Anomalous aortic origin of the pulmonary arteries: Case series and literature review

Salvatore Agati[1], Carlos Guerra Sousa[2], Felice Davide Calvaruso[1], Rosanna Zanai[1], Ivana Campanella[1], Daniela Poli[1], Alfredo Di Pino[1], Luca Borro[3], Fiore Salvatore Iorio[4], Massimiliano Raponi[5], Robert H Anderson[6], Simone Reali[1], Andrea De Zorzi[1], Aurelio Secinaro[7]

[1]Mediterranean Congenital Heart Center – Bambino Gesù – San Vincenzo Hospital, Taormina, Italy, [2]Department of General Surgery, Hospital Santo Tomas, Panamá, [3]Department of Clinical Directors, Bambino Gesù Children's Hospital IRCCS, Rome, Italy, [4]Department of Pediatric Cardiology and Cardiac Surgery, Bambino Gesù Children's Hospital IRCCS, Rome, Italy, [5]Medical Director, Bambino Gesù Children's Hospital IRCCS, Rome, Italy, [6]Emeritus Professor, Royal College of London, London, UK, [7]Department of Imaging, Bambino Gesù Children's Hospital IRCCS, Rome, Italy

ABSTRACT

Anomalous origin of the pulmonary arteries from the ascending aorta is a rare, but severe clinical entity necessitating a scrupulous evaluation. Either the right or the left pulmonary arteries can arise directly from the ascending aorta while the other pulmonary artery retains its origin from the right ventricular outflow tract. Such a finding can be isolated or can coexist with several congenital heart lesions. Direct intrapericardial aortic origin, however, must be distinguished with origin through a persistently patent arterial duct. In the current era, clinical manifestations usually become evident in the newborn rather than during infancy, as used to be the case. They include respiratory distress or congestive heart failure due to increased pulmonary flow and poor feeding. The rate of survival has now increased due to early diagnosis and prompt surgical repair, should now be expected to be at least 95%. We have treated four neonates with this lesion over the past 7 years, all of whom survived surgical repair. Right ventricular systolic pressure was significantly decreased at follow-up. Our choice of treatment was to translocate the anomalous pulmonary artery in end-to-side fashion to the pulmonary trunk. Our aim in this report is to update an Italian experience in the diagnosis and treatment of anomalous direct origin of one pulmonary artery from the aorta, adding considerations on the lessons learned from our most recent review of the salient literature.

Keywords: Direct reimplantation of pulmonary artery, fetal diagnosis, neonatal diagnosis, persistent ductus arteriosus, pulmonary artery and neonatal urgency, Tetralogy of Fallot

INTRODUCTION

When either the right or right or left pulmonary artery arises directly from the ascending aorta, then of necessity the pulmonary arteries themselves are discontinuous, with the other artery retaining its origin from the right ventricle. It follows, therefore, that the lesion does not represent half of a common arterial trunk. Estimated incidence of direct aortic origin of a pulmonary artery is about 0.1%[1,2] although Liu *et al.* recently reported a prevalence of 0.33% in a single-center experience.[3] Direct aortic origin of the right pulmonary artery is much more common than the left one since its first description in 1868,[4] about 300 cases have been reported in literature, most of them in relation to surgical correction. As already emphasized, it is inappropriate to describe the entity as "hemitruncus" since there are separate aortic and pulmonary valves. In this report, we describe

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Address for correspondence: Dr. Salvatore Agati, Director Congenital Heart Surgery, Mediterranean Congenital Heart Center, Bambino Gesù, Taormina, Italy. E-mail: sasha.agati@opbg.net

our experience with four neonatal cases encountered over the last 7 years. We reviewed 45 surgical articles from the PubMed and clinical key databases. Of these, 9 were retrospective series,[3,5,6] whereas 36 were case reports.[7-12] The overall surgical experience described is based on 137 patients, of whom 50 were neonates. The most common associated defect, reported in 70 cases, was patency of the arterial duct. We aimed to clarify the classification of the lesion and to provide a better understanding of the underlying morphogenesis.

METHODS

Between January 2011 and April 2018, four consecutive neonates underwent surgery at the Centro Cardiologico Pediatrico del Mediterraneo, Taormina, Italy, for correction of direct intrapericardial aortic origin of a pulmonary artery.

Clinical records were reviewed to document clinical features, operative procedures, and perioperative courses. All patients had a preoperative echocardiogram. Computed tomography and magnetic resonance imaging were used in the last two patients to confirm the diagnosis and optimize the surgical planning. The preoperative imaging tools permitted us to establish the site of origin of the anomalous pulmonary artery, the presence of additional intracardiac defects, the estimated right ventricular pressure, and the presence or absence of right ventricular dysfunction. Surgical correction consisted of direct implantation of the anomalous pulmonary artery, as described in the literature.[13] Briefly, through a standard median sternotomy approach, we dissected and mobilized the aorta and the left and right pulmonary arteries. On cardiopulmonary bypass, the anomalous artery was snugged to prevent pulmonary overcirculation and loss of perfusion pressure than was separated from the ascending aorta without aortic cross-clamping, with the primary suture of the aortic origin. In the case of anomalous origin of the right pulmonary artery, extensive mobilization of the intrapericardial branches beyond the lobar artery permitted attachment in posterior fashion to reach the pulmonary trunk in the absence of tension, using a continuous 6-0 PDS suture. Similar mobilization was used so as to reimplant the left pulmonary artery. Postoperative all patients received dopamine (range: 5–10 mcg/kg/min and mean infusion time: 74 h) and inhaled nitric oxide (20–30 ppm/h, mean 49 h) with mean intubation time of 34 h. All received postoperative oral sildenafil (range: 2 mg/kg/day), suspended at 3-month follow-up. Table 1 reports demographics, preoperative details, and follow-up.

Case 1

A 3 kg symptomatic 10-day-old male baby was referred with a diagnosis of congestive heart failure. During the physical examination, a systolic murmur was audible at the left parasternal border, along with an intense second heart sound. Oxygen saturations were measured at 95% in the right arm, 90% in the right leg, 92% in the left arm, and 88% in the left leg. The patient was diaphoretic and tachypneic. The electrocardiogram (ECG) showed sinus rhythm, QRS left-axis deviation, and hybrid pattern from V1 to V5. Chest X-ray showed cardiomegaly, pulmonary cone dilation, and increased pulmonary flow. The echocardiogram showed a bicuspid aortic, severe isthmic coarctation with a closed arterial duct, and a systolic pulmonary pressure of 100 mmHg. The coarctation was resected using an extended end-to-end anastomosis. On the 2nd postoperative day, due to the persistence of elevated systolic pressure and an inability to extubate the patient, a new transthoracic echocardiography revealed intrapericardial aortic origin of the right pulmonary artery [Figure 1]. Surgical correction was undertaken on the same day, and after delayed sternal closure, there was a satisfactory postoperative evolution. The

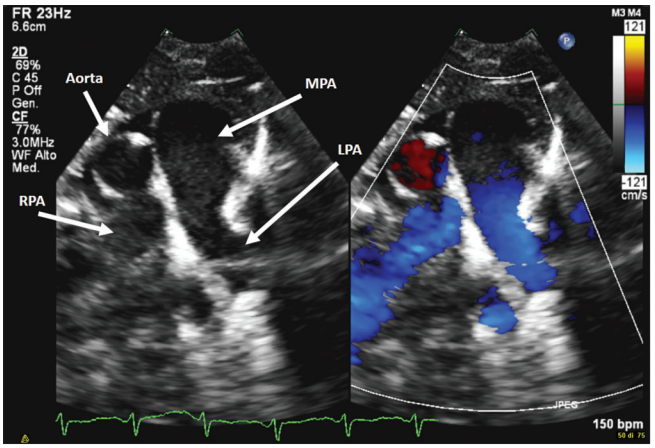


Figure 1: Preoperative echo short-axis parasternal view. MPA: Main pulmonary artery, LPA: Left pulmonary artery, RPA: Right pulmonary artery, aorta

Table 1: Demographics and follow-up

Case	Age at the diagnosis	Weight (kg)	Type	Prenatal diagnosis	Associate anomalies	Symptoms	Age at surgery (days)	Outcome	Follow-up
1	10 days	3	AORPA	No	CoAo	CHF	14	Alive	7 years
2	2 days	2.5	AOLPA	No	No	Tachypnea, failure in thrive	22	Alive	6 years
3	3 days	3.6	AORPA	No	PDA, PFO	Emergency	4	Alive	3 years
4	22 days (34-week GA)	2.1	AORPA	No	No	Emergency	23	Alive	3 months

AORPA: Anomalous origin of the right pulmonary artery, AOLPA: Anomalous origin of the left pulmonary artery, PDA: Persistent ductus arteriosus, CoAo: Aorta coarctation, PFO: Persistent foramen ovale, GA: Gestational age, CHF: Congestive heart failure

pulmonary systolic pressure was subsequently measured by echocardiography at 35 mmHg. The patient, at 7-year follow-up, is asymptomatic, well grown, with no gradient across the reimplanted pulmonary artery [Figure 2] and normal systolic pulmonary pressures.

Case 2

A 2.4 kg 2-day-old male baby was referred to our intensive care unit because of the finding of a systolic murmur on auscultation of the chest. The patient was asymptomatic, with a normal pattern in the ECG. Transthoracic echocardiography revealed a persistently patent oval foramen and a right aortic arch with intrapericardial aortic origin of the left pulmonary artery. The systolic pulmonary pressure was measured at 85 mmHg. At catheterization, it was confirmed that the aortic arch was right sided, with a closed arterial duct, hypertrophy of the right ventricle, and the right pulmonary artery arose in isolation from pulmonary trunk. On the 22nd postnatal day, due to increasing of tachypnea and reduction in breastfeeding, the patient underwent surgical correction. The postoperative course was uneventful, and the patient was discharged with a systolic pulmonary pressure of 40 mmHg. After 6 years of follow-up, he remains asymptomatic, with normal systolic pulmonary pressures.

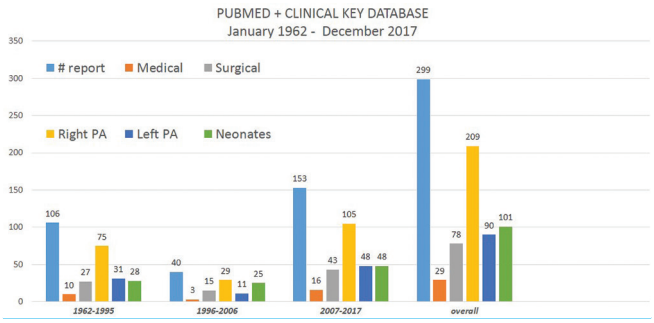
Case 3

A 3.6 kg 3-day-old female baby with diaphoresis and polypnea was transferred to our intensive care unit with the suspicion of transposed arterial trunks. During the physical examination, the oxygen saturation was measured at 90%, and a continuous murmur was heard in the right infraclavicular region. The ECG showed sinus rhythm, right axis deviation of the QRS complex, a prominent R wave in lead V1, and significant enlargement of the right heart chambers. The chest X-ray showed cardiomegaly and increased pulmonary flow. Transthoracic echocardiography revealed intrapericardial aortic origin of the right pulmonary artery, along with an oval fossa defect and persistent patency of the left-sided arterial duct. Systolic pulmonary pressure was calculated at 84 mmHg. The findings were confirmed by a computed tomographic scan [Figure 3]. The patient deteriorated, and emergent surgical correction was performed. During the postoperative period, the patient suffered a pulmonary hypertension crisis, which required treatment with nitric oxide and pulmonary vasodilators. She was successfully discharged on the 14th postoperative day. After 20-month follow-up, she required percutaneous angioplasty due to stenosis at the site of reimplantation of the right pulmonary artery. After 3-year follow-up, she is asymptomatic, with a pulmonary arterial pressure of 35 mmHg.

Case 4

A 22-day-old male baby, born prematurely at 34-week gestation, was transferred from a regional neonatal intensive care unit after diagnosis of intrapericardial aortic origin of the right pulmonary artery. Weighing 2.1 kg at birth, he was severely desaturated and bradycardic. He was losing weight despite gavage nutrition. During his

Table 2: Overall reported cases on PubMed and clinical key from January 1962 to December 2017



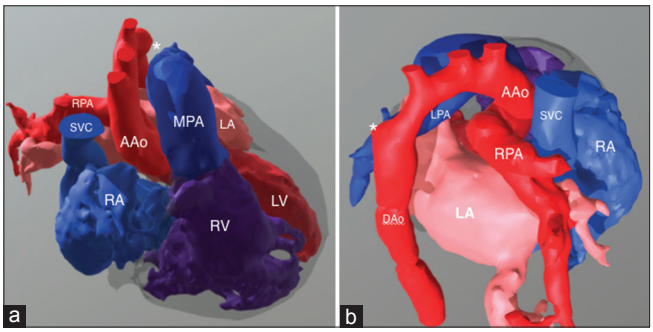


Figure 3: Software-aided anatomical segmentation of the heart and great vessels based on postiodinate contrast computed tomography images shows three-dimensional anatomy of Case number 3: a) disconnected branch pulmonary arteries with the right pulmonary artery originating from the mid-ascending aorta (in red) and left pulmonary artery from the main pulmonary artery (in blue); b) the left-sided aortic arch isthmus is enlarged probably in keeping with "ductal ampulla" (asterisk) due to left ductus arteriosus ligament in this position

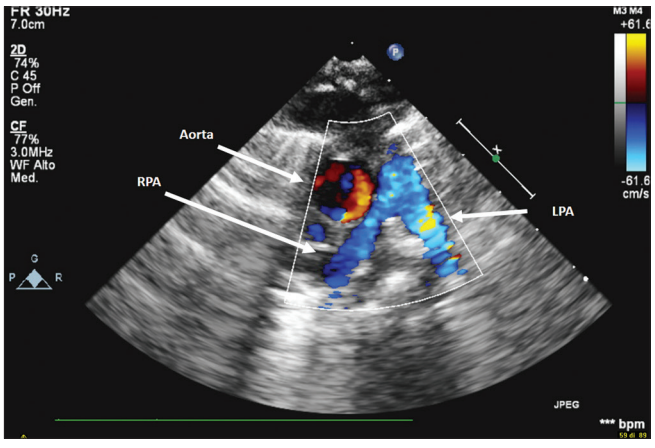


Figure 2: Seven-year follow-up – echo short-axis parasternal view. Aorta, RPA: Right pulmonary artery, LPA: Left pulmonary artery

Table 3: Clinical outcome of the surgical reported cases

Type	Author/ years	Cases	AORPA/ AOLPA	Type of surgery	CPB/ neonates	Diagnosis	Time of surgery median days	Associate anomalies	Outcome
Case report over all 36 patients	2007-2017	36	18/18	23 DR, 13 Dacron, Gore-Tex, PA banding, VSD closure, RMBTS	26/16	25 TTE/2 TEE 4 fetal, 17 angio-CT 16 angiocatheter	60	8 PDA, 8 TOF	1 death
Multiple cases	Nathan/2007 (USA)	16	14/2	11 DR 5 PA	16/10	16 TTE, 5 catheterism; 3 patients missed diagnosis on TTE	84	13 PDA	1 death
	Kajihara/2008 (Japan)	8	8/0	7 DR 1 PTFE	7/6	8 TTE/8 catheterism	35	7 PDA, 1 ARSA, 1 CoA, 1 ASD	All alive (8)
	Erdem/2010 (Turkey)	7	1/6	7 DR	7/4	7 TTE, 2 catheterism	44	3 PDA, IAA 1, 1 APW	All alive (7)
	Amir/2010 (Israel)	12	10/2	8 DR 3 PA	12/8	12 TTE 6 catheterism	49	12 PDA, 1 VSD 2 PFO,1 CoA	1 death
	Talwar/2014 (India)	11	5/6	7 DR 1 pericardial 1 saphenous 1 RPA band with innominate LPA shunt 1 innominate to RPA shunt without bypass	9/0	11 TTE 11 catheterism	2190	9 TOF	2 deaths
	Vasquez/2015 (Mexico)	5	3/2	5 PA	0/0	4 TTE 3 angio-CT 2 catheterism	4, 8 years (1752 days)	2 PDA, 1 IAD, 1 SVSOA, 1 SOLPA	1 death
	Liu/2015 (China)	19	17/2	14 DR	14/7	19 TTE	3 months median (4 days-21 years)	12 PDA 1 TOF 11 ASD, 1 ARSCA, 1 LAA + RDA + ARSCA, 2 VSD, 1 CoA, 2 APSD, 1 ASD	14 been operated and alive, the others 5 no described
	Yang/2015 (China)	11	7/4	6 DR 3 anastomosed to the MPA with an aortic ring 1 aortic flap 1 aortic flap and PA	11/0	11 TTE 11 catcheterism 11 angio-CT	382 days (12, 7 months)	4 PDA, 3 TOF, 4 VSD, 5 ASD, 2 APW, IAA 1	1 death
	Cho/2015 (South Korea)	12	8/4	7 DR, 5 PA	12/5	12 TTE 6 catheterism 5 angio-CT	152 days	9 PDA 2 TOF 2 CoA,4 VSD, 5 PFO, 1 ASD, 1 MAPCAS	1 death

CPB: Cardiopulmonary bypass, AORPA: Anomalous origin of the right pulmonary artery, AOLPA: Anomalous origin of the left pulmonary artery, DR: Direct reimplantation, PA: Pericardial augmentation, TTE: Transthoracic echocardiography, ASD: Atrial septal defect, MAPCAS: Multiple anomalous pulmonary collaterals arteries, APW: Aortopulmonary windows, ARSCA: Anomalous right subclavian artery, APSD: Aortopulmonary septal defect, IAA: Interrupted aortic arch, LPA: Left pulmonary artery, RPA: Right pulmonary artery, MPA: Main pulmonary artery, TEE: Transesophageal echocardiography, PDA: Persistent ductus arteriosus, CoA: Aorta coarctation, VSD: Ventricular septal defect, PFO: Persistent foramen ovale, LAA: Left aortic arch, SOLPA: Stenosis of origin of the left pulmonary artery, IAD: Interatrial defect, SVSOA: Subvalvular stenosis of the aorta, CT: Computerized tomography, PTFE: Polytetrafluorethylene, ARSA: Aberrant right subclavian artery, TOF: Tetralogy of Fallot, RMBTS: Right modified blalock taussig shunt

stay in the neonatal intensive care unit, transthoracic echocardiography revealed a right ventricular pressure of 100 mmHg. Magnetic resonance imaging then confirmed the aortic origin of the right pulmonary artery, with a closed arterial duct [Figure 4]. At arrival in our department, he was transferred promptly to the operating room for surgical correction. During the postoperative course, he suffered one episode of ventricular fibrillation, which was successfully treated and developed a postcardiotomy pericardial effusion. He was discharged on the 27th postoperative day with normal weight gain. At 3-month follow-up, the right ventricular pressure is 30 mmHg.

DISCUSSION

Although the entity has often been described as a "hemitruncus," the presence of separate aortic and

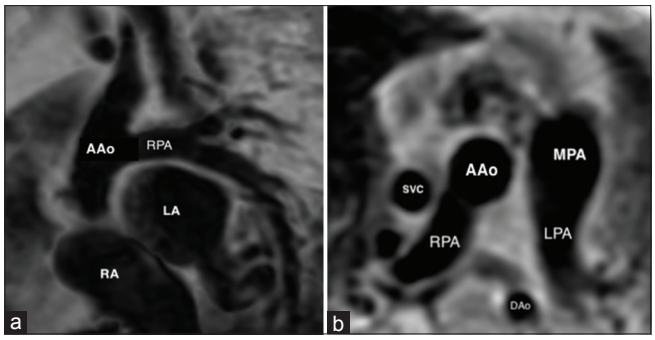


Figure 4: Sagittal (panel a) and axial (panel b) magnetic resonance imaging views acquired on Case number 4 using black-blood sequences clearly show disconnected branch pulmonary arteries with the right pulmonary artery arising from the mid-ascending aorta. Left pulmonary artery is in continuity with the anatomical main pulmonary artery

pulmonary valves shows this to be an inaccurate description.[14] The intrapericardial origin of the aorta also distinguishes the lesion from discontinuous pulmonary arteries, with ductal origin of the pulmonary artery lacking direct origin from the right ventricle. The lesions can simply be described in terms of intrapericardial aortic origin of one pulmonary artery, with further description provided of any associated lesions.[15] Several theories have been offered in terms of morphogenesis. One suggesting an error in development of cells migrating from the neural crest to the fifth or sixth aortic arches[16] can be discounted simply because there is no fifth pharyngeal arch seen during normal development. Unequal partitioning of the "conotruncus" by eccentric coalescence of the right and left outflow tract ridges[17,18] can similarly be discounted since the outflow cushions separate the intermediate and proximal parts of the developing outflow tract. An eccentric growth the protrusion of the dorsal wall of the aortic sac, which forms the aortopulmonary septum, offers a much more realistic explanation.[19]

Our review of the literature reveals ongoing confusion in the description and reporting of clinical examples of intrapericardial aortic origin of one pulmonary artery. Our interrogation of the chosen databases has provided details of 299 patients reported from 1962[13] January to December, 2017 [Table 2]. Seventy-three percent (78/107) of these reports are dedicated to surgical correction. It transpires that 60% of the reported patients were male, with anomalous origin of the right pulmonary artery being much more frequent than the left. Neonatal correction has increased dramatically over the past two decades, with 63% of reported patients being treated as neonates over the period from 1996 to 2006. Ongoing surgical reports over the past 10 years provide details of 137 patients [Table 3]. Direct implantation is the surgical technique of choice in all patients and may be performed with (87%) or without CPB. Transthoracic echocardiography remains the gold standard for the diagnosis although we failed to diagnose our first patient when using this technique. A few reports now describe fetal diagnosis.[10,19-22] Our own experience confirms that despite severe perinatal symptoms, surgical correction in the neonatal period provides excellent short- and long-term outcomes. Pulmonary hypertension, nonetheless, remains one of the most common causes of the death in premature and newborn, with or without congenital heart disease. Our experience shows that intrapericardial aortic origin of one pulmonary artery must be considered in all cases of perinatal pulmonary hypertension and can become a surgical emergency if not early detected.

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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