A New Approach to Surgical Correction of Double Outlet Right Ventricle with Remote Interventricular Communication

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Central Picture Legend:

Virtual dissection showing the remoteness of the inlet interventricular communication.

Central message:

Challenges remain when correcting double outlet right ventricle with a remote interventricular communication. We created a new communication, with excellent hemodynamic results.

Keywords:

Double outlet right ventricle, non-committed interventricular communication, mirror-imaged heart. Septomarginal trabeculation
Introduction:

Significant challenges are faced by the surgeon in achieving biventricular repair when, in the setting of double outlet right ventricle, the interventricular communication is remote from the arterial roots. This is known to occur in up to one-fifth of patients. (1) Having recently encountered a patient in this setting, we were able to construct an intraventricular baffle redirecting the left ventricle to the aortic root, having created a new outlet alongside the existing non-committed communication. The parents of the patient provided informed written consent for the publication of this data (IRB February 3, 2024. No#: Moh/CSR/CR/23/40).

Case report:

A one-year-old boy, known to have complex congenital cardiac anomalies, was referred to our center for evaluation. Transthoracic echocardiography (Video clip) revealed a mirror-imaged heart with double outlet right ventricle and bilateral infundibulums. The aortic root was anterior and left-sided relative to the pulmonary root. There was severe pulmonary valvar stenosis, with dysplastic leaflets, and subvalvar obstruction. The interventricular communication was perimembranous, and opened to the inlet of the right ventricle, remote from the arterial roots.

To aid in surgical planning, we made a contrast computed tomographic scan (Video clip), followed by modeling and printing (Figure 1a-c & video clip) of the three-
dimensional dataset. Intraoperative evaluation confirmed the location of the interventricular communication. It was remote from the aorta, with the leaflets and cords of the tricuspid valve covering its upper margin. We noted, however, the potential to create a new interventricular communication in the subpulmonary outflow tract (Figure 2a-b & video clip). Having created the new outlet, we placed a baffle in the right ventricle so as to direct both communications into the aortic root. Confirming the right ventricle to be of adequate size, we then placed a 16 millimeter Contegra conduit to the pulmonary arteries. Although the patient had moderate tricuspid regurgitation, with diastolic right ventricle dysfunction, this improved with medical management such that the patient could be discharged from hospital on the fourteenth postoperative day.

Subsequent postoperative evaluation after two months, including additional three-dimensional modeling (Figure 2c-d & video clip), revealed an unobstructed pathway from the left ventricle to the aorta. A four-dimensional magnetic resonance imaging scan demonstrated that three-quarters of flow from the left ventricle was across the newly created interventricular communication (Figure 2e).

**Comment:**

The relationship of the interventricular communication, when both arterial trunks arise exclusively from the right ventricle, depends on its location within the right ventricle relative to the arterial roots, and the length of the infundibulums. (2)
In this setting, it can be difficult to determine the precise anatomy using cross-sectional echocardiography. The use of three-dimensional modeling and printing has proven its value in surgical planning. (3) Virtual dissection of the volume rendered three-dimensional computed tomographic datasets also permits visualization of the intracardiac anatomy in a contextually appropriate manner. (4) Such virtual dissection in our patient (Figure 1e-f& central image) showed a defect that opened to the inlet of the right ventricle inferior to the caudal limb of the septomarginal trabeculation. The presence of tricuspid-to-mitral fibrous continuity confirmed its perimembranous nature, permitting us to infer that the conduction axis would be located postero-inferiorly. (5) Despite the remote nature of the defect, however, it was aligned with the subaortic outflow tract. To commit the defect to the aorta, nonetheless, would have required its extensive enlargement, along with detachment of some of the cords supporting the leaflets of the tricuspid valve. We deemed that such an approach might have mandated the use multiple patches (1,6,7), with the potential risk of re-interventions on the tricuspid valve, heart block, and obstruction of the outlet from the left ventricle. Enlarging the defect towards the subpulmonary outflow tract, and addition of an arterial switch, (8) was deemed inappropriate due to the stenosis of the pulmonary valve. We decided, therefore, to create an additional defect opening from the left ventricle to the subpulmonary infundibulum, having shown the right ventricle to be sufficiently large to
accommodate a baffle connecting both arterial roots to the left ventricle, while still permitting construction of a right ventricle-to-pulmonary arterial conduit.

The need to creating or enlarge an interventricular communication arises when the defect is the outlet for the morphologically left ventricle. (9) To our knowledge, ours is the first deliberate attempt to create an additional communication as an alternative surgical approach when correcting double outlet right ventricle with a remote interventricular communication. Our postoperative scan using four-dimensional magnetic resonance imaging showed that the maneuver created a more direct route to the aorta compared to the original inlet perimembranous defect. Theoretically, it would have been plausible simply to close the original interventricular communication, retaining only the new communication as the left ventricular outlet. This approach may be considered in future cases, although we would be wary that the new defect, having exclusively myocardial borders, might reduce spontaneously in size.
Figure legends

Figure 1:

Three dimensional modeling, showing the view from the right ventricle (panels a and b), reveals the location of the perimembranous interventricular communication in relation to the orifice of the tricuspid valve and the aortic root. The coronal view (panel c) shows the interventricular communication, despite its remoteness, to be directly below the aortic root. Panel d shows a comparable morphological specimen. The defect again opens to the inlet of the right ventricle, yet is directly below the aortic root. We have previously described this defect as being sub-aortic. We now recognize that, as in the current case, it is remote, albeit still potentially related to the aorta. The oval shows the site of creation of the new communication. Virtual dissections of the three-dimensional dataset (panels e and f) show the internal anatomy as viewed from the left and right sides of the ventricular septum.

Figure 2: Pre-operative modelling permitted us to anticipate the site of a new interventricular communication, which would open to the
subpulmonary outlet tract. The panels show its site as seen from the right (panel a) and left (panel b) ventricles, along with the planned suture line (yellow lines). Abbreviations: 1) tricuspid valve, 2) the original interventricular communication, 3) the new interventricular communication (marked in red in panel b). Postoperative modelling showing the subsequent anatomy and the flow from both interventricular communications to the aorta, again as seen from the right (panel c) and the left (panel d) ventricles (TV: tricuspid valve, RV: right ventricle, MV: mitral valve, LV: left ventricle). Magnetic resonance imaging permitting four-dimensional analysis of flow (panel e). Of the flow, three-quarters was through the new interventricular communication (white arrow), rather than the original defect (yellow arrow).

Video clip legend

The video clip, with its commentary, summarizes the case report and the three-dimensional modeling that demonstrates the preoperative anatomy, the surgical planning, and the postoperative anatomy. It also includes the preoperative echocardiography CT scan images and the postoperative 4D flow MRI.

References


Supplementary material
References:


