Growth of the lateral tunnel in patients who underwent a total cavopulmonary connection at less than 5 years of age

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Abstract

Objective: A lateral tunnel-total cavopulmonary connection (LT-TCPC) using a right atrial (RA) free wall is the first choice of treatment for patients with a small body weight in this institute. Whether the growth of the LT is appropriate or not according to the growth of the body remains controversial. In addition, the optimal initial diameter of an LT is unknown. The purpose of this study was to verify the growth of the LT.

Methods: Ninety-one patients of a total of 267 TCPC cases underwent an LT-TCPC at less than 5 years of age in this institute between March 1991 and June 2008. The data on 47 of the 91 patients, which were available to investigate the LT growth, were retrospectively reviewed. The mean age at LT-TCPC was 37 ± 11 months (16—57 months). The mean body weight at TCPC was 12.4 ± 2.4 kg (7.6—20.0 kg). The initial LT diameter was determined with Hegar’s sizer of the estimated half-pulmonary arterial (PA) diameter, which is a diameter that results in half of the dimension of the normal pulmonary valve. The measured maximum LT diameter (mm) divided by the estimated half-PA diameter (mm) was considered as the LT index. The size of the LT was evaluated using either echocardiography or angiography.

Results: The mean follow-up period was 7.4 ± 3.5 years (1.6—13.5 years). The LT index was initially 2.0 ± 0.7 (1.3—4.5), 2.0 ± 0.4 (1.3—3.2) at 1 year after operation, 2.1 ± 0.5 (1.5—3.2) at 5 years after operation, 1.9 ± 0.4 (1.5—2.8) at 10 years after operation and 2.1 ± 0.5 (1.6—2.5) at 13 years after operation, respectively.

Conclusions: LT growth suitable for the body growth can be expected. Although there was some variation in the initial LT diameter, the LT index tended to converge at 2.0 with growth.

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1. Introduction

Since Fontan and Baudet first described their procedure for the correction of tricuspid atresia in 1971, the principles of the Fontan procedure have been applied to all forms of functional univentricular heart defects [1]. After several modifications of this operation, the total cavopulmonary connection (TCPC), which was first reported by de Leval et al. [2], has become the standard method for the Fontan procedure because it provides better venous haemodynamics [3] and is less arrhythmogenic [4] than the other Fontan modifications. In the original TCPC method, a lateral tunnel (LT) was created using a halved intra-atrial polytetrafluoroethylene (PTFE) graft; however, some modifications have been implemented in this institute. Initially, an LT is created only using an autologous atrial wall as previously reported [5]. Second, a standardised method for determining the initial size of an LT is used to prevent postoperative abnormal dilatation of the LT, which will cause an intratunnel thrombus and will become arrhythmogenic. The autologous intra-atrial tunnel has a potential for growth, which is a very important feature, especially in patients with low body weight; this is because an extracardiac conduit TCPC (EC-TCPC) is not often suitable in such patients due to the limitation of the available extracardiac graft size. However, no evidence is available to determine the appropriate growth of an LT created with an autologous atrial wall that is obtained in the midterm to long-term follow-up. The purpose of this study was to describe the actual growth of the LT created with an autologous atrial wall. This study was conducted in patients less than 5 years of age because the Fontan procedure is usually completed when a patient is less than 5 years of age and also because the growth of the LT is especially important in this patient population.
2. Materials and methods

2.1. Patients

The medical and surgical records of patients with congenital heart disease who underwent TCPC in Okayama University Hospital between March 1991 and June 2008 were retrospectively reviewed. A total of 267 patients underwent the Fontan procedure in this study period, and a lateral tunnel TCPC (LT-TCPC) was used in 117 of the 267 patients. Ninety-one of the 117 patients were less than 5 years of age. The data of the LT growth in midterm and long-term period were available from 47 of the 91 patients, and these data were evaluated.

2.2. Surgical procedures

The LT-TCPC was performed using a conventional continuous-flow cardiopulmonary bypass and mild-to-moderate hypothermia. An LT was created with an autologous atrial wall using polypropylene running sutures. First, a longitudinal atriotomy was made, which was similar to the Senning procedure (Fig. 1(a)). This incision was carefully separated from the sulcus terminalis, and then the lateral flap of the anterior atrial wall was sutured down onto the atrial septum posterior to the atrial septal defect using Hegar’s dilator as a guide (Fig. 1(b)). The LT diameter was determined using Hegar’s dilator of the estimated half-pulmonary arterial diameter, which is a diameter that results in half of the dimension of the normal pulmonary valve [6]. The terminal crest was carefully separated from the suture line. The orifice of the coronary sinus was placed in the atrial side of the heart. A small PTFE patch with a fenestration of 2.7—4.0-mm diameter was placed in the LT if a fenestration was required (Fig. 1(c)). After creating the LT, the atrium was closed by suturing the medial flap of the anterior atrial wall and anterior portion of the LT (Fig. 1(d)).

2.3. Data collection and measurements

This study was approved by the Institutional Research Ethics Board at the Okayama University Hospital, and patient consent was waived. The data collected from the medical record review included patient demographics, cardiac diagnosis, clinical condition, surgical data and postoperative and follow-up clinical status, and contained catheter and echocardiographic data. A value, which was calculated from a measured maximum LT diameter (mm) divided by the estimated half-pulmonary arterial diameter (mm), was considered to be the LT index. The size of the LT was evaluated with echocardiography or angiography; then, the maximum value of the LT diameter was used for the evaluation. Continuous data were expressed with these mean values and the standard deviations or these median values and the range as appropriate.

3. Results

3.1. Patient characteristics

In all, there were 26 females and 21 males. The chief diagnoses of these patients were hypoplastic left heart syndrome in 11 patients, tricuspid atresia in nine patients, single left ventricle except for tricuspid atresia in six patients, mitral atresia in three patients, single right ventricle except for mitral atresia in four patients, Ebstein’s anomaly in two patients and others in 12 patients. One patient demonstrated an asplenia. The mean age of these patients was 37 ± 11 months (range, 16—57 months). The mean body weight was 12.4 ± 2.4 kg (range, 7.6—20.0 kg). A preoperative catheter examination was performed in all of these patients and revealed the mean SaO2 to be 82 ± 6% (range, 66—97%), while the average mean pulmonary arterial pressure was 11.2 ± 2.9 mmHg (range, 5.0—18.0 mmHg); the median pulmonary arterial index (Nakata index) was 248 (140—550), the mean pulmonary arterial resistance was 1.74 ± 0.58 Wood unit m² (range, 0.63—3.08 Wood unit m²) and the mean systemic ventricular ejection fraction was 64 ± 11% (range, 41—86%). The mean estimated half-pulmonary arterial diameter at the operation was 8.5 ± 1.0 mm (range, 6.0—11.0 mm). There was no preoperative atrial tachyarrhythmia in these patients. The median cardiopulmonary bypass time was 105 min (range, 53—223 min), and the median aortic cross-clamp time was...
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systemic ventricular pressure was 5.3 mmHg. The LT index was quite constant throughout the follow-up period despite of the body growth. The LT index tended to become smaller and a smaller LT index was apt to become a little bit larger or stay unchanged.

3.2. Postoperative outcomes

All patients survived the operation. The median follow-up time was 7.4 years (range, 1.6—13.5 years). There was one late death due to sepsis of unknown cause. No patient experienced EC-TCPC conversion. Early atrial tachyarrhythmia occurred in three patients (6.9%), and it resolved during the hospital stay in two of these three patients. Late atrial tachyarrhythmia was observed in three patients (6.9%). One of these three patients had asplenia. One patient with asplenia (2.3%) underwent pacemaker implantation due to sick sinus syndrome. Three patients (6.9%) experienced protein-losing enteropathy, and one patient (2.3%) experienced plastic bronchitis. Four patients (9.3%) experienced surgical re-intervention in the late postoperative period: thrombectomy in the LT and a plasty of the left pulmonary artery in one patient, plasties of the ascending aorta and the inferior vena cava and the pulmonary artery. One patient also had a thrombus in the LT.

3.3. Growth of the LT

Fig. 2 shows the growth of the patients’ body weight and changes in the LT index. The LT tunnel diameter was measured in 30 patients immediately after the operation, in 33 at 1 year after the operation, in 26 at 3 years after the operation, in 23 at 5 years after the operation, in 11 at 7 years after the operation, and in three at 13 years after the operation. The patients’ body weight was initially 12.4 ± 2.4 kg (range, 7.6—20.0 kg), 14.5 ± 2.5 kg (range, 9.9—18.5 kg) at 1 year after operation, 17.5 ± 3.0 kg (range, 11.6—23.0 kg) at 3 years after operation, 22.8 ± 7.2 kg (range, 13.8—50.0 kg) at 5 years after operation, 26.0 ± 6.7 kg (range, 15.6—37.6 kg) at 7 years after operation, 35.7 ± 13.8 kg (range, 24.0—68.0 kg) at 10 years after operation and 47.0 ± 7.6 kg (range, 39.0—54.0 kg) at 13 years after operation, respectively. The LT index was initially 2.0 ± 0.7 (range, 1.3—4.5), 2.0 ± 0.4 (range, 1.3—3.2) at 1 year after operation, 2.0 ± 0.5 (range, 1.2—3.4) at 3 years after operation, 2.1 ± 0.5 (range, 1.5—3.2) at 5 years after operation, 1.8 ± 0.4 (range, 1.4—3.1) at 7 years after operation, 1.9 ± 0.4 (range, 1.5—2.8) at 10 years after operation and 2.1 ± 0.5 (range, 1.6—2.5) at 13 years after operation, respectively. The actual LT diameter was initially 17.0 ± 4.5 mm (range, 10.0—30.0 mm), 18.0 ± 3.8 mm (range, 11.5—27.1 mm) at 1 year after operation, 19.9 ± 5.0 mm (range, 12.6—32.0 mm) at 3 years after operation, 22.8 ± 5.7 mm (range, 13.9—36.5 mm) at 5 years after operation, 21.5 ± 2.9 mm (range, 18.7—29.4 mm) at 7 years after operation, 24.5 ± 5.6 mm (range, 17.7—36.0 mm) at 10 years after operation and 29.8 ± 6.3 mm (range, 23.0—35.4 mm) at 13 years after operation, respectively.

In addition, the actual changes of the LT index in each patient are shown in Fig. 3. There was an obvious tendency for the LT index to converge to around 2.0. The initial LT index was more than 3.0 in three patients; however, these LT indexes tended to decrease. Therefore, no abnormal dilatation was observed even in these three patients. On the other hand, no restriction of the LT was observed even in patients with a relatively small initial LT index.

4. Discussion

The method applied for establishing an LT is different from the original method described by de Level et al. at some points. First, they used a large interior PTFE patch to create an LT; however, in the current series, the LT was created only...
with the autologous atrial wall as reported previously [5]. A very small fenestrated PTFE patch is applied if a fenestration was required (Fig. 1(c)). Second, a Hegar’s dilator of the normal estimated half-pulmonary arterial size is used as a guide to create an LT. There has been no standardised method for creating an LT so far. Therefore, it has depended on the surgeon’s experience and judgement. The current method allows even an inexperienced surgeon to create an LT with relatively constant diameter and with smooth streamline. Although 18 of the 33 patients, who underwent postoperative catheter examinations, demonstrated a slight degree of turbulence of the venous blood stream, which always occurred on the most inferior site of the LT probably because of the size discrepancy between the inferior vena cava and the LT, no pressure gradient was observed in 31 of the 33 patients and no tendency of expansion of the LT was found in any of the 33 patients.

This is the first study, which reported the actual postoperative midterm to long-term growth of LT, which is created with the autologous atrial wall. Interestingly, the average of the LT index did not change throughout the follow-up period. In fact, there are some variations in the initial LT index. The LT indices ranged from 1.3 to 4.5 immediately after the operation. However, a larger LT index tended to become smaller and a smaller LT index was apt to become a little bit larger or stay unchanged. As a result, the LT index showed an obvious tendency to converge to around 2.0. The appropriate growth of the LT can be expected in the midterm to long-term follow-up if the patient maintains a good Fontan circulation. An extracardiac LT with pedicled pericardium may become an alternative procedure of LT-TCPC. Some authors have reported that an LT created with pedicled pericardium can grow while also preserving the tubular morphology of the LT and maintaining good haemodynamics [7,8]. However, there has so far been no report regarding the long-term outcome of LT created with a pedicled pericardium and which describes its growth in detail.

The incidences of the early and late atrial tachyarrhythmias in this study were both 6.9% with mean follow-up period of 7.3 years. The incidence of early and late tachyarrhythmia was 9.9% and 8.8%, respectively, among all of the 91 patients who underwent LT-TCPC before 5 years of age at this institute (unpublished data). The incidence of tachyarrhythmias may be slightly underestimated because neither a Holter electrocardiogram (ECG) nor an exercise ECG is conducted as a routine follow-up of arrhythmia in this institute. However, these incidences of atrial tachyarrhythmias seem to be lower than those of other reports [9–12]. The relatively restricted initial LT diameter in this study may contribute to the low incidence of the postoperative atrial tachyarrhythmia. In addition, extreme care is applied with regard to the suture line while creating an LT. First, the use of sutures carefully avoided the terminal crest because anchoring the LT to the terminal crest promotes the development of atrial flutter [13]. Second, there were few suture lines near or around the sinus node. After a right atriotomy, the lateral edge of the anterior atrial flap was sutured onto the atrial septum to create an LT. The upper limit of the suture line was relatively far from the sinus node, which also may contribute to prevent postoperative atrial tachyarrhythmias [14,15]. Third, the median longitudinal atriotomy, such as the Senning procedure, was performed and was kept far away from the sulcus terminalis. These methods prevented injuries to the longitudinal atrial conduction [13,16]. These modifications may have contributed to the low incidence of the atrial tachyarrhythmias.

Only one patient had an early thrombo-embolic event and no late thrombo-embolic event was observed in this study. One patient experienced a right atrial thrombus 2 weeks after the LT-TCPC and underwent a right atrial thrombectomy. The incidence of thrombo-embolic event was 2.2%, among all 91 patients who underwent LT-TCPC before 5 years of age at this institute (unpublished data). Although the use of anticoagulation for patients who undergo a TCPC has been controversial, warfarin has been routinely administered for all of the patients who underwent a TCPC. However, if a patient has proven to maintain an excellent Fontan circulation based on a postoperative catheter examination, which is usually performed 1 year after operation, warfarin can be converted to aspirin or ceased if required.

There was a concern that an EC-TCPC may be thrombogenic because of the use of artificial graft in the venous circulation; however, no study has found that an EC-TCPC was more thrombophilic than an LT-TCPC in midterm to long-term follow-ups. The risk of thrombo-embolism is more closely related to the suboptimal haemodynamics or an underlying coagulopathy than the type of Fontan procedure itself [17,18]. In addition, Lardo et al. revealed that an EC-TCPC had superior venous haemodynamics in comparison to an LT-TCPC [19]. Therefore, although an LT-TCPC had been the first choice in this institute, an EC-TCPC is the first-line strategy at present. However, there are still some advantages in the use of an LT-TCPC. First, the TCPC can be completed even in the patients with small body size. If the patients have atrioventricular valve regurgitation, newly developing arterio-pulmonary collateral arteries or pulmonary arteriovenous shunt, then the TCPC should be completed when the children are as young as possible to prevent deterioration of these conditions. A PTFE graft with a diameter of at least 16 mm was used for the EC-TCPC. In addition, the body sizes of the patient’s parents should also be carefully considered when selecting the graft size because a 16- to 18-mm PTFE graft may be too small if the patient’s body size becomes bigger than an average Japanese person in the future. In addition, there is some risk of pulmonary venous obstruction when an EC-TCPC is performed for a small child. Second, an LT-TCPC will be stronger against infection than an EC-TCPC. Patients with a past history of mediastinitis or other risk factors for infection may thus be good candidates for an LT-TCPC.

An important limitation of this study is that this study was based on retrospective non-randomised research. Therefore, the data collection of the LT growth was not integrated and the collected data had many deficits. However, as shown in Fig. 3, there was an obvious tendency to growth of the LT and the methods used for constructing the LT were quite consistent during the study period. Therefore, the results of this study and subsequent conclusions, which were obtained, are thought to be reliable.

In conclusion, an LT growth suitable for the body growth can be expected. Although there were some variations in the initial LT diameter, the LT index tended to converge at around 2.0 with growth.
I'm not a mathematician, but it seems to me you make two major assumptions here. You used normal estimated PA diameter in your formula, and you also expected normal PA growth after Fontan completion. As we know, this may not necessarily be the case. Can you comment on the validity of your formula?

Dr Fuji: Theoretically, if a patient has pulmonary branches with normal half-pulmonary arterial diameter, the pulmonary arteries must be capable of enough cardiac output. So we think the use of the normal half-pulmonary arterial diameter is very reasonable because patient can be all right without trouble.

Dr Tsang: I have a surgical question. The longitudinal right atriotomy was similar to the Senning procedure. The lateral aspect of your atrial wall was sutured down onto the atrial septum posterior to the ASD using a Hegar.

In the case of one-stage TCPC, there was a lot of atrial tissue for the tunnel. In the case of staged TCPC, there might not be a lot of atrial tissue for your right atrial lateral tunnel, especially in the case of staged Norwood with complete atrial septectomy.

Can you tell us how your technique evolved to compensate the possible lack of atrial tissue for your right atrial lateral tunnel?

Dr Fuji: If we have some kind of difficulties for creating the lateral tunnel, we use the extracardiac TCPC.

Dr Tsang: Another surgical question. Your right atrial lateral tunnel got an awful lot of right atrial tissue exposed to high systemic venous pressure. Does that explain the high incidence of early and late arrhythmia in this group of patients?

Dr Fuji: If the patient has a high pulmonary arterial pressure, we think the extracardiac TCPC is better because, for maintaining the good lateral tunnel shape for long-term period, we need a good pulmonary condition and good ventricular function. If not so, the patient sometimes suffers from dilation of the lateral tunnel.

Dr Tsang: One final and quite simple question. In the current era, what is your first-line strategy in a small child who needs a completion of TCPC?

Dr Fuji: If the patient's body weight is 10 kg or more, generally, extracardiac TCPC is our first-line. However, recently the number of patients who need early Fontan completion has been increasing. Patient with hypoplastic left heart syndrome, ativoventricular valve regurgitation moderate or more, pulmonary arteriovenous fistula, or significant collateral right-to-left shunt, should undergo early Fontan completion before the condition deteriorates. We usually use lateral tunnel in patients less than 2 years of age.

But I said before, good pulmonary and cardiac condition is necessary.

Dr Sano: I'm going to add to the answers to your question

Number one is that we adopted the normal PA size from Kirklin's formula. According to Kirklin's formula, we estimate normal lateral tunnel size is half PA size. There are two major pathways to RA, SVC and IVC, so we think a half PA size may be enough to drain IVC blood.

Second, most of the lateral tunnel was used when we did not adopt the two-stage Fontan, so that the right atrium was quite good sized, and the free wall of the right atrium was quite good quality. If the free wall of the right atrium is small and quality is no good, we use extracardiac TCPC.

Third, most of the arrhythmias occurred in the patient with heterotaxy syndrome. Previously, we tried to do lateral tunnel in almost all patients. In these patients, some have developed arrhythmias, so we changed our policy since then. But other than heterotaxy patients, no arrhythmias occurred.

Dr C. Pizzaro (Wilmington, Delaware): I don't know if I missed this, but did you ever consider to include in your measurements the cross-section of the inferior venae cava orifice and then compare that to the opening where you created the upper anastomosis of the lateral tunnel to the central pulmonary arteries? In essence, to provide an unobstructed pathway, the tunnel needs to accommodate the IVC flow.

As we know, this might be a more reliable way to establish if the tunnel is adequate for flow.