# Anatomically corrected malposition of the great arteries

Kalimuddin Aziz, Najma Patel

## Abstract

A rare case of anatomically corrected malposition of the great arteries with bilaterally absent conus and ventricular septal defect is presented. Embryogenic mechanism responsible for the malformation, diagnostic problems of imaging and the increased risk posed for intraventricular surgical repair of associated defect are discussed here.

**Keywords:** Malposition, Corrected malposition of great arteries, Conotruncal rotation, Anatomically corrected malpositions, Cardiogenesis of malpositions.

## Introduction

Anatomically corrected transposition of the great arteries, later to be termed anatomically corrected malposition (ACM) of the great arteries,[1] is a rare malformation. In this condition, the transposition of the great arteries is not present and the great arteries are normally connected to their appropriate ventricles. The abnormality lies in the inter-arterial relationship which is of parallel vessels. In case of the D-loop ventricles, the great vessels are in L-malposition, and in L-loop ventricles, the great arteries are in D-malposition.[1-4] We report here a case of ACM with bilaterally absent conus, which is an even more rare association.[4,5]

## Case Report

An eight-month-old boy, one of a pair of twins, born of full-term, normal delivery, weighing below the 3rd percentile, was referred to a paediatric cardiologist due to the presence of a murmur, recurrent chest infection and failure to gain weight. The other twin had no cardiac problems. On examination, the heart and respiratory rates were normal.

Clinical examination showed normal first heart sound. The second heart sound was loud with close splitting, suggesting pulmonary arterial hypertension. There was a grade 2/6 pansystolic murmur at the left lower sternal border. The chest x-ray showed upright apex, moderate cardiac enlargement, computed tomography (CT) ratio of

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Department of Paediatric Cardiology, National Institute of Cardiovascular Diseases, Karachi, Pakistan.
**Correspondence:** Kalimuddin Aziz. Email: heart@cyber.net.pk

0.6 and increased pulmonary vascularity (Figure-1.A). Pulmonary arterial prominence was not present at the usual position, and the right border below the superior vena cava position showed a prominent, rounded bulge which, in retrospect, was interpreted as the enlarged main pulmonary artery. Preoperative echocardiographic imaging showed situs solitus, and D-loop ventricles with the pulmonary artery to the right of aorta, i.e. L-malposed (Figure-1.B-1.D). The great vessels were normally connected and transposition was not present. There was no conal muscle under the aorta or the pulmonary artery. Mitral-aortic valve and pulmonary-tricuspid valve continuity was imaged. The pulmonary and aortic annuli were contiguous and both vessels' valve annuli were at the same level. There was a large, doubly-committed ventricular septal defect. The main pulmonary artery was somewhat anterior and to the right, and the aorta was to the left in parallel pattern. Cardiac catheterisation was
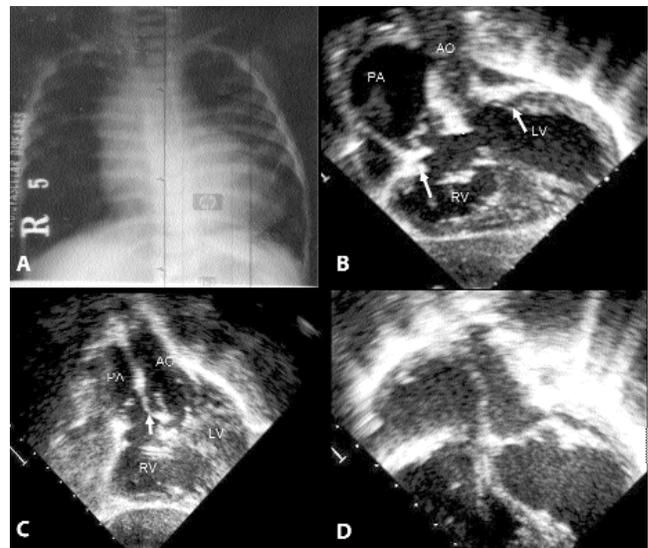


**Figure-1. A** shows a chest x-ray with prominent right upper border due to right-sided location of the pulmonary artery. Figure 1.B shows a two dimensional subxiphoid long axis view with parallel great vessels arising deep down from D-loop ventricles. Pulmonary artery (P) is on the right of the aorta (A) and the great vessels are side by side at their roots. A pulmonary-tricuspid and mitral-aortic continuity is present (arrows). The conus is bilaterally absent and a doubly-committed ventricular septal defect is present (D). Figure 1.C shows ventricular septal defect that has been closed surgically (arrow). Figure 1.D shows a four chamber apical view with tricuspid valve attachment to the septum at a lower location than the mitral valve indicating that the right ventricle is on the right side of the left ventricle (D-loop ventricles).
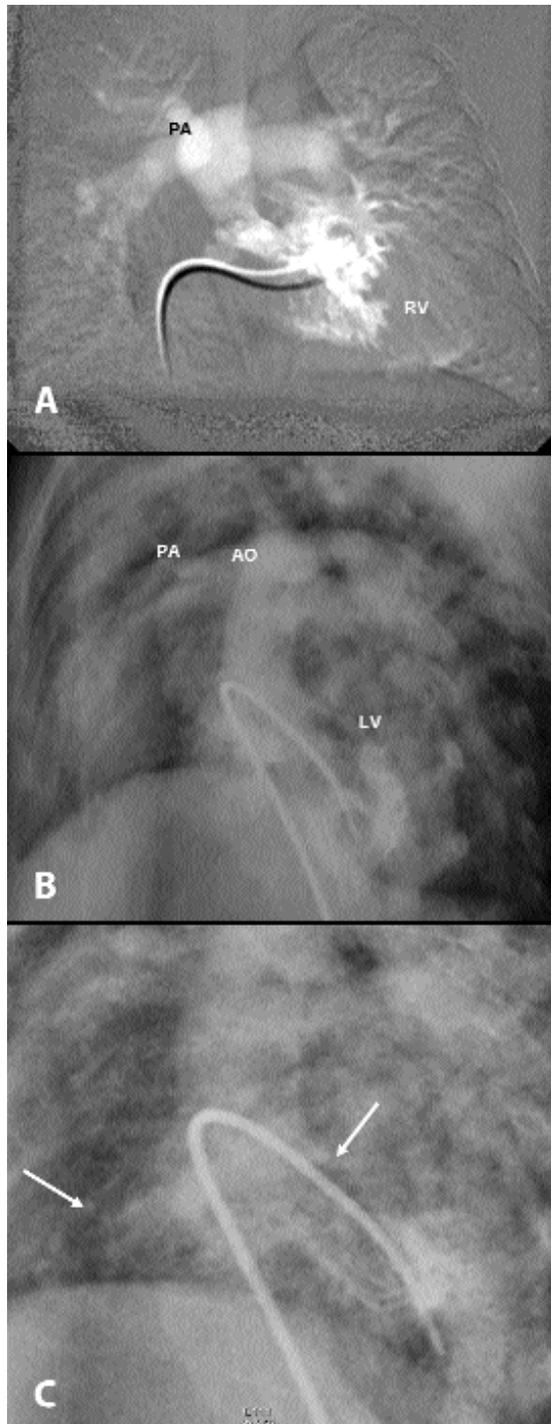
**Figure-2. A-B:** Cineangiograms. Figure 2.A shows that the pulmonary artery (PA) is arising from the right ventricle (RV) while in Figure 2.B, it is arising in a left anterior oblique projection; the aorta(AO) is arising from the left ventricle (LV) and the pulmonary artery is parallel to the aorta (AO) . Note the abnormal position of the pulmonary root and the right ventricular outflow tract. The pulmonary artery arises straight up from the right ventricle and subpulmonary conus is not present under the pulmonary artery or the aorta. Figure 2.C: Left ventricular angiogram shows normal origin of coronary arteries from the aorta (arrows).

undertaken to clarify the diagnosis. Angiography confirmed situs solitus and D-loop ventricles, that is, the right-sided right ventricle and the concordant atrioventricular connections, the great vessels, were in L-malposition, but the ventriculoarterial connections were concordant, i.e. normally connected (Figure-2.A-B); the subarterial conus was bilaterally absent. Angiogram showed that the left and right coronary arteries arose normally from the aorta, and the anterior descending artery came from the left coronary artery (Figure-2). Haemodynamics showed pulmonary arterial hypertension and a large left-to-right shunt. The child underwent closure of the ventricular septal defect through the tricuspid valve; no abnormal coronary artery crossing the right ventricular outflow tract was found. Intraoperatively, the diagnosis of ACM and bilaterally absent conus was confirmed.

## Discussion

ACM is a rare malformation and bilaterally absent conus is an even more rare association. The L-malposition of the great arteries occurs on D-loop ventricles, and the D-malposition of the great arteries occurs on L-loop ventricles. No transposition was present because the great arteries were normally connected to their respective ventricles.

Two basic types of ventriculoarterial and intra-vessel relations have been described in ACM:[1] D-loop ventricles and L-malposition of the great arteries, as in our case; and second, L-loop ventricles and D-malposition of the great arteries. These types can occur in situs solitus, situs inversus or asplenia syndrome,[1,3] levocardia or dextrocardia. A myriad of associated defects including juxtaposed atrial appendages, tricuspid atresia in ventricular septal defects, arch hypoplasia, and coarctation of aorta, various conal abnormalities such as hypoplastic conus, bilaterally present or absent under the pulmonary artery and/or the aorta, have been reported.[1-5] The association of bilaterally absent conus, as in our case, is rare.[4]

The inter-arterial and ventriculoarterial relationship develops after a complex set of events.[6-8] In normal hearts, the great vessels are such that the pulmonary artery is superior, left and anterior due to the persistence and continued growth of subpulmonary conus, and the aorta is posterior, inferior and right to the pulmonary artery[1] due to the absorption of the subaortic conus. Thus, the right ventricular outflow tract crosses the aorta anteriorly connected to the pulmonary artery, and the absorption of the subaortic conus connects the aorta to the left ventricle and mitral-aortic continuity.

After the septation of the conotruncus, the aorta lies to

the right of the pulmonary artery in parallel, above the right ventricle. The most important mechanism which normally brings the aorta posterior and above the left ventricle is an 80-110 degree counter-clockwise rotation of the conotruncus looking from the ventricles downstream.[7]

The subaortic conus is absorbed and the subpulmonary conus growth brings the pulmonary artery superiorly to the left of aorta so that the right ventricular outflow tract crosses the aorta anteriorly.

Our invoking of over-rotation of the conotruncus as a cause of ACM is based on observations on human embryos as well as experimental animals.

The normal 110 degree conotruncal rotation (Figure-3.A-B) would produce normally positioned great arteries, while a 90 degree rotation produces anterior pulmonary artery and posterior aorta. Less than 90 degree counter-clockwise rotation of the conotruncus would lead to a gradual left and anterior positioning of the pulmonary artery, and a right and posterior shift of the aorta. Approximately 180 degrees of over-rotation would produce reversal of the starting position of the great arteries, that is, aorta to the left and pulmonary artery to the right, side by side and parallel with each other above their related ventricles, as seen in anatomically corrected malpositions.

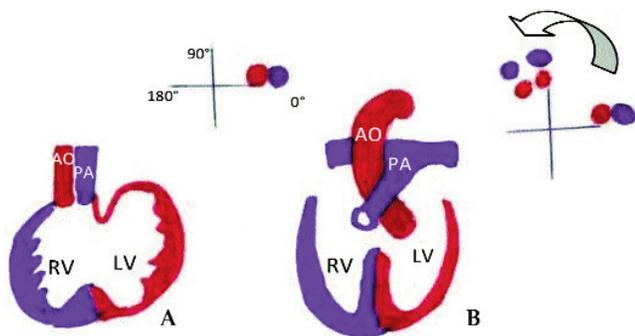Rotational abnormalities of the conotruncus such as



**Figure-4. A-B (S, D, L):** Anatomically corrected malposition in D-loop. Figure 4.A: After septation of the truncus, the great arteries are side by side above the right ventricle (RV); the aorta (red) is to the right and the pulmonary artery (blue) is to the left. We hypothesise that the conotruncus rotates counter-clockwise beyond the normal 110 - 150 degree rotation to approximately 180 degrees, which would place the pulmonary artery (PA) above the right ventricle and the aorta (AO) towards the left ventricle (LV) in parallel or in L-malposition relationship. Figure 4.B: Dilatation of the right ventricle and shifting of the AV canal to the right brings the aorta above the left ventricle. Further position of the great vessels would depend upon the absorption of the sub-great arteries conus.

arrest of the rotation in experimental animals, produces a double outlet right ventricle. The transposition of the great arteries is caused by arrested conotruncus rotation. Conal muscular abnormalities such as shortened conus have been associated with arrested conotruncal rotation in animal experimentation. The evidence of conotruncal rotation and rotational abnormalities is well-established in recent studies.[11,12]

Over-rotation of the conotruncus in L-loop ventricles leads to D-malposition, and over-rotation in D-loop



**Figure-3. A-B:** Normal positioning of the great arteries. Figure 4.A: After the septation of the truncus in D-loop, the aorta (red) is located to the right of the pulmonary artery (blue). Both great vessels are side by side above the right ventricle (RV). Figure 4.B: When no rotation occurs, the aorta remains above the right ventricle as in double outlet right ventricle. After a 90 degree counter-clockwise rotation of the conotruncus (arrow), the PA (blue dot) is anterior and the AO (red dot) is posterior. A 110-120 degree counter-clockwise rotation however, would bring the pulmonary artery anterior and to the right, and the aorta posteriorly, which would shift the PA above the right ventricle (RV) and the AO towards the left ventricle (LV). Development of the right ventricle and shifting of the atrioventricular canal to the right ventricle and absorption of the subaortic conus further brings the aorta above the left ventricle and mitral-aortic valve continuity is seen.
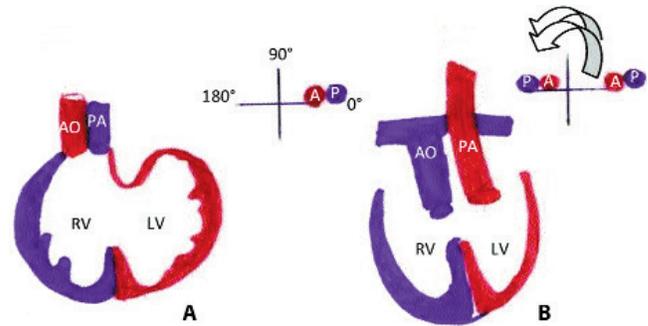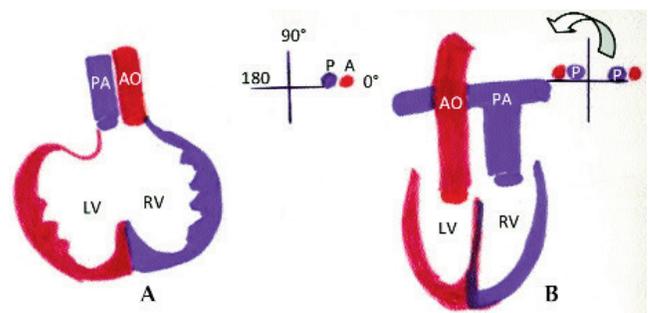


**Figure-5. A-B (S, L, D):** Anatomically corrected malposition in L-loop. Figure 5.A: After septation of the truncus, the great arteries are side by side above the right ventricle. The aorta (red) is to the left and the pulmonary artery (blue) is to the right. We hypothesise that the conotruncus rotates counter-clockwise beyond the normal 110-150 degrees to approximately 180 degrees, which would place the pulmonary artery(PA) above the right ventricle (RV) and the aorta closer to left ventricle (LV), in parallel or in D-malposition relationship. Figure 5.B: Right ventricular dilatation and shifting of the AV canal. If absorption of the subaortic conus occurs, it would bring the aorta above the left ventricle and aortic-mitral continuity would be seen. The presence and size of subaortic conus varies in anatomically corrected malpositions.
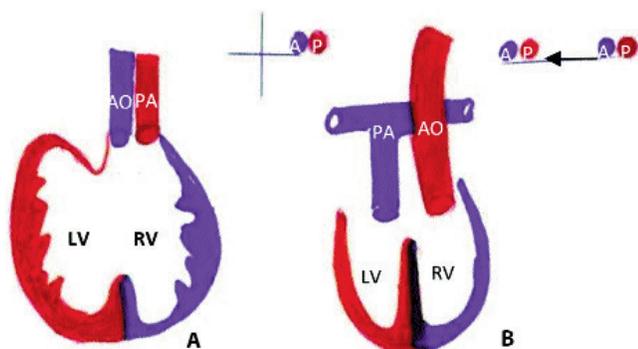
**Figure-6. A-B (S, L, L):** L or corrected transposition position. Figure 6.A: After septation of the truncus, the great arteries are side by side above the right ventricle (RV). The aorta (red) is to the left and the pulmonary artery (blue) is to the right above the right ventricle. We hypothesise that the conotruncus does not rotate at all, which would keep the pulmonary artery (PA) to the right towards the left ventricle (LV) and aorta (AO) to left above the right ventricle (RV), which itself is left-sided. Figure 6.B: With the absorption of the subpulmonary conus and right ventricular dilatation, the pulmonary artery descends into the left ventricle, producing mitral-pulmonary continuity; the aorta remains above the right ventricle with its subaortic conus.

ventricles leads to L-malposition types of ACM (Figure-4.A-B and Figure-5.A-B).

The arrested conotruncal rotation in D-loop ventricles is well-established in experimental animal studies, and it has been shown to result in complete transposition. Therefore by extending the same logic, L-loop ventricles arrested conotruncal rotation would create corrected transposition (Figure-6.A-B).[9-11] ACMs must be differentiated from corrected transposition of great arteries because it can pose difficulties for the surgeon to correct the intra-cardiac defects due to the position of the right coronary artery.[12]

## Conclusion

The increasing reports of ACM of the great arteries suggest that the anomaly may not be as rare as it is thought to be. More powerful imaging modalities such as CT angiography and magnetic resonance imaging are being used to diagnose ACM and therefore increasing reports of such cases are coming forward.

## References

1. Van Praagh R, Van Praagh S. Anatomically corrected transposition of the great arteries. Br Heart J 1976; 29: 112-9.
2. Van Praagh R ,Durnin RE, Jockin H, Wagner HR, Korns M, Garabedian H, et al. Anatomically corrected malposition of the great arteries (S,D,L). Circulation 1975; 51: 20-31.
3. Freedom R, Harrington DP. Anatomically corrected Malposition of the great arteries Report of 2 cases,one with congenital asplenia frequent association with juxtaposition of atrial appendages. Br Heart J 1974; 36: 207-15.
4. Anderson RH, Becker AE, Losekoot TG, Gerlis LM. Anatomically corrected malposition of great arteries. Br Heart J 1975; 37: 993-1013.
5. Loyo SY , Desai A Q , Sharma S. "Anatomically corrected malposition" a rare case with bilateral absence of complete subarterial muscular infundibulum. Int J Cardiol 1991; 30: 131-4.
6. Bajolle F, Zaffran S. Kelly RG, Hadchouel J, Bonnet D, Brown NA, et al. Rotation of the myocardial wall of the outflow tract is implicated in the normal positioning of the great arteries. Cir Res 2006; 98: 421-8.
7. Goor DA, Dische R, Lellihei CW. The cono truncus I its normal inversion and absorption. Circulation 1972; 46: 375-84.
8. Restivo A, Piacentini G, Placidi S, Saffirio C, Marino B. Cardiac outflow tract: a review of some embryogenetic aspects of the conotruncal region of the heart. Anat Rec A Discor Mol Cell Evol Biol 2006; 288: 936-43.
9. Chuaqui BG, Bersch W. The formal genesis of the transposition of the great arteries. J Virchows Arch A Pathol Pathol Anat1973; 358: 11-34.
10. Lomonico MP, Moore GW, Hutchins GM. Rotation of junction of the outflow tract and great arteries in the embryonic human heart. Anat Rec 1986; 216: 516-44.
11. Bostrum MP, Hutchins GM. Arrested rotation of the out flow tract may explain double out let right ventricle. Circulation 1988; 77: 1258-65.
12. Colli Am, Deleval M, Sommerville J. Anatomically corrected transposition of the great arteries, diagnostic difficulties and surgical repair of associated defects. Am J Cardiol 1985; 11: 1367-72.