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CASE IMAGE

Abnormal origin of right pulmonary artery from the ascending aorta in an infant ("*Hemitruncus*")

Alexandros Tsoutsinos[1] | **Ioannis Germanakis**[2] | **Meletios Kanakis**[3] | **George Samanidis**[4] | **Stefanos Despotopoulos**[1] | **Theofili Kousi**[5] | **Spyridoula Katsilouli**[6] | **Olga Karapanagiotou**[6] | **Dimitrios Bobos**[1]

[1]Department of Paediatric Cardiology and Adult Congenital Heart Disease, Onassis Cardiac Surgery Centre, Athens, Greece

[2]Department of Paediatrics, University Hospital Heraklion, School of Medicine, University of Crete, Heraklion, Greece

[3]Department of Paediatric Cardiac Surgery and Congenital Heart Surgery, Onassis Cardiac Center, Athens, Greece

[4]1st Department of Adult Cardiac Surgery, Onassis Cardiac Surgery Center, Athens, Greece

[5]Department of Anaesthesiology, Onassis Cardiac Surgery Center, Athens, Greece

[6]Department of Radiology and Computed Tomography, Onassis Cardiac Center, Athens, Greece

Correspondence
Alexandros Tsoutsinos, Department of Paediatric Cardiology and Adult Congenital Heart Disease, Onassis Cardiac Surgery Center, Athens, Greece.
Email: tsutsi@otenet.gr

Key Clinical Message

Anomalous origin of right pulmonary artery from the ascending aorta is a rare congenital malformation and it needs surgical management. Consequences of this condition affect lead to pulmonary hypertension and severe pulmonary vascular disease.

Abstract

Anomalous origin of right pulmonary artery from the ascending aorta is a rare congenital heart malformation that results in early infant mortality affecting the right pulmonary artery more than the left. These patients are at risk for the early development of significant pulmonary hypertension. The surgical management during the early period of life is imperative.

KEYWORDS

anomalous pulmonary artery, ascending aorta, congenital heart disease, hemitruncus arteriosus, pulmonary hypertension

Anomalous origin of right pulmonary artery from ascending aorta (AORPA) is a rare congenital heart malformation with a prevalence of 0.33% in patients with congenital heart disease.[1–3] The term "*hemitruncus arteriosus*" is described for the same disease, but is considered inadequate, because unlike—truncus arteriosus—two arterial valves aortic and pulmonary are present.[1]

AORPA is 4 to 8 times more common than the anomalous origin of the left pulmonary artery (AOLPA).[2] Both conditions may be associated with other abnormalities including ventricular, atrial septal defects, tetralogy of Fallot, interrupted aortic arch, aortopulmonary window, isthmic hypoplasia, and patent ductus arteriosus (PDA).[2] Embryologically, malformations of the 5th and 6th aortic

arches are implicated. Diagnosis is usually established by echocardiography. Cardiac computed tomography (CT) and magnetic resonance imaging (MRI) with 3D reconstruction are used for confirmation of suspected cases and better preoperative planning.[1–3]

The pathophysiology resulting from this condition affects both lungs and leads to pulmonary hypertension and severe pulmonary vascular disease. AORPA is a lethal condition and 70% of untreated patients are dead by 6 months of age and 80% by 1 year.[2,3]

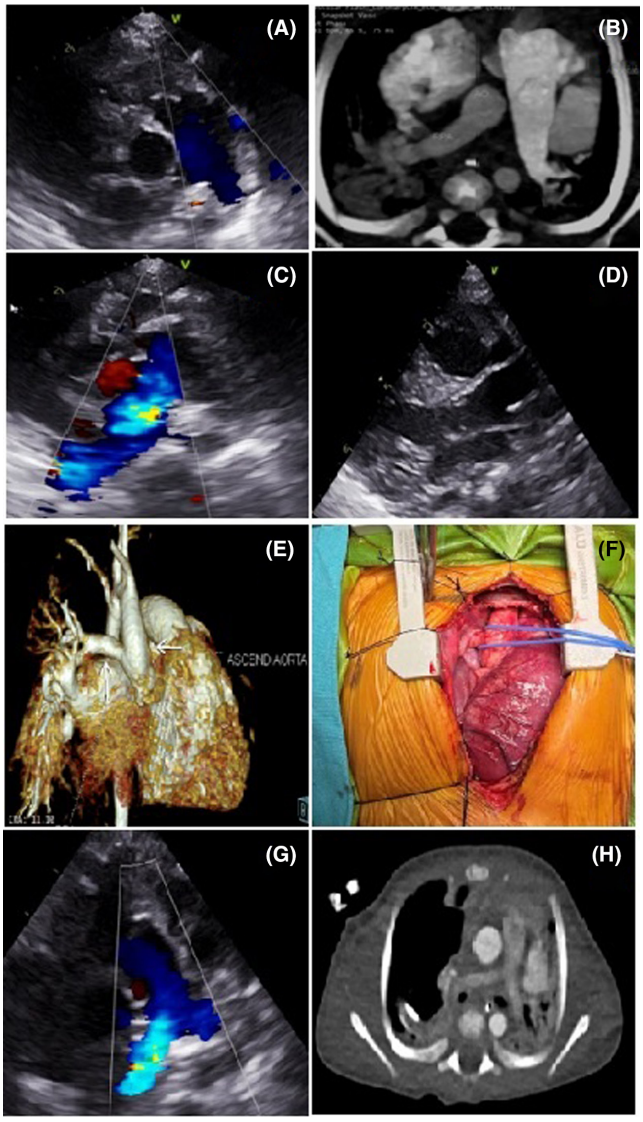

FIGURE 1 (A): Short axis echocardiographic view depicting the main pulmonary artery (MPA). Only the left pulmonary artery (LPA) originates from MPA. (B): Long axis view showed a vessel that arose lateral and posterior from the ascending aorta. (C): Short-axis view showed a flow in the location of the right pulmonary artery but no vessel connected to the trunk of the pulmonary artery. (D): CT angiography showed the connection ascending aorta and right pulmonary artery. (E): CT reconstruction showed the complete anatomy of malformation. (F): Intraoperative image. (G): Postoperative echocardiography showing the good flow pattern through the RPA. (H): Postoperative CT showing the good surgical result. RPA is connected to MPA.

A 2-month-old male infant (weight: 4.1 kg) was referred to hospital with failure to thrive, tachypnea, and cyanosis. Room air saturation was 77%, chest X-ray showed cardiomegaly and consolidation of the right upper lobe, heart rate was 160/min, and the liver was palpable 2 cm below the costal margin, with cold extremities. In the blood sample pH was 7.33 and lactic acid 8.8. The infant was intubated and inotropic support was started. Transthoracic echocardiography showed the trunk of the pulmonary artery and the left pulmonary artery, but the right pulmonary artery could not be visualized especially from the short axis view (Figure 1A,B). The diagnosis of abnormal origin of the RPA from the ascending aorta laterally was suspected (Figure 1C) and confirmed by CT angiocardiography. (Figure 1D,E). There was also significant dilatation of the right ventricle, which compressed the left ventricle in systole. Tricuspid insufficiency showed increased pressure with an estimated right systolic pressure of 70–75 mmHg.

The infant underwent a median sternotomy. Severe right ventricular dilatation was prominent and the right pulmonary artery originated from the lateral aspect of the ascending aorta (Figure 1F). PDA was ligated and under cardiopulmonary bypass the right pulmonary artery reimplanted to the main pulmonary artery. A patent foramen ovale of 3 mm was deliberately left due to the existing high pulmonary artery pressure. Postoperatively the right pulmonary artery could be identified to the expected anatomic location (Figure 1G,H). Inhaled NO started just after its arrival to ICU and stopped on postoperative day 11. Postoperative course was stormy due to the existed pulmonary hypertension and weaning process from the ventilator remained unsuccessful for several times. Finally, the infant extubated on postoperative day 25 and discharged from the hospital on day 40.

AORPA is a rare congenital heart malformation and it needs surgical management. Pathophysiologically, a large left-to-right shunt from the aorta to the pulmonary circulation and then to the left atrium is created. The contralateral lung is therefore subjected to the entire right ventricular output. This anomaly must be corrected the soon as possible because can lead to irreversible results.[4]

AUTHOR CONTRIBUTIONS

Alexandros Tsoutsinos: Investigation; writing – original draft. **Ioannis Germanakis:** Investigation. **Meletios Kanakis:** Investigation; methodology; visualization; writing – original draft. **George Samanidis:** Visualization; writing – original draft. **Stefanos Despotopoulos:** Investigation. **Theofili Kousi:** Investigation. **Spyridoula Katsilouli:** Investigation. **Olga Karapanagiotou:** Investigation. **Dimitrios Bobos:** Writing – original draft.

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DATA AVAILABILITY STATEMENT
The data that support the findings of this study are available on request from the corresponding author.

CONSENT
Written informed consent was obtained from the parent of the children to publish this report in accordance with the journal's patient consent policy.

ORCID
Meletios Kanakis https://orcid.org/0000-0002-9708-0523
George Samanidis https://orcid.org/0000-0001-6355-1932

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