Truncus Arteriosus Communis

Unusual Case Associated with Transposition

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SUMMARY A child with truncus arteriosus communis, characterized by the posterior origin of an individualized pulmonary trunk is presented. This relationship between the great arteries is unusual in truncus arteriosus communis and the spatial orientation resembles that seen in transposition of the great vessels. A brief discussion is proposed about a proper terminology in this type of complex anomaly.

VARIETIES OF COMMON TRUNCUS have been previously discussed in the literature. For classification purposes, the length of the main pulmonary trunk and its point of origin from the common trunk have been most frequently utilized. It has been suggested that the type of common truncus, with a persisting segment of the pulmonary artery (type I of Collett and Edwards), is embryologically derived from the partial failure of completion of truncal septation. Depending upon the length of the main pulmonary artery segment and its position with reference to the aortic portion of common trunk, it might be possible to identify truncus arteriosus in which the aorta and pulmonary artery remnants are in the position usually identified as transposition of the great arteries. This type of great vessel arrangement has not been reported in truncus arteriosus.

The present case report is illustrative of what we believe to be the simultaneous presence of "common truncus" and transposition of the great vessels.

Case Report

A five-year-old child was admitted to Texas Children's Hospital for evaluation of congenital heart disease. He was essentially asymptomatic, but known to have a complicated heart anomaly from previous venous angiographic study.

On physical examination the child was well developed and had no signs of congestive heart failure. The blood pressure was 90/60 in both arms. There was evidence of mild cardiomegaly with a right ventricular heave palpable at the left lower parasternal area. A grade 2 systolic ejection murmur began immediately following an ejection click and was heard best in the pulmonary area. The second heart sound was single. No diastolic murmurs or sounds were heard. The electrocardiogram was interpreted as regular sinus rhythm with evidence of right ventricular hypertrophy (fig. 1). By X-ray examination, the heart was slightly enlarged, without selective chamber enlargement. The aortic arch was on the left side. The vascular pedicle was narrow. The pulmonary vascular shadows were large near the mediastinum, but small near the periphery of the lungs (fig. 2).

Heart catheterization data are presented in table 1. A large ventricular septal defect was seen in the angiograms below a single overriding semilunar valve. A single arterial vessel of short length emerged from the heart and divided into two vessels; one with the characteristics of an ascending aorta and one with those of a pulmonary artery. The main pulmonary artery arose posteriorly from the common trunk and had a 2 cm long undivided segment that was obscured by the ascending aorta in the postero-anterior projection and was seen to be completely posterior to the aorta in the lateral projection (fig. 3).

The final diagnosis was truncov-conal septal defect (common truncus arteriosus) with transposition of the divided portion of the great vessels and pulmonary vascular obstructive disease.

In view of the high pulmonary resistances (ratio of pulmonary to systemic resistances equal to 0.78) this child was not considered a suitable candidate for corrective surgery.

Discussion

In most anatomic specimens of common truncus arteriosus, type I of Collett-Edwards, the longer the main pulmonary artery trunk, the more lateral and anterior is its

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origin from the common trunk. Similarly, when there is a very short pulmonary artery trunk, the origin is more often from the posterior wall of the truncus, a finding that makes the differentiation of type I from type II (Collett-Edwards) clinically difficult. This was not the finding in the present case, in which a relatively long main pulmonary artery trunk originated posterior and did not cross the aortic trunk in the lateral view.

This unusual heart stimulates a review of possible embryologic defects that might have caused it.

Two hypothesis have been suggested to explain common truncus. The traditional hypothesis states that the only developmental error is the incomplete septation of the trunco-conus. The second hypothesis states that pulmonary infundibular atresia occurred at the same time that the truncus and its valve failed to divide.

It is our belief that there are two embryologic faults in this heart. It seems reasonable to believe that in this case, truncus arteriosus resulted from failure of completion of the trunco-conal septum. Further, the spatial arrangement of the great vessels is such that, had trunco-conal septation been completed, the arteries would have been in the typical position of transposed great arteries (figs. 4, 5).

This view, transposition of the great arteries with trunco-conal septal defect, is further supported by the clinical findings of narrow superior mediastinum and absence of a prominent left pulmonary artery shadow. Further the

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<td>Left atrium</td>
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<td>Pulmonary vein</td>
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**Figure 1.** Electrocardiogram on admission, showing signs of right ventricular hypertrophy.

**Figure 2.** Chest roentgenogram showing mild cardiomegaly, increased hilar pulmonary vascular markings, decreased at the periphery.
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Figure 3. Retrograde aortogram in the frontal (3a) and lateral (3b) projections, showing the unusual origin of the pulmonary trunk posterior to the common arterial trunk.

Figure 4. Diagrammatic representation of normally crossed great vessels with complete (left side) and incomplete (right side) truncal septation. (a = frontal view; b = lateral view). This refers to the usual form of truncus arteriosus communis.

Figure 5. Diagrammatic representation of transposed great vessels with complete (a) and incomplete (b) truncal septation. Panel b is the case of the present report.
hemodynamic observation that blood in the pulmonary artery contained more oxygen than that in the aorta is contrary to that in the usual trunco arteriosus, but similar to that in transposition of the great vessels.

We believe that the recognition of these variants is conceptually important, even though the practical importance may be limited. Surgically it may be of some relevance. The pulmonary trunk posterior to the aorta would require a variation of the Rastelli technique for correction.

Hallerman and colleagues reported in their series of 27 cases of trunco arteriosus type I (Collett-Edwards), eight cases in which the pulmonary trunk arose posteriorly from the undivided trunco. Unfortunately the authors do not clarify the length of the pulmonary trunk in these cases. It is conceivable that most of them had a short trunk, as expected in the case of an isolated anomaly of the trunco-septal. A case reported by Testelli as interrupted aortic arch and common trunco appears to be similar to our case with the aorta completely anterior, immediately above the trunco valve, as in transposition complexes.

The possibility of other trunco-septal anomalies, such as transposition, co-existing with common trunco, is further evidence of the inadequacy of the traditional terminology "truncus arteriosus communis persists." The recently suggested terminology of trunco-septal defect reflects the need for description of additional identifying character-istics in order to avoid confusion, namely extension of the defect, unequal partition, abnormal spiraling of the trunco-septal, inversion of the bulboventricular loop, lateral positions of the trunco-conus, and associated malformations of the aortic arch. From this perspective, the classification of this case as transposition of the great vessels in "common trunco" is easily understood and conveys an image of the abnormality not made clear by the simple diagnosis of truncus arteriosus.

References
Truncus arteriosus communis. Unusual case associated with transposition.
P Angelini, A L Verdugo, J P Illera and R D Leachman

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