Double outlet both ventricles: A rare ventriculoarterial connection

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Video clip is available online.

Double outlet both ventricles (DOBV) is an exceedingly rare ventriculoarterial connection wherein both arterial trunks override the interventricular crest, producing a ventricular arrangement seemingly connected equally to both arterial trunks. Understanding of the basic pathology and accurate preoperative diagnosis are prerequisite for a successful biventricular repair.

CLINICAL SUMMARY

A 3-month-old male patient weighing 5 kg was referred to us with symptoms of heart failure and cyanosis. Informed consent was obtained from the parents for the publication. Two-dimensional (2D) echocardiogram showed a large interventricular defect with a prominent outlet septum. As the echocardiography probe was swept from one end of the interventricular communication to the other, the commitment of the aortic root and the pulmonary root shifted from the left ventricle to the right ventricle (Video 1). A 3-dimensional (3D) model showed the conal septum perpendicular to the interventricular septum. Commitment of both ventricles to the aorta and the pulmonary artery confirmed the diagnosis of DOBV (Video 2).

The defect was approached through a longitudinal right ventriculotomy. There was a large interventricular communication that lacked a well-defined circumferential margin and extended from the septal leaflet of the tricuspid valve to the lateral extent of the pulmonary valve. The aortic valve was separated from the pulmonary valve by the conal septum. The aortic valve and the pulmonary valve were separated from the atrioventricular valves. A triangular patch carved from tanned autologous pericardium was used to close the part of the interventricular communication to partition the left ventricle from the pulmonary trunk. This

CENTRAL MESSAGE

DOBV is an abnormal ventriculoarterial connection, wherein biventricular repair is feasible with proper diagnosis and surgical planning.

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effectively creates a double outlet right ventricle with sub-aortic communication. A second larger curved patch was then used to tunnel the left ventricle to the aortic root. By using 2 patches, one can avoid twisting of the patch in the middle, which otherwise may occur with a single patch placed on the tortuous margins of the interventricular communication (Video 3).

Intraoperative TEE showed no residual shunts and unobstructed inflow and outflow tracts with good biventricular function (Video 4). The patient had an uneventful postoperative period.

DISCUSSION

Double-outlet ventricles are abnormal ventriculoarterial connections in which both arterial roots arise entirely or predominantly from one of the ventricles. These malformations are usually associated with an interventricular defect that forms the sole outlet from the other ventricle. In an extremely rare form, the defect is doubly committed, and the outlet septum is rudimentary or fibrous, thereby committing the defect to either arterial root, termed as DOBV.1

The various clinical manifestations are failure to thrive, cyanosis, feeding difficulties, and recurrent respiratory tract infections. The diagnosis can be readily established by 2D echocardiography.2 Three-dimensional echocardiography or virtual 3D modeling and 3D-printed models are the potential tools in surgical planning. In our case, the use of 3D-printed model certainly helped in surgical planning.

Iyer and colleagues2 reported 7 cases of DOBV; 6 patients underwent successful interventricular septation through the right atrium using 2 patches with promising early and midterm outcomes. A transventricular approach enabled us to define the entire margin of interventricular communication to perform an accurate repair. Biventricular repair is feasible in patients with DOBV. Definitive diagnosis and appropriate surgical planning are necessary for successful surgical outcome.

References