We describe the case of a newborn with 22q11 deletion and an unusual form of common arterial trunk with the main pulmonary artery arising anteriorly at the level of the common arterial valve sinus.

Case reports: We report the case of a term newborn male who presented to our institution with a prenatal diagnosis of congenital heart disease. His mother was a 29-year-old woman with 22q11 deletion who was evaluated in the high-risk perinatal clinic after an abnormal fetal ultrasound. The initial fetal echocardiogram performed at 26 weeks of gestation showed a common arterial valve overriding a large subarterial ventricular septal defect, with no significant stenosis or regurgitation of this valve. The pulmonary arteries were not clearly identified. Subsequent fetal evaluation showed what appeared to be a main pulmonary artery arising anteriorly to the ascending aorta, very proximal to the common arterial valve, at the level of the arterial valvar sinus (Figure 1; Movie 1). No infundibulum or separate right ventricular outflow tract or second semilunar valve could be visualized. Prenatal counseling was given toward the likely diagnosis of a truncus arteriosus.

After an uneventful term delivery with an Apgar score of 8/8, the patient was empirically started on prostaglandin infusion until a truncus arteriosus with interrupted aortic arch was suspected (Figure 2). An echocardiogram within the first few hours of life demonstrated a single outflow tract with a trileaflet semilunar valve overriding the crest of the ventricular septum and a large subarterial ventricular septal defect. It was confirmed that the main pulmonary artery arose anterior to the aorta at the level of the arterial valvar sinus (Figure 3A through 3D; Movies IIA through IID), coursing anterior to the ascending aorta, and arching up above the aortic arch before diving posteriorly and branching into left and right pulmonary arteries. Crisscrossing of the pulmonary arteries was noted (Figure 5; Movie IV). The aortic arch was rightward and intact, with an aberrant left subclavian artery. There was no ductus arteriosus, which made the diagnosis of truncus arteriosus likely. Chromosome analysis revealed the patient to have 22q11 deletion.

On day of life 7, the patient underwent complete surgical repair (Figure 6). The main pulmonary artery was transected from the ascending aorta, and the aortopulmonary communication was closed with a bovine pericardial patch. A 12-mm Contegra graft (Medtronic, Minneapolis, Minn) was then anastomosed between the right ventricle and the transected main pulmonary artery, just proximal to its bifurcation. A secundum atrial septal defect was partly closed with a running Prolene suture (Ethicon, Inc, Somerville, NJ), leaving a 3-mm...
atrial septal defect. The patient had an uneventful postoperative course and was discharged home at 2 weeks of age.

Since the middle of the previous century, truncus arteriosus has been the subject of several classifications, commonly delineated on the basis of the origins of the pulmonary arteries. The accepted classification systems include those devised by Collett and Edwards,1 the van Praagh classification,2 and a modification of the Collett and Edwards classification developed by the Society of Thoracic Surgeons.3 To the best of our knowledge, despite the multitude of nomenclatures, there has not been a demonstration at autopsy or a discussion regarding anterior origin and course of the main pulmonary artery giving rise to both branch pulmonary arteries from the sinus of the common trunk. Bensky and Velvis4 reported an unusual case of what they called a truncus arteriosus with a hypoplastic main pulmonary artery arising anteriorly from the ascending aorta about 12 mm above the level of the truncal valve, giving rise to a left pulmonary artery. A small right pulmonary artery arose directly from underneath the ascending aorta. The single arterial valve was quadricuspid, with mild stenosis and moderate insufficiency, which rendered the diagnosis of common arterial trunk.4 Although this case is similar to the patient seen at our institution with the anterior location of the main pulmonary artery, in our patient, the pulmonary trunk arose significantly more proximal to the truncal valve. Despite its unusual origin, in the present case, the main pulmonary artery segment was not traced to the outflow tract of the right ventricle anterior to the aorta, which makes this malformation consistent with the diagnosis of truncus arteriosus in which the common arterial trunk gives rise to the pulmonary, systemic, and coronary arteries.

Our case of a newborn with a common arterial trunk is unusual given the origination of the pulmonary artery in a very proximal position at the level of the sinuses of the common arterial valve.

Disclosures

None.

References

Figure 3. A, Parasternal long-axis 2D echocardiographic image demonstrating the main pulmonary artery arising from the truncal artery at the level of the common arterial valve sinus. The common arterial valve is seen overriding the ventricular septal defect. LA indicates left atrium; LV, left ventricle; RV, right ventricle; MPA, main pulmonary artery; and Aorta, ascending aorta beyond the origin of the pulmonary artery. B, 2D and color Doppler echocardiography in the parasternal long axis demonstrating the main pulmonary artery arising from the ascending aorta just at the level of the common arterial valve sinus. The common arterial valve is seen overriding the ventricular septal defect. C, 2D and color Doppler echocardiography in the parasternal short axis. This image demonstrates the truncal valve leaflets and the small anterior pulmonary artery. D, High parasternal short-axis image demonstrating the pulmonary artery and its branches anterior to the ascending aorta.
Figure 4. Lateral angiograph in the proximal portion of the aorta demonstrating the main pulmonary artery arising anteriorly from the ascending aorta at the level of the common arterial valve and arching up above the transverse arch.

Figure 5. Angiography in the distal main pulmonary artery (right) shows crisscross pulmonary artery branches arising above the transverse arch.

Figure 6. Intraoperative photo after opening of the patient’s chest. The pulmonary artery can be seen arising anterior and very proximal on the truncal artery.
Anterior Origin of the Main Pulmonary Artery From the Arterial Valvar Sinus: Unusual Truncus Arteriosus
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