

A rare case of common arterial trunk with coarctation of the aorta: a case report

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Background

A common arterial trunk is a relatively uncommon type of congenital heart defect. The anomaly is caused by an incomplete conotruncal septation. Arch anomalies, such as interruption, are associated with 10–20% of cases. We present a rare case of common arterial trunk with coarctation of the aorta and patent ductus arteriosus (PDA).

Case summary

A term baby who was discovered to have a murmur on examination, for evaluation of the murmur an echocardiography was performed on Day 2 of life, which revealed the diagnosis of a common arterial trunk and coarctation of the aorta. The baby was given prostaglandin and intubated. Due to his poor general condition, he underwent an emergency pulmonary artery branch banding. He needed another 5 days in the intensive care unit to be stabilized before undergoing full repair.

Discussion

Our patient has a common arterial trunk with a tricuspid competent truncal valve. The trunk is subdivided further into ascending aorta and main pulmonary artery. The pulmonary artery provided two branches and a PDA that connected to the descending aorta. The aortic branching pattern was as usual, there was an area of tight coarctation and posterior shelf just after the origin of the left subclavian artery before the descending aorta–PDA junction. The presence of coarctation may be considered as the left side of the spectrum of morphological changes toward the formation of aortic arch interruption (Type 4 Van Praagh).

Keywords

Common arterial trunk • Congenital heart anomalies • Aortic coarctation • Case report

ESC Curriculum

2.1 Imaging modalities • 2.2 Echocardiography

Learning points

- Truncus arteriosus associated with coarctation of the aorta is a very rare condition, which can be diagnosed by echocardiography.
- Some of the persistent truncus arteriosus cases are not included in the current classifications.
- Bilateral pulmonary artery band may be required before complete repair in case of haemodynamic instability of these cases.

Introduction and background

A common arterial trunk is a relatively uncommon type of congenital heart defect. Arch anomalies, such as interruption, are associated with 10–20% of cases.¹ The aorta, pulmonary arteries, and coronary arteries are all supplied by a single great artery that emerges from the

base of the heart. The anomaly is caused by an incomplete conotruncal septation. Around the 32nd day of embryological development, a developmental arrest of conotruncal septation occurs. The truncal vessel carries blood from both ventricles.² The pulmonary arteries branch from this vessel, which is located generally above the coronary ostium. Aortopulmonary and interventricular defects are thought

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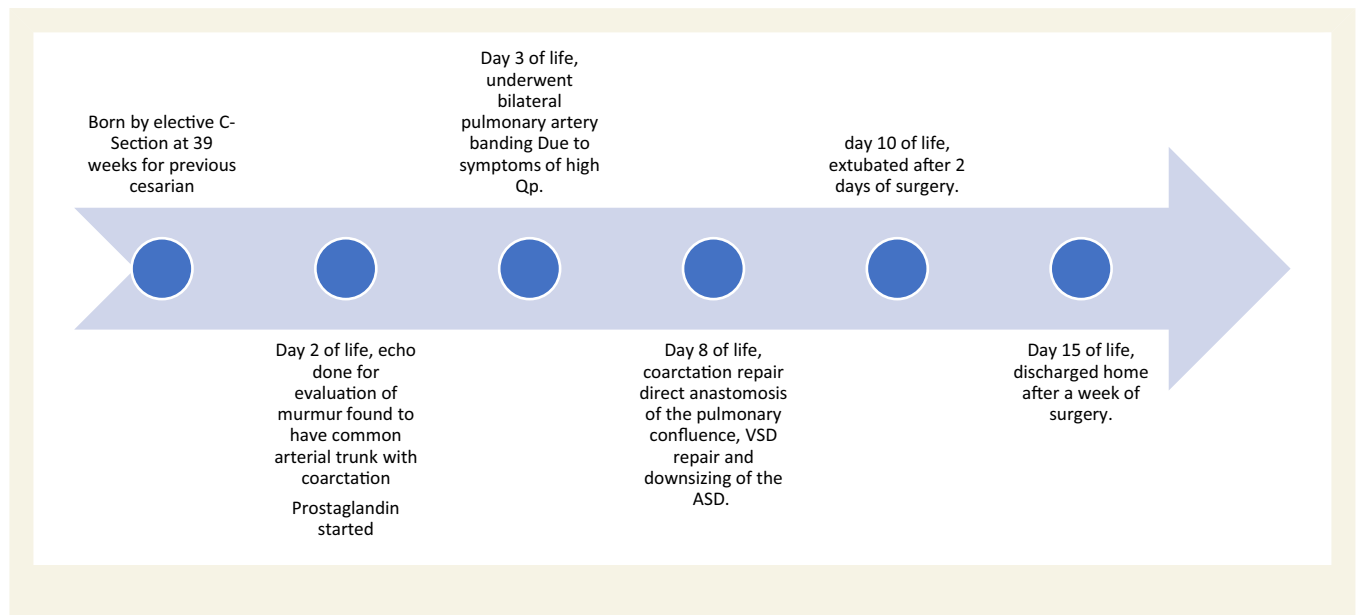
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to be caused by the conotruncal septal defect. The ductus arteriosus is not required to support foetal circulation because both systematic and pulmonary flows originate from the common trunk. As a result, the ductus arteriosus is either absent or diminished in patients with a normal calibre aortic arch.² We present a rare case of the common arterial trunk with coarctation of the aorta and patent ductus arteriosus (PDA). Because of the patient's poor clinical status, palliation with bilateral pulmonary artery branch banding and Prostaglandin E1 infusion was required. When haemodynamics improved, the complete repair could be completed a few days later.

Due to his poor general condition, he underwent an emergency pulmonary artery branch banding, as a complete repair would have carried a high risk. The patient's haemodynamics improved significantly after banding, with improved blood gases and urine output. He needed another 5 days in the intensive care unit to be stabilized before undergoing full repair. The surgical findings confirmed the anatomy described by echocardiography, as shown in the diagram below (*Figure 1*).

Coarctation segmented resection was performed during the surgery. An end to side anastomosis was then performed between the descending aorta and arch, as well as a direct anastomosis of the pulmonary con-

Timeline



Case summary

A term baby who was delivered via elective caesarean section at 39 weeks due to a previous caesarean section. Mother had normal 13 and 20 weeks foetal scans. As his father had operated transposition of the great arteries, the mother was referred to foetal cardiology but she declined cardiac scan. He was born in good health, with no need for resuscitation, but was later discovered to have a murmur on examination.

The patient was then admitted to the neonatal intensive care unit and on the second day of life, transthoracic echocardiography was performed which revealed the diagnosis of a common arterial trunk with coarctation of the aorta, large ASD secundum, large 7 mm VSD with truncal valve overriding the septum, and tricuspid truncal valve with no stenosis and mild regurgitation (*Figure 1*). The baby was given prostaglandin and intubated before being transferred to a tertiary-care paediatric cardiac intensive care unit. He developed metabolic acidosis, decreased urine output, episodes of bradycardia, rising lactate, and hypotension shortly after admission due to pulmonary over circulation with poor systemic perfusion, resulting in multiple organ failure.

fluence. A large patch of core matrix was then fashioned over the anterior aspect of the right ventricular to pulmonary artery anastomosis. Ventricular septal defect and atrial septal defect were repaired.

The clinical course was uneventful, with no major complications. Extubation was done after 48 h and the baby was discharged home within 7 days of surgery.

Discussion

According to Van Praagh, Colette, and Edwards classifications (*Figure 2*),³ the common arterial trunk is divided into four types.⁴ Van Praagh defined Type A4 as having coexisting aortic anomalies (hypoplasia or interruption). While aortic arch anomalies were not involved in Colette and Edwards classifications.^{3,4}

Russell *et al.*⁵ recently proposed an alternative and simplified method for categorizing common arterial trunk, based on the examination of 28 autopsied hearts at Chicago Memorial Hospital. The authors defined either aortic or pulmonary dominance in 20 and 8 specimens, respectively. Only when pulmonary dominance was found, the

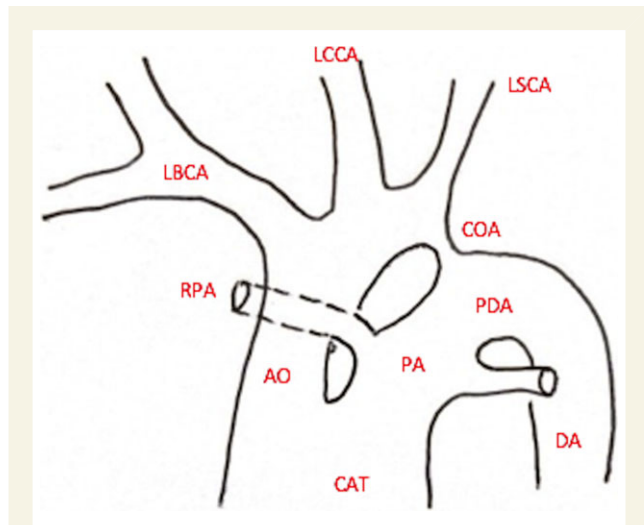


Figure 1 A diagram demonstrating the anatomy of the anomaly. CAT, common arterial trunk; PDA, patent ductus arteriosus; AO, ascending aorta; PA, pulmonary artery; DA, descending aorta; LSCA, left subclavian artery; LCCA, left common carotid artery; LBCA, left brachiocephalic artery; RPA, right pulmonary artery branch; COA, coarctation of the aorta.

aortic component of the trunk was observed to be hypoplastic and the ductus supplied the majority of flow to the descending aorta.⁵

Our patient has a common arterial trunk with a tricuspid competent truncal valve. The trunk is subdivided further into ascending aorta and main pulmonary artery (Figures 1 and 3). The pulmonary artery provided two branches and a PDA that connected to the descending aorta (Figure 4). The aortic branching pattern was as usual, there was an area of tight coarctation and posterior shelf just after the origin of the left subclavian artery before the descending aorta–PDA junction

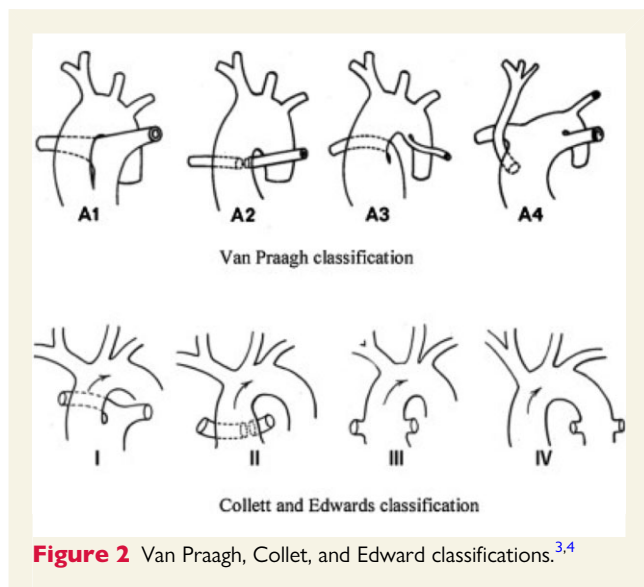


Figure 2 Van Praagh, Collet, and Edward classifications.^{3,4}

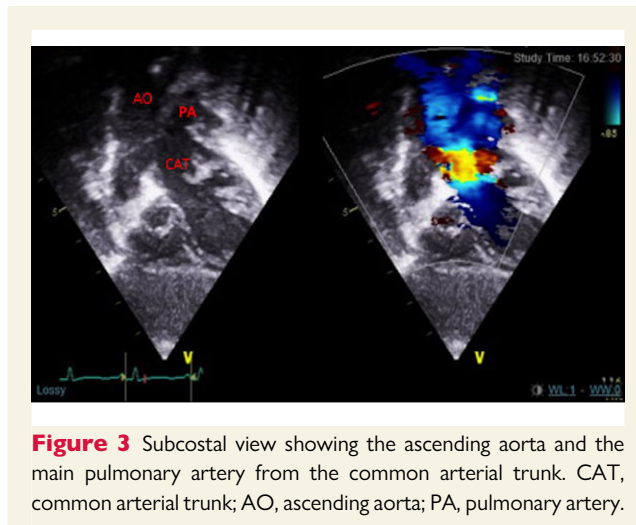


Figure 3 Subcostal view showing the ascending aorta and the main pulmonary artery from the common arterial trunk. CAT, common arterial trunk; AO, ascending aorta; PA, pulmonary artery.

(Figures 1, 5, and 6). Z-scores for aortic dimensions were calculated (ascending -2.5 , transverse -6 , isthmus -6 , descending -1).

Although the ascending aorta and pulmonary artery are both normal in size, with a PDA that is not usually seen in the common arterial trunk, the presence of coarctation may be considered as the left side of the spectrum of morphological changes toward the formation of aortic arch interruption (Type 4 Van Praagh).

Following an analysis of various pieces of the literature, it was rare to find a clear description of a similar case. For instance, in the above-mentioned paper published by Russell *et al.*⁵ among the 28-specimen analyzed, one case only had severe aortic coarctation. However, the anatomy of this case was not described in detail to compare to ours.

Chen *et al.* conducted a retrospective review of 50 consecutive patients with truncus arteriosus who underwent anatomical repair between July 2004 and July 2014. One patient was reported to have coarctation of the aorta, which represented 2% of the cases.⁶

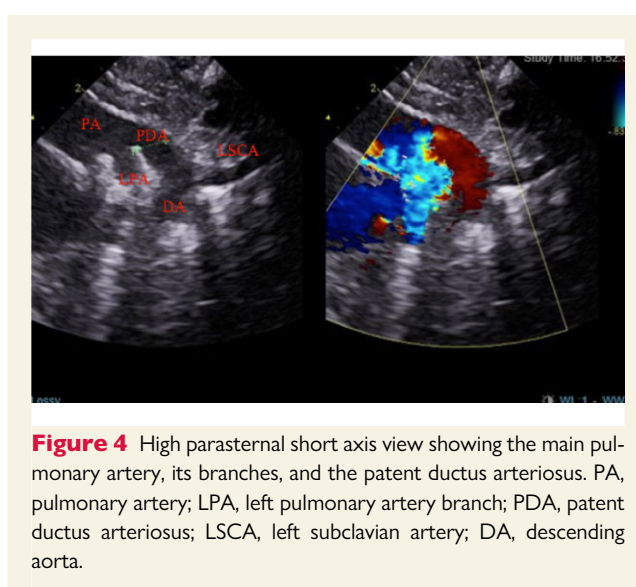


Figure 4 High parasternal short axis view showing the main pulmonary artery, its branches, and the patent ductus arteriosus. PA, pulmonary artery; LPA, left pulmonary artery branch; PDA, patent ductus arteriosus; LSCA, left subclavian artery; DA, descending aorta.

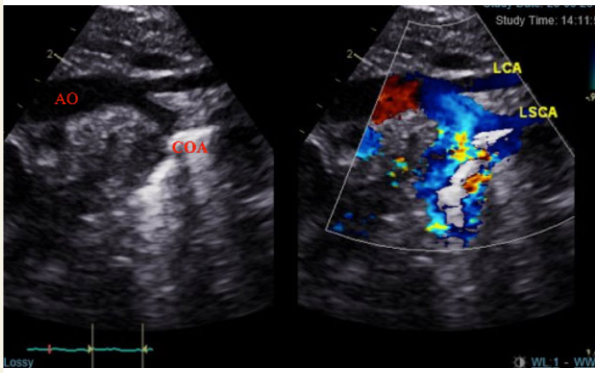


Figure 5 Suprasternal view showing the aortic arch and its branches with the coarctation site after the left subclavian artery. AO, ascending aorta; COA, coarctation site; LCA, left common carotid artery; LSCA, left subclavian artery.

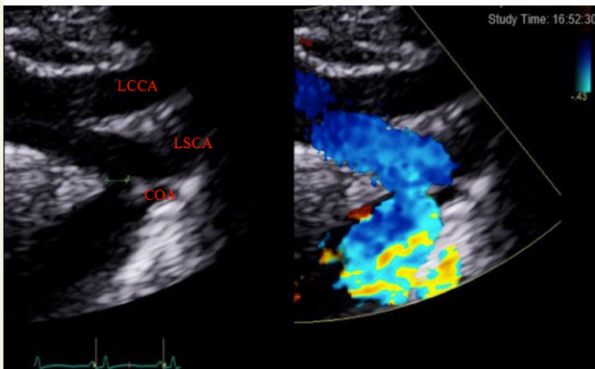


Figure 6 Zoomed suprasternal view showing the coarctation site. LCCA, left common carotid artery; LSCA, left subclavian artery; COA, coarctation site.

A series of 25 cases of truncus arteriosus examined post-mortem were studied retrospectively in a study conducted by Ceballos *et al.* Two cases involved an aortic arch interruption. In 10 cases, there was moderate hypoplasia of the distal aortic arch, but no true coarctation was found.⁷

Brown *et al.* studied 60 patients who had common arterial trunk physiologically corrected. Interrupted aortic arch was the most common cardiac condition in six patients, followed by coronary artery anomalies in six, non-confluent pulmonary arteries in four, and a supra-cardiac anomalous pulmonary venous return in one. There were, however, no cases of coarctation.⁸

Conclusion

We reported a case of a common arterial trunk associated with coarctation of the aorta, which was managed surgically. We believe that it represents a detailed description of a very rare anomaly.

Lead author biography



Dr Mohamed Abdelaal started his career in paediatric cardiology in 2005. This long experience enabled him to be exposed to many rare congenital heart diseases. He is interested in diagnosing and reporting rare congenital heart malformations.

Supplementary material

Supplementary material is available at *European Heart Journal – Case Reports* online.

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Slide sets: A fully edited slide set detailing these cases and suitable for local presentation is available online as Supplementary data.

Consent: The authors obtained a written consent from the patient’s parents for the publication of data in accordance with COPE guidelines.

Conflicts of interest: The authors whose names are listed immediately below certify that they have NO affiliations with or involvement in any organization or entity with any financial interest (such as honoraria; educational grants; participation in speakers’ bureaus; membership, employment, consultancies, stock ownership, or other equity interest; and expert testimony or patent-licensing arrangements), or non-financial interest (such as personal or professional relationships, affiliations, knowledge, or beliefs) in the subject matter or materials discussed in this manuscript.

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