Double-outlet right ventricle with non-committed ventricular septal defect

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Abstract

Objective: The term non-committed was used to define hearts in which the VSD was anatomically related to, or was close to, neither great vessel, being separated from both by considerable muscle. We report our experience of the surgical management of this subset, considered being of particular surgical relevance. Methods: Between January 1987 and December 1997, 23 patients having double-outlet right ventricle (DORV) with non-committed VSD underwent biventricular repair. Nine (39%) had undergone previous palliation. The median age was 20 months and the median weight was 8.5 kg. Two main types of repair were used: intraventricular baffle repair (n = 21) and arterial switch operation with VSD to pulmonary artery baffle (n = 2). At repair, 12 (52%) patients required concomitant VSD enlargement. In two other patients presenting with restrictive inlet VSD associated with tricuspid attachments, crossing the subaortic pathway biventricular repair was abandoned at operation. Results: There were two hospital deaths (9%, 70% CL: 3±19%). Eight patients (35%, 70% CL: 23±48%) underwent nine reoperations, six for subaortic stenosis. No late death occurred. At last visit, all patients were asymptomatic and only two had cardiac medication. Conclusions: The biventricular repair of DORV with non-committed VSD is feasible in the vast majority of cases with comparable results to other subsets of DORV. After repair, the subaortic region is at risk for development of subaortic stenosis. © 1999 Elsevier Science B.V. All rights reserved.

Keywords: Congenital; Heart septal defects; Ventricular; Double outlet right ventricle

1. Introduction

Since Kirklin described the first successful correction of double-outlet right ventricle (DORV), surgical repair has been extended to more complex forms of the malformation [1]. In 1972, Lev and Bharati introduced their classification according to the anatomic position of the VSD as related to the arterial trunks [2]. The term 'non-committed' (or remote) was used to define hearts in which the ventricular septal defect (VSD) was anatomically related to, or was close to, neither great vessel, being separated from both by considerable muscle. DORV with non-committed VSD has challenged surgeons throughout the modern era of congenital heart surgery [3–5]. This subset was known to have a poor outcome and higher risk for reoperation, and was frequently treated by an univentricular repair [6]. To our knowledge, this is the first serial report on the surgical management of this subset.

1.1. Anatomic considerations

In this series, the VSD was defined as non-committed on the basis of both preoperative echocardiographic data and, above all, intraoperative surgical inspection. Non-committed is not an anatomic definition: it was used to define the location of the VSD in cases with DORV in which the VSD was distant from both arterial valves [2]. This subset includes DORV with an atroventricular (AV) canal type, inlet (muscular) or trabecular VSD. Also, it exists DORV with conoventricular (perimembraneous) VSD distant to the great arteries because of the length of the subarterial conus. In those cases, echocardiography (subcostal view) shows the left ventricular outflow directed to the right ventricle instead of one of the great arteries. To avoid confusion, we suggest to define these defects as non-committed when the distance between the VSD and the arterial valves was at least equal to the matched aortic valve diameter of the particular patient (Fig. 1) [7].
2. Methods

Between January 1987 and December 1997, 23 patients having DORV with non-committed VSD underwent biventricular repair. The diagnosis and anatomic findings were based on a combination of echocardiography, angiography and surgical inspection.

The median age was 20 months (range: 50 days to 10 years) and the median weight was 8.5 kg (range: 3.5±24 kg). Nine patients (39%) had undergone previous palliation: seven patients had pulmonary artery banding, four associated with coarctation repair, and, two patients had modified Blalock-Taussig shunt. Twenty patients (87%) had at least one associated cardiac anomaly (Table 1).

The biventricular repair consisted in intraventricular baffle repair in 21 patients (91%). In all patients, the repair was performed through right ventriculotomy. Eleven patients required anterior or anterocaudal VSD enlargement, two patients had partial resection of the conal septum and one had both. In four of the nine patients presenting with the straddling or the abnormal attachments of tricuspid chordae precluding baffle construction, the previously reported principles of myocardial flap tailoring techniques were performed [8]. The pulmonary stenosis was treated by an infundibular patch in four patients, and by a transannular patch in one. Three patients required right ventricular outflow conduit repair. One patient had a valved conduit to avoid the section of a left anterior descending coronary artery crossing the infundibulum. One other patient had a reparation a l’etage ventriculaire (REV) procedure with the interposition of a valved conduit in pulmonary position [9]. Finally, one patient with L-malposition of the great arteries received a right ventricle to pulmonary artery conduit because of the presence of pulmonary stenosis and the natural course of right coronary artery excluding the insertion of a transannular patch. In four patients presenting without initial pulmonary stenosis, in order to avoid the development of an intraventricular obstruction due to the baffle, the right ventriculotomy was closed by means of a patch.

In two patients (9%), related to the great artery malposition (anterior aorta), it was easier to tunnelize the VSD to the pulmonary artery; the procedures were then completed by an arterial switch. The VSD was approached through right ventriculotomy in both, and both required infundibular muscle dissection.

During the study period, in two other patients, the biventricular repair was abandoned after surgical intracardiac exploration. Those patients constituted a small subgroup having a restrictive inlet VSD associated with tricuspid attachments crossing the subaortic pathway: one was converted to univentricular repair and the second patient, excluded from Fontan operation criteria because of high pulmonary vascular resistance, was left with his balanced pulmonary artery banding (Fig. 2).

3. Results

Two hospital deaths occurred (9%, 70% CL: 3–19%). The cause of mortality was low cardiac output in both, and was associated with intrapulmonary hemorrhage in one. Two patients developed postoperative atroventricular block and required pace-maker implantation (9%, 70% CL: 3–19%). One patient was treated for wound infection. At discharge echocardiography, six patients (29%, 70% CL: 18–42%) represented > 25 mmHg (maximum) left ventricle to aorta gradient. The follow-up was complete in 19 of

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<tr>
<td>Straddling AV valves b</td>
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<tr>
<td>Subaortic stenosis</td>
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a AV, atioventricular; CAVC, common atrioventricular canal; PA, pulmonary artery.
b Or abnormal tricuspid chordae attachments.
21 survivors (90%, 70% CL: 79–97%) at a median interval of 58 months.

No late mortality occurred. Eight patients (35%, 70% CL: 23–48%) underwent nine reoperations. The cause was subaortic stenosis in six (26%, 70% CL: 16–39%). The median delay between the two operations was 53 months (range: 10–96 months). Three of them had > 25 mmHg (maximum) left ventricle to aorta gradient immediately after the repair. In five, the previously described technique of extended septoplasty was successfully performed and in the remaining subaortic fibromuscular resection was enough to relieve the stenosis [10]. In addition, one patient required two reoperations for tricuspid valve repair and right ventricle outflow valve valvulization. One other patient who developed valve regurgitation underwent aortic valve replacement 6 years after the repair. No mortality occurred in reoperated patients. At last visit, three patients (16%, 70% CL: 7–29%) still require cardiac medication.

4. Discussion

The construction of an intracardiac baffle to tunnel the LV to the aorta using the VSD as the egress of the LV is the preferred surgical approach for the repair of the majority of the forms of DORV and also of d-transposition of great arteries with a VSD and pulmonary stenosis. The same principle is applied to Taussig–Bing hearts when an ASO is indicated: the VSD is tunnelized to the pulmonary artery and repair is completed by an ASO. On the other hand, reconstruction of the right ventricular outflow, if necessary, requires a variety of surgical techniques, including the Lecompte manoeuvre for the REV operation, infundibular dissection with outflow patch enlargement and extracardiac conduit insertion using homograft or synthetic valved conduit.

This study emphasizes the importance of detailed definition of the intracardiac anatomy. Although in our recent series [11] the location of the VSD did not influence operative outcome, the classification of DORV according to the VSD site introduced by Lev and Bharati remains an essential surgical-anatomic distinction. In our experience, the VSD location was so relevant that it guided the surgical strategy, namely surgical technique and timing for definite repair. In the presence of two adequate, even slightly imbalanced ventricles, the following anatomic criteria helped to determine the surgical management: great artery relationship, the presence or absence of pulmonary stenosis and other major cardiac anomalies.

The term non-committed was first used by Lev et al. in 1972 [2]. In 1975, Zamora, Moller and Edwards employed the term ‘remote’ to define these defects [12]. Van Praagh et al. also used the term ‘uncommitted’ [7]. Both terms were used to describe the presence of a ‘considerable’ distance...
between the VSD and the outflow tracts supporting the aorta and the pulmonary trunk rather than being anatomical definitions. Thus, this subgroup of DORV included patients with AV canal type VSD, with trabecular VSD (inlet or not), but also patients with conoventricular VSD distant to the arterial valves (Fig. 1). Our data correspond to the series of previous publications on the morphologic features of this anomaly. Van Praagh et al. presented 32 postmortem cases of DORV with mainly conotrunkal malformations [7]. Eleven of them had non-committed VSD, including 10 conoventricular and one muscular VSD. Eighteen specimens in Stellin et al. included 12 perimembranous VSD with inlet extension, three AV canal type VSD, two perimembranous juxtapulmonary VSD (classified as surgically non-committed) and one inlet muscular VSD [5]. In this series, the incidence of the need for VSD enlargement

Fig. 3. Intraoperative transventricular view of a heart having DORV with non-committed conoventricular VSD. (A) The VSD is considerably distant from both arterial valves. (B*) baffle construction almost always requires parietal band section. The insertion of the baffle patch includes both interrupted pledget-supported and continuous prolene sutures.
and/or conal septal resection in order to construct an unobstructed left ventricle outflow was particularly high (57%). In those patients, the VSD was enlarged anteriorly or antero-caudally, and a large connection between the left ventricle and the baffle tunnel was obtained. Also, infundibular muscle resection, particularly in the presence of a well-developed parietal band creating a potential subaortic stenosis, was almost always required. In addition, with the two patients presenting with type C AV canal type VSD, in nine patients the atrioventricular valve straddling or the abnormal tricuspid valve chordae attachments on the outlet septum were observed. A variety of surgical techniques were performed [8]. In four patients, the abnormal tricuspid chordae were mobilized by tailoring myocardial flaps which secondarily were reimplanted at the edges of the baffle patch; no evidence of postoperative tricuspid valve regurgitation was observed in those patients. In this series, although there was a high rate of AV valve insertion anomalies, the overriding of the later was not observed [13,14]. At repair, the conventional anatomical landmarks of the conduction axis were considered. The sutures were placed rather at the right ventricular aspect of the VSD rim and on the leaflet tissue of the tricuspid valve, when necessary. However, two patients (9%) developed postoperative complete AV block and required permanent pace-maker implantation. Fig. 3 represents the intraoperative transventricular view of a heart with DORV and non-committed conoventricular VSD, before (A) and after (B) baffle construction.

In all patients, the intracardiac baffle was constructed through right ventriculotomy. Although right ventriculotomy may result in arrhythmogenic potential of the scar, we believe that the adequate intraventricular repair of DORV and non-committed VSD almost always requires right ventriculotomy [15]. In this series, late rhythm disorders related to right ventriculotomy were not observed. Although the Fontan operation has been proposed as a solution to the complex forms of this anomaly, it was always our policy to push the indications of biventricular repair as far as possible [16,17]. Since the functional state of patients following the Fontan operation deteriorates with time, we elected to primarily perform the biventricular repair even in the very complex forms [18]. It is an obvious fact that patients who underwent biventricular repair of DORV with non-committed VSD constitute a high risk group for need of reoperation for secondary subaortic stenosis. At the present time, one-quarter of the patients of this series has been reoperated on. We recently reported our experience on subaortic stenosis after biventricular repair for DORV [10]. The reoperation for subaortic stenosis should not only aim to relieve the obstruction, but also to streamline the left ventricle outflow pathway; ‘extended septoplasty’ constituted an adapted technique to treat this complication. We performed this operation in five of this series of patients, with excellent early results. The data allows us to conclude that a certain percentage of patients having DORV and non-committed VSD require a sort of two-stage management: the biventricular repair and the reoperation for the progressively developing subaortic stenosis relief which can be treated by a standardized technique with excellent results.

References

Appendix A. Conference discussion

**Dr J. Van Son (Leipzig, Germany):** Since you have a 21% rate of subaortic obstruction after intracardiac baffle repair for double-outlet right ventricle with non-committed ventricular septal defect, what is your cut-off point in terms of age for this type of repair? I presume that you attempt to postpone the operation beyond the 1st year of life in order to be able to construct a relatively large baffle.

**Dr Belli:** Yes, when it is possible to wait. Thus, it is our policy to push the indications of biventricular repair as far as possible. In this series, one-fourth, maybe it will be one-third later, needed reoperation. This could be realized with good results and without mortality. We can conclude that a certain percentage of patients required a two-stage management: the first was the repair and the second was the treatment of the secondary subaortic stenosis. Thus, in our experience, this is preferable to an univentricular management.

**Dr Van Son:** Are you sure about that? In my experience and that of others, such as the Great Ormond Street group, there are situations in which the short- and long-term survival after a complex biventricular repair, as such for double-outlet right ventricle with a non-committed ventricular septal defect, has certainly been far less optimal than after univentricular palliation. Although the Fontan principle is clearly a palliative one, the incorporation of an often poorly functioning right ventricle with associated potential problems such as subaortic obstruction is also palliative in nature and the wisdom of this biventricular approach can be questioned. Why did you not preferably apply univentricular palliation in the patient cohort with high potential for subaortic obstruction?

**Dr Belli:** As recently reported, we believe also that the follow-up after Fontan repair in terms of mortality and quality of life deteriorates with time. On the other hand, in almost all patients who had developed or not, subaortic stenosis are growing well with no cardiac medication nor rhythm disturbance.

**Dr K. Yamani (Mecca, Saudi Arabia):** The question I wanted to ask, how can you avoid the conduction system, injuring the conduction system in this kind of operation?

**Dr Belli:** At operation or at reoperation?

**Dr Yamani:** At the operation.

**Dr Belli:** When the VSD is muscular inlet the risk of AV block is probably higher. But the other subsets of non-committed VSD don’t represent a particular risk compared to the other locations of VSD in double-outlet right ventricle.

**Dr Yamani:** Did you entertain any AV block passed off in this kind of operation?

**Dr Belli:** In this series, we had only two patients who required pacemaker implantation; one of them died because of low cardiac output and pulmonary complications. The frequency was not so significant compared to the other patients who underwent DORV repair with VSD enlargement.

**Dr J. Moll (Lodz, Poland):** I found you had tricuspid insufficiency in those patients which had chordae transferred. And I did another technique. I did incision in the patch and put the chordae through the patch. And I found if very useful, with no tricuspid regurgitation. Of course, it’s impossible to do it in all patients, but in this case it was very useful.

**Dr Belli:** We used the technique that you described. We also used other techniques to avoid chordae. In four patients there was no other solution than doing this particular technique. We observed then, at reoperation, two of them with tricuspid chordae in their initial place without any valve regurgitation. So, this allows us to present the technique.

**Dr B. Maruszewski (Warsaw, Poland):** In the previous sessions, we discussed the specifically good performance of patients with two ventricles after Fontan operations. I asked the question and haven’t heard the answer, may I ask you again, what are the exclusion criteria, for these patients from biventricular repair? Which patients would you exclude from this operation with your experience and reoperation rate?

**Dr Belli:** We had 25 patients who were on the operating table for biventricular repair. And I believe the presence of components, together, such as inlet restrictive VSD with the tricuspid chordae crossing the outflow, also with the necessity of doing subaortic resection of a parietal band, should be an indication, maybe, to abandon doing the biventricular repair because this becomes a high-risk procedure. However, the patient who had Fontan repair with chordae straddling or abnormal attachments, developed very soon AV valve incompetence, he had the mechanical valve and later, he required a transplantation. So it’s a good example to insist to do biventricular repair for these cases.