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Brief Report

Successful extensive enlargement of a non-committed ventricular septal defect in double outlet right ventricle

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Abstract We performed an arterial switch operation in a patient with double outlet right ventricle with non-committed ventricular septal defect, and abnormal insertion of the tension apparatus of the tricuspid valve which produced moderate tricuspid regurgitation. This required extensive enlargement of the ventricular septal defect between the attachments of the cords of the tricuspid valve so as to create the interventricular rerouting that made possible the arterial switch operation. Postoperatively, we produced a straight, unobstructed, left ventricular outflow tract, improved the extent of tricuspid regurgitation, and achieved low right atrial pressures. Enlargement of the interventricular communication can set the scene for biventricular repair in this particular subset of patients with both arterial trunks arising from the morphologically right ventricle.

Keywords: Interventricular rerouting; arterial switch operation; biventricular repair; surgery

BIVENTRICULAR REPAIR OF DOUBLE OUTLET right ventricle when the interventricular communication, or ventricular septal defect, is non-committed still represents a challenging and controversial procedure.[1,2] If biventricular repair is attempted, problems have been encountered such as late obstruction of the left ventricular outflow tract due to the length of the interventricular tunnel, tricuspid valvar dysfunction, and obstruction to right ventricular inflow, and the risk of reoperation is high.[1,3,4] The procedure is more difficult when there are abnormal insertions of the tension apparatus of the tricuspid valve, these abnormalities increasing the risks of leaving an obstructed interventricular tunnel, and promoting the likelihood of tricuspid valvar dysfunction. In a patient having anomalous attachments of the tendinous cords supporting the tricuspid valve to the superior margin of the interventricular communication, we enlarged the defect extensively in an antero-superior direction between the attachments of the tricuspid cords, and created an interventricular tunnel which, subsequent to an arterial switch operation, served to produce a biventricular repair. In this report, we describe and discuss our procedure.

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Case report

Our patient was noted to have a heart murmur soon after birth, and underwent extended end-to-end repair for coarctation of the aorta and banding of the pulmonary trunk at the age of 16 days, the diagnosis made at the hospital providing initial surgery being double outlet right ventricle with non-committed ventricular septal defect and aortic coarctation. Cardiac catheterization at the age of 1 year and 3 months revealed left and right ventricular end-diastolic volumes of 290 and 274 per cent of normal, respectively, and a severe sub-aortic stenosis of 6 millimetres diameter produced by a circumferential muscular ridge (Fig. 1a). At the age of 1 year and 8 months, weighing 8.7 kilograms, he was referred to our hospital for further surgical management. An echocardiogram then showed a left ventricular end-diastolic volume of 145 per cent of normal, left ventricular ejection fraction of 55 per cent, sub-aortic stenosis, mild aortic regurgitation, and moderate tricuspid valvar regurgitation. As we considered that a functionally univentricular repair using the Damus–Kaye–Stansel procedure would have courted the risk of long-term atrioventricular valvar

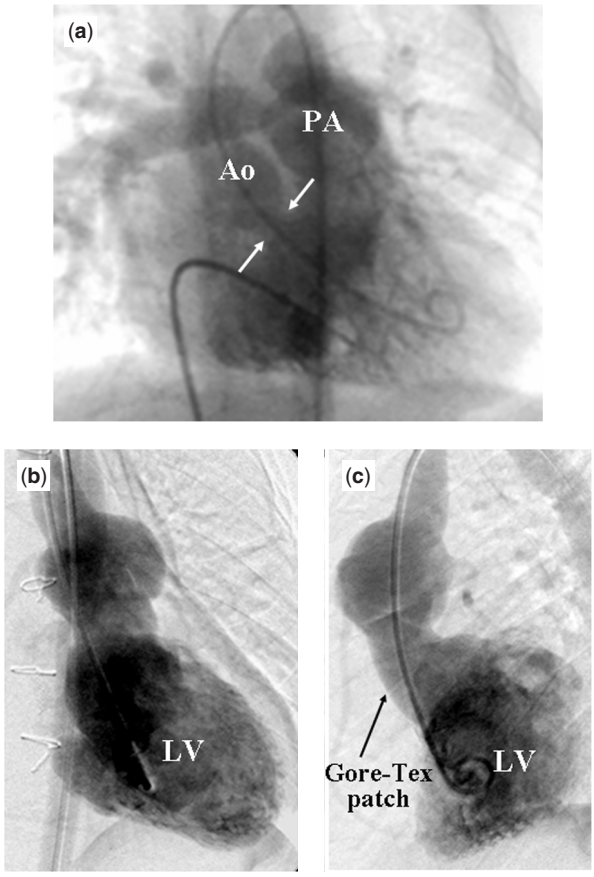

Figure 1.
*(**a**) Preoperative angiogram, (**b**) postoperative angiogram in frontal view, and (**c**) postoperative angiogram in lateral view. Ao: aorta; PA: pulmonary artery; LV: left ventricle; Arrow: 6 millimetres sub-aortic stenosis.*

regurgitation owing to the moderate regurgitation already present across the tricuspid valve, we opted to attempt biventricular repair with interventricular rerouting.

We proceeded to surgery using cardiopulmonary bypass with moderate hypothermia. The interventricular communication, of 13 millimetres diameter, was perimembranous, being roofed by fibrous continuity between the leaflets of the mitral and tricuspid valves, and opened exclusively to the inlet of the morphologically right ventricle. The tendinous cords supporting the leaflets of the tricuspid valve, inserted to the superior margin of the defect, obstructing the pathway to the sub-aortic area (Fig. 2a). We found tight muscular sub-aortic stenosis, with the aortic valve barely visible from our transatrial approach. Although the sub-pulmonary infundibulum was distant from the defect, it seemed feasible that, if we made an incision in the crest of the muscular septum antero-superiorly between the attachments of the cords supporting the leaflets of the tricuspid valve, we would be able to create a pathway from the

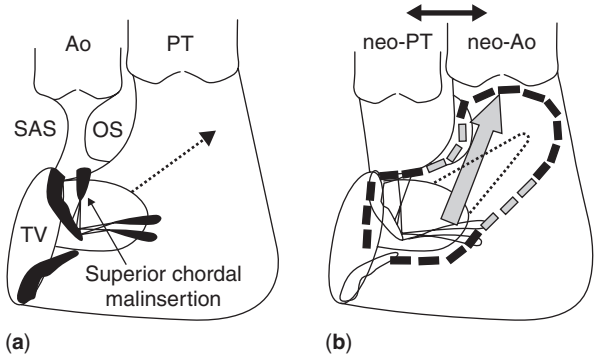

Figure 2.
*Operative scheme. (**a**) Anomalous insertions of the tendinous cords supporting the tricuspid valve to the superior margin of the ventricular septal defect obstructed the route to the sub-aortic area and potentially prevented incision of the muscular ventricular septum. The dotted arrow shows the line of our planned incision. (**b**) Despite the potential caveats, it proved possible extensively to enlarge the ventricular septal defect. The dotted line shows the area of enlargement. The arrow with the diagonal line shows the new route from the left ventricle to the aorta after the arterial switch procedure. Diagonal square: pledgets on the left ventricle; Black square: pledgets on the right ventricle; Ao: aorta; PT: pulmonary trunk; SAS: sub-aortic stenosis; OS: muscular outlet septum; TV: tricuspid valve.*

interventricular communication to the mouth of the sub-pulmonary infundibulum without disturbing the anomalous tendinous cords. By following this strategy, we succeeded in enlarging the defect anteriorly by 7 millimetres, continuing our incision until we reached the free wall of the left ventricle (Fig. 2a). We then constructed an interventricular tunnel of Gore-tex, working transatrially and through a short transverse right ventricular incision. As part of the reconstruction, we incised the muscular outlet septum, shifting it towards the native sub-pulmonary area by placing the patch on the native aortic side. We reinforced the cut margins of the ventricular septum by placing our sutures transmurally, thus preventing the development of an intramural haematoma and, at the same time, creating an adequate left ventricular outflow tract (Fig. 2b). Our interventricular patch of Gore-Tex was designed to minimize bulging, particularly at the inlet portion of the right ventricle, and also to avoid any obstruction within the newly created pulmonary channel. We used a pledgetted mattress suture at the base of the septal leaflet of the tricuspid valve to avoid atrioventricular block. As the aorta and pulmonary trunk were positioned side by side, it was also necessary to perform the arterial switch procedure, albeit without the need for anterior translocation of the pulmonary trunk (Fig. 2b). The muscular tissues that had produced obstruction in the initially sub-aortic area were resected through the old aortic valve, which became the pulmonary valve after the switch procedure, and the new

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pulmonary root was enlarged using autologous pericardium. Tricuspid valvoplasty was performed by approximating the zone of apposition between the antero-superior and septal leaflets.

The clinical course postoperatively was uneventful. At a month after the operation, catheterization showed a straight and unobstructed left ventricular outflow tract, improved tricuspid regurgitation, and right atrial pressures of 6 millimetres of mercury (Fig. 1b,c). The electrocardiogram showed sinus rhythm and complete right bundle branch block. When seen in the out-patient clinic at his referring centre, his condition was noted to be excellent, with no limitation of physical activities. Investigations revealed left ventricular end-diastolic dimensions of 118 per cent of normal, left ventricular ejection fraction of 49 per cent, and normal right ventricular pressures 16 months after the operation.

Discussion

The essential feature of our procedure was an appropriate and adequate incision made in the muscular ventricular septum sufficient to prevent injury to the cords supporting the tricuspid valve, and at the same time avoiding the creation of left ventricular outflow stenosis. Having assessed the nature of attachment of the tendinous cords in our patient, we deemed it possible to enlarge the defect extensively between the cords and towards the sub-pulmonary infundibulum. Although the pulmonary infundibulum was distant from the original defect, it proved possible to create a straight and widespread pathway from the left ventricle to the outflow tract, which became the new sub-aortic outlet subsequent to the arterial switch procedure. At the same time, we were able to improve the initial tricuspid valvar regurgitation by performing a tricuspid valvoplasty.

Ventricular dysfunction is always a concern when incisions have been made in the muscular ventricular septum. A study from Tokyo Women's Medical University, however, showed that enlargement of the ventricular septal defect decreases the risks of producing obstruction to the left ventricular outflow tract, and yet does not induce left ventricular dysfunction.[5] In fact, investigations of our patient 16 months after the operation showed that left ventricular volume was decreased, and left ventricular contraction was maintained, although the ejection fraction had decreased from 55 per cent preoperatively to 49 per cent postoperatively due to the large akinetic area produced by the internal conduit.

When assessing the optimal treatment for patients with double outlet right ventricle with non-committed interventricular communication, Delius et al.[6] opined that the risks in the short and intermediate term of biventricular repair might outweigh the potential long-term disadvantages of functionally univentricular repair. In two large series reported for surgical correction of double outlet right ventricle, nonetheless, the non-committed type of defect did not emerge as a risk factor for early or late death, and 83[3] and 94[4] per cent of patients, respectively, required no cardiac medication. Furthermore, Belli et al.[4] concluded that biventricular repair was almost always feasible when there were two adequate ventricles. Although we acknowledge the need for additional supportive data, we believe that the long-term functional results of biventricular repair are superior to those obtained following functionally univentricular repair. Especially in our patient, functionally univentricular repair carried the risk of progressive regurgitation across the systemic morphologically tricuspid valve. The good postoperative left ventricular function, coupled with normal right ventricular and low right atrial pressures, presage a better outcome over the long-term than the Fontan operation. Thus, we suggest that even if it proves necessary to carry out extensive enlargement of the interventricular communication, biventricular repair still provides the best surgical option for patients with double outlet right ventricle and non-committed interventricular communications.

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