

Prenatal Detection of Truncus Arteriosus by Ultrasound

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SUMMARY. This report describes the echocardiographic findings of truncus arteriosus, right aortic arch, and persistent left superior vena cava draining into the coronary sinus in a 32-week-old fetus, in whom the diagnosis was confirmed by autopsy.

KEY WORDS: Prenatal diagnosis — Fetal echocardiography — Truncus arteriosus

Persistent truncus arteriosus is a rare cardiovascular malformation with an incidence of 1%–4% in autopsy series [3, 6]. The anomaly is characterized by a single arterial vessel arising from the base of the heart, which gives origin to the coronary, pulmonary, and systemic arterial circulations [4, 5].

Case Report

A healthy 26-year-old woman, gravida 2, was referred at 32 weeks of gestation. On obstetric ultrasonography, the fetus was noted to have bilateral hydronephrosis, bony demineralization, and enlargement of the heart and great vessels.

Fetal echocardiography demonstrated the heart on the left side of the chest. There was a large ventricular septal defect and a large single overriding great artery, which gave rise to the pulmonary arteries and the aorta (Fig. 1, top). The truncus arteriosus was thought to be type 1 [3] based on the origin of the pulmonary arteries (Fig. 1, bottom). A left superior vena cava was seen to drain into the coronary sinus, but the aortic arch was not detectable on that side. The arch could be identified on the contralateral side and therefore was assumed to be a right-sided arch (Fig. 2). Pulsed Doppler examination displayed mild tricuspid regurgitation.

Amniocentesis showed an abnormal chromosomal karyotype with trisomy 13 translocation. Decreased fetal movement was followed by fetal death at 37 weeks of gestation. A stillborn male infant was delivered with stigmata of the trisomy 13 including cleft palate, polydactyly and clinodactyly, microtia and microphthalmia. At autopsy, bilateral renal dysplasia with hydronephrosis, left subdural hematoma, and Meckel's diverticulum were found. The malformation of the heart was identified as a truncus arteriosus type 1 with right-sided aortic arch and left superior vena cava to coronary sinus; the foramen ovale was widely patent (Fig. 3).

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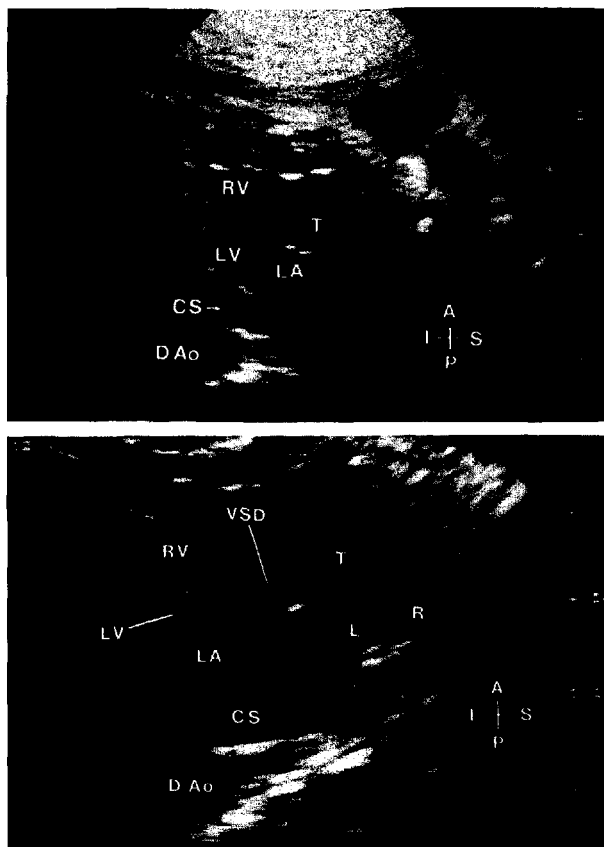


Fig. 1. (Top) Long axis view of the fetal heart. The truncus arteriosus (T) can be seen as a single great artery overriding a ventricular septal defect. Behind the left atrium (LA), the enlarged coronary sinus (CS) is visualized. (Bottom) After magnification of the frame and angulation of the transducer toward the fetal left shoulder, the common origin of the left and right pulmonary arteries (L, R) from the truncus (T) is demonstrated. A, anterior; DAo, descending aorta; I, inferior; L, left; LA, left atrium; LV, left ventricle; P, posterior; R, right; RV, right ventricle; S, superior; and VSD, ventricular septal defect.

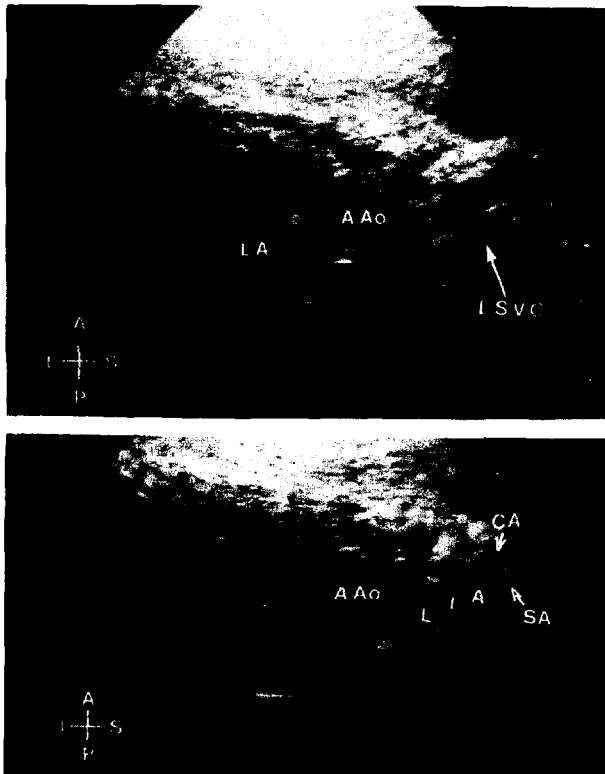


Fig. 2. (Top) Sagittal body plane demonstrating the left superior vena cava (*LSVC*) draining into the coronary sinus behind the left atrium (*LA*). (Bottom) Just to the right of the left superior vena cava, the length of the ascending aorta (*AAo*), the left innominate artery (*LIA*), and its division into left carotid (*CA*) and left subclavian artery (*SA*) are displayed. The *LIA* indicates the presence of a right-sided aortic arch. *A*, anterior; *I*, inferior; *P*, posterior; and *S*, superior.

Discussion

Truncus arteriosus has been detected previously in utero [1], but no images of the abnormality were displayed. We found the malformation was imaged best in long-axis planes, where the ventricular septal defect, the truncal override, and the origin of the well-developed pulmonary artery from the truncus were identified. Slight clockwise rotation of the transducer showed the absence of a right ventricular outflow tract. This finding, coupled with the size and origin of the pulmonary artery, allowed differentiation from tetralogy of Fallot.

In truncus arteriosus there is a high incidence of associated lesions. In autopsy series of truncus arteriosus, a right aortic arch was found in 19%–23%, and a persistent left superior vena cava in 4%–9% [2, 6]. Conversely, in our postnatal series, among 103

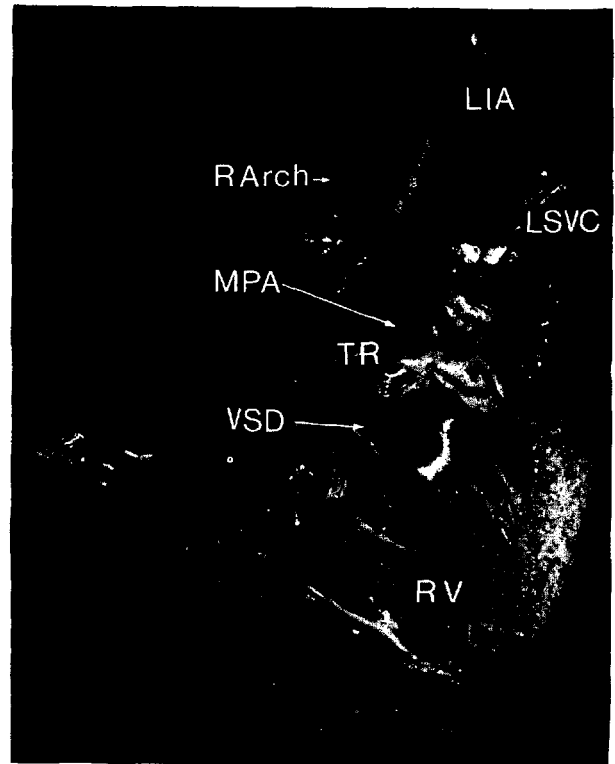


Fig. 3. Heart specimen viewed from the anterior aspect; the right ventricle (*RV*) is opened. The truncus arteriosus (*TR*) arises in top of the ventricular septal defect (*VSD*), which is v-shaped and framed by the septomarginal trabecula. The origin of the main pulmonary artery (*MPA*) from the truncus can be seen just above the tricuspid truncal valve. More cranially, the first vessel to head and neck is a left-sided innominate artery (*LIA*), thus indicating a right aortic arch (*RArch*). A left superior vena cava (*LSVC*) is also demonstrated.

patients found to have a persistent left superior vena cava by echocardiography, five children were found to have truncus arteriosus. The finding of a right aortic arch and left superior vena cava–coronary sinus connection in utero was similar to the postnatal appearance of these anomalies. The innominate artery and its branching into carotid and subclavian arteries was identified on the left side running close and parallel to the left superior vena cava. This configuration indicated mirror-image head and neck vessels and, therefore, the presence of a right aortic arch. The arch was found far to the right with respect to the position of the left superior vena cava. Conversely, a left arch would have been situated in close proximity to the left superior vena cava.

We conclude that the echocardiographic diagnosis of a truncus arteriosus is possible in the fetus,

and that the detection of such a defect is an indication for referral to a center with facilities for cardiac care of the newborn infant.

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References

1. Allan LD, Crawford DC, Anderson RH, Tynan MJ (1984) Echocardiographic and anatomical correlations in fetal congenital heart disease. *Br Heart J* 52:542-548
2. Bharati S, McAllister HA, Rosenquist GC, Miller RA, Ta-
tooles CJ, Lev M (1974) The surgical anatomy of truncus
arteriosus communis. *J Thorac Cardiovasc Surg* 67:501-510
3. Freedom RM (1984) Truncus arteriosus. In: Freedom RM,
Culham JAG, Moes CAF (eds) *Angiocardiography of congen-
ital heart disease*. Macmillan, New York, pp 437-452
4. Thiene G, Bortolotti V, Galluci V, Terribile V, Pellegrino PA
(1976) Anatomic study of truncus arteriosus communis with
embryologic and surgical consideration. *Br Heart J* 38:1109-
1123
5. Van Mierop LHS, Patterson DF, Schnarr WR (1978) Patho-
genesis of persistent truncus arteriosus. *Am J Cardiol* 41:755-
762
6. Van Praagh R, Van Praagh S (1965) The anatomy of common
aorticopulmonary trunk (truncus arteriosus communis) and
its embryologic implications: a study of 97 necropsy cases.
Am J Cardiol 16:406-426