Mid-term results of bidirectional cavopulmonary anastomosis and hemi-Mustard procedure in anatomical correction of congenitally corrected transposition of the great arteries†

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Abstract

OBJECTIVES: The Senning or Mustard procedure combined with the arterial switch operation (ASO) (± VSD and no left ventricular (LV) outflow tract obstruction) or the Rastelli operation (VSD and LV outflow tract obstruction) has become the preferred strategy over conventional repair as it is thought to prevent long-term dysfunction of the right ventricle (RV). More recently, hemi-Mustard rerouting of blood from the inferior vena cava to the RV in combination with bidirectional cavopulmonary anastomosis (BCPA) has been adopted by some centres for potential benefits over the classic atrial switch procedure. The aim of this study was to analyse our experience with hemi-Mustard and BCPA as part of an anatomical repair of congenitally corrected transposition of the great arteries (CCTGA) in selected patients.

METHODS: Between 2004 and 2011, eight patients underwent hemi-Mustard/BCPA with the Rastelli operation (n = 6) or ASO (n = 2). The median age was 2.9 (range: 1.2–9.1) years. Positional anomalies were present in 75% of the patients. Both patients with ASO had dysplastic and insufficient tricuspid valves. In the Rastelli group, four patients had previously received shunts followed by BCPA in one patient. In the ASO group, both patients underwent pulmonary artery banding initially.

RESULTS: There was one in-hospital death and no late mortality. Two patients received a pacemaker. One patient from the Rastelli group required conduit change 6 years later. At the mean follow-up of 4.5 years, six and one patients are in NYHA classes I and II, respectively; six patients showed good biventricular function, while one had LV dysfunction. Systemic venous obstruction and sinus node dysfunction were not observed, and BCPA was functioning well in all patients.

CONCLUSIONS: Hemi-Mustard/BCPA is useful in anatomical repair of CCTGA in selected patients. When compared with the classic atrial switch operation, it is technically easier which makes it especially helpful in atrio-apical discordance; it unloads an RV with limited size or function, and avoids complications related to the upper limb of the classic atrial switch procedure. Mid-term results of this approach are favourable. Further follow-up is needed to prove long-term benefits.

Keywords: Congenital heart defects • Congenitally corrected transposition • Discordant atroventricular connection • Mustard procedure • Arterial switch operation • Rastelli operation

INTRODUCTION

Congenitally corrected transposition of the great arteries (CCTGA) is characterized by discordant atroventricular and ventriculo-arterial connections. It is often associated with other anomalies: ventricular septal defect (VSD), left ventricular outflow tract obstruction (LVOTO), Ebstein-like malformation or valvar dysplasia of the tricuspid valve (TV) and positional heart anomalies, e.g. dextrocardia and mesocardia in situs solitus, or levocardia in situs inversus. Late outcomes after conventional repair retaining a right ventricle (RV) in systemic position were unfavourable particularly due to dysfunction of the RV and the TV [1].

Many centres have now adopted anatomical repair allowing the left ventricle (LV) to support the systemic circulation. Several groups have reported excellent short- and mid-term results with this approach [2–4]. The anatomical repair comprises of two techniques based on the presence or absence of the LVOTO: the
double switch procedure (atrial and arterial switch operation (ASO)) in the absence of LVOTO or atrial switch plus Rastelli operation when LVOTO is present. The atrial switch procedure can be either a Senning or a Mustard operation.

Recently the benefits of a modified atrial switch procedure (hemi-Mustard procedure rerouting blood from the inferior vena cava to the TV in combination with a bidirectional cavopulmonary anastomosis (BCPA)) have been suggested by some authors [5–7]. We have used this approach in selected patients to simplify the repair when exposure was limited in positional anomalies, when the RV size or function was reduced either pre- or postoperatively, and/or to unload an insufficient and/or dysplastic TV. We have analysed the mid-term results of hemi-Mustard/BCPA as part of anatomical repair of CCTGA.

PATIENTS AND METHODS
Between 2004 and 2011, eight non-consecutive patients (two females and six males) received hemi-Mustard/BCPA as part of the anatomical repair of CCTGA. The median age was 2.9 years (range: 1.2–9.1 years) and the median weight was 14 kg (range: 10–24 kg). Positional heart anomalies were present in six patients (five had dextro- or mesocardia in combination with situs solitus, whereas one patient had levo cardia associated with situs inversus). Two patients with absent LVOTO underwent an ASO procedure, while the remaining six patients had either pulmonary stenosis (three patients) or pulmonary atresia (three patients) and underwent the Rastelli procedure. Both patients from the ASO group had insufficient Ebstein-like TV with a smaller RV cavity, and they underwent pulmonary artery banding (PAB) initially. The PA bands were left in place for 7 and 12 months, respectively. Four patients from the Rastelli group received a systemic to pulmonary shunt (two patients had two consecutive shunts) followed later by BCPA in one of them. All patients had a VSD, i.e. multiple (one patient), conoventricular (six patients) and conal (one patient) type. One patient had bilateral superior vena cava. Two patients (both from the Rastelli group) had an abnormal coronary artery anatomy, i.e. single anterior origin (one patient) and single posterior origin (one patient). One patient with an ASO had a mitral valve cleft. One patient from the Rastelli group had poor RV and LV function and severe tricuspid regurgitation preoperatively. All patients were in normal sinus rhythm the day before complete repair. Table 1 lists the patients’ preoperative data. All patients underwent preoperative heart catheterization to assess their suitability for BCPA.

Surgical technique
We aim to have a minimum body weight of 10 kg when performing anatomical correction of CCTGA. At the time of complete repair all the procedures were done using cardiopulmonary bypass (CPB) under moderate hypothermia. Previous shunts, bands and ductus were closed or taken down. Upon clamping the aorta and administration of cold crystalloid cardioplegia, a hemi-Mustard procedure was performed initially; the atrial septum was completely excised, the coronary sinus was unroofed and a PTFE baffle was constructed to divert blood flow from an inferior vena cava and a coronary sinus to a TV and a RV.

In the Rastelli group (Fig. 1), a PTFE intraventricular baffle connecting the LV with the aorta was fashioned via a right ventriculotomy. The VSD was not enlarged in any of the patients. The main pulmonary artery was detached from the LV and the proximal stump including the pulmonary valve was closed. A 16–20-mm bovine jugular vein conduit (Contegra, Medtronic, Inc., USA) was used to connect the RV with the main or branch pulmonary artery based on the underlying anatomy (the conduit was placed to the left of the aorta in all the Rastelli patients).

In the ASO group (Fig. 2), VSDs were closed with a xenopercardial patch via mitral or aortic valves. In one patient, an additional mid-muscular VSD was closed primarily and in the same patient tricuspid and mitral valvuloplasty was also performed. TV repair consisted of the closure and invagination of the anterosuperior commissure, while mitral valve plasty involved cleft closure and chordal shortening. Then, a conventional ASO was performed.

When deemed necessary (in seven out of eight patients), the right atriotomy was enlarged with a xenopercardial patch to avoid pulmonary venous obstruction. A BCPA (bilateral in one patient) including ligation of the azygos vein was performed at the end of the operation on the beating heart. Upon weaning from CPB, all the patients underwent TEE during modified ultrafiltration. The median CPB time was 245 (224–321) min, and the median aortic cross-clamp time was 157 (132–211) min. The chest was closed primarily in all but two patients.

Follow-up
The follow-up is complete for all patients. The mean follow-up was 4.5 (0.2–6.8) years. Clinical status was evaluated according to NYHA classification. Cardiac anatomy and the function of valves and ventricles were assessed by means of echocardiography and classified semi-quantitatively.

RESULTS
Early results
There was one in-hospital death. This patient from the ASO group developed progressive LV dysfunction and endocarditis, and died eventually from Aspergillus sepsis 77 days postoperatively. Two patients (one patient with an ASO, one patient with Rastelli) developed a complete heart block and received a permanent pacemaker. Two patients (Rastelli group) had a transient postoperative complete heart block with full recovery. Two patients (Rastelli group) were treated medically for supraventricular tachycardia. One patient (Rastelli group) with poor preoperative RV function and severe tricuspid regurgitation developed RV failure and renal failure requiring temporary peritoneal dialysis. Four patients had pneumonia (three patients with Rastelli and one patient with an ASO). Three Rastelli patients developed pleural effusion. The superior vena cava syndrome was notably not observed in any of the patients. The median length of ventilation support was 6 (1–54) days. The median length of ICU stay was 10 (2–84) days.

Late results
One patient from the Rastelli group needed a conduit change six years postoperatively. The same patient underwent radiofrequency ablation for medication-resistant atrial flutter.
At the last follow-up, six patients (five patients with Rastelli, one patient with an ASO) showed good biventricular function. One Rastelli patient with preoperative RV and LV failure remained with moderate LV dysfunction and was treated with ACE-inhibitors and diuretics. Systemic or pulmonary venous obstruction, sinus node dysfunction, atrial baffle leak, residual VSD or aortic regurgitation was not observed in any of the patients. Six patients were in NYHA class I and one patient (Rastelli group) in NYHA class II. Table 2 lists the postoperative patient data.

**DISCUSSION**

The long-term outcome of patients with CCTGA who underwent the conventional repair is unfavourable, and anatomical
correction is currently considered the preferred strategy [1, 4, 8, 9]. Despite its complexity, anatomical correction can be performed with low morbidity and mortality [2–4, 8, 10, 11].

Some authors have reported hemi-Mustard/BCPA as part of anatomical repair of CCTGA [5–7]. This 1.5-ventricle repair unloads an RV that can be small-sized either preoperatively (e.g. in patients with an Ebstein-like TV) or postoperatively (particularly after the Rastelli procedure where the intraventricular baffle may occupy a significant part of the RV) and dysfunctional either preoperatively or postoperatively (particularly after the Rastelli procedure due to ventriculotomy, intraventricular baffle and post-bypass diastolic dysfunction). Small RV size has been reported as a cause of mortality in Rastelli patients, while RV dysfunction related to a right ventriculotomy has been implicated as cause of long-term pleural effusion [12, 13]. Hemi-Mustard/BCPA reduces the volume load of a commonly dysplastic and insufficient TV and may contribute to the long-term preservation of its function upon anatomical repair [14]. Hemi-Mustard/BCPA facilitates the atrial switch part of the operation, thus reducing cross-clamp and bypass times that are known risk factors for death in these patients [4]. Moreover, it shortens an atrial baffle suture line, thus possibly reducing the risk of atrial baffle leaks (which is particularly true for positional heart anomalies with limited atrial exposure) and late atrial arrhythmias being the most frequent late morbidity [3, 15]. Finally, hemi-Mustard/BCPA eliminates complications related to the upper limit of the classic atrial switch procedure, such as sinus node dysfunction and systemic venous obstruction [16, 17]. Six of our 8 patients had atrio-apical discordance, and we strongly feel that hemi-Mustard/BCPA greatly facilitates the atrial part of anatomical repair.

Recently, Malhotra et al. published favourable mid-term results of using hemi-Mustard/BCPA as part of anatomical repair of CCTGA during a 15-year period. The authors implied that the above benefits might have reflected into excellent survival (>98%) and functional status (>90% of the patients in NYHA class I), and improved conduit longevity in Rastelli patients. BCPA-related complications were transient and rare, and no complications related to the modified atrial baffle were seen. The authors also identified subjects with elevated pulmonary vascular resistance and infants <4 months as those not being eligible for this type of repair [14].

Gaies et al. reported a higher rate (33%) of neurological injury in Rastelli patients. This might have been caused by impaired cerebral perfusion due to high central venous pressure and low systemic pressure as a result of RV dysfunction. In patients with inadequate RV or TV, therefore, the authors suggested the creation of an atrial baffle fenestration or addition of BCPA to prevent neurological complications [4].

On the other hand, Barron et al. discouraged the use of this approach in all CCTGA patients because of limited functional capacity and the lack of transvenous pacing options and access for re intervention when compared with the classic atrial switch procedure [18].

At our institution, we have used hemi-Mustard/BCPA as part of anatomical repair of CCTGA in selected patients only. These patients either had small RV (six patients from the Rastelli group) or an insufficient, Ebstein-like TV (two patients with ASO). In addition, most of our patients (75%) had positional heart anomalies. All were older than one year and did not have elevated pulmonary vascular resistance.

Reddy et al. suggested that possible long-term complications of BCPA (progressive cyanosis due to decreased upper body flow and pulmonary artery changes, e.g. arteriovenous fistulae, aortopulmonary collaterals, systemic venous collaterals and poor pulmonary artery growth) will be neutralized by antegrade flow to the main pulmonary artery [6]. This was confirmed in all our patients. Moreover, the combination of BCPA and the antegrade pulmonary blood flow has not led to the superior vena cava syndrome in any of them.

The pulmonary venous atrium was augmented with a xenopericardial patch in seven out of eight patients to prevent pulmonary venous obstruction.

In spite of using a technique to avoid the conduction system during VSD closure and atrial baffle construction, two out of eight patients had developed complete heart block and received a permanent pacemaker. This is consistent with a previous series suggesting an unpredictable course of the conduction system in CCTGA patients without preoperative electrophysiological mapping [19].

There is an increased risk of LV deterioration in patients requiring LV training by means of PAB [20]. Two patients from the ASO group received PAB to control congestive heart failure (caused by a large VSD) and not for LV training as proposed by

### Table 2: Postoperative patient data

<table>
<thead>
<tr>
<th>Patient</th>
<th>ICU (d)</th>
<th>VS (d)</th>
<th>Complications</th>
<th>Redo</th>
<th>Reintervention</th>
<th>Death</th>
<th>Last FU</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>84</td>
<td>54</td>
<td>RVF, CHB, PD</td>
<td>PM/CC</td>
<td>RFA</td>
<td>No</td>
<td>Moderate LV dysfunction, good RV function, NYHA II</td>
</tr>
<tr>
<td>2</td>
<td>4</td>
<td>2</td>
<td>PE</td>
<td></td>
<td></td>
<td>No</td>
<td>Good biventricular function, NYHA I</td>
</tr>
<tr>
<td>3</td>
<td>77</td>
<td>77</td>
<td>LVF, CHB, airway infection, chylothorax, endocarditis, sepsis</td>
<td></td>
<td></td>
<td>Yes</td>
<td>Poor LV function, good RV function, death</td>
</tr>
<tr>
<td>4</td>
<td>2</td>
<td>1</td>
<td>Transient CHB, pneumonia</td>
<td>PM/</td>
<td>LVTE</td>
<td>No</td>
<td>Good biventricular function, NYHA I</td>
</tr>
<tr>
<td>5</td>
<td>8</td>
<td>3</td>
<td>Transient CHB, pneumonia</td>
<td></td>
<td></td>
<td>No</td>
<td>Good biventricular function, NYHA I</td>
</tr>
<tr>
<td>6</td>
<td>6</td>
<td>4</td>
<td>Pneumonia, PE</td>
<td></td>
<td></td>
<td>No</td>
<td>Good biventricular function, NYHA I</td>
</tr>
<tr>
<td>7</td>
<td>11</td>
<td>8</td>
<td>PE</td>
<td></td>
<td></td>
<td>No</td>
<td>Good biventricular function, NYHA I</td>
</tr>
<tr>
<td>8</td>
<td>16</td>
<td>10</td>
<td>Pneumonia</td>
<td></td>
<td></td>
<td>No</td>
<td>Good biventricular function, NYHA I</td>
</tr>
</tbody>
</table>

ICU: length of ICU stay; d: days; VS: length of ventilation support; FU: follow-up data; RVF: right ventricular failure; CHB: complete heart block; PD: peritoneal dialysis; PM: pacemaker implantation; CC: conduit change; RFA: radiofrequency ablation; LV: left ventricle; RV: right ventricle; NYHA: New York Heart Association Classification; PE: pleural effusion; LVF: left ventricular failure; LVTE: left ventricle thrombectomy.
some authors [21]. Postoperatively, LV function was preserved in all our patients. In one patient with poor preoperative LV function, ventricular dysfunction improved later.

Pressure and volume unloading of an RV has led to improved RV function and better TV performance in our patient cohort.

Our clinical study has several limitations. The number of included patients is small and the data were analysed retrospectively. Postoperative echocardiographic assessment is semiquantitative only.

In conclusion, mid-term results of hemi-Mustard/BCPA as part of the anatomical repair of CCTGA in selected patients are favourable. Further follow-up is needed to prove the long-term benefits and to disclose potential drawbacks of this strategy.

Conflict of interest: none declared

REFERENCES


APPENDIX. CONFERENCE DISCUSSION

Dr M. Kostolny (London, UK): The concept described in this presentation is a modified atrial switch procedure, or a hemi-Mustard procedure, with claimed benefits of unloading the right ventricle, avoiding superior vena cava complications, and simplifying procedures in centres with limited exposure to the Senning operation. This is an extension of the concept of the so-called partial biventricular repair introduced by Drs Hanley and Van Andsell. In my opinion, the Senning still represents the gold standard, as it relies on defined anatomical landmarks and with each layer of the Senning procedure having an impact on the subsequent layer. Its superiority compared with the Mustard procedure has been clearly demonstrated in large, long-term studies.

My first question relates to the fact that this is a series of non-consecutive patients, and I would like to ask how many other patients there were and what the selection criteria were for using this particular modification. My second question is about the potential drawbacks of the Glenn procedure. Perhaps you could expand on this.

Dr Sojak: In response to your first question, unfortunately we don’t have any clear selection criteria. This is a non-consecutive series of patients with congenitally corrected transposition of the great arteries, so it means that four of the patients we had in the group received the conventional double-switch atrial switch and Rastelli procedure. Those patients had normal right ventricles and tricuspid valves. But in our series, based on preoperative echo and catheterization data, we decided to do a modified atrial switch procedure because of a small right ventricle. In the other two patients who received the double-switch procedure, they clearly had a small right ventricle and a small and dysfunctional tricuspid valve. This is the reason why we did a modification of the atrial procedure.

In response to your second question, the previous authors have described risk groups in terms of receiving modified atrial switch procedures. These were patients who were younger, small infants, and patients with high pulmonary vascular resistance. Actually, in our group, all of the patients were older than one year, so this was not a problem, and they did not have increased pulmonary vascular resistance, as they had been catheterized and had effective pulmonary stenosis or bands in place, so we didn’t notice any problems in this sense.

Dr O. Raisky (Paris, France): As you probably saw this morning, in Paris, for single ventricle, we love cavopulmonary anastomosis and antegrade blood flow. Sometimes we have a huge systolic wave in the cavopulmonary anastomosis. We deal with this by doing moderate right PA banding. Have you observed this in your series?

Dr Sojak: We are aware of these results, but we have never used this technique. Actually, the postoperative echo series showed normal flow in the bi-directional Glenn even though there was antegrade flow from the pulmonary arteries.