Type A4 Truncus Arteriosus with Quadricuspid truncal valve (QTV): A rare cause of infantile heart failure

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Abstract

Truncus arteriosus is an exceedingly rare congenital heart disease involving conotruncal septum of developing heart. Clinical presentations vary depending upon associated anomalies. Surgical management is extremely challenging and differs with anatomic subtypes. We present a case of Truncus arteriosus – Vaan Praagh type A4, quadricuspid truncal valve with moderate truncal stenosis and regurgitation, type B interrupted aortic arch who underwent bilateral pulmonary artery banding.

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Abstract: Truncus arteriosus is an exceedingly rare congenital heart disease involving conotruncal septum of developing heart. Clinical presentations vary depending upon associated anomalies. Surgical management is extremely challenging and differs with anatomic subtypes. We present a case of Truncus arteriosus – Vaan Praagh type A4, quadricuspid truncal valve with moderate truncal stenosis and regurgitation, type B interrupted aortic arch who underwent bilateral pulmonary artery banding.

Background: Truncus arteriosus (TA) is a rare congenital heart defect where failure of conotruncal septum development leads to failure of separation of two great arteries. Neural crest cells fail to migrate to distal bulbus cordis of developing heart to form the aorticopulmonary septum (spiral septum). Prevalence of TA is 76 per million live births, representing 1.19% of all congenital heart disease.[1] Prevalence of interrupted aortic arch is about 3 per million live births.[2] The combination of type B interrupted aortic arch with truncus arteriosus, also classified as Vaan Praagh type A4, is extremely rare consisting 12% of all cases of truncus arteriosus.[3] However association with quadricuspid truncal valve in Vaan Praagh type A4 truncus arteriosus is not yet reported in literature.

Case: A 4-day-old baby-girl with history of a positive anomaly scan at 7 month of gestation, was delivered at 35 weeks of gestation. After birth there was no cyanosis, spells, no breathing or feeding difficulty, no features suggestive of heart. Clinical examination showed 93% oxygen saturation in room air with a constant ejection click, grade 3 ejection systolic murmur at aortic area. Transthoracic echocardiogram (TEE) showed large sub-arterial ventricular septal defect**(Figure 1A)** , single outflow with quadricuspid truncal valve**(Figure 2)** with mild-moderate truncal regurgitation due to non-coaptation of leaflets and moderate truncal stenosis **(Figure 1B-1D, Figure 2),** type B interrupted aortic arch with bilateral pulmonary arteries originating separately from ductal arch**(Figure 3A, 3B).** Cardiac CT showed a common trunk continuing as ductal arch after giving right brachiocephalic trunk and left common carotid artery, type B interrupted aortic arch, and both left and right pulmonary arteries originating from posterior and inferior aspect of ductal arch with no branch pulmonary stenosis **(Figure 4A-4C)** . Hence the diagnosis of Truncus arteriosus – Vaan Praagh A4 associated with quadricuspid truncal valve with moderate truncal stenosis and mild-moderate truncal regurgitation was made (schematic depiction in**Figure 5)** . We suggested Truncal repair with truncal valve repair for the patient. However, the patient developed hemodynamic instability during surgical repair immediately after anesthesia induction. Hence bilateral pulmonary artery banding was done and staged repair of truncus was planned. On follow up she had failure to thrive and functioning PA band **(Figure 6)** . Cardiac CT was planned to reassess pulmonary artery growth and planned for definitive repair.

Discussion: Truncus arteriosus (TA) is a conotruncal abnormality where the ascending aorta and pulmonary trunk fail to separate during development of the fetus, characterized by single arterial outlet from the heart overriding the ventricular septum and supplying coronary, pulmonary and systemic circulations. Very often it is associated with valvular abnormality. Most often truncal valve is tricuspid followed by quadricuspid or bicuspid which lead to either stenosis or regurgitation hemodynamically. Almost all patients have non-restrictive sub-arterial ventricular septal defect with absence of conal septum. Prevalence of TA is 76 per million live births, and represents 1.19% of all congenital heart disease. DiGeorge syndrome and chromosome 22q11 deletion have a well-established relation with the anomaly.[4] The original system by Collett and Edward, divided truncus arteriosus into 4 types from Type I to IV.[5] This classification was further modified

by Van Praagh.[6] Both the classification systems are briefed in **Table 1** . Our patient falls under Van Praagh Type 4A because of presence of type B interrupted aortic arch and both pulmonary arteries arising separately from ductal arch. Our patient additionally had quadricuspid truncal valve with moderate truncal stenosis with mild -moderate truncal regurgitation. Verma et al reported 3 cases of Type A4 TA, none of which had been reported to have quadricuspid truncal valve.[7] As the presentation is variable at different ages from childhood to adult, early diagnosis and surgical repair is crucially important for survival. Corrective surgery for common arterial trunk with interrupted aortic arch is challenging and preoperative planning is crucial. In surgical repair, pulmonary arteries are detached from parent trunk and connected to right ventricle by a valved homograft. Associated critical anomalies like interrupted aortic arch or obstructive lesions (like truncal valve) are repaired in same setting. Conduit stenosis or regurgitation, branch pulmonary artery stenosis, neo-aortic truncal valve insufficiency or stenosis, VSD patch leak, and aortic arch obstruction are amongst repair related complications. We could not perform definitive surgery due to hemodynamic instability upon induction by anesthetic agents forcing us to do palliative surgery in the form of bilateral PA banding.

Conclusion: Truncus arteriosus – Vaan Praagh A4 is extremely rare and its association with QTV has not been documented till date. Managing such patients is extremely challenging, often requiring staged palliative surgery. We possibly report the first case of Truncus arteriosus – Vaan Praagh A4 associated with QTV and suggest more sensitization among cardiologists.

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Conflict of Interest: None

Figure Legends:

Figure 1. A. Modified A4C view showing large sub-arterial VSD, B. Turbulence in outflow tract at valve level, C. Moderate truncal stenosis with peak gradient of 62mmHg, D. Mild-moderate truncal regurgitation seen on same view

Figure 2. Quadricuspid truncal valve in fully closed state, note the central AR jet through non-coapting leaflets [* denotes all leaflets]

Figure 3. A. Modified ductal view showing dilated ductal arch with sequential RPA (red arrow), LPA (orange arrow) branching, B. Ductal arch giving rise to RPA and LPA

Figure 4. A. Branching pattern in CT angiogram (LAO section) and 3D reconstructed CT (posterior view) correlation (Note that ascending aorta is small and type B interrupted giving only right brachiocephalic artery and left common carotid artery); B. Expected normal arch and existing ductal arch shown; C. Cardiac CT coronal section showing right brachiocephalic artery and left common carotid artery arising from hypoplastic ascending aorta.

Figure 5. Schematic depiction of anatomy of the index case.

Figure 6. Post bilateral pulmonary artery banding showing significant gradients with functioning PA band

Table 1 : Classification systems for truncus arteriosus.

Collete and Edward classification	Collete and Edward classification	Van Praagh classi
Type I	Both PA arising from a short pulmonary trunk	**Type A1**
Type II	Separate origin of the PA from the posterior aspect of the truncus	**Type A2**
Type III	Separate origin of the PA from the lateral aspect of the truncus	**Type A3**
Type IV	A pseudo truncus (now considered a form of PA atresia with VSD)	**Type A4**

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