Clinical results of arterial switch operation for double-outlet right ventricle with subpulmonary VSD

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Abstract

Objective: An arterial switch operation is considered a good alternative for the repair of double-outlet right ventricle (DORV) with atrioventricular concordance connection and subpulmonary ventricular septal defect (VSD) when intraventricular rerouting is not feasible. The clinical results of an arterial switch operation with ventricular septal defect closure for this anomaly were studied.

Methods: Between 1986 and 1997, 27 patients ranging from 10 days to 5 years of age (mean 0.4 years) underwent an arterial switch operation with ventricular septal defect closure for the correction of double outlet right ventricle with subpulmonary VSD. The 50% rule was used to define double-outlet right ventricle. Arch anomalies were associated in nine cases, and were corrected either previously or simultaneously. A subarterial muscle resection was performed in 14 without any subsequent stenosis of the ventricular outflow tract. The relationship of the great arteries was mostly anteroposterior in 15 and mostly side by side in 12. The left coronary artery (main trunk or circumflex artery) courses behind the pulmonary artery in 15/27 (six/15 in the anteroposterior relation and ten/12 in the side by side relation). The Lecompte maneuver was used to reconstruct the pulmonary artery in all but five cases with a side by side relationship of the great arteries.

Results: There was one operative death (3.7%) and three late deaths. The actuarial survival rate was 83–88% at 9 years. Right ventricular outflow tract obstruction including peripheral pulmonary stenosis developed in seven cases operated on in the early era. The reoperation free rate was 46–56% at 9 years.

Conclusion: Although double-outlet right ventricle with subpulmonary VSD has complex features, including an aortic arch obstruction and coronary artery anomalies, an optimal definitive surgical repair using an arterial switch operation can be performed safely with a thorough understanding of this variable anomaly. The prevention of right ventricular outflow tract obstruction at the time of an arterial switch operation may thus help improve the rate of late morbidity.

Keywords: Arterial switch operation; Double-outlet right ventricle; Taussig-Bing anomaly; Subpulmonary ventricular septal defect; Lecompte maneuver

1. Introduction

Although various surgical options have been employed to manage double outlet right ventricle (DORV) with subpulmonary ventricular septal defect (VSD) [1–11], which is also called the Taussig-Bing anomaly, anatomic repair is now considered to be the choice of treatment for this type of anomaly [4,6–12]. An anatomic repair means that the morphologically left ventricle is connected to the aorta while the morphologically right ventricle is connected to the pulmonary artery, without the use of any tubular prosthetic materials. This type of correction is achieved by either an intraventricular repair [1,2,4,5,7,8,10] or an arterial switch operation [4,7,8,10]. The intraventricular repair by tunneling left ventricular flow to the aorta through VSD is thought to have various advantages over an arterial switch operation, including no need for a reimplantation of the coronary artery and a reduced risk for late development of aortic incompetence. However, the indications of the intraventricular repair for DORV with subpulmonary VSD are...
quite limited, according to the internal cardiac morphology [7–12].

The arterial switch operation with ventricular septal defect closure seems to be feasible in a more substantial number of patients than the intraventricular repair [10], especially in the cases with anteroposterior relationship of the great arteries [4,7,8,10]. In this article, we evaluated the clinical results of the arterial switch operation for DORV with subpulmonary VSD, including the late results with a follow-up as long as 9.9 years.

2. Patients and methods

The definition of the DORV is followed by the 50% rule, namely one great artery arises completely from the right ventricle and the second more than 50% from the right ventricle, irrespective of their supporting structure [13]. Between 1986 and 1997, 38 patients underwent a total correction of the DORV with subpulmonary VSD. Among them, 27 patients from 10 days to 5.4 years of age (mean 0.4 years) underwent an arterial switch operation with ventricular septal defect closure (Table 1). Of these patients, 22 patients (81%) were aged 3 months or less. Their body weight ranged from 2.5 to 16 kg (mean 4.3 kg), and they consisted of 17 boys and ten girls. Except for the early era, our strategy for the repair of DORV with subpulmonary VSD is an anatomic repair, namely an intraventricular repair or an arterial switch procedure. An intraventricular repair is proposed when the great arteries relation is side by side, the tricuspid-pulmonary valve distance is not less than the aortic valve diameter, and there is no abnormal insertion of tricuspid valve chordae to the outlet septum. Otherwise, the arterial switch procedure is recommended.

The associated anomalies are listed in Table 1. Among the 27 patients, nine patients (33%) had arch anomalies, a coarctation of the aorta in five patients and an interrupted aortic arch in four patients. The relationship of the great arteries was more or less anteroposterior in 15 patients and side by side in 12.

Eight patients had undergone previous palliative procedures (Table 2). All patients with a coarctation of the aorta underwent a two-stage repair, while all patients except for one with an interrupted aortic arch had a one-stage repair.

The coronary anatomy according to Shafer’s classification [14] is depicted in Table 3. In the cases with anteroposterior relation of the great arteries, Shafer type 1 (the most common pattern in complete transposition; the left coronary artery from the sinus 1 and the right coronary artery from the sinus 2) was dominant and found in nine patients. In the remainder, the left main trunk (Shafer type 7a in two patients) or left circumflex artery (Shafer type 2a in four patients) courses behind the pulmonary artery (40%). While the left main trunk or left circumflex artery courses behind the pulmonary artery in 80% of the cases with side by side relation of the great arteries (Shafer type 2a in two, type 4 in two, type 7a in two and type 9 in four patients). The remaining two patients had a Shaher type 3a coronary artery (single coronary artery from the sinus 1).

Subaortic stenosis was diagnosed in three patients preoperatively. Abnormal conal insertion of the tricuspid valve chordae was found in five patients. Peripheral pulmonary artery stenosis which requires the plasty was found in one patient, and restrictive VSD was recognized in two patients.

2.1. Operative management

All patients underwent an operation through a mid-sternotomy using cardiopulmonary bypass with moderate hypothermia, except for the patients with an interrupted aortic arch in which deep hypothermia with the circulatory arrest of the lower body was employed. Myocardial protection was done with cold hyperkalemic crystalloid cardioplegic solution containing 5% albumin.

The VSD was closed through a right ventriculotomy or right atriotomy to introduce the left ventricular blood to the pulmonary artery (Table 2). Two patients had multiple ventricular septal defects, one of which was found postopera-
In another patient, three ventricular septal defects were closed with one patch. A resection of the outlet septum with/without ventriculo-infundibular fold was performed in the most recent 14 patients. A patch enlargement of the right ventricular outflow tract to relieve the stenosis was additionally performed in two patients. The enlargement of the ventricular septal defect was required in two patients. Among five patients with an abnormal conal insertion of the tricuspid valve chordae, one patient required a reattachment of the chordae. In the remainder four patients, the ventricular septal defect had been able to close without a reattachment of the chordae.

After closure of the VSD, an arterial switch was performed including a translocation of the coronary arteries. The Lecompte maneuver was used for the reconstruction of the pulmonary artery in all but five cases with side by side relation of the great arteries, in which the original Jatene procedure was employed to prevent pulmonary stenosis. In the subset with side by side relation of the great arteries, intraoperative conversion from the Lecompte maneuver to the original Jatene procedure due to pulmonary stenosis was done in two patients, while an intraoperative conversion of the original Jatene procedure to the Lecompte maneuver due to coronary artery trouble was performed in one patient. A reconstruction of the pulmonary artery was done using the xenopericardium in the early era, while an autologous pericardium or direct anastomosis (modification of the Pacifico’s method [15]) was used in the most recent cases.

A repair of an interrupted aortic arch, including a re-do procedure was done in four patients simultaneously. One patient also required plasty of the peripheral pulmonary artery simultaneously.

The mean follow-up of 3.3 ± 2.8 years (up to 9.9 years) was achieved in all but two survivors. The actuarial survival and freedom from reoperation rates were calculated by the Kaplan–Meier method. The statistical analysis was carried out using Stat View (Abacus Concepts, Berkeley, CA) on a Macintosh computer.

### 3. Results

#### 3.1. Operative results

Myocardial ischemia due to coronary artery kinking developed in three cases. The cause of coronary artery kink-
ing was a technical error in one patient with anteroposterior relation of the great arteries and coronary distribution with Shaher type 1, and the coronary artery kinking was resolved by a further dissection of the coronary artery. In another patient with side by side relation of the great arteries and coronary distribution with Shaher type 9, the original Jatene procedure was initially performed. The left main trunk of the coronary artery running behind the pulmonary artery was compressed between the neo-aorta and neo-pulmonary artery. This problem was resolved by an intraoperative conversion of arterial switch from the original Jatene procedure to the Lecompte maneuver. The last patient had side by side relation of the great arteries and coronary distribution with Shaher type 4. In this patient, an intraoperative conversion from the Lecompte maneuver to the original Jatene procedure was done due to the difficulty of the reconstruction of the pulmonary artery with the Lecompte maneuver. Poor perfusion of the left circumflex artery running between the neo-aorta and neo-pulmonary artery was recognized after the completion of the arterial switch procedure. A further dissection of the left circumflex artery appeared to improve the coronary perfusion, however, the patient died on the day of operation due to a low cardiac output. Operative death only occurred in this patient (3.7%).

3.2. Late results

A reoperation was necessary in seven survivors and no operative mortality occurred. Relief of a right ventricular outflow tract obstruction was necessary in two patients who had been operated on in the early era, and had not undergone a resection of the outlet septum at the time of the arterial switch operation. One of them underwent a mitral valve replacement and plasty of the ascending aorta simultaneously. Anastomotic stenosis of the pulmonary artery with/without peripheral pulmonary stenosis developed in five patients who underwent an arterial switch procedure with the Lecompte maneuver (three in the anteroposterior relation and two in the side by side relation of the great arteries) in the early era. One of them underwent a concomitant operation to relieve supra-aortic stenosis. Another patient required an implantation of a permanent pacemaker. A re-coarctation of the aorta was recognized in another patient, and was successfully repaired by balloon angioplasty. The freedom rate from reoperation at 9 years was 46 ± 20% (Fig. 1).

Significant aortic incompetence developed in three cases. One patient with moderate aortic incompetence developed pneumonia 6 months after the arterial switch operation and died suddenly. In the remaining two patients, the grade of aortic incompetence remained moderate at 6 months and 4.5 years after the arterial switch operation, respectively. The later patient had also undergone pulmonary artery banding before the arterial switch operation.

Residual VSD was recognized in one patient with multiple VSD, which had not been diagnosed before the arterial switch operation. As the residual VSD is thought to be insignificant, the patient did not require a reoperation.

There were three late deaths. The causes of late death were myocardial infarction due to occlusion of the left coronary artery 6 months after the operation in one patient, acute heart failure for some unknown reason 5 months after the operation in one, and sudden death in one already described. The actuarial survival rate at 9 years was 83 ± 8%.

4. Discussion

The DORV with subpulmonary VSD has complex features including the intracardiac structures, the relationship of the great arteries, the coronary artery anatomy and the aortic arch obstruction.

The intracardiac structures and the relationship of the great arteries both play an important role in selecting the type of anatomic repair. We performed an intraventricular repair in one patient with anteroposterior great artery relation (Patrick–McGoon operation [1]). In this patient, a right outflow tract obstruction developed and a reoperation to relieve this obstruction was thus required. Recently, the left ventricular outflow tract obstruction with a peak pressure gradient 25 mmHg developed in this patient, who has thus been followed carefully. Thereafter, our indications for an intraventricular repair are only limited in cases with side by side relation of the great arteries similar to those of many other surgeons [4,8,10]. For the intraventricular repair with side by side relation of the great arteries (Kawashima operation [2]), the distance of the tricuspid-pulmonary valve should be big enough in order to avoid any subsequent left ventricular outflow tract obstruction. The optimal distance of the tricuspid-pulmonary valve for intraventricular repair seems to differ for each surgeon [4,7,8,10,12]. Our criteria for this distance is greater or equal to the aortic valve diameter, and five patients who had no abnormal insertion of the tricuspid chordae to the outlet septum underwent the Kawashima operation. As a result, our recent criteria for intraventricular repair is quite limited and this procedure is only indicated in cases with side by side relation of the great arteries, when the distance of the tricuspid-pulmonary valve is greater or equal to the aortic valve diameter, and with no abnormal insertion of the tricuspid chordae to the outlet septum. For all other patients, the arterial switch procedure with ventricular septal defect closure is recommended.

In the early era, we did not recognize the importance of a resection of the outlet septum in the arterial switch procedure, which thus resulted in a subsequent right ventricular outflow tract obstruction in two cases. Fortunately, we have not seen this problem after introducing a vigorous resection of the outlet septum with/without a combined resection of the ventriculo-infundibular fold.
In the arterial switch procedure, a decision must be made regarding whether or not to perform the Lecompte maneuver. We performed the Lecompte maneuver in all cases with anteroposterior relation of the great arteries and in seven out of 12 with side by side relation of the great arteries. Anastomotic and/or peripheral pulmonary stenosis had developed and had to be repaired in five cases among them. Postoperative pulmonary stenosis is now known to be the most common complication after an arterial switch operation for the transposition of the great arteries [16–18]. A refined analysis revealed the risk factors of postoperative pulmonary stenosis to include: the use of foreign materials to close the defect of the excised coronary button (xenopericardium), over-stretching of the left pulmonary artery by the neo-aorta after the Lecompte maneuver, and so on [17].

To manage this problem, we now dissect the pulmonary artery as much as possible to avoid kinking. The reconstruction of the pulmonary artery is done without any foreign materials if possible, or with the autologous pericardium if no direct anastomosis is possible [17]. Patch plasty of the left pulmonary artery with autologous pericardium would be adopted if necessary. Although many surgeons seem to prefer the Lecompte maneuver even in cases with side by side relation of the great arteries [10], reconstruction of the pulmonary artery without a translocation of the pulmonary artery (the original Jatene procedure) might be a useful alternative.

The translocation of the coronary arteries without any obstruction of coronary flow is mandatory for the arterial switch procedure. When the relationship of the great arteries is anteroposterior, the most common coronary pattern was Shaher type 1 (or the so-called transposition of the great arteries type coronary). When the relationship of the great arteries is side by side, various types of coronary patterns are observed, and the left coronary artery (main trunk or circumflex artery) courses behind the pulmonary artery quite frequently. These results were compatible to those of other reports [8,19–21]. Intraoperative coronary trouble was observed in three patients, one due to technical problems and the trouble was solved by a re-arrangement of coronary translocation. In the other two patients, the coronary trouble was caused by the compression of the coronary artery from the pulmonary artery placed behind the neo-aorta (the original Jatene procedure) in cases with side by side relation of the great arteries. One of these cases was managed by a translocation of the pulmonary artery in front of the neo-aorta (Lecompte maneuver), while the other patient died due to a low cardiac output. Although we consider the original Jatene procedure to be a useful alternative to the Lecompte maneuver in cases with side by side relation of the great arteries to prevent pulmonary stenosis, great care should be paid when the left coronary artery courses behind the pulmonary artery. Kurosawa et al. [22] reported difficulties in cases with side by side relation of the great arteries managed by the Lecompte maneuver. He speculated that, in such cases, the new aorta must be forced beneath the pulmonary artery and may thus compress the left coronary artery that courses behind the new aorta. Fortunately, we have not encountered any such complications in our series.

The future possible development of aortic incompetence is a disadvantage of the arterial switch operation to the intraventricular repair [10,17,19]. In our series, aortic incompetence remained less than moderate in most cases, and no aortic valve replacement was required. However, in our experience with 226 arterial switch operations, previous pulmonary artery banding is supposed to be a risk factor for aortic incompetence [17] and two patients also underwent aortic valve replacement. A further long-term follow-up is required before any definite conclusions can be made regarding this matter.

The association of arch anomalies is frequently seen in this anomaly [8,10,11,21]. A repair of the aortic arch obstruction in a one-stage or in two-stage procedure remains controversial, although the one-stage procedure has recently become increasingly popular [11,23]. Our strategy for the coarctation complex is a two-stage repair using subclavian flap aortoplasty with/without pulmonary artery banding, while our strategy for the interrupted aortic arch complex is a one-stage repair. According to these strategies, all patients with coarctation of the aorta in this series had a two-stage procedure, while all patients except for one with an interrupted aortic arch had a one-stage procedure. Based on our results in this series, we consider our strategy to be satisfactory, however, we are also presently considering the use of a one-stage repair in patients with coarctation complex.

In conclusion, the double-outlet right ventricle with subpulmonary VSD has complex features, including an aortic arch obstruction and coronary artery anomalies. An optimal definitive surgical repair using an arterial switch operation can be performed safely with a thorough understanding of this variable anomaly. The prevention of a right ventricular outflow tract obstruction during an arterial switch operation may improve the occurrence of late morbidity.

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References


When you have a neonate 1 week of age, what is your policy? You believe it’s better to use pericardium? Do you think it’s better to use the autologous pericardium to fill up the defect. Then you believe that staged operation for coarctation is better than one-stage completion of operation?

Dr. Moll: Two-stage repair for the coarctation of the aorta and one-stage repair for the interrupted aortic arch.

Dr. Moll: Then you believe that staged operation for coarctation is better than one-stage completion of operation?

Dr. Moll: What was the technique of connection of the pulmonary artery? Do you use a patch?

Dr. Moll: Yes, to pulmonary.

Dr. Moll: Two-stage repair for the interrupted aortic arch. Two-stage repair for the coarctation of the aorta and one-stage repair for the interrupted aortic arch.

Dr. Moll: Because usually in these cases the pulmonary artery is very big and you can do it easy.

Dr. Moll: As the pulmonary artery is big and you can do it directly, I am asking why do you do it? Do you think it’s better to use pericardium? Do you prefer to use the autologous pericardium.

Dr. Moll: No. We prefer to use the direct anastomosis, of course.

Dr. Moll: I agree with that, yes.

Dr. Moll: I obviously agree with your opinion in performing arterial switch operations for this type of anatomy. When you have a neonate 1 week of age, what is your policy? You believe there is still a role in pulmonary artery banding or would you do a primary full repair?

Dr. Moll: We have no experience with such a young age. Our youngest patient is 10 days old, but I think ...

Dr. Moll: We would perform the arterial switch operation.

Dr. Quaeghebeur: Well, 10 days old, fine.

Dr. Moll: We would perform the arterial switch operation.

Dr. Quaeghebeur: I agree with that, yes.