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**Anomalous right pulmonary artery from the aorta**

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## Abstract

Anomalous origin of the right pulmonary artery is a rare form of congenital heart disease. It usually presents in early infancy with heart failure and rapid development of pulmonary hypertension. There are about 131 cases reported in the literature and we report one such case.

**MeSH:** right pulmonary artery, aorta, pulmonary hypertension, patent ductus arteriosus, heart defects, congenital

## Introduction

Anomalous right or left pulmonary artery arising from the aorta are relatively rare congenital heart disease entities. There are several reports, mostly describing anomalous right pulmonary artery arising from the posterior aspect of the ascending aorta.[1] The clinical presentation is in early childhood with congestive cardiac failure and with the onset of early pulmonary hypertension. In some, there is an absence of cardiac failure or a very short abbreviated period of failure followed by the development of pulmonary vascular disease.

This rare disease is thought to come about because of failure of migration of the embryonic branch pulmonary artery to reach the pulmonary trunk end of the truncoaortic sac before septation occurs. There are known associations with an anomalous right pulmonary artery (RPA): patent ductus arteriosus (PDA), interrupted aortic arch (IAA), aortopulmonary window (AP window) are frequently encountered Tetralogy of Fallot (TOF) tends more to be associated with a left anomalous pulmonary artery (LPA) arising from the aorta. It has also been reported that LPA arising from the aorta is more associated with chromosome 22 deletions (as compared to RPA arising from the aorta), in addition to TOF, a right aortic arch or both (TOF and R aortic arch).[1]

We present a case of an anomalous (RPA) arising from the aorta (Ao) in a 13 year old Chinese boy.

## Case report

This boy was followed up from the age of two months. At the time of his presentation, his mother related that at birth he was slightly tachypnoeic but fed well when compared to his siblings. The family only sought treatment at two months because of the onset of cyanosis. An echocardiogram at the time diagnosed a hemitruncus – a single trunk arising from the heart with the LPA being supplied by the PDA, and the RPA arising from the ascending aorta. Another paediatric cardiologist from a different institution also concurred. Unfortunately, his mother was told that at that time no repair was feasible for the child in Malaysia and he was treated conservatively. A review echocardiogram was performed at 11 years of age, as he represented with haemoptysis . The echo revealed a pulmonary trunk (MPA) in the normal position with a LPA arising from the MPA, and a PDA. The RPA was seen to come off the aorta, hence, a cardiac catheterization was arranged to confirm the diagnosis and also for measurement of pulmonary pressures. Unfortunately, his pulmonary pressures were systemic in both the MPA (130/76) and LPA. His RPA was presumed to have the same pressures as the ascending Ao (AAo 130/78) and was not entered. The picture of the pulmonary vasculature was also of irreversible pulmonary hypertensive changes (see figures 1,2). He has subsequently been placed on calcium channel blockers, as his MPA pressures decreased slightly with the administration of 100% oxygen. He is presently cyanosed, clubbed but asymptomatic.

Figure 1 An NIH catheter was placed in the MPA and an angiogram performed revealed the MPA, LPA and descending Ao (that was connected by the PDA)..

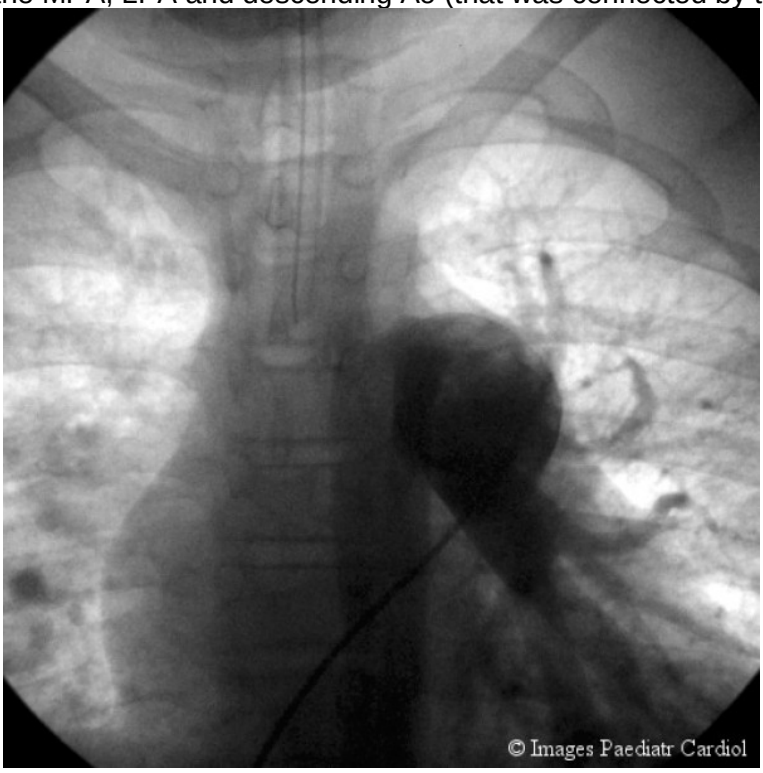
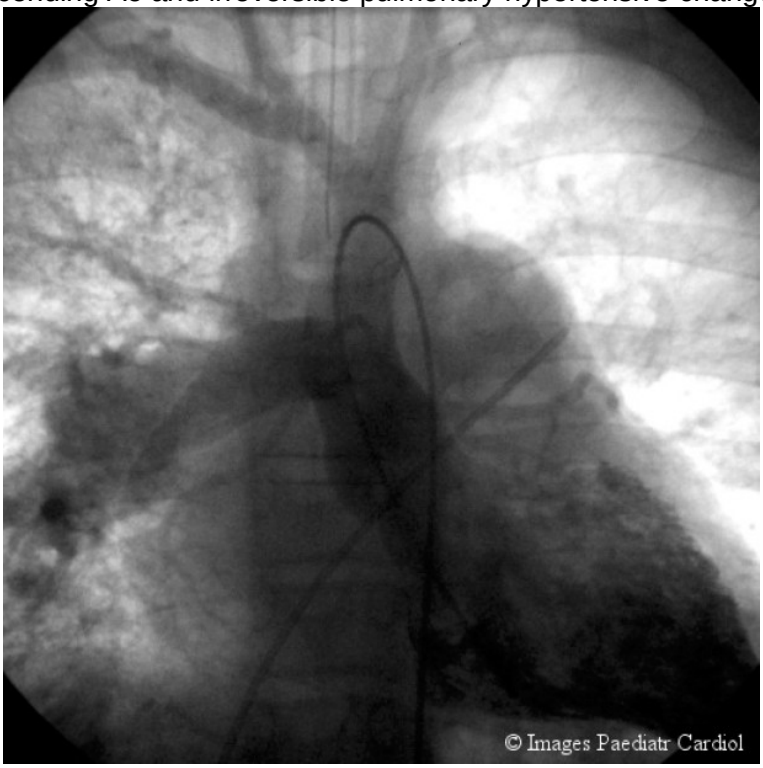


Figure 2 An LV gram showing the RPA arising from the posterior part of the ascending Ao and irreversible pulmonary hypertensive changes.




## Discussion

Pulmonary artery arising from the aorta was first described by Fraentzel in 1868 and since that time, about 131 cases have been reported.[2,3] This entity is rare, and can involve LPA or RPA or both.[4] The RPA originates anomalously more commonly than the LPA.[2] There are a number of associations that arise commonly in this anomaly including TOF, AP window, and IAA. The association of chromosome 22 deletions has also been described in both anomalous LPA and RPA.[2] In this case, the anomaly was associated with a PDA.

Pulmonary hypertension has been described as starting early in most of the cases reported, and intervention had been mainly carried out in infancy, although there are rare cases of non-operated survivors in adulthood with apparently normal pressures in the lungs.[5] This case also illustrates that echocardiography can diagnose these lesions. If the diagnosis is in doubt, angiography should be performed early.

Various types of surgical techniques are employed for the repair of the anomalous pulmonary arteries, with the most frequently employed being direct anastomosis. Alternative techniques include end to end anastomosis with a synthetic graft, interposition with a homograft patch, aortic flap, etc.[2] The results in the neonatal period have been promising, but due to the rarity of this lesion, it is difficult to predict the optimal timing or the best repair technique.

In conclusion, anomalous RPA from the aorta is a rare congenital heart disease, and necessitates thorough investigation as early surgical repair is associated with good outcomes.

## References

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