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Prenatal diagnosis of truncus arteriosus using multiplanar display in 4D ultrasonography

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Abstract

Prenatal diagnosis of truncus arteriosus with two-dimensional (2D) sonography requires expertise in fetal echocardiography. Indeed, truncus arteriosus shares with tetralogy of Fallot and pulmonary atresia with a ventricular septal defect (VSD) the sonographic finding of a single arterial trunk overriding a VSD. The diagnosis of truncus arteriosus can be confirmed when either the main pulmonary artery or its branches are visualized arising from the truncus itself. This requires sequential examination of multiple scanning planes and a process of mental reconstruction of their spatial relationships. The advantage of multiplanar imaging in three-dimensional and four-dimensional ultrasonography is that it allows for the simultaneous visualization of three orthogonal anatomic planes, which can be very important in diagnosing cardiac abnormalities. We report, first, a case of truncus arteriosus diagnosed *in utero* where the multiplanar display modality provided important insight into the differential diagnosis of this conotruncal anomaly, and then, review the diagnosis of truncus arteriosus on ultrasound.

Keywords

prenatal diagnosis; ultrasound; heart; fetus; fetal; conotruncal; 3D ultrasound; STIC; cardiac defect; congenital

INTRODUCTION

Truncus arteriosus is a congenital heart defect in which a single arterial trunk exits the heart by way of a single arterial valve, and gives rise directly to the coronary, systemic and one, or both, pulmonary arteries.[1] This conotruncal anomaly is the result of an incomplete septation (persistence) of the distal portion of the cardiac outflow tract of the embryonic heart into the aorta and the pulmonary artery.[2] Truncus arteriosus is a cyanotic congenital cardiac anomaly characterized by increased pulmonary blood flow.[3] Early surgical repair in the neonatal period carries low morbidity and mortality[4–12] and prevents the long term sequelae of pulmonary over-circulation and heart failure.[12,13]

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We report the prenatal diagnosis of a case of truncus arteriosus using ultrasound, and describe the insights provided by novel three-dimensional (3D) imaging techniques in the differential diagnosis of this conotruncal anomaly. Moreover, we review the classification system of truncus arteriosus and summarize the chromosomal anomalies, genetic syndromes, and cardiac and non-cardiac defects that can be associated with this condition. Finally, we briefly discuss the clinical presentation at birth and major prognostic factors.

CASE REPORT

A 17-year-old G1P0 woman was referred to our unit at 23 weeks of gestation because a complex fetal cardiac anomaly was observed in a previous scan done elsewhere. Two-dimensional (2D) sonography of the outflow tracts identified a single, large vessel containing a single valve and overriding a ventricular septal defect (VSD) (Figure 1). Multiplanar display of a volume dataset obtained with four-dimensional (4D) ultrasound demonstrated that the aortic arch and both pulmonary arteries arose from this large vessel (Figure 2). Therefore, the prenatal diagnosis of truncus arteriosus type A2 in the Van Praagh classification[14] was made. On parental request, a termination of pregnancy was performed, and a stillborn neonate weighing 490 grams was delivered vaginally. The autopsy confirmed the sonographic findings of a single large vessel overriding a VSD, consistent with the prenatal diagnosis of truncus arteriosus type A2 in the Van Praagh classification (separate but adjacent origin of the pulmonary arteries from the arterial trunk) (Figure 3). Additional findings included: a left superior vena cava, abnormal lung lobation, facial anomalies (low set and posteriorly rotated ears, hypertelorism, downward slanting of the palpebral fissures, long philtrum, midfacial hypoplasia with a small mandible), and limb anomalies (rocker-bottom feet, bilateral hypoplasia of the fifth digits, clinodactyly). The postnatal karyotype was normal (46 XY) and there was no 22q11.2 deletion found (DiGeorge/Velocardiofacial syndrome).

DISCUSSION

Truncus arteriosus is a conotruncal anomaly which accounts for 1% of all congenital heart diseases detected in prenatal life.[15] Natural fetal wastage makes the intrauterine incidence even higher.[16] Both sexes are equally affected,[17,18] although a predominant distribution in females has been reported in one series.[19]

Truncus arteriosus belongs to the group of ectomesenchymal tissue migration abnormalities that are due to anomalous migration of neural crest cells through the branchial arch vessels during cardiogenesis.[20] This heart defect occurs when there is incomplete septation (persistence) of the truncus arteriosus (the distal portion of the cardiac outflow tract of the embryonic heart tube) into the pulmonary artery and the aorta.[2] The morphologic characteristics of a common arterial trunk are: 1) the common arterial trunk itself; 2) a common arterial orifice; and 3) a VSD in the outflow region (common outlet).[21] During the embryonic period, defective separation at the level of the aortic sac, the ventriculo-arterial junction, and the outlet, respectively, may account for these defects.[21] The VSD is usually located in the infundibular region, immediately beneath the truncal valve. However, this defect can be absent (type B, common aorticopulmonary trunk),[14,22–24] or very small.[25] The single valve at the origin of the truncus is often malformed, stenosed, or incompetent. In most cases, the ductus arteriosus is small, present as a ligamentous remnant or absent (it is uncommon to find a moderate or large ductus).[18,26]

Classification of truncus arteriosus

Collet and Edwards[27] classified the truncus arteriosus anomaly according to the anatomic origin of the pulmonary arteries and to the spatial relationship between these vessels (Table

I). In 1965, Van Praagh and Van Praagh proposed another anatomical classification which also takes into account the presence or absence of a VSD in association with the truncus arteriosus (Table II).[18] The frequencies of the four anatomic types according to the classification proposed by Van Praagh and Van Praagh[18] (Table II) were evaluated in a series of 100 cases.[17] Type A1 was the most frequent (47–50%), while type A3 was the least frequent (2–8%).[17]

Proper distinction between type A1 and A2 defects (Van Praagh classification) may be difficult even with angiographic and anatomical evaluations, if the defective aorticopulmonary septum is very short. Thus, it has been proposed that type A1 and A2 defects should be merged into one group.[17] In addition, truncus type IV (based on Collett and Edwards classification) resembles, in its description, pulmonary atresia with a VSD with major aortopulmonary collaterals and, thus, it is probably a misnomer. More recently, members of the Society of Thoracic Surgeons - Congenital Heart Surgery Database Committee, and representatives from the European Association for Cardiothoracic Surgery, have proposed a unified truncus arteriosus nomenclature system for use in a surgical database, based on the revised classification proposed by Van Praagh[14] in 1976 (Table III).[28] This new nomenclature has been proposed based on the observations that: 1) description of the origins of the left and right pulmonary branches from the common arterial trunk, as imaged by angiography and echocardiography, is frequently inconsistent with the appearance during surgery; 2) even when preoperative studies suggest the presence of a partially formed aorticopulmonary septum, the surgeon can rarely isolate and band a pulmonary artery segment; and 3) rare cases of truncus arteriosus without a VSD (in the classification of Van Praagh) are often associated with either conjoined aortic and pulmonary semilunar valves, or a large pulmonary valve and aortic atresia; thus, they may not represent true cases of truncus, but rather cases of a large aorticopulmonary window or of aortic atresia.[28]

Prenatal diagnosis of truncus arteriosus

Conotruncal anomalies, such as truncus arteriosus, can be prenatally diagnosed with accuracy in up to 80% of cases when multiple sonographic planes, Doppler color flow mapping and pulsed Doppler interrogation are used.[29] Experience in fetal echocardiography is a requirement for the prenatal assessment, since the four-chamber view, essential in cardiac prenatal screening, is unremarkable in up to 40–50% of cases in which there is a major congenital heart defect.[30,31] Moreover, abnormalities in the four-chamber view are present in only 30% of cases of conotruncal anomalies.[32] Prenatal diagnosis of truncus arteriosus with fetal echocardiography has been reported as early as the first trimester (using high resolution transvaginal ultrasound),[33] at 16 weeks of gestation in a fetus with increased nuchal translucency and bilateral echogenic foci,[34] and at 18 weeks of gestation in a fetus with intrauterine growth restriction.[35] However, most cases are diagnosed between 20 and 25 weeks.[15,16,32,36–40]

The role of novel sonographic imaging techniques in the differential diagnosis of truncus arteriosus

Visualization of a single arterial vessel overriding a VSD (rather than a distinct aorta and pulmonary artery) during examination of the outflow tracts, is a common finding of three cardiac defects: truncus arteriosus, tetralogy of Fallot and pulmonary atresia with VSD.[41] In tetralogy of Fallot and pulmonary atresia with VSD, a distinct pulmonary artery exiting from the right ventricle is anatomically present; however, the degree of pulmonary stenosis/atresia may be such that it is not recognizable by ultrasound, and only a single vessel (aorta) can be identified arising from the fetal heart. Thus, differentiating truncus arteriosus from these two other congenital heart defects relies on the sonographic demonstration that the

main pulmonary artery, or at least one of its branches, arise from the single arterial trunk. [41] In a recent observational study conducted at three referral centers, the prenatal echocardiographic diagnosis of truncus arteriosus was made in 24 cases, and confirmed by either autopsy or at postnatal echocardiography or surgery in 23 cases.[16] The remaining neonate had a pulmonary atresia with VSD, which was erroneously diagnosed *in utero* as a truncus arteriosus.[16]

Because truncus arteriosus is a complex conotruncal anomaly and has multiple subtypes, the prenatal diagnosis requires sequential examination of multiple scanning planes and a process of mental reconstruction of their spatial relationships, which demands expertise and knowledge in fetal echocardiography. Such a process, however, can be facilitated by novel display modalities in 3D and 4D ultrasound, including multiplanar display, tomographic ultrasound imaging (TUI) and power Doppler rendering. The advantages of 3D and 4D over 2D sonography in obstetrics have been described.[42–46] Two-dimensional sonography traditionally relies on standard anatomic planes for a thorough examination of the fetal heart. However, 3D and 4D sonography can facilitate the visualization of these planes, further simplify the examination of the fetal heart, and can potentially reduce operator dependency that is characteristic of 2D fetal echocardiography. By reslicing volume datasets of the fetal heart (which can only be obtained with 3D and 4D sonography), sonographic planes can be easily obtained.[47]

Multiplanar imaging in 3D and 4D ultrasonography is a display modality that allows for the simultaneous visualization of three orthogonal anatomic planes (transverse, sagittal and coronal). An imaging tool, referred to as the “reference dot”, can be used to identify and track anatomic structures in these planes. For example, placement of the reference dot in the sagittal view of the common trunk, at the origin of the right pulmonary artery and aortic arch (Panel B of Figure 2), allowed the visualization of the left pulmonary artery arising from the common trunk in the transverse and coronal planes (Figure 2, panels A and B, respectively). Although the left pulmonary artery was visualized by 2D sonography, the confirmation of its nature, and also that of the right pulmonary artery and aortic arch, was accomplished by placing the reference dot on these vascular structures in the multiplanar display, and by rotating the volume dataset along the x- and y-axes (“Spin” technique).[48] It has also been demonstrated that multiplanar display may help in the evaluation of abnormal vascular connections.[49]

TUI is another new display modality available in 3D and 4D ultrasonography that allows simultaneous visualization of up to eight parallel anatomical planes. TUI has been used for the examination of the fetal heart[47,50–53] and other fetal organs.[54] A recently described novel algorithm combining Spatio-Temporal Image Correlation (STIC) and TUI allowed the simultaneous visualization of the four-chamber view, three-vessel and trachea view, and both outflow tracts.[47] This algorithm allowed for visualization of the standard planes for fetal echocardiography in most fetuses with and without heart defects. The application of this algorithm to a 4D volume dataset from a normal fetus and that from the fetus with truncus arteriosus are displayed in Figures 4a and 4b, respectively. In figure 4b, a large VSD is observed in the four-chamber view (panel D), and a single vessel overriding the VSD is observed in panel C. The cross-section of the aortic root was not visualized in the short axis view (Figure 4b panel A, and supplementary video clip 1).

B-flow is a new display modality in 4D ultrasound that digitally enhances signals from weak blood reflectors from vessels and, at the same time, suppresses strong signals from surrounding tissues. This display modality may have less signal drop out when the ultrasound beam is perpendicular to the vessel.[55] The use of B-flow in the case presented

herein demonstrated that this imaging modality provided important insight into the location of the left pulmonary artery and aortic arch (Figure 5 and supplementary video clip 2).

Rendered images of 4D volume datasets obtained with power Doppler have been previously used to demonstrate the crisscrossing of the great arteries in a normal fetus, and the absence of this crisscrossing in a fetus with transposition of the great arteries.[56] Video clip 3 displays the rendered images of a volume dataset obtained with power Doppler in the current case of truncus arteriosus. During diastole, the rendered Doppler signals are visualized entering both ventricles, and during systole, the power Doppler signal indicates that only one vessel arises from both ventricles. Thus, power Doppler rendering may help in the diagnosis of truncus arteriosus. Therefore, we found that the use of 3D/4D sonography provided important insight into the differential diagnosis of truncus arteriosus.

Association with chromosomal anomalies and genetic syndromes

Chromosomal anomalies are detectable in 8.7% of cases of truncus arteriosus.[16] Despite the reported sporadic association with trisomy 18,[57] duplication of 8q,[58,59] terminal deletion of 7q,[60] and mosaicism for ring chromosome 22,[35] the most frequent chromosomal abnormality in fetuses with truncus arteriosus is microdeletion of 22q11, detectable in up to 40% of cases when a cytogenetic examination is performed.[16,34,61–64] In fact, conotruncal heart defects are the most common cardiovascular malformations seen in the presence of 22q11 deletion.[65] Moreover, monosomy 22q11 has been associated with distinct phenotypes affecting the derivatives of the third and fourth branchial pouches, such as: DiGeorge sequence, velocardiofacial syndrome, conotruncal anomaly face syndrome, facial dysmorphic features, CHARGE syndrome (coloboma of the eye, heart defects, atresia of the nasal choanae, retardation of growth and/or development, genital and/or urinary abnormalities, ear abnormalities and deafness), and cases of isolated complex cardiovascular malformations.[66,67] Therefore, a prompt examination for extracardiac manifestations on prenatal sonography is indicated in these cases.

Nonsyndromic conotruncal heart defects, including isolated truncus arteriosus anomaly, have been proposed to occur sporadically,[68] or as a result of a complex and multifactorial inheritance,[69] where the effects of one or more genes interacts with environmental factors.[70,71] The familial occurrence of truncus arteriosus supports a potential genetic basis for this anomaly. Indeed, several individuals of a family can be affected by this congenital heart defect,[72,73] and a higher frequency of truncus arteriosus has been reported among siblings.[74–76] Furthermore, monozygotic[77] and dizygotic[78] twins concordant for truncus arteriosus, as well as a case of double truncus arteriosus in thoracopagus twins,[79] have been reported. A monogenic-oligogenic inheritance has been proposed for conotruncal anomalies such as truncus arteriosus,[69,80,81] and pedigree analysis of families with a history of first cousin marriages and truncus arteriosus have revealed compatibility with an autosomal recessive inheritance.[72] Recurrence rates of congenital cardiac malformations in families of index cases of truncus arteriosus range from 6.6% in the absence of associated cardiac malformations, to 13.6% in the presence of complex truncus arteriosus.[82] Finally, truncus arteriosus and other conotruncal defects have been experimentally induced in fetuses of rats exposed to teratogens,[83] and an association has been proposed between maternal exposure to alcohol, dyes, lacquers, paints, upper respiratory infections during the first trimester, and conotruncal anomalies in the developing fetus.[71]

Association with other cardiac and non-cardiac defects

The prevalence of associated heart and great vessel anomalies in fetuses with truncus arteriosus is 35%.[16] The spectrum of aortic arch anomalies associated with persistent truncus arteriosus includes: interruption of the aortic arch (9–19% of cases), and left (58–

66%) or right-sided (21–34%) aortic arch.[14,17] The combination of truncus arteriosus with interrupted aortic arch is limited to case reports and small series,[36,84–91] but carries a high early mortality and high risk of re-interventions in survivors.[92] The association of truncus arteriosus with double aortic arch is rare.[93–96] A high incidence of coronary ostial and arterial abnormalities (44–49%) has also been described in the presence of truncus arteriosus.[14,17] While in most cases these are not of functional importance,[14,17] they can contribute to a high operative mortality rate, and may be a cause of late sudden death.[97] Truncus arteriosus has been described in association with tricuspid atresia/absence of right atrioventricular connection,[98–103] atrioventricular septal defects,[104] atrioventricular canal,[105,106] single ventricle,[76,107–109] and pulmonary atresia.[110,111]

DiGeorge syndrome (facial, palate and ear anomalies, hypocalcemia, and defective thymic-dependent cellular immunity) is present in 30% of patients with truncus arteriosus.[112] Other anomalies such as holoprosencephaly,[40] splenic agenesis/asplenia syndrome,[113] polysplenia syndrome,[105] diaphragmatic hernia, and esophageal atresia with distal tracheoesophageal fistula[114] have been reported with truncus arteriosus.

Clinical presentation in the neonatal period

Truncus arteriosus, like transposition of the great arteries and total anomalous pulmonary venous return, is considered a cyanotic congenital cardiac anomaly characterized by increased pulmonary blood flow.[3] Pulmonary blood flow, in contrast, is reduced in tetralogy of Fallot and tricuspid atresia, the other two major congenital cyanotic cardiopathies.[115] In neonates affected by truncus arteriosus, cyanosis is not a constant feature. It can be minimal, intermittent, or even absent, because the large pulmonary flow itself may guarantee the resting arterial oxygen saturation to be close to normal.[116] However, if the cardiac defect is unrecognized or left untreated, the increasing amount of mixed blood perfusing the pulmonary circulation leads to an increase in pulmonary vascular resistance and to cardiac heart failure, of which cyanosis is a constant feature.[116,117] Widened pulse pressure, bounding pulses, [117] as well as auscultatory murmurs[116] are additional clinical features. Failure to thrive, respiratory infection or distress, and congestive heart failure[116,118] may manifest later according to the severity of the condition, and particularly, if the diagnosis and appropriate treatment are delayed. However, some cases are only diagnosed during adulthood.[119–122]

Prognostic factors

Predictors of poor neonatal outcome in the presence of truncus arteriosus include: severe truncal valve regurgitation, truncal valve stenosis, concomitant interrupted aortic arch, coronary abnormalities, associated anomalies, prematurity, birthweight <2.5–3.0 kilograms, as well as surgical variables (timing and type of surgical repair).[6,15,118,123–125] Truncal valve regurgitation is the result of the presence of a single semilunar valve, which is frequently dysmorphic, dysfunctional or both.[126] Persistency of moderate-severe insufficiency after surgical treatment reduces short and long term survival.[126] Neonates with truncal valve stenosis (defined as a Doppler velocity ≥ 2 m/sec across the valve on the postnatal echocardiogram) have a higher risk of early sudden death and can be potentially identified by prenatal Doppler velocimetry of the truncal valve.[15] Prenatal assesment of these risk factors may assist in a more individualized parental counseling.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

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Figure 1.

A single large vessel arises from the left and right ventricles. A thin vessel, subsequently confirmed to correspond to the left pulmonary artery, originates from the left side of this arterial trunk. RV: right ventricle; LV: left ventricle; CT: common arterial trunk; LPA: left pulmonary artery; SVC: superior vena cava; S: fetal spine.

Figure 2.

3D multiplanar display imaging of the common arterial trunk. Panel A: in this transverse plane the left pulmonary artery is seen arising from the common arterial trunk. Panel B: in this sagittal plane the right pulmonary artery and the aortic arch are seen arising from the common arterial trunk. Panel C: coronal view, in which both left and right pulmonary arteries are demonstrated to originate from the common trunk. Ao: aorta; CT: common arterial trunk; R PA: right pulmonary artery; L PA: left pulmonary artery; D Ao: descending aorta.

Figure 3.

In the pathology specimen, the left pulmonary artery originates from a common arterial trunk (TA) arising from the fetal heart. RV: right ventricle; TA: truncus arteriosus; LPA: left pulmonary artery; R: right; L: left.

Figure 4.

a. A novel algorithm using Tomographic Ultrasound Imaging (TUI) allows for the simultaneous visualization of the short axis of the aorta in panel A, the three vessel and trachea view in panel B, the long axis of the left outflow tract in panel C and the four-chamber view in panel D.

b. The application of this algorithm to the present case of truncus arteriosus demonstrates a large ventricular septal defect (VSD) in the four-chamber view in panel D. Panel C displays a large vessel overriding the VSD. Of note, the root of the aorta is not visualized in panel A, which corresponds to the short axis of the heart. This indicates that the anatomic orientation of the common arterial trunk is similar to that of a normal pulmonary artery. Panel B displays the common trunk giving rise to the left pulmonary artery (LPA) and the aortic arch (Ao arch).

Figure 5.
B-flow imaging displaying the common truncus (CT) and some of its branches, including the aortic arch (Ao Arch) and the left pulmonary artery (LPA); the image is seen from the left. D Ao: descending aorta, BT: brachiocephalic trunk.

Table I

Classification of Collett and Edwards

Type	Description
I	A short main pulmonary artery, arising from the posterior side of the truncus arteriosus, bifurcates into right and left pulmonary arteries
II	Two pulmonary arteries arise from the back of the truncus, adjacent to one another
III	Two pulmonary arteries arise from the posterolateral sides of the truncus, far from each other
IV	The pulmonary arteries are absent, and lungs are supplied by systemic pulmonic collateral arteries derived from the descending aorta

Table II

Classification of Van Praagh and Van Praagh *

Type	Description
1	The A-P septum is partly formed, thus a main pulmonary artery segment is present
2	The A-P septum is entirely absent, hence there is no discrete main pulmonary artery and both pulmonary arterial branches arise directly from the truncus. Their proximity to one another is not specified.
3	One branch pulmonary artery arises from the truncus, and the other branch pulmonary artery has either a ductal or descending aorta origin (collateral arteries)
4	Underdevelopment of the aortic arch (the aortic isthmus displays hypoplasia, coarctation, atresia, or absence) associated with a large patent ductus arteriosus

* The Van Praagh and Van Praagh classification also specifies the presence (type A) or the absence (type B) of a ventricular septal defect. Thus, each case of truncus arteriosus is assigned a nomenclature that includes a letter and a number.

A–P: aorticopulmonary septum.

Table III

Truncus arteriosus nomenclature

Modified Van Praagh Classification	Van Praagh and Van Praagh	Collett and Edwards
Truncus arteriosus with confluent or near confluent pulmonary arteries (large aorta type)	Type A1, A2	Type I, II, III
Truncus arteriosus with absence of one pulmonary artery (large aorta type with absence of one pulmonary artery)	Type A3	
Truncus arteriosus with interrupted aortic arch or severe coarctation (large pulmonary artery type)	Type A4	