Left atrial isomerism: biventricular repair

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Abstract

Objective: Biventricular repair of hearts with left atrial isomerism often necessitates complex atrial and ventricular baffle procedures. We analysed our experience with an accent on baffle techniques. Methods: From 1997 until 2008, 12 patients (four male) with left atrial isomerism received biventricular repair. Their median age at surgery was 9 (range: 1—24) months. Four patients had dextrocardia. Nine patients presented with left superior vena cava, three with absent right superior vena cava, five with unroofed coronary sinus and nine others with inferior vena cava interruption with (hemi)azygos continuation. Anomalous pulmonary venous drainage was present in three patients. Eight had a monoatrium. Atrioventricular septal defect (AVSD) occurred in six (complete AVSD in two). One patient with complete AVSD had right pulmonary agenesis with long segment tracheal stenosis. Multiple VSDs presented in one whereas three patients had double-outlet right ventricle (DORV) (one with borderline LV hypoplasia). Two had previous pulmonary artery banding. Complex intra-atrial baffle constructions were performed in seven patients. Complete AVSDs were corrected using two patches and all other AVSDs had one patch repair. Multiple VSDs were closed directly. DORV patients had intraventricular tunnel repair. Results: No early mortality occurred. Median follow-up was 54 (range: 2—134) months. One patient with complete AVSD and pulmonary agenesis died late after tracheal repair. Four patients needed five re-operations (closure of residual ASD (one), relief of left (two) or right (two) ventricular outflow obstruction, pulmonary artery branch plasty (one)). There was no atrial baffle stenosis. Four received a pacemaker. All survivors are in NYHA class I. Conclusions: Survival and functional status of left isomerism patients after biventricular repair is good. Complex repairs with atrial or ventricular baffles are frequent. Arrhythmias were common and pose a concern late after repair.

1. Introduction

Patients presenting with atrial isomerism (visceroatrial heterotaxy syndrome) characteristically show complex cardiac and extracardiac anomalies. Two subtypes of atrial isomerism are recognised: left atrial isomerism (LAI) and right atrial isomerism (RAI), characterised by atrial appendages on both sides of the body having the appearance of morphologically left atrial appendage and right atrial appendage, respectively. Cardiac anomalies most commonly associated with atrial isomerism include anomalous systemic and pulmonary venous connections, anomalies in atrioventricular (most commonly atrioventricular septal defect — AVSD) and anomalies in ventriculoarterial connections (most commonly of double-outlet right ventricle (DORV) type) [1]. As a consequence, for many patients presenting with atrial isomerism, the only surgical option is single ventricle palliation. In recent years, methods for surgical biventricular repair have been developed. When attempting a biventricular repair in these patients one is often faced with creating complex intra-atrial baffles or intraventricular tunnels, complicating repair.

This article presents a study of our surgical experience with biventricular repair in left atrial isomerism patients, with an accent on intra-atrial baffle repair.

2. Materials and methods

Between 1997 and 2008, 32 patients with left atrial isomerism who underwent surgical treatment were identified from our surgical database. Diagnosis of left atrial isomerism was established preoperatively via ultrasound investigation using definitions published by Huhta et al. and through direct examination of the atrial appendages during surgical procedures [2]. Of these 32 patients with left atrial isomerism, 12 received biventricular repair at our centre. Four (33.3%) of these 12 patients were male. All others had single ventricle palliation.

Of these 12 patients, two had previous pulmonary artery banding. Seven patients received complex intra-atrial baffle constructions, whereas one patient underwent surgery for...
complex tracheal stenosis. The median age at biventricular repair was 9 (range: 1—24) months. The median follow-up was 54 (range: 2—134) months.

2.1. Anatomy

Besides left atrial isomerism, our patients exhibited the following anatomical anomalies. Ventricular mass was predominantly right sided (dextrocardia) in five of our patients, with four of these patients having the cardiac apex pointing to the right. One of these patients with right lung agenesis had the apex pointing to the left.

An AVSD was frequent in this group with two complete AVSDs (defined by a common valvar orifice, no or few attachments of the bridging leaflets to the ventricular septal crest, and a potential for shunting on atrial and ventricular levels). Two intermediate-type AVSDs (defined as having two separate valvar orifices, with fibrous attachments to the ventricular septal crest and a potential for shunting on ventricular but mostly on atrial level) and two partial AVSDs (defined by two valvar orifices, bridging leaflets firmly fused at and to the ventricular septal crest and minimal potential for shunting on ventricular level). Multiple muscular VSDs presented in one patient.

Eleven patients had balanced ventricles; the topology was right-handed in eight and left-handed in four patients. One of the patients had borderline hypoplastic left ventricle.

Ventriculoarterial connections were concordant in nine and discordant DORV type in three patients. All DORV patients had a side-by-side relationship of the great arteries with a right-sided aorta and a peri-membranous VSD with sub-aortic extension. In one of these patients, there was no outlet septum and therefore there was a doubly committed VSD. All patients with DORV had separate atroventricular junctions guarded by normal tricuspid and mitral valves. Type and mode of atroventricular and ventriculoarterial connections are represented in Table 1.

We found different anomalous systemic and pulmonary venous connections in our group of patients (Fig. 1.). A left superior vena cava was present in nine patients, an interrupted inferior vena cava with (hemi)azygos continuation was observed in nine patients, an unroofed coronary sinus in five: four of these on the left side and one on the right side and an absent right superior vena cava was present in three. The liver veins drained in one patient to the left-sided atrium and in the middle of the monoatrium in another three. In eight, the ASD was so large or remnants of the atrial septa were either so small or not present we could claim a true monoatrium.

Bilateral left bronchus morphology was observed in all. Multiple spleens were present in nine whereas in the remaining three patients this was not reliably documented. It can be argued that in terms of pulmonary venous connections an anomalous pulmonary venous connection is impossible in left atrial isomerism. For that reason, we intent to describe the pulmonary venous connections as accurately as possible.

One patient had an anomalous pulmonary venous drainage with the right superior pulmonary vein draining separately to the right-sided atrium, and the other three pulmonary veins draining via a vertical vein to the right-sided atrium. In two other patients with dextrocardia, the left-sided pulmonary veins drained antero-superiorly in the left-sided atrium with the right-sided veins draining posteriorly and more towards the midline of the right-sided atrium. In three patients, we found the pulmonary veins draining via a common orifice to the appropriate sided (pulmonary) atrium.

One patient had right lung agenesis with long segment tracheal stenosis. One patient had a persistent ductus arteriosus (PDA) as a solitary anomaly.

2.2. Surgical repair

All except one patient underwent their correction through cardiopulmonary bypass, with standard aortic and bicaval cannulation, mild-to-moderate hypothermia and St. Thomas cardioplegia repeated every 30 min if needed.

The remaining patient underwent closure of PDA through a left anterior thoracotomy.

In all cases, a biventricular repair was accomplished with the morphological left ventricle as the systemic ventricle.

Two patients had previous pulmonary artery banding. One had an AVSD and the second a DORV with borderline hypoplastic left ventricle. In case of complete AVSD, a two-patch technique was used, whereas in partial or intermediate-type AVSD, a single-patch technique was used.

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Table 1

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<thead>
<tr>
<th>Type of atrioventricular connection</th>
<th>Left isomerism (n = 12)</th>
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<tbody>
<tr>
<td>Biventricular (mixed)</td>
<td>12</td>
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<tr>
<td>Mode of atrioventricular connection</td>
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<tr>
<td>Two separate and perforate valves</td>
<td>6</td>
</tr>
<tr>
<td>Common AV valve (AVSD)</td>
<td>6</td>
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<tr>
<td>Ventricular topology</td>
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<tr>
<td>Biventricular with right hand topology</td>
<td>8</td>
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<tr>
<td>Biventricular with left hand topology</td>
<td>4</td>
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<tr>
<td>Ventriculoarterial connections</td>
<td></td>
</tr>
<tr>
<td>Concordant</td>
<td>9</td>
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<tr>
<td>Double-outlet right ventricle</td>
<td>3</td>
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In all AVSDs, the 'cleft' in the left atrioventricular (AV) valve was always closed.

In all patients with DORV an intraventricular tunnel repair was performed. In one patient with DORV, anomalous pulmonary venous connection and borderline hypoplastic left ventricle was first thought not to be suitable for biventricular repair and was put on a single ventricle palliation track with a bidirectional Glenn shunt. Later he underwent a successful biventricular repair. The Glenn anastomosis was taken down, a right ventricle to pulmonary artery continuity was re-established, the DORV was treated by an intraventricular tunnel repair and an intra-atrial baffle for blood redirection to appropriate ventricles was created.

2.3. Atrial septation

Due to complex and numerous systemic and pulmonary venous anomalies, a complex atrial baffle construction was needed in seven cases. In all cases, a bovine xenopericardial patch was used for baffle construction. In some cases, more than one patch was needed to create an unobstructed atrial baffle. All systemic venous blood (caudal veins in normal position or 'unroofed' left superior vena cava (LSVC) and hepatic veins) were redirected to the right ventricle and the pulmonary circulation while at the same time directing all pulmonary venous blood to the left ventricle and systemic circulation. In patients who had AVSD as the major associated anomaly, we created atrial baffles from patches used for atrial side of the repair. Fig. 2 depicts types of complex atrial baffles used in correcting our patients. As can be seen in Fig. 2(A—C), in cases of 'unroofed' LSVC (or right superior vena cava — RSVC) the patch needs to be placed in such a way that blood from the LSVC drains to the systemic venous atrium. In this way, the atrium is septated in obliquely with a risk of obstructing the blood flow in both atria or making one atrium too small. When this risk was considered too high, for example when the posterior midline of the atrium and the orifice of the LSVC are relatively farther apart, we chose to reconstruct the coronary sinus with a xenopericardial patch and use a separate patch to septate the atrium. The pulmonary veins now drain over the newly constructed coronary sinus baffle to the mitral valve (Fig. 2F). In cases of anomalous pulmonary venous connections (as described above, all pulmonary venous connections other than that of all pulmonary veins entering the posterior aspect of the pulmonary venous atrium), it is sometimes difficult to bring pulmonary venous blood unobstructed to the systemic ventricle. In Fig. 2 cases D and E, we observe patients with dextrocardia and anomalous pulmonary venous connection, which may complicate atrial septation. In one case we were able to achieve septation using only one long obliquely placed patch, whereas in the second case the distance between both lung veins was considered as being too long, so here initially a baffle was created connecting all pulmonary veins and draining them towards the pulmonary venous atrium and subsequently another patch was used to accomplish final septation of the atrium into a systemic and a pulmonary venous compartment. In another patient with anomalous pulmonary venous connection (G), we were able to direct pulmonary venous blood to the left ventricle using one large obliquely placed patch.

Fig. 2. Atrial baffles. Schematic representation of atrial baffles. (A—C) Cases with AVSD with 'unroofed' LSVC, where atrial septation was achieved with the use of obliquely positioned patch used for the atrial side of the AVSD repair. (D and E) Two cases with anomalous pulmonary venous connections are shown. In one case septation was achieved by using two patches (D). The first patch was used to connect the pulmonary veins draining them towards the pulmonary venous atrium and the second for septating the atrium. In the second case (E) were able to achieve septation by placing one oblique patch. (F) Septation with reconstruction of coronary sinus is shown. (G) A case with anomalous pulmonary venous connections and DORV is shown. Septation was achieved with the use of oblique patch, this patient also needed an intraventricular tunnel repair.

3. Results

There were no early deaths. One patient with complete AVSD and pulmonary agenesis and tracheal stenosis died late after a second tracheal repair, due to multi-organ failure caused by sepsis. This patient had first undergone repair of long segment tracheal stenosis and pulmonary artery banding, later followed by successful biventricular repair of a complete AVSD.

3.1. Re-operations

Four (33.3%) patients needed five re-operations. One patient with AVSD and a complex atrial baffle construction had to be re-operated 2 months later due to atrial baffle patch dehiscence. Two patients with DORV (one with borderline hypoplastic LV after PA banding and another with TAPVD) needed relief of recurrent or residual right ventricular outflow tract obstruction (RVOTO) 4 and 5 months after repair, respectively. One of these two patients needed a revision and plasty of pulmonary artery branch after 1 month, while at the same time left ventricular outflow tract obstruction (LVOTO) resection was done. One patient with complete AVSD had sub-aortic stenosis resection 26 months after first repair.

3.2. Arrhythmia

Eight (66.6%) patients had arrhythmias since repair. Six patients experienced arrhythmias early after repair, we
observed complete AV block in three patients, transitional higher-degree AV block in two patients and periods of premature ventricular contractions in one patient. Three patients with complete AV block needed pacemaker implantation; all had AVSD correction. Two patients developed complete AV block late after repair 5 months and 5 years after repair, respectively. One of these patients had pacemaker implantation. The patient who developed AV block after 5 years had no intracardiac surgery. No spontaneous AV block, sinus node dysfunction or any other arrhythmias were reported prior to repair.

3.3. Follow-up

Follow-up was available for all patients. Median follow-up was 54 (range: 2–134) months. Echocardiography results were available for all patients. Four (33.3%) patients had residual lesions, moderate left AV valve insufficiency, moderate left ventricular outflow tract obstruction and residual apical VSD. All patients showed good left and right ventricular function, with shortening fraction between 30% and 41%. There is no atrial baffle stenosis present. All patients are in NYHA class I and without any cardiac medication at last follow-up.

4. Discussion

Patients presenting with heterotaxy syndrome are characterised by numerous cardiac and extracardiac anomalies. In recent years, advances in operative and pre- and postoperative care have improved outcomes in these patients, especially in single ventricle palliation patients [3]. Recent studies suggest similar survival rates in patients treated by biventricular repair or single ventricle palliation, yet only 70% of patients undergoing single ventricle palliation are in NYHA class I compared to 98% of patients treated with biventricular repair [4,5].

In this article, we have studied our experience with biventricular repair in patients diagnosed with left atrial isomerism. In recent reports, authors have reported their ability to perform biventricular repair in 30–50% of patients presenting with left atrial isomerism [6–8]. We were able to perform biventricular repair in 12 (37.5%) out of 32 patients with left atrial isomerism. When attempting a biventricular repair in these patients, numerous systemic and pulmonary venous connection anomalies (Fig. 1) may complicate the repair, and complex atrial baffles are often needed for successful atrial partition [12]. All systemic venous blood needs to be redirected to the right ventricle and the pulmonary circulation, while ensuring that all pulmonary blood flow goes to the left ventricle and systemic circulation. In the presence of LSVC with unroofed coronary sinus, redirecting systemic venous blood can be challenging. This could be achieved either by placing a long oblique patch that had to be sutured directly underneath the ostium of the LSVC in the roof of the atrium, or by reconstructing a new coronary sinus and thereafter septating the atrium with a separate patch. A similar problem arises in the case of associated anomalous pulmonary venous connection. Pulmonary venous blood needs to be baffled towards the left ventricle, which we were able to achieve using a single, oblique, properly trimmed patch or multiple patches (Fig. 2). It was possible to create unobstructed atrial baffles redirecting blood flow to the appropriate ventricles in all of our patients, with last follow-up showing no obstruction of these atrial baffles. We observed patch dehiscence, with subsequently a haemodynamically important residual ASD, needing surgical repair in one patient. In this patient, the initial patch was augmented by a second patch at first surgery. All of our patients are in NYHA class I at last follow-up; however, they were not free of re-operations, as mentioned in Section 3.

The reported incidence of arrhythmia in patients with left atrial isomerism after surgical repair is high [5]. Some authors reported an incidence of arrhythmia up to 50% [8]; many of these patients needed postoperative pacemaker implantation. Arrhythmias after repair were also common in our group of patients. Eight patients developed arrhythmias after surgery. Three of them needed pacemaker implantation early after repair, and one late after repair. All pacemakers were implanted due to complete AV block. All patients needing pacemaker implantation had an AVSD. Complex cardiac lesions in left isomerism patients include abnormalities of conduction system: absence, hypoplasia or abnormal location of the sinus node. Abnormalities of AV conduction system seem to be connected to associated abnormalities of AV connection and ventricular architecture [9]. Studies suggest that AV block is more common (between 7% and 39%) in left isomerism patients and is 4 times more common in patients with an associated AVSD [10]. Studies also suggest that progressive slowing of atrial rhythm is typical of left isomerism patients [11]. One of our patients developed AV block 5 years after surgery for PDA, no intracardiac surgery was done in this patient. All patients tolerated arrhythmias well after biventricular repair and, in spite of pacemaker implantation, are doing well. Remarkably enough, we have not encountered any preoperative AV block or sinus node dysfunction in this group.

In this study, we have seen that biventricular repair in left isomerism patient is feasible and functional results are good. We feel that biventricular repair is usually in the presence of normal sized ventricles, AV valves and outflow tracts. When these criteria are not met, we would perform a univentricular palliation in these patients.

References

Appendix A. Conference discussion

Dr C. Pizarro (Wilmington, Delaware, USA): Anatomic complexity and variability constitute the hallmark of heart disease associated with heterotaxy. Therefore, it is difficult to decide which is the best management option when confronted with this entity.

This decision is underpinned by the underlying anatomy, the presence of extracardiac anomalies, and institutional preference or bias. However, despite our best efforts, late morbidity and mortality remain a considerable issue.

The incidence of biventricular repair in this series falls within the 30% to 40% range reported by larger series; however, this figure can vary depending on the criteria set to undertake biventricular repair.

I noticed that you exclude patients with pulmonary stenosis or pulmonary atresia from consideration for the biventricular repair track, and unbalanced AVSD has been reported as a risk factor for mortality in large series, so this can be the reason for arrhythmias.

What criteria, echo, MRI, for example, would you use to offer biventricular repair in the patients with borderline forms of unbalanced AVSD? Secondly, the outcome of these patients in terms of survival and functional class appears to be quite good; however, this is at the expense of a substantial rate of reinterventions. Keeping in mind that patients managed towards single ventricle represent the most unfavourable end of the spectrum, can you tell us what number of patients did undergo single ventricle palliation during the same time period, and what is the functional outcome of those, particularly those who underwent univentricular repair, so I have no data in this regard.

Nevertheless, we need to know that these patients also exhibit anomalies in the sinus node with hypoplasia or malposition and malformations of the AV conduction system. So this can also be the reason for arrhythmias.

We have seen in two of our patients that arrhythmias occurred late after repair, in one five months and in the other five years after the repair. And it is common or, let’s say in these patients we can see a progressive slowing of the atrial rhythm and eventually they develop AV block without any surgery.

Dr Stellin: Yes. I agree.

Dr Vodiskar: Of course, when doing complex atrial baffles, there are numerous suture lines in the atrium. At the same time I wonder whether your patients would have been better off being treated by a bicaval pulmonary anastomosis, rather than rerouting the blood with baffles within the common atrium.

Dr Vodiskar: First, I would like to answer the second question. We unfortunately did not study the other two-thirds of the population who underwent univentricular repair, so I have no data in this regard.

Nevertheless, we would evaluate each patient separately to decide whether biventricular repair is possible, usually by the means of echo. We would, of course, use standard measurements of size of the ventricles and of the AV valves and ventricular outflow tracts.

What was the —

Dr Pizarro: The first question is largely regarding which criteria you would use in the patients who are in the middle of the spectrum regarding biventricular repair single ventricle?

Dr Vodiskar: I don’t have a definite answer to that question. We had one patient with a hypoplastic or borderline hypoplastic left ventricle with a double-outlet right ventricle who was first put on the univentricular palliation track, but later we decided to convert to biventricular repair. He underwent successful biventricular repair.

Nevertheless, he needed two re-operations due to right ventricular outflow tract obstruction and pulmonary artery branch obstruction, and he later also developed left ventricular outflow tract obstruction.

So I can’t offer you any guidelines. I think that every patient needs to be evaluated separately, and then the decisions are made on the basis of associated morphology or size of the ventricles.

Dr G. Stellin (Padova, Italy): In your experience there is 66% of arrhythmia incidence at your follow-up.

Dr Vodiskar: Yes.

Dr Stellin: I wonder whether the arrhythmias are somehow related to the suture lines inside the atrium. At the same time I wonder whether your patients would have been better off being treated by a bicaval pulmonary anastomosis, rather than rerouting the blood with baffles within the common atrium.

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