Results of an extracardiac pericardial-flap lateral tunnel Fontan operation

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Abstract

Objective: Extracardiac pericardial-flap lateral tunnel Fontan operation has the theoretical advantage of growth potentiality of the extracardiac tunnels. The mid-term results of this technique and morphologic change of the lateral tunnel were studied. Methods: Clinical data were reviewed in 42 patients who underwent extracardiac pericardial-flap lateral tunnel Fontan operation between November 1993 and December 2004. The age was 2.8 ± 1.5 years and the body weight was 12.3 ± 3.2 kg. Extracardiac tunnel was constructed using the pedicled pericardium with the base undetached. By reviewing the follow-up cardiac angiograms (2.3 ± 1.4 years postoperatively), ratios of diameter and cross-sectional area of the lateral tunnel to those of inferior vena cava were obtained. Results: There were 4 surgical mortalities (10%). Postoperative morbidity included prolonged pleural effusion in 5 patients and heart block in 1 patient. Follow-up was possible in 37 patients and the follow up duration was 3.8 ± 2.2 years. There were two late deaths due to ventricular dysfunction and sudden death of unknown causes. Two patients required reoperation due to subaortic stenosis and stenosis between inferior vena cava and lateral tunnel. In one patient, bradyarhythmia was observed but there was no thromboembolic complication. Follow-up anteroposterior and lateral diameter ratio were 1.1 ± 0.5 and 1.2 ± 0.5. The cross-sectional area ratio was 2.6 ± 2.3. In 5 patients, fusiform dilatation of the lateral tunnel was observed, but in the remaining patients, the lateral tunnel preserved tubular morphology with good hemodynamics. Conclusions: Extracardiac pericardial-flap lateral tunnel Fontan operation is relatively simple and feasible even in patients with previous median sternotomies. The mid-term results were acceptable, and the lateral tunnel demonstrated a tendency to preserve its tubular shape. However in some patients, dilatation of the pericardial-flap tunnel was observed during follow up. Longer follow-up is required to determine the morphologic changes of the lateral tunnel and the value of this technique.

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Keywords: Congenital heart defect; Hemodynamics; Pericardium; Outcome assessment; Fontan operation

1. Introduction

Fontan-type operation has been performed for patients with a functional single ventricle. To improve the circulatory function of the Fontan operation, minimizing the energy loss of the systemic venous flow through the pulmonary artery is very crucial. For this reason, constructing a uniform tubular structure between the vena cavae and pulmonary artery is proven to be hydrodynamically more efficient than a right atrium to pulmonary artery connection type Fontan operation [1—3]. Various techniques were proposed. Puga et al. [4] developed the intraatrial lateral tunnel technique in which a prosthetic patch was used to make a tubular structure in the lateral wall of the atrium. With this technique, the systemic venous flow can maintain its laminar flow through the tunnel and better hemodynamics with low central venous pressure could be obtained [1,5]. However, prosthetic materials, a large incision on the atrial wall and cardiac arrest are required for the procedure. Hvass et al. [6] introduced a new method of constructing an extracardiac lateral tunnel using an autologous pericardial flap. Because baffle is constructed just with the autologous tissue, there is a theoretical advantage of growth of the lateral tunnel, a lesser tendency for infectious and thromboembolic complications [7]. However, no sufficient data are available for the long-term results of pericardial-flap extracardiac lateral tunnel Fontan operation and the morphological change of the lateral tunnel.
tunnel. To assess the mid-term clinical results and to define the growth and morphologic changes of the pericardial-flap lateral tunnel, we reviewed the clinical data of our patients.

2. Materials and methods

Between November 1993 and December 2004, 42 patients underwent extracardiac lateral tunnel Fontan operation using autologous pericardial flap at our institute. The clinical data of these patients were reviewed. The age at Fontan operation was 2.8 ± 1.5 (1.2–6.9) years and 20 patients were younger than 2 years. The mean body weight was 12.3 ± 3.2 (8–22) kg.

The intracardiac anomalies for Fontan operation were tricuspid atresia, pulmonary atresia with intact ventricular septum, and various types of functional single ventricles (Table 1). Heterotaxia syndrome was included in 13 patients, and 3 patients had systemic venous anomaly of interrupted inferior vena cava (IVC) with azygous continuation and separate hepatic venous drainage directly to the atrium.

Forty patients had undergone 64 palliative procedures before the Fontan operation (Table 2) including 36 bidirectional cavopulmonary shunt operations. Preoperative arterial oxygen saturation was (SaO2)83 ± 6%. Perioperative cardiac rhythm was normal sinus in 39 patients. Supraventricular tachyarrhythmia was combined in two patients, and intermittent ventricular tachycardia was observed in one patient. Atrioventricular valve regurgitation of more than a moderate degree was combined in 7 patients. Significant brachial pulmonary arterial stenosis was combined in 14 patients, and mean pulmonary arterial index was 220 ± 79.

2.1. Operative technique

Operations were performed by median sternotomy under mild hypothermic cardiopulmonary bypass. Cannulation of the ascending aorta was done in a standard fashion. The venous drainage cannulae were inserted directly to the superior vena cava (SVC) and IVC. Aortic cross-clamping and cardiac arrest was used if intracardiac procedures were required. Basically the surgical technique described by Gundry and associates [7] was used. The pericardium on the inferior vena cava or hepatic vein side was mobilized, with care taken not to damage the phrenic nerve. The pericardium was incised at its superior extent in a perpendicular direction toward the superior vena cava and its inferior extent toward the IVC. The postero-omeral wall intact (Fig. 1). The proximal margin of the incision was oversewn to the undivided posterior wall with a 5-0 polypropylene suture, closing the atrium. The inferior edge of the pericardial flap was sutured to the edge of the partially divided IVC, and this suture line was continued for a short distance onto the atrial free wall. Next, the inferior wall of the ipsilateral pulmonary artery was opened and the posterior lip of the opened pulmonary artery was sewn to the dome of the atrium. The superior margin of the pericardial flap was sewn to the anterior lip of the pulmonary artery incision. Then this suture line was continued along the anterior free margin of the pericardial flap toward the IVC, suturing the margin of the flap onto the atrial free wall so that the extracardiac lateral tunnel can be constructed with a hemicylindrical shape of pericardial flap with the atrial wall as its floor. In all cases the pericardium was adequate to permit a baffle construction, so no prosthetic material was used. For the patients who were high risks candidate for Fontan operation and with high postoperative central venous pressure, fenestration was created with a punch on the right atrial wall between the lateral tunnel and the lumen of the atrium. For the patients who had not undergone bidirectional cavopulmonary connection, the SVC was divided at its junction with right atrium and anastomosed to the incision on the superior aspect of pulmonary artery in the end-to-side manner.

Pulmonary arterial angioplasty was done in 14 patients. For the patients whose pulmonary arterial stenosis was localized to the branch ipsilateral to the pericardial flap, the pulmonary arterial enlargement was done at the time of constructing the baffle by extension of the pulmonary

<table>
<thead>
<tr>
<th>Table 1</th>
<th>Anatomic diagnoses</th>
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<tr>
<td>Tricuspid atresia</td>
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</tr>
<tr>
<td>Pulmonary atresia with intact ventricular septum</td>
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<td></td>
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<tr>
<td>Double outlet right ventricle</td>
<td>5</td>
<td></td>
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<tr>
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<td>Other complex functional single ventricle</td>
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Table 2

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<th>Procedure</th>
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<th>%</th>
</tr>
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<tbody>
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<td>86</td>
</tr>
<tr>
<td>Systemic-to-pulmonary artery shunt</td>
<td>22</td>
<td>52</td>
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<tr>
<td>Pulmonary artery reconstruction</td>
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<td>12</td>
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<tr>
<td>Extension of ventricular septal defect</td>
<td>3</td>
<td>7</td>
</tr>
<tr>
<td>Repair of atrioventricular valve</td>
<td>3</td>
<td>7</td>
</tr>
<tr>
<td>Pulmonary artery banding</td>
<td>2</td>
<td>5</td>
</tr>
<tr>
<td>Repair of coarctation of aorta</td>
<td>2</td>
<td>5</td>
</tr>
</tbody>
</table>

Fig. 1. Operative technique. (A) The pericardial flap was prepared and the anterior half of the inferior vena cava (IVC) was incised and its proximal margin was oversewn to the undivided posterior wall. (B) The pericardial flap was sutured on the edge of partially divided IVC and the suture line was continued on the right atrial wall to the incision on the pulmonary artery to complete the extracardiac lateral tunnel.
arterial incision across the stenotic portion and covering it with a pericardial flap of largely tailored anteriosuperior end.

For atrioventricular valve insufficiency, valve repair was done in 3 patients, and replacement with mechanical valve prosthesis was done in 1 patient. For a patient with tricuspid atresia with transposition of the great arteries, the bulboventricular foramen was extended to relieve the possible left ventricular outflow tract stenosis. Mean cardiopulmonary bypass time was 110 ± 46 min and aortic cross-clamp time was 63 ± 37 min.

2.2. Postoperative management and assessment of growth of the lateral tunnel

All patients were anticoagulated with warfarin to a target prothrombin time international normalized ratio of 2 to 3. Warfarin anticoagulation was maintained for six months and switched to antiplatelet therapy unless there was arrhythmia, ventricular dysfunction or poor Fontan hemodynamics. Echocardiograms were taken at least once a year and a catheterization was performed at one year after operation. The morphologic change of the lateral tunnel was evaluated by analyzing the angiograms and computer tomogram taken 6 months or later after operation. The lateral and anteroposterior diameters of the lateral tunnel were measured and compared with those of inferior vena cavae. The diameter of the inferior vena cava was measured just below the anastomosis with the extracardiac lateral tunnel. The diameter of the lateral tunnel was measured mid-portion between the anastomoses with pulmonary artery and IVC in principle. If the lateral tunnel showed dilatation or stenosis, the widest or smallest diameter was measured.

2.3. Statistical analysis

Continuous data are presented as the mean value ± standard deviation. The pressure measured at the SVC and IVC was compared using paired t-test. Difference was considered significant when the p value was 0.05 or less.

3. Results

3.1. Early results

There were four surgical mortalities (10%). Three patients died of Fontan circularity failure. They had fenestration but postoperatively showed low cardiac output with high central venous pressure and low arterial oxygen saturation. Take-down of Fontan circulation was performed in 2 patients but they did not recover. The other patient died of congestive heart failure probably due to perioperative myocardial infarction. Postoperative echocardiogram revealed severe hypokinesis ventricular anterior wall motion. In these mortality cases, intraoperative and postoperative echocardiogram revealed no evidence of stenosis of pulmonary and systemic venous drain. Thus in no case did the use of the pericardial flap technique appear to have influenced mortality.

The mean immediate postoperative central venous pressure of the survivors was 15 ± 3 mmHg, and the median intensive care unit stay was 4 days. Arrhythmia was found in 5 patients after operation. Temporary supraventricular tachycardia was observed in 1 patient. In 3 patients, bradycardia associated with sinus node dysfunction was observed, but all patients recovered normal sinus rhythm at discharge. Complete atrioventricular block was developed in a patient whose atrioventricular valve was replaced with mechanical prosthesis. The postoperative course of this patient was also complicated by ventricular dysfunction. The patient had right ventricular type single ventricle with common atrioventricular valve and had undergone atrioventricular valve repair at the time of bidirectional cavopulmonary connection, but the atrioventricular valve regurgitation aggravated to moderate to severe degree and the ventricle was moderately enlarged. Postoperatively, ventricular function deteriorated and the patient showed acute renal failure requiring temporary peritoneal dialysis and prolonged pleural effusion. A permanent pacemaker was implanted on postoperative day 18 and ventricular function was slightly improved. The patient was discharged on postoperative day 60 with symptoms of congestive heart failure.

Neurologic complication was found in a patient who had an episode of seizure postoperatively, but the patient was discharged from the hospital without any neurological sequelae. Five patients had prolonged pleural effusion more than 2 weeks. There was no operation-related infection or phrenic nerve palsy.

3.2. Follow-up results

Among the 38 survivors, 37 patients were followed-up except one and the follow-up duration was up to 11.7 years (3.8 ± 2.2 years). There were 2 late mortalities. The patient who showed ventricular dysfunction and complete heart block died of congestive heart failure at postoperative 6 months. About 3 months after discharge, he was readmitted and died of ventricular dysfunction and complications of congestive heart failure. The other late mortality case died of an unknown cause 2 years after Fontan operation. The patient underwent Fontan operation at the age of 6.5 years and had prolonged pleural effusion postoperatively. The patient maintained normal sinus rhythm and good functional class during the 2 years follow-up. Cause of death was not confirmed because information regarding the death could not be obtained from the caregiver.

Two patients required reoperations during the follow-up. A patient, who had undergone Fontan operation at 4.9 years old, underwent reoperation due to a stenosis at the anastomosis between the inferior vena cava, and the stenotic anastomosis site was enlarged with patch after 4.4 months after Fontan operation. The other one was a patient with tricuspid atresia with transposition of the great arteries who had undergone Fontan operation at 1.4 years old. The patient required reoperation to enlarge the bulboventricular foramen due to subaortic stenosis 2.1 years later. The operation was completed without any complications such as phrenic nerve injury or damage to the conduit.

Echocardiogram was taken for all patients and cardiac catheterization and angiogram was taken in 18 patients. On angiograms and echocardiograms, swirling of the blood flow
was not observed in the lateral tunnel and there was no evidence of pulmonary vein stenosis in any patients. No intracardiac or intra baffle thrombi was found with cardiac imaging studies, and no thromboembolic complication was reported. Newly developed arrhythmia was found in one patient who showed bradyarrhythmia. The functional class was NYHA I or II in 34 patients and NYHA III in two patients.

3.3. Hemodynamic evaluation and the growth of extracardiac lateral tunnel

Cardiac catheterization and angiograms were taken in 18 patients postoperatively 6 months or later. The interval between Fontan operation and follow-up angiograms was 2.3 ± 1.4 years. The pressure measured at the SVC was 15.1 ± 3.4 mmHg, and at the IVC was 16.1 ± 4.2 mmHg, and there was no statistical difference.

The IVC drainage flow was studied with the anteroposterior and lateral projection of angiogram. The angiograms revealed wide unrestricted anastomoses between the baffle and the IVC. There was no stenosis between the lateral tunnel and the pulmonary artery. The angiogram also revealed unobstructive venous flow to both pulmonary arteries. Follow-up angiogram of a patient who required reoperation due to anastomosis site stenosis, also showed unstenotic pathway.

We observed the morphologic feature of the lateral tunnel and the blood flow through the lateral tunnel on the anteroposterior and lateral projection of the angiogram. In most cases, the lateral tunnel had a uniform tubular shape (Fig. 2), and the blood flow of the IVC drained to the pulmonary without turbulence. The extracardiac lateral tunnel showed some fusiform dilatation in 5 patients (Fig. 3), but narrowing of the lateral tunnel on its course was not observed.

The anteroposterior diameter ratio of the lateral tunnel to IVC was 1.1 ± 0.5 and lateral diameter ratio was 1.2 ± 0.5 (Fig. 4). The cross-sectional area ratio of lateral tunnel and inferior vena cava was 2.6 ± 2.3. In 5 patients, the cross-sectional area of the lateral tunnel is marked enlarged (Fig. 5). But, there were no patients showing narrowing of the lateral tunnel.

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Fig. 2. Angiogram of the inferior vena cava and extracardiac lateral tunnel 5.3 years after pericardial-flap extracardiac lateral tunnel total cavopulmonary connection. (A) Anteroposterior view and (B) lateral view showing smooth-walled tubular structure leading from inferior vena cava to pulmonary artery.

Fig. 3. Angiogram 12 months after pericardial-flap extracardiac Fontan operation. (A) anteroposterior and (B) lateral view showing dilated extracardiac lateral tunnel.

Fig. 4. Distribution chart of the anteroposterior and lateral diameter ratio of extracardiac pericardial-flap tunnel to inferior vena cava.

Fig. 5. Distribution chart of the cross-sectional area ratio of extracardiac pericardial-flap lateral tunnel to inferior vena cava.
4. Discussion

Recently, extracardiac conduit total cavopulmonary connection is adopted by many surgeons for repair of functional single ventricle and polytetrafluoroethylene (PTFE) vascular graft is commonly used as a conduit [8—11]. By using a relatively simple technique of interposing a vascular graft between the IVC and the pulmonary artery, a very uniform tubular structure can be easily made and the hydrodynamical efficiency can be optimized. There is less risk of obstruction of pulmonary venous drainage or atrioventricular valve inflow and better long-term results are expected than other technical modification by reduction of atrium-related complications such as arrhythmia and thromboembolism, because no portion of the atrium is exposed to the high pressure Fontan circuit and the suture line on the atrium is minimized.

Considering the fact that the conduit is a delivering pathway of the IVC blood to the pulmonary artery, for maximal hydrodynamic efficacy, the conduit should maintain the diameter of the IVC. However if conduit is used, due to the lack of growth potential, relatively large sized conduit is used at the time of Fontan operation to avoid a reoperation. If size discrepancy between the conduit and IVC is high, turbulent flow develops at their junction, and it induces hydrodynamic energy loss, thus hydrodynamic benefit of the total cavopulmonary connection is reduced. The upper limit of critical conduit to IVC diameter ratio is approximately 1.5, without a substantially negative hydrodynamic consequence [12]. Generally the IVC diameter of the normal population ranges 20—24 mm, and it usually reaches 60—80% of adult value at the age of 2—4 years when the Fontan operation is usually preformed [13—15]. So, in many cases 18—20 mm or larger diameter conduit can be used [11,16,17]. If the Fontan candidate’s physique is small or Fontan operation is planned younger than the usual age, we should abide by the hydrodynamic disadvantage of size discrepancy of IVC and the large sized conduit or reoperation should be considered later instead of using adequate size of conduit for the patient’s physique.

On the contrary, by using a pericardial flap, the lateral tunnel can be designed to match the size of the IVC for each patient. Therefore, if the lateral tunnel constructed with the viable tissue has an adequate tendency to grow, pericardial-flap lateral tunnel technique has an advantage of providing the ideal conduit size at the time of Fontan operation, and avoiding the requirement for reoperation for conduit replacement. Actually, in some studies, the continued growth of extracardiac lateral tunnel was observed [7,18]. In our study, we studied the growth of the lateral tunnel by comparing its size measured in the angiogram with that of the IVC. Considering the fact that the lateral tunnel is a pathway of the IVC blood flow to the pulmonary artery, we think, it is more reasonable to compare the size of lateral tunnel with that of the IVC rather than comparing it with patient’s body surface area or height. In the review of the angiograms of our patient group, fusiform dilatation of the lateral tunnel was observed in some patients. However, in most patients, the lateral tunnel showed a tendency to preserve a relatively uniform tubular structure. In our cohort, one patient required reoperation due to anastomosis site stenosis between the lateral tunnel and the IVC. But, in this case, it can be assumed that the anastomosis site stenosis was induced by the technical problem at Fontan operation, not by the insufficient growth of the pericardial flap, because the stenosis was observed from the immediate postoperative period and reoperation was done just after 4.4 months later. This can be prevented with technical vigilance during the procedure. It is not clear that the enlargement of the lateral tunnel is caused by growth of the viable pericardium or dilatation of atrial wall which constitutes some of the posteroomedial wall of lateral tunnel, by high central venous pressure. In many patients, the cylindrical shape of the lateral tunnel was preserved and this finding suggests the growing of living tissue rather than passive dilatation. But, we also noted fusiform-shaped lateral tunnel during follow-up, so passive dilatation of the lateral tunnel cannot be excluded from the mechanism. Because the observation of the lateral tunnel morphology is cross-sectional, we do not know if the dilatation was due to the design of the conduit at the time of surgery or progressive dilatation. To define the fate of the lateral tunnel, serial follow-up for a longer period is required. This will be very important because if the pericardial flap lateral tunnel continues to dilate over time, later problems that can be seen in atrio-pulmonary connection cannot be avoided and it will show that this technique has no advantage over the extracardiac conduit technique.

The pericardial flap technique is a relatively simple and feasible procedure. To use the pericardial flap, adequate size of pericardium to construct the lateral tunnel should be obtained and the surface should be smooth to prevent thrombi formation. In our experience, autologous pericardium was good enough to be used even in patients who had previous sternotomies. No patient in this cohort required any prosthetic material to construct the lateral tunnel. And in no patient was initially planned pericardial flap technique given up and replaced with other technique due to the inadequate pericardial tissue. On the contrary, largely tailored pericardial flaps could be used for angioplasty if there was an ipsilateral pulmonary artery branchial stenosis. It is advised to leave sufficient pericardial tissue at the time of palliative procedure through median sternotomy for the candidates of Fontan operation to enhance the feasibility of pericardial flap technique. Our patients received anticoagulation for six months, then antplatelet therapy. No patient had thromboembolic complication, and no evidence of thrombus was found in the serial imaging studies. So, we think previous sternotomy is not a contraindication to this technique.

While tailoring the pericardium, incision should be made on the pericardium anteroposterior direction near the phrenic nerve. Care should be taken not to injure the phrenic nerve because diaphragm palsy is associated with increased morbidity after Fontan operation [19]. The phrenic nerve can be easily observed from the pleural aspect of the pericardium, so its injury can be prevented by limiting the extension of pericardial incision not too close to the phrenic nerve and minimizing usage of electrocautery near the phrenic nerve. An effort has to be made to place sutures on the pericardium in a careful fashion and to avoid any full-thickness sutures near the phrenic nerve. With these cautions, there was no postoperative phrenic nerve palsy,
either in a series of 54 patients of Kavarna and associates [20], or in our cohort.

Mid-term clinical outcome of pericardial-flap extracardiac lateral tunnel Fontan operation was acceptable. The functional status of the patients was NYHA class I or II in almost all patients except for two, and the echocardiogram and cardiac catheterization demonstrated good hemodynamics. Although there were 2 late mortalities that were rather early, in 1 patient this technique does not affect the outcome, but in the other patient, its relation is unclear.

One of the possible complications of this technique is pulmonary venous obstruction ipsilateral to the pericardial-flap tunnel. The pulmonary vein could be compressed by the relatively high-pressure lateral tunnel. None of our patients showed any evidence of pulmonary vein compression but this possible complication should be checked postoperatively and during follow-up, and this will be more important for patients with dilated lateral tunnel.

Another demerit of this technique is the suture lines placed on the atrium. It has been reported that the incidence of atrial flutter is associated with long suture line on the right atrium [21]. In our cohort, supraventricular arrhythmia was observed in just one patient during the follow-up. However, longer observation is required because mean follow-up duration of 3.8 years was too short to draw conclusions about late postoperative arrhythmias or other late-phase event.

In conclusion, extracardiac pericardial-flap lateral tunnel Fontan operation is a relatively simple and safe technique. It is feasible even in patients who had undergone previous median sternotomies. Mid-term results were acceptable and the lateral tunnel constructed with pericardial flap demonstrated a tendency to preserve its tubular shape. However, in some patients, dilatation of the pericardial-flap baffle was observed during follow up. Longer follow-up is required to determine the growth of the lateral tunnel and the value of this technique.

References


**Dr Park**: During the bidirectional Glenn shut, I preserve the right side of the pericardium more, and we use the Gore-Tex membrane to prevent severe adhesion. When I do the completion Fontan, I look at the pericardium. If the pericardium was thickened, I resect it and use the supplementary bovine pericardium. Another point is that because the lower part and the upper part of pericardium are different, even lateral length of the pericardium is important to construct the tunnel. In almost of all cases, I can make a lateral tunnel with their own pericardium.

**Dr Ziemer**: So there has not been any phrenic paralysis in your series?

**Dr Park**: No, I have not encountered any issues with phrenic nerve paralysis. However, in order to avoid such complication, I have advocated superficial bites and avoiding any deep sutures around this region.