Results of the double switch operation for congenitally corrected transposition of the great arteries

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Abstract

Background: Congenitally corrected TGA (CC-TGA) is characterized by discordant atrioventricular and ventriculo arterial connections. In absence of right ventricular outflow tract obstruction (RVOTO), repair by atrial and arterial switches remains a challenging procedure for which long term follow-up is uncertain. Methods: From 1995 to 2007, 20 patients (median age: 26 months) with CC-TGA had double switch procedure. Segmental anatomy was {SLL} in all patients, dextrocardia in two patients, mesocardia in two patients. Ventricular septal defect was present in 17 patients, aortic coarctation in 2 patients and interrupted aortic arch (IAoA) in 1 patient. Five patients had tricuspid valve regurgitation. Six patients had AV blocks, 4 patients had pacemaker implantation prior to repair. Pulmonary artery banding was performed in 17 patients, for congestive heart failure (14 patients) or left ventricular retraining (3 patients). Three patients, including one patient with IAoA had primary repair. After LV retraining, repair was performed when indexed LV mass to LV volume ratio was above 1.5. A median follow-up of 60 months was achieved in all. Results: There were no deaths. Postoperative pacemaker implantation was required in four patients. Reoperation for Senning obstruction was necessary in one patient, and pacemaker battery replacement in another patient. One patient had mild neoaortic insufficiency, two had mild tricuspid regurgitation and two had mild mitral regurgitation. All were in NYHA I—II. Actuarial survival at 10 years was 100% and freedom from reoperation at 5 and 10 years were 93% and 77.4%, respectively. Conclusion: Double switch for CC-TGA without RVOTO can be performed with no mortality and low morbidity. Since these results seem to last for several years, it should be considered as the optimal procedure.

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1. Introduction

Congenitally corrected transposition of the great arteries (CC-TGA) is associated with discordant connection between atria and ventricles and between the ventricles and the great arteries. CC-TGA most often occurs with situs solitus of the atria and abdominal viscera with {SLL} segmental anatomy, but also occurs in situs invs with {IDD} segmental anatomy. It accounts for less than 1% of congenital heart diseases [1]. Patients with AV discordance usually have an associated cardiac abnormality such as a ventricular septal defect (VSD), morphologic left ventricle outflow tract obstruction (LVOTO) or atresia, and, various degrees of Ebstein’s-like malformation of the tricuspid valve (TV).

Malposition of the cardiac apex and conduction defects are common [2,3]. The variety of associated anomalies dictates the symptoms, considerably affects the prognosis and indicates the need and timing of surgical operation in the young population [4,5]. Several multicentric studies have shown the decreased longevity in the natural history of patients with CC-TGA [6,7]. Surgical approaches for CC-TGA included physiologic and anatomic repair. In patients who underwent a physiologic procedure, the morphologic right ventricle (mRV) remains supporting the systemic circulation with disappointing late results [8–10]. The anatomic repair or double switch (DS) is a challenging procedure: it allows the LV connection to the aorta by an arterial switch and the atrioventricular concordance established by an atrial switch procedure. The success of DS procedure depends on the ability of the left ventricle to support the systemic circulation. That can be obtained after retraining by means of pulmonary artery banding (PAB) [11]. The aim of the study was to evaluate both early and late outcome after DS procedure.
Excluded from this series are cases of CC-TGA with obstruction to pulmonary blood flow.

2. Materials and methods

Between 1995 and 2007, 20 patients who presented with CC-TGA and without RVOTO underwent DS procedure. There were 15 males and 5 females. The median age was 26 months (ranges 6—151 months) and the median weight was 11 kg (ranges 5 to 42 kg). All patients had preoperative two-dimensional echocardiography Doppler studies, and 18 had cardiac catheterization. All patients had situs solitus {SLL} segmental anatomy, 16 patients presented with levocardia, 2 with dextrocardia, and 2 with mesocardia. The most frequent associated anomaly was VSD, present in 17 patients (85%).

Five patients had tricuspid valve incompetence. Two of them presented an Ebstein-like anomaly. Associated cardiac lesions and surgical data are listed in Table 1.

Before procedure, six patients developed spontaneous complete heart block; among them four had pacemaker implantation prior to DS procedure.

Seventeen patients (85%) underwent PAB to control congestive heart failure and to reduce excessive pulmonary blood flow in 14 patients and to retrain of the left ventricle in the other 3 patients. For the 3 patients (1, 4 and 6 years old) who had an indication for LV retraining, a second procedure was required to tighten the banding. The mean time elapsing from first banding to final repair was 14 months with a range of 11—43 months. Three patients, including one with interrupted aortic arch (IAoA) had primary repair. Two patients with initial aortic coarctation underwent surgical palliation with PAB.

The management of CC-TGA in children is presented by an algorithm (Fig. 1).

2.1. Surgical technique

All PAB procedures were performed through median sternotomy except in two patients; where the procedure was performed through left thoracotomy for the convenient aortic coarctation repair.

The double switch approach was made through a median sternotomy incision. The repair of the VSD was tranatrial in nine, through the aorta in six and through the right ventricle in the other 3 patients.

Closure of the defect through the right atrium and mitral valve generally presents an excellent exposure. Tricuspid valve regurgitation was not addressed.

A Senning procedure was performed in all cases. Two patients with initial aortic coarctation underwent surgical palliation with PAB.

The management of CC-TGA in children is presented by an algorithm (Fig. 1).

![Fig. 1. Algorithm for the management of CC-TGA in children.](https://academic.oup.com/ejcts/article-abstract/35/5/879/465151/28-March-2022)
maneuver was used in all cases. Four patients had delayed sternal closure.

2.2. Statistical analysis

The data were imported into Stat View and presented as median with range. The actuarial method was used including confidence intervals.

3. Results

3.1. Early results

There was no hospital death.

One patient required early reoperation for pulmonary artery bifurcation stenosis and residual VSD at ten post-operative days. Another patient had reoperation for pulmonary venous pathway thrombosis on the seventh day.

Among the 20 patients, 6 patients presented preoperatively with complete heart block (CHB). Four had pacemaker implantation prior to DS procedure and two patients recovered full sinus rhythm after DS.

Four other patients developed permanent AV block following repair and required pacemaker implantation (two during the operation, one at 10th day post-op and one at 18th day post-op).

Operative and postoperative data are listed in Table 2.

3.2. Survivors

Follow-up information was complete for all patients. The median interval between the surgical repair and the last follow-up was 60 months, (range from 1 to 12 years).

Routine follow-up was conducted by referral pediatric cardiologists and consisted of assessment of clinical status, electrocardiography (ECG), Holter monitor and echo Doppler studies.

3.3. Late results

Late reoperation was performed in two patients:

One patient underwent repair to repair pulmonary venous pathway stenosis nine months later. Another patient needed a pacemaker revision 84 months after DS procedure.

At the time of the last follow-up, 17 patients were in NYHA functional class I, and 3 patients were in class II.

Actuarial survival at 10 years was 100% (95% confidence intervals, 0—17%) and freedom from reoperation at 5 and 10 years were 93% (95% confidence intervals, 0.79 —1) and 77.4% (95% confidence intervals, 0.47—1), respectively.

Two patients had mild tricuspid valve regurgitation. One patient developed mild neoaortic insufficiency and two patients mild de novo left atrioventricular valve regurgitation.

At echocardiographic studies all patients had normal biventricular function.

One patient developed a severe systemic ventricular dysfunction 9 months after operation. The coronary angiography was normal, echocardiography showed no valvular

<table>
<thead>
<tr>
<th>Patient</th>
<th>Time of follow-up</th>
<th>Functional class</th>
<th>Echocardiographic lesions</th>
<th>Reoperation (delay in months)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>144</td>
<td>I</td>
<td>None</td>
<td>None</td>
</tr>
<tr>
<td>2</td>
<td>144</td>
<td>I</td>
<td>None</td>
<td>None</td>
</tr>
<tr>
<td>3</td>
<td>148</td>
<td>I</td>
<td>Mild MR</td>
<td>None</td>
</tr>
<tr>
<td>4</td>
<td>132</td>
<td>I</td>
<td>None</td>
<td>None</td>
</tr>
<tr>
<td>5</td>
<td>98</td>
<td>II</td>
<td>Mild TR</td>
<td>None</td>
</tr>
<tr>
<td>6</td>
<td>96</td>
<td>II</td>
<td>Mild neoaao R</td>
<td>Pacemaker revision (84)</td>
</tr>
<tr>
<td>7</td>
<td>68</td>
<td>I</td>
<td>Mild MR</td>
<td>None</td>
</tr>
<tr>
<td>8</td>
<td>65</td>
<td>I</td>
<td>None</td>
<td>None</td>
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<td>9</td>
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</tr>
<tr>
<td>11</td>
<td>44</td>
<td>I</td>
<td>Mild TR</td>
<td>None</td>
</tr>
<tr>
<td>12</td>
<td>39</td>
<td>I</td>
<td>None</td>
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<td>I</td>
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<tr>
<td>19</td>
<td>10</td>
<td>I</td>
<td>None</td>
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</tr>
</tbody>
</table>

MR: mitral regurgitation; TR: tricuspid regurgitation, neoAo R: neoaortic regurgitation.
dysfunction. Outcome after 6 months was favorable under medical treatment. Ejection fraction of the left ventricle was increased from 34% to 60%.

Postoperative patients profiles are listed in Table 3.

4. Discussion

Isolated CC-TGA without significant associated cardiac anomalies is an uncommon entity. Only 1—2% of CC-TGA has no coexisting anomalies [13].

The long-term outcome of patients with CC-TGA after classic surgical approach was unsatisfactory [8,9,10]. Despite its complexity anatomical repair or double switch procedures represents today a good alternative in the management of CC-TGA surgery. Some advantages are particularly notable in terms of improving right ventricle (RV) and tricuspid valve function combined with low morbidity and mortality, and the fact that the left ventricle (LV) becomes the systemic ventricle [14].

For KARL et al., initial risk of double switch repair is similar to that of classic repairs, and long-term outlook is likely to be better [5]. On the other hand, Shin’oka et al., in a retrospective study in 2007 on 189 patients had concluded no differences observed between long-term survival rates of patients who underwent conventional and anatomical repair [15].

The age of our population was young, as in most series, with better results [3,16—19]. Some of them underwent double switch as a primary procedure [5,20,21]. In presence of associated hemodynamically significant VSD, primary repair can be discussed early in life. It avoids potential deterioration of the pulmonary valve after PAB. However, in this series, patients who presented with cardiac failure during the neonatal period and even later were more likely to undergo a 2-stage management with initial PAB. This was obviously due to the limited experience with the double switch as well as Senning procedure early in life.

In this series, three patients underwent morphologic LV retraining. PAB was performed at 1, 29 and 35 months of age and the repair, consecutively 12, 43 and 17 months later. Two of them required procedure for tightening of the banding in order to improve LV retraining. The evaluation of the LV was based on the echocardiography LV mass/volume ratio and the septal geometry as well as the LV pressure cardiac catheterization. In our practice, the criteria for LV retraining were based on our experience with rapid 2-stage management of simple TGA and have evolved with time. Earlier, a right to left septal geometry and a myocardial mass index: 35 G/m² was considered the inferior limit to indicate retraining [22]. Since 2000, the indication was based on the indexed LV mass/volume ratio. The LV myocardial mass was calculated according to the formula issued by the American society of Echocardiography. The measurements of the LV diameter were performed in long axis view. The LV volume was calculated through frozen end diastolic image of the LV in apical two- and four-chamber view indexed to the body surface area (BSA). After LV retraining, repair was performed when indexed LV mass to LV volume ratio was above 1.5. That has allowed us to perform primary ASO in simple TGA until 6 months of age with good outcome.

Although repair was uneventful, the older patient who underwent retraining developed severe LV dysfunction 9 months postoperatively which progressively recovered under medical treatment.

During the study period, a 4-year-old boy with severe tricuspid valve regurgitation underwent two retraining procedures with PAB until he reached suprasystemic LV pressure. Although tricuspid valve regurgitation diminished significantly in time, neither LV mass increase nor septal geometry became favorable to plan a double switch procedure (Fig. 2)

Bove et al. indicate that older patients are less likely to undergo satisfactory left ventricular retraining [14]. However, the upper age limit for LV training is not yet defined. Brawn et al. are not in favor of retraining patients over 15 years of age [23]. Quinn et al. showed that these patients were more likely to have deterioration of LV function at an earlier stage [3]. Retraining appears to be justified in presence of significant tricuspid valve regurgitation and should be performed as soon as possible, if necessary revision of the PAB is also mandatory. For asymptomatic CC-TGA, the justification for the procedures with the aim to reach anatomical repair needs to be well established.

Although PAB was identified as risk factors for the development of pulmonary root dilatation and subsequent neoaoctic valve regurgitation [17,23], the large experience with ASO allowed us to deal with this condition: in early and long term evaluation, any of the study patients presented more than mild aortic valve regurgitation.

Complete heart block is frequent in patients with CC-TGA. The estimated incidence of complete heart block is 2% of patients per patient-year after diagnosis, and 40% of the

Fig. 2. Echocardiographic assessment of congenitally corrected transposition of the great arteries with severe TR. (A) A 4-year-old, before surgical management with severe TR. (B) A 7-year-old, after two PAB procedure LV remains unprepared, however, TR became moderate and the patient asymptomatic.
patients develop complete heart block during a 20-year period [24]. Several studies showed that the course of the conduction system is abnormal in patients with CC-TGA [25]. Indication for the implantation of a permanent pacemaker should not be restrictive because the incidence of sudden cardiac deaths in these patients is higher. Loss of AV synchrony synchronisation can contribute to LV dysfunction because of ineffective contraction. Then, cardiac resynchronisation therapy can be necessary for the treatment of cardiac failure [17]. Recently, all patients were carrying 3 electrodes for sequential three chambers stimulation.

The Senning procedure carries a risk of atrial arrhythmias late after surgery [14]. This was associated with systemic right ventricle and appeared more likely in adulthood. Longer follow-up is necessary in order to determine whether the patients are going to develop atrial arrhythmias later on after the double switch procedure or not.

The prognosis in patients with CC-TGA is related to the integrity and function of their tricuspid valve [4,18]. Five patients presented with severe tricuspid valve regurgitation (TVR). Significant regression of the tricuspid valve integrity and function of their tricuspid valve [4,18]. Five switch procedure or not.

This improvement was related to the modification of the septal geometry [18]. Tricuspid valve dysplasia associated with severe regurgitation constitutes a principal indication for PAB in patients with CC-TGA early in life.

5. Conclusion

This study revealed that the double switch repair of CC-TGA can be performed with a significant improvement in the incidence of the in-hospital mortality.

Follow-up and an assessment of the functional status of these patients were satisfactory, particularly for whom presented with associated VSD.

References


Appendix A. Conference discussion

Dr P. Vouhe (Paris, France): I would like to begin this discussion by asking you several questions.

First, regarding the patients with double discordance and VSD, I have two questions. The first one is what are your criteria of choice between primary repair and pulmonary banding? And then, the second question, in patients in whom you do a pulmonary banding, what is the ideal age for a complete repair?
Dr Sarris: Well, to answer your question, I think my first approach to this patient depends on the size of the patient, on the size of the heart. An arterial switch operation is not an issue as well at closure of the VSD. My concern is about the atrial switch operation, whether this is a Senning operation or a Mustard operation in case of dextrocardia. I think I will do a primary repair without any pulmonary artery banding if I consider that I have enough room to perform a safe atrial switch, which means by 5 kg, I think we have done patients right away without palliative procedure.

Now, those patients who underwent pulmonary artery banding prior to the double switch operation, I think if we do the pulmonary artery banding in the early infancy, at 1 year of age we can proceed for the double switch operation. Dr Vouhe: Now, moving to the patients without VSD, with double discordance and intact septum. In your decision tree, I see that you advocate banding when there is tricuspid regurgitation or complete heart block, and that you advocate only medical surveillance when there is nothing else. As you probably know, in our unit in Neckar, we perform a banding in all patients even if they have no regurgitation, in the hope to delay tricuspid regurgitation and maybe to train the left ventricle if it becomes necessary one day to do a double switch. What is your opinion about this kind of prophylactic banding?

Dr Sarris: I'm waiting for your results.

Dr Vouhe: Very good. You will see these results in a few years. And my last question, it is a little bit out of the scope of this presentation. It is dealing with patients with double discordance, VSD and left ventricular outflow tract obstruction. Do you also advocate some kind of anatomic repair, Rastelli or Senning, or do you leave some place for conventional repair?

Dr Serras: No. Our approach in these patients with left ventricular outflow tract obstruction is to achieve an anatomic repair, and either by combined Senning and Rastelli procedures or as we have recently read the paper of Brizard on the possibility of doing a Nikaidoh operation in this subset of patients. If we cannot proceed for anatomic repair, we prefer to go for a Fontan-type repair.

Dr A. Conns (Liverpool, United Kingdom): Our experience in Liverpool is still limited by the fact, as you probably know, that Liverpool is only one hour drive from Birmingham, so almost all the patients are referred to Dr Bill Brawn, which is much better for the cardiologist, for the patients and for us. Nevertheless, we have four patients in our protocol, only because we use for the pulmonary artery banding the device which is the FloWatch allowing for an adjustable progressive banding with telemetric control. And I've seen that the pulmonary artery banding is in the central position of your algorithm.

My comment is the following: We use the FloWatch to obtain a progressive adaptation of the ventricle, because we are allowed to have a progressive tightening of the band over weeks with echocardiography monitoring. Then, the device can stay in place for a long time, up to 2-3 years, allowing the required release of the banding if the patient is outgrowing the band. This allows us to maintain all the options open for both univentricular type of repair or biventricular type of repair. As a matter of fact, among the four patients followed in these last 3 years, only 1 went to repair and it was one-and-one-half ventricle type of repair, as the option you mentioned before.

My question is the following: If you could have the device allowing a progressive adaptation and also the possibility to stay in place for years, would you change your protocol?

As a final comment, a disclosure: I don't get a penny for a single device FloWatch implanted.

Dr Serras: We have come to use in the lab the FloWatch apparatus, and one of my colleagues was also considering to use this material in our unit. However, there are two drawbacks with this material. First, it seems to be too large for our small patients and takes too much place in the chest, according to me. Second, we are very happy with our protocol, so why change?

Dr G. Sarris (Athens, Greece): I have one question that pertains to the age of the patients. Your median age is nice and low. The question is, what would your approach be for a patient who is referred late, perhaps in their late teens or even in adulthood, symptomatic, in a situation of intact ventricular septum.

Dr Sarris: I think that you are talking a patient with intact ventricular septum.

Dr Sarris: Yes.

Dr Serras: of course, because if there is a VSD, this patient is probably Eisenmenger and it is too late to intervene.

Dr Sarris: Yes.