Anatomically corrected malposition of great arteries: A nidus for the left ventricular outflow tract obstruction

Sir,

We would like to bring to your notice an interesting case of anatomically corrected malposition of great arteries (ACMGA) presented with left ventricular outflow tract obstruction (LVOTO) few years after transcatheter closure of perimembranous ventricular septal defect (PmVSD).

A 3-year-old boy diagnosed to have moderate PmVSD with ACMGA underwent transcatheter closure using 6/4 Amplatzer duct occluder II (ADO II) from retrograde (arterial) approach.[1] The child was given Aspirin 3 mg/kg for 6 months and kept on regular follow-up.

Transthoracic echocardiography on 2-year follow-up showed developing LVOTO due to hypertrophy of subaortic conus, which progressed to severe degree with a peak gradient of 70 mmHg [Figure 1a]. The obstruction was 1 cm below the previously deployed ADO II device. Computerized tomography angiography was done which confirmed the findings of LVOTO [Figure 1b]. The child underwent surgical resection of LVOTO under transesophageal guidance [Figure 1d]. Postoperative assessment revealed no gradient in the LVOT. The child is asymptomatic, and no recurrence of LVOTO seen during the follow-up.

The embryological basis for this condition was described by Goor et al. as isolated inversion of conus which results in inversion of conal musculature together with the great arteries.[2] The posterior left ventricle connects to the anterior aorta resulting in an elongated LVOT as seen in the angiogram of our case [Figure 1c]. This pushes the pulmonary artery (PA) posterior and right to the aorta. The abnormal spatial orientation of PA with respect to the right ventricle makes it difficult for the catheter to enter PA from the right atrium. There are four types of ACMGA described by Van Praagh. Type 1 (S, D, L) situs solitus, d-loop ventricle, left and anterior aorta; Type 2 (S, L, D) situs solitus, l-loop ventricle, right and anterior aorta; Type 3 (I, L, D) situs inversus, l-loop ventricle, right and anterior aorta; and Type 4 (I, D, L) situs inversus, d-loop ventricles, left and anterior aorta.[3] The most common type of ACMGA is Type 1, as in our case. Types 1 and 3 are physiologically corrected, whereas Type 2 and Type 4 have transposition physiology. In all types of ACMGA, the aorta is anterior to the PA and supported by a muscular subaortic infundibulum despite ventriculoarterial concordance. As a result, there is a high incidence of subaortic obstruction in this condition. Other associated anomalies are ventricular septal defect (VSD), right ventricular outflow tract obstruction, right ventricular
hypoplasia, juxtaposed atrial appendage, and right aortic arch. VSD, right arch, and juxtaposed appendage were present in our case.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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