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Original Article

Anomalous origin of one pulmonary artery from the ascending aorta

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Abstract We describe the surgical repair in three infants presenting with one pulmonary artery arising from the ascending aorta, the other artery arising normally from the right ventricle via the pulmonary trunk. Repair consisted of reimplantation of the anomalous pulmonary artery to the pulmonary trunk, in association with repair of associated intracardiac malformations. All patients survived the surgical procedures, and were discharged in stable clinical condition. Subsequently, two of the three patients developed stenosis at the surgical anastomosis relatively early after the initial procedure, and underwent reoperation. Although survival after operation is now expected for this malformation, reports of late results are lacking. Larger numbers of operations are needed before we can reach definitive conclusions.

The origin of one branch pulmonary artery from the ascending aorta in the presence of a pulmonary valve and main pulmonary artery is a very rare congenital cardiac anomaly. *Patients and methods:* Between January 1995 and June 2003, 3 infant girls presented with the origin of one branch artery from the ascending aorta, while the other pulmonary artery originated from the pulmonary trunk which was in continuity with the right ventricular outflow tract. The pulmonary artery that arose from the right ventricle was left in 2 and right in 1 patient. *Results:* At the age 13, 48 and 62 days respectively, the patients underwent surgical repair consisting with reimplantation of the anomalous pulmonary artery branch to the pulmonary trunk in association with repair of intracardiac malformations. There were no hospital deaths. Postoperative complications included: prologed intubation in two patients (10 and 16 days), low output syndrome in 1 patient, cardiac tamponade in 1 patient and seizures in 1 patient. All patients were discharged in good clinical condition. There have been no late deaths. Subsequently, two of the three patients developed stenosis at the surgical anastomosis relatively early after the initial procedure, and after unsuccessuful balloon dilation, underwent surgical reoperation. *Conclusions:* Although operative survival is now possible for this malformation, reports of late results are lacking. Two of the three patients developed stenosis at the surgical anastomosis relatively early after surgery. Larger numbers of operations are necessaries to reach definitive conclusions.

Keywords: Hemitrucus; persistent patency of arterial duct; congenital heart disease

ORIGIN OF ONE PULMONARY ARTERY FROM THE ascending aorta, with the other artery taking its origin from the right ventricle via the pulmonary trunk, is a rare congenital cardiac malformation.[1–5] Although often called "hemitruncus", this name is misleading, since the anomaly has neither development nor anatomic relationships to the common arterial trunk. Unlike the common trunk, the essence of anomalous origin of one pulmonary artery is that there are two separate outflow tracts, each guarded by a separate arterial valve. In the commonest variant, it is the right pulmonary artery that arises from the ascending aorta, at or just above the sinutubular junction. More rarely, the left

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pulmonary artery can take origin from the ascending aorta in about the same location.[4,5] In another form, the right pulmonary artery arises more distally from the ascending aorta, near the origin of the brachiocephalic artery. There is disagreement whether this type should be included with the usual variant, since the right pulmonary artery may take origin from a right-sided arterial duct rather than the ascending aorta. It is rare for the anomalously arising right pulmonary artery also to be associated with other structural heart defects, except for persistence of the arterial duct. Aorto-pulmonary window, and more rarely tetralogy of Fallot, have been reported in a few cases. Conversely, origin of the left pulmonary artery from the ascending aorta is invariably associated with tetralogy of Fallot, a right aortic arch, or both.[6] These differences in the underlying anatomical substrate suggest different developmental mechanisms for these two anatomical variants. Although surgical repair and survival have been reported in a few series of patients with this defect, to the best of our knowledge nothing has been reported concerning survival and outcomes in the long term. In this report, we describe three patients with anomalous origin of one pulmonary artery from the ascending aorta who underwent a successfully one-stage repair, and have been followed up from 1.5 to 8 years.

Patients and methods

Between January 1995 and June 2003, we treated 3 infant girls who presented with origin of one pulmonary artery from the ascending aorta, the other pulmonary artery originating from the right ventricular outflow tract via the pulmonary trunk. The patients were aged 13, 48 and 62 days at presentation. The diagnosis was made by cross-sectional echocardiography in all, and confirmed at cardiac catheterization in the first two patients. It was the left pulmonary artery that arose from the aorta in our first and third patients, and the right pulmonary artery in the other. Both patients with anomalous origin of the left pulmonary artery also had tetralogy of Fallot, while the other patient had tetralogy of Fallot with pulmonary atresia, the intrapericardial pulmonary arteries being supplied by systemic-to-pulmonary collateral arteries. All three patients had a right aortic arch, and all were found to have microdeletions of chromosome 22-q11.

Results

Surgical findings and hospital course

Patient #1. A 13-day-old infant, weighing 3.5 kilograms, presented with mild cyanosis, an arterial saturation of oxygen being 90 percent in room air, and mild tachypnoea. At surgery, we confirmed the anomalous origin of the left pulmonary artery from the ascending aorta in association with tetralogy of Fallot. There was an unrestrictive malalignment ventricular septal defect, severe valvar and subvalvar pulmonary stenosis, and hypoplasia of the ventriculopulmonary junction and the pulmonary trunk. The right pulmonary artery, of 5 millimetres diameter, originated from the pulmonary trunk, while the left pulmonary artery, of 7 millimetres diameter, took its origin from the ascending aorta just above the aortic valve (Fig. 1). Surgical correction consisted in a transatrial closure of the ventricular septal defect using a patch, and transjunctional reconstruction of the right ventricle outflow tract using a patch of autologous pericardium. The left pulmonary artery was detached from the ascending aorta with a cuff of aortic wall and anastomosed end-to-side to the pulmonary trunk, using a continuous running suture of non-resorbable synthetic monofilament. The site of the anastomosis to the pulmonary trunk was further enlarged with autologous pericardium. The oval foramen was left partially open. Cardiopulmonary bypass time was 123 minutes, cross-clamp time 53 minutes, and circulatory arrest lasted for 16 minutes. The postoperative course was complicated by prolonged ventilation over a period of 10 days, and clonic seizures lasting for 5 days, treated successfully

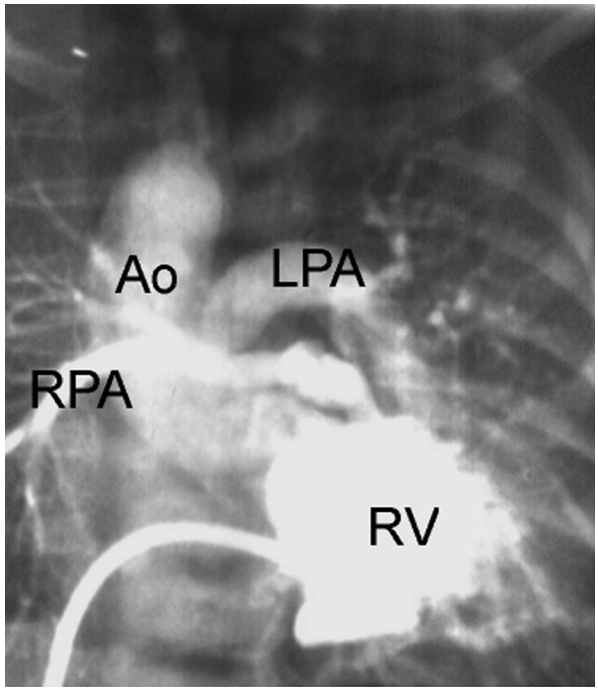

Figure 1.
The cineangiogram, in antero-posterior projection, from our first patient showing anomalous origin of the left pulmonary artery (LPA) from the ascending aorta (Ao). The right pulmonary artery (RPA) takes origin from the infundibulum of the right ventricle (RV).

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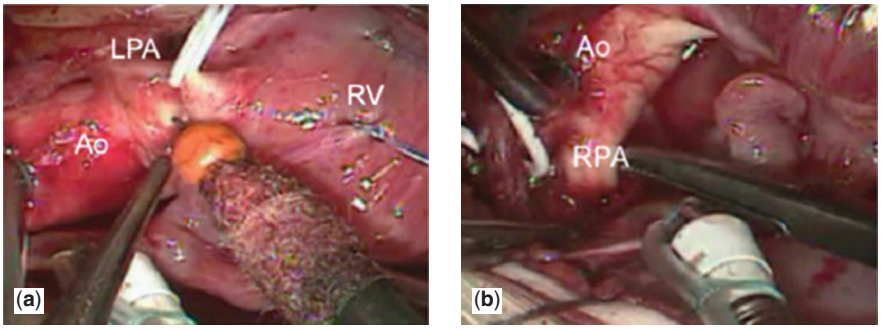

Figure 2.
*An intraoperative view in our second patient of the origin (**a**) of the left pulmonary artery (LPA) from the infundibulum of the right ventricle (RV), and (**b**) the right pulmonary artery (RPA) from the ascending aorta (Ao).*

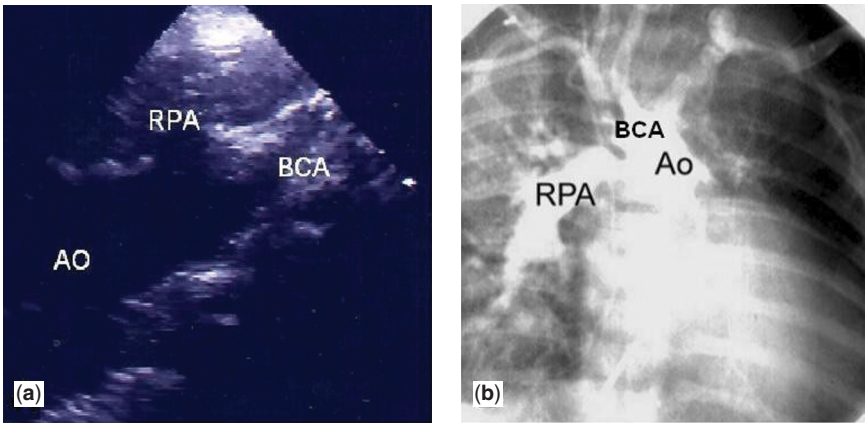

Figure 3.
*Cross-sectional echocardiography (long-axis view) (**a**) from our second patient showing distal origin of the right pulmonary artery from the ascending aorta close to the brachiocephalic artery (BCA). The cineangiogram (**b**) shows origin of the right pulmonary artery from the ascending aorta. Other abbreviations as for Figures 1 and 2.*

with phenilydatoine. Cranial ultrasound revealed an ischaemic area in the left parietal and thalamic region, probably due to air embolism, and diffuse cerebral oedema that subsequently resolved, the cause of this complication never being identified.

Patient #2. A 48-day-old infant weighing 3.6 kilograms presented with mild cyanosis, the arterial saturation of oxygen being 91 percent in room air. At surgery (Fig. 2), we confirmed the echocardiographic and angiographic diagnoses (Fig. 3) of anomalous origin of the right pulmonary artery from the ascending aorta in association with tetralogy of Fallot and pulmonary atresia, noting anatomic but not functional continuity of the pulmonary trunk and left pulmonary arteries with the right ventricular infundibulum. There was an unrestrictive ventricular septal defect, but the muscular outlet septum was absent, there being continuity between the leaflets of the aortic and pulmonary valves. The left pulmonary artery, of 2.5 millimetres diameter, originated from the pulmonary trunk, while the right pulmonary artery, of 5 millimetres diameter, took its origin from the ascending aorta, just above the sinutubular junction. Two small systemic-to-pulmonary collateral arteries arose from the descending aorta and supplied the left pulmonary artery. We made a transventricular incision of 15 millimetres at infundibular level, through which we closed the ventricular septal defect using a patch. We detached the right pulmonary artery from the ascending aorta, and anastomosed it in end-to-side fashion with the pulmonary trunk using a synthetic non-reasorbable monofilament. Continuity between the right ventricular outflow tract and the pulmonary arteries was then established using a 12 millimetre heterograft valved conduit. Because of suprasystemic pressure in the right ventricle at the end of cardiopulmonary bypass, we fenestrated the patch used to close the ventricular septal defect, making an opening of 5 millimetres diameter. The oval foramen was left open. Cardiopulmonary bypass lasted for 230 minutes, cross-clamp time was 50 minutes, and the circulation was arrested for 13 minutes. The postoperative course was complicated by low cardiac output and transient anuria. The infant recovered with supportive therapy, and was discharged from the intensive care unit on the fifth postoperative day.

Patient #3. A 62-day-old premature infant, born at 30 weeks gestation weighing 950 grams, presented with mild cyanosis, having an arterial saturation of oxygen of 86 percent in room air. The infant was also tachypnoeic and dyspneic. At surgery, performed at 9 weeks of age when the infant weighted 2.7 kilograms, we confirmed the cross sectional echocardiographic diagnosis (Fig. 4) of anomalous origin of the left pulmonary artery from the ascending aorta in association with tetralogy of Fallot. The ventricular septal defect was typically unrestrictive and malaligned, and there was severe valvar and subvalvar pulmonary stenosis. The ventriculo-pulmonary junction was 3 millimetres in diameter, while the

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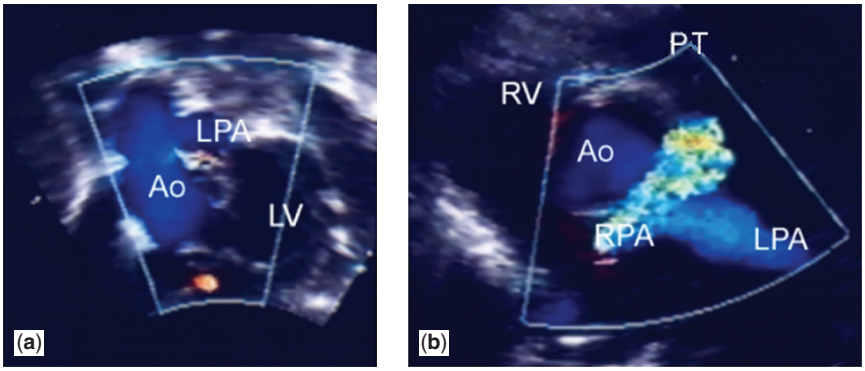

Figure 4.
*The cross-sectional echocardiography, in subcostal view (**a**), in our third patient shows origin of the left pulmonary artery from the ascending aorta. The short-axis view (**b**) shows the different origins of the pulmonary arteries, the left from the ascending aorta and the right from the right ventricle through the pulmonary trunk (PT). Other abbreviations as for previous figures.*

pulmonary trunk had a diameter of 4 millimetres. The right pulmonary artery, of 3 millimetres diameter, took origin from the pulmonary trunk, while the left pulmonary artery, of 4 millimetres diameter, originated from the ascending aorta at the sinutubular junction. A patent right arterial duct connected the concavity of the right aortic arch to the right pulmonary artery. Surgical correction consisted in transatrial closure of the ventricular septal defect with a patch, combined transatrial-transpulmonary myotomy of the muscular bands obstructing the right outflow tract, and transjunctional reconstruction of the right ventricle outflow tract with a pulmonary homograft monoleaflet patch. The left pulmonary artery was detached from the ascending aorta and anastomosed in end-to-side fashion to the pulmonary trunk using a continuous running suture of synthetic non-resorbable monofilament. The oval foramen was left open. Cardiopulmonary bypass lasted for 158 minutes, cross-clamping for 69 minutes, and the circulation was arrested for 37 minutes. The postoperative course was complicated by cardiac tamponade on the fourth postoperative day, leading to exploration and drainage of the pericardial space, without any finding of active bleeding, and prolonged ventilation for 16 days.

There were no hospital deaths and all 3 patients were discharged home in stable clinical conditions.

The postoperative clinical and cross-sectional echocardiographic findings at discharge are shown in Table 1.

Table 1. Postoperative clinical and echocardiographic data at discharge after the first operation.

Pt	SaO_2 (%)	ΔP RVP/RAP† (mmHg)	ΔP PT/RPA (mmHg)	ΔP PT/LPA (mmHg)
#1	90	25	15	15*
#2	91	90	75*	57
#3	99	48	25	30*

Abbreviations: ΔP: pressure gradient in millimetres of mercury (mmHg); RVP: right ventricular pressure; RAP: right atrial pressure; RPA: right pulmonary artery; LPA: left pulmonary artery; PT: pulmonary trunk.
†Indirect calculation of the pressure in the right ventricle by measuring the pressure gradient between the right ventricle and the right atrium due to tricuspid valve regurgitation and adding the value of the pressure in the right atrium (5–10 mmHg).
*Anastomosed branches

Follow-up

Complete clinical and echocardiographic follow-up was obtained in all 3 patients at 18, 30, and 104 months after initial surgery, respectively. All patients are still alive, asymptomatic, and in stable clinical condition at the most recent follow-up. The latest cross-sectional echocardiographic and lung scan data are shown in Table 2.

Reinterventions

Our first patient required percutaneous dilation of the left pulmonary artery 27 months after surgery, with transient improvement, but required surgical reoperation for stenosis at the origin of the left pulmonary artery 48 months after the initial surgery because of a gradient of 55 millimetres of mercury across the site of stenosis (Fig. 5a), along with recurrent obstruction within the right ventricular outflow tract. Repair consisted of incision of the postero-inferior division of the parietal extension of the muscular outlet septum, resection of an additional anomalous muscle band crossing the right ventricular outflow tract, and replacement of the patch initially used to reconstruct right ventricular outflow tract with a larger homograft patch. Included in the repair was augmentation of the anastomosis of the left pulmonary artery to the pulmonary trunk.

Our second patient underwent three attempts at percutaneous dilation of the site of anastomosis of the right pulmonary artery at 2, 8, and 16 months after surgery. Due to stenosis of the homograft conduit, and bilateral stenosis of the pulmonary arteries (Fig. 5b), the patient underwent surgical reoperation 30 months after the initial repair. Reoperation consisted of enlargement of the right ventricle outflow tract with homograft material, which was extended

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Table 2. Echocardiographic and lung scan data at the latest follow-up.

Pt	Time after correction (time after reoperation) (months)	RVP+RAP (mmHg)	ΔP RV/PT (mmHg)	ΔP PT/RPA (mmHg)	ΔP PT/LPA (mmHg)	Lung scan (%)	
						Left	Right
#1	104 (56)	44	10	20	30	45	55
#2	30 (1)	56	8	42	23	53	47
#3	18	35	10	15	13	43	57

Abbreviation: RVOT: right ventricle outflow tract. Other abbreviations as for Table 1

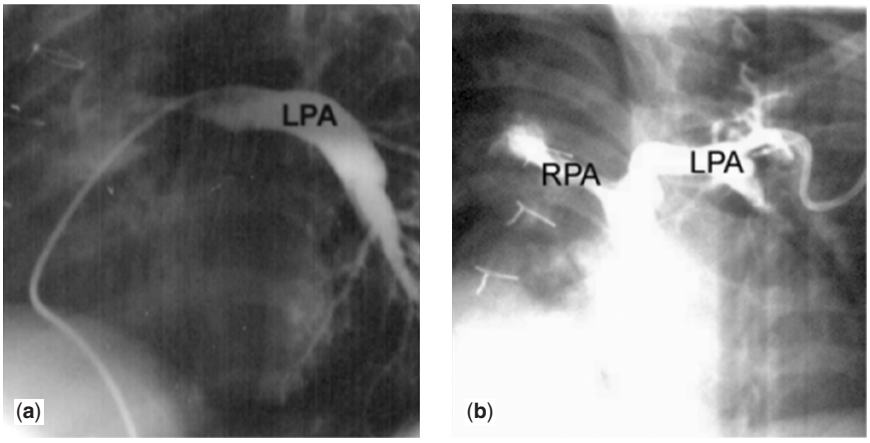

Figure 5.
*Cineangiograms before reinterventions, in antero-posterior view showing (**a**) stenosis at the level of the anastomosis of the reimplanted left pulmonary artery in our first patient, and (**b**) stenosis at the level of the anastomosis of the reimplanted right pulmonary artery in our second patient.*

to augment the origin of the right and left pulmonary arteries. Our third patient has not required any additional intervention.

Discussion

We have described 3 patients having anomalous origin of one pulmonary artery from the ascending aorta in association with tetralogy of Fallot and a right aortic arch. In two of the patients, multiple procedures were needed to correct stenosis of the anastomosis between the pulmonary artery and the pulmonary trunk, but all 3 patients are now in good clinical condition at mid-term follow-up.

This particular congenital cardiac malformation must be repaired early to avoid pulmonary vascular disease. Indeed, it has been shown that histological features of pulmonary vascular disease can be seen as early as the first month of life with this defect.[6]

Since the first successful surgical correction reported in 1961,[7] various surgical techniques have been proposed for the repair of this rare anomaly.[1,6,8] The morphology, however, is variable, and experience with each suggested technique is limited.[4,5] The selection of a primary anastomosis versus interposition of a synthetic graft has not appeared to affect either the operative mortality or survival at 1 year.[8] Direct anastomosis of the anomalous pulmonary artery to the pulmonary trunk, nonetheless, seems to be the preferred procedure.[1,9] Although surgical repair and survival have been reported in a few series of patients with this defect,[6,8,10] long term survival and late outcomes, to the best of our knowledge, have not. In our small series, two of the three patients developed stenosis of the surgical anastomosis. In these patients, we had used two different techniques to create the anastomosis, using a cuff of aortic tissue in the first patient, and using a valved heterograft conduit without a cuff in the second patient. In both, we employed a continuous non-resorbable suture to create the anastomosis.

It is unclear if the anastomotic obstruction is a common feature of aortic origin of one pulmonary artery, or if it is more likely in the setting of tetralogy of Fallot. Interventional procedures failed to provide long-lasting relief of the obstruction, and surgical reintervention was undertaken in both patients. Long-term follow-up of these patients is essential to permit timely reintervention, albeit that, in our experience, larger numbers of operations are necessary to reach definitive conclusions.

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References

1. Fyler DC. Origin of a pulmonary artery from the aorta (hemitruncus). In: Nadas (ed.). Pediatric Cardiology, Henley & Belfus, Inc., Philadelphia, 1993, pp 697–699.
2. Jacobs JP, Quintessenza JA, Gaynor JW, Burke RP, Mavroudis C. Congenital heart surgery nomenclature and database project: aortopulmonary window. Ann Thorac Surg 2000; 69: 44–49.
3. Kirklin JW, Barratt-Boyes BG. Truncus arteriosus. In: Cardiac Surgery, 2nd edn. Churchill Livingstone Inc., New York, 1993, Ch. 38, pp 1131–1151.
4. Gerlis LM, Ho SY, Smith A, Anderson RH. The site of origin of non confluent pulmonary arteries from a common arterial trunk or from the ascending aorta: its morphological significance. Am J Cardiovasc Pathol 1990; 3: 115–120.
5. Duncan WJ, Freedom RM, Olley PM, Rowe RD. Two-dimensional echocardiography identification of hemitruncus: anomalous origin of one pulmonary artery from ascending aorta with the other pulmonary artery arising normally from right ventricle. Am Heart J 1981; 102: 892–896.
6. Kutsche LM, Van Mierop LH. Anomalous origin of a pulmonary artery from the ascending aorta: associated anomalies and pathogenesis. Am J Cardiol 1988; 61: 850–856.
7. Armer RM, Shumaker HB, Klatte EC. Origin of the right pulmonary artery from the ascending aorta. Report of a surgically corrected case. Circulation 1961; 24: 662–668.
8. Fontana GP, Spach MS, Effman EL, Sabiston DR Jr. Origin of a pulmonary artery from the ascending aorta. Ann Surg 1987; 206: 102–113.
9. Penkoske PA, Castaneda AR, Fyler DC, Van Praagh R. Origin of pulmonary artery branch from ascending aorta. Primary surgical repair in infancy. J Thorac Cardiovasc Surg 1983; 85: 537–545.
10. Sechtem U, Jungehulsing M, de Vivie R, Mennicken U, Hopp HW. Left hemitruncus in adulthood: diagnostic role of magnetic resonance imaging. Eur Heart J 1992; 13: 1040–1044.