Abnormal origin of the left pulmonary artery from the descending aorta and heterotaxy syndrome: an undescribed phenotypic association

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Abstract

Extensive screening in a newborn with prenatal suspicion of VACTERL syndrome identified an anomalous origin of the left pulmonary artery from the descending aorta with an arterial duct and left aortic arch, and normal intra-cardiac anatomy. Other anatomical anomalies suggested heterotaxy syndrome. At one-month-old, re-implantation of the 3.5 mm left pulmonary artery was performed by direct tension-low anastomosis. Post-operative course was complicated by severe left pulmonary atelectasis, and the patient died 20 days later.

Case report

A full-term male newborn who had prenatal suspicion of VACTERL syndrome (oesophageal atresia, anal atresia, right kidney agenesis, ectopic pelvic left kidney) without cardiac anomalies. We report here such a case in association with heterotaxy syndrome.

Anomalous origin of the left pulmonary artery from the aorta is extremely rare.1 In most cases, the anomalous origin of the left pulmonary artery originates from the ascending aorta and is strongly associated with tetralogy of Fallot, right aortic arch and anomalous subclavian artery.1,2 To the best of our knowledge, only 3 cases of anomalous origin of the left pulmonary artery arising from the descending aorta have been previously reported.3–5 None of them had associated cardiac anomalies.

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Received: 26 December 2019 Revised: 10 May 2021 Accepted: 13 May 2021 First published online: 4 June 2021

Keywords:
CHD; heterotaxy syndrome; anomalous left pulmonary artery; descending aorta; vascular surgery
left bronchus by local oedema. Post-operative CT imaging confirmed a small-calibre, but patent re-implanted left pulmonary artery far away from the left main bronchus. Despite selective right intubation and optimal medical treatment, the patient presented a severe infection and necrosis of the left lung and died 20 days after the surgery.

Discussion

Anomalous left pulmonary artery from the descending aorta is exceedingly rare. The three cases reported so far share common characteristics: left-sided aortic arch, anomalous origin of the left pulmonary artery origin from the ipsilateral wall of the descending aorta at a significant distance from the left subclavian artery, patent arterial duct connecting the pulmonary trunk to the under surface of the left aortic arch in two cases and no associated intra-cardiac lesions. Our case is very similar, with a patent arterial duct, and in addition, a left superior caval vein to coronary sinus. These cases are very different from the more frequent, albeit rare, anomalous origin of the left pulmonary artery from the ascending aorta.

Various embryologic hypotheses have been reported about the constitution of anomalous origin of branch pulmonary arteries from either right or left aortic arch. Developmentally, central pulmonary arteries have a dual origin: the pulmonary trunk originates from the embryonic outflow tract, while the right and left pulmonary arteries originate from the ventral pharyngeal mesenchyme (splanchnic plexus) connecting within the aortic sac to the pulmonary arch, formerly named sixth aortic arch. Intra-pulmonary arteries develop within the lung buds and connect to the dorsal aorta through the intersegmental arteries, which disappear when the connection with the central pulmonary arteries occurs. On the left side, the left pulmonary artery derives from the proximal part of the left pulmonary arch, while the distal part gives rise to the arterial duct. Various hypotheses have been raised for anomalous origin of one pulmonary artery from the ascending aorta, the most prevalent being an asymmetric development of the aortic sac. This hypothesis is not applicable for the anomalous origin of left pulmonary artery from descending aorta. This rare anomaly could be due to a lack of development of the left pulmonary arch, total when no duct is present, or partial, involving only the proximal part of the arch, like in our case and others. Another subject of interrogation is the nature of the vessel connecting the descending aorta and the intra-parenchymal left pulmonary artery. If this anomaly results from lack of development of the left pulmonary arch, this vessel should be of another origin, like in tetralogy of Fallot with pulmonary atresia. In this setting, major aortopulmonary collateral arteries originating from the descending aorta are considered persistent intersegmental arteries. In our case like in other cases described, the argument against this anomalous vessel being a major aortopulmonary collateral artery is that it does supply the entire left pulmonary arterial tree, while major aortopulmonary collateral arteries, if they also enter in the hilum, usually supply a lobe or a segment and not the entire lung. However, some cases with tetralogy of Fallot with pulmonary atresia with one lung supplied by a single collateral artery arising from the descending aorta have been published.

Figure 1. CT imaging showing the left pulmonary artery (LPA) arising from descending aorta. Three-dimensional frontal CT angiogram with LPA (*) arising from the lateral surface of the descending aorta (DAo)

Figure 2. CT imaging showing intra-thoracic laterality defects. (a) Two-dimensional frontal CT angiogram with bilateral morphologically left bronchi (late first branching of both bronchi), RB = right bronchus, LB = left bronchus. (b) Three-dimensional CT imaging showing similar external shape of the two atrial appendages; RAA = right atrial appendage; LAA = left atrial appendage; Ao = aorta; PA = pulmonary artery; RV = right ventricle; LV = left ventricle.
The anatomical differences regarding associated anomalies in anomalous origin of the left pulmonary artery from the ascending aorta and anomalous origin of the left pulmonary artery from the descending aorta may reflect a different embryological background. Anomalous origin of the left pulmonary artery from the ascending aorta is commonly associated with tetralogy of Fallot and right aortic arch, which could be related to disturbances in neural crest cells migration. Indeed, anomalous origin of the left pulmonary artery from the ascending aorta was described in patients with DiGeorge syndrome and Waardenburg syndrome.\(^1\)

On the contrary, the rare cases reported of anomalous origin of the left pulmonary artery from the descending aorta display no associated intra-cardiac lesions and have a left aortic arch, despite a similar alleged mechanism, namely the absence of proximal pulmonary arch.

The diagnosis of heterotaxy in our case was made according to Lin’s criteria, based on the association between: abdominal situs abnormality (midline liver), spleen abnormality (asplenia), left bronchial isomerism, bilateral superior vena cava and intestinal malrotation, despite the absence of intra-cardiac defects.\(^2\) The definition of heterotaxy and the very term “heterotaxy” itself are still subject to many controversies. According to the IPCCC ICD-11 nomenclature, heterotaxy is defined as “a congenital malformation in which the internal thoraco-abdominal organs demonstrate abnormal arrangement across the left-right axis of the body”, right and left isomerisms are defined as variants of heterotaxy, and “heterotaxy does not include normal and mirror-imaged arrangements of the internal organs (situs solitus or inversus)”.\(^3\)

According to Van Praagh, heterotaxy is defined as “an abnormal symmetry of certain viscera and veins (lungs, liver, caval veins) and situs discordance between various organ systems and between the various segments of the heart”.\(^4\) Although some anatomical studies claimed that atrial appendages were uniformly isomeric in the setting of heterotaxy, several other reports demonstrate that this is not always the case.\(^5\) We cannot affirm in our case that there was isomerism of the pectinate muscles, even if their external appearance looked similar on the CT scan, but the presence of lateralised atrial appendages would not preclude heterotaxy anyway.\(^6\)

Moreover, the absence of intra-cardiac defects does not rule out the diagnosis of heterotaxy, as it is now recognised that 10–16% of heterotaxy patients have no CHD.\(^7\) The fact that the electrocardiogram was normal with a normal P wave axis does not rule out heterotaxy neither, as assessed by Wren et al.\(^8\) Some authors have defined a CHD in the setting of heterotaxy as “any intracardiac lesion or an abnormality of venous returns to the heart”.\(^9\) For all these reasons, the diagnosis of heterotaxy, made on the association of left bronchial isomerism, abdominal organs laterality defects and bilateral superior caval vein, is unequivocal in our patient. Dis Harmony between the arrangement of the bronchi and the abdominal organs is relatively frequent (20% in the series by Yim)\(^10\), but the association of left bronchial isomerism and asplenia is rare. All these considerations are in favour of an analytic approach based on accurate description of bronchi, abdominal organs and cardiac features independently, including the venous and arterial connections.\(^11\)

We have no embryologic explanation to propose for the unusual association between anomalous origin of the left pulmonary artery from the descending aorta and heterotaxy syndrome found in our patient.

Complete diagnosis and anomalous origin of the left pulmonary artery origin analysis are not always easy by echocardiography alone. CT angiogram and three-dimensional reconstruction were very helpful in our case to detail the lesions and guide the surgeon.

Overall, the surgical repair strategies for the re-implantation of the anomalous origin of the left pulmonary artery to the pulmonary trunk show good long-term results despite the late high risk of post-operative restenosis across the anatomicostic site. When the distance between anomalous origin of the left pulmonary artery and pulmonary trunk is large, cardiopulmonary bypass is required during the procedure. Although early repair avoids pulmonary hypertension and irreversible pulmonary vascular disease, outcome of our patient was complicated by ventilation issues related to post-operative oedema compressing the left bronchus and severe bronchectasis.

In conclusion, we present a rare case of anomalous origin of the left pulmonary artery from the descending aorta, associated with a particular form of heterotaxy, with dis harmonious patterns of bronchi and abdominal organs, bilateral superior caval veins and no intra-cardiac lesions. This complex and unusual case reinforces the growing consensus that all the still pending controversies about heterotaxy should be resolved by the accurate description of each thoracic and abdominal organ independently, including the segments of the heart and the venous and arterial connections.\(^12\)

Financial support. This research received no specific grant from any funding agency, commercial or not-for-profit sectors.

Conflict of interest. None.

Ethical standards. This article does not contain any studies with animals performed by any of the authors.

References


