubular arterial duct (RPDA) connecting to the RPA. A complete balloon occlusion of the RPDA (d) is confirmed on an aortogram before assessing the pressures. LPA: Left pulmonary artery, RPA: Right pulmonary artery, LIA: Left innominate artery

restrictive left-sided arterial duct. The phase-contrast imaging could be tweaked to quantify the shunt by analyzing pulmonary venous return from all the individual pulmonary veins and systemic venous return from the superior and inferior caval veins; but such low-velocity flow calculations are not well validated.[5] The additional challenges in a young child are movement and breathing artifacts, claustrophobia, and lack of universal availability of magnet-compatible monitoring tools for a sedated child. As phases of respiration significantly alter the pulmonary and systemic venous return, confusions exist regarding use of multi-average free breathing sequences versus single breath-hold sequence.[6] Very fast heart rates in sedated children may affect the temporal resolution of the phase contrast images. Lung perfusion scintigraphy is another technique employed to assess differential lung perfusion, but limitations exist in delivery of the radionuclide tracer to an isolated pulmonary artery.[7]

Temporary balloon occlusion is one of the tools to decide operability in hypertensive arterial ducts associated with confluent pulmonary arteries.[4,8] We resorted to balloon occlusion of the right arterial duct to assess the operability.[8] Even though there are no guidelines regarding the quantum of reduction of pulmonary artery pressures during balloon occlusion, experts consider a 20%–25% fall in pulmonary artery systolic pressures and diastolic pressure ratio <0.5 as indicators of operability in patients with confluent pulmonary artery.[4,8] In single-lung circulation associated with disconnected pulmonary artery, operability may be indicated even with a minimal fall in pulmonary pressures on balloon occlusion. Even though the right lung received the entire systemic venous return during balloon occlusion of the duct, there was a significant fall in right pulmonary artery pressures. A fall in pulmonary artery pressures with an increase of aortic pressures accompanied by relative bradycardia due to Nicodalani-Branham effect demonstrated operability.[9] This information was very reassuring and permitted us to proceed with unifocalization. If the right pulmonary artery pressures had remained at systemic levels during balloon occlusion, we would have anticipated a stormier postoperative course characterized by pulmonary hypertension and left lung hyperperfusion injury. In such cases, nonselective pulmonary vasodilators such as sildenafil or bosentan could aggravate hyperperfusion in the left lung.[10] Inhalational nitric oxide may be preferred as it matches segmental perfusion to the ventilation contrary to the other vasodilators that fail to maintain a match between perfusion and ventilation. After unifocalization, if the elevated vascular resistance of right lung leads to hyperperfusion lung injury and hemorrhagic edema of the left lung, nonselective pulmonary vasodilators such as sildenafil or bosentan could aggravate hyperperfusion in the left lung. As the ventilation is impaired in the edematous left lung, inhalational nitric oxide will preferentially act on the right lung and will provide a match between the segmental perfusion and ventilation.[10]

CONCLUSIONS

When severe unilateral pulmonary arterial hypertension results from different separate sources of perfusion to the two lungs, practical difficulties exist in calculating differential and total pulmonary vascular resistance between the two lungs. Fick oximetry has limitations to identify differential pulmonary flows to both lungs. Even though alternative methods such as perfusion scintigraphy and magnetic resonance imaging help in identifying differential lung perfusion, their use is not practical in patients with multifocal lung perfusion. A careful clinical assessment combined with astute interpretation of imaging and hemodynamic data is needed to decipher operability in such complex cardiac anatomies.

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 initials 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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