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# The optimal Fontan connection: a growing extracardiac lateral tunnel with pedicled pericardium.

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# SURGERY FOR CONGENITAL HEART DISEASE

## THE OPTIMAL FONTAN CONNECTION: A GROWING EXTRACARDIAC LATERAL TUNNEL WITH PEDICLED PERICARDIUM

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*Objective:* **The concept of a lateral tunnel for the Fontan operation is now widely accepted. Most lateral tunnels are constructed intraatrially with the use of aortic crossclamping. Construction of extracardiac lateral tunnels with the use of homografts or other nonviable tubes eliminates aortic crossclamping but lacks growth potential in length or width. The native pericardium, which is "sealed" posteriorly along the pulmonary artery, atrium, and inferior vena cava, could be turned down onto the right atrium to form a viable extracardiac lateral tunnel.** *Methods:* **We designed and successfully constructed extracardiac lateral tunnels using viable autologous pericardium, pedicled on its lateral blood supply, in 19 patients aged 9 months to 5 years. All patients had a previous Glenn shunt; five patients had dextrocardia and a midline inferior vena cava. The patients' inferior vena cava–right atrial connection was opened transversely and the right atrial opening was sutured to its back wall, keeping the eustachian valve in the inferior vena cava. The underside of the right pulmonary artery was opened longitudinally; its inferior edge was sewn to the adjacent pericardial reflection. Any "pocket" or depressions in the posterior pericardium along the pulmonary veins were closed with running suture. Two incisions were made in the right pericardium down to the phrenic nerve parallel to the inferior vena caval and pulmonary arterial openings. This pedicled pericardium was trimmed and sewn as a roof to the upper edges of the inferior vena cava and pulmonary artery openings and then sewn longitudinally along the unopened right atrial wall, completing the viable extracardiac lateral tunnel. Although no fenestrations were used, these could be made during construction, or more significantly, owing to the lack of thick walled structures, in the catheterization laboratory in the postoperative period.** *Results:* **All 19 patients had respiratory/cardiac pulsations in the pulmonary arteries owing to the compressible lateral tunnel. At follow-up of up to 2½ years, all tunnels are growing and no obstructions have occurred.** *Conclusion:* **The viable autologous pericardial extracardiac lateral tunnel can be constructed without cardiac ischemia, can be fenestrated in the postoperative period, and forms a compressible, nonthrombogenic conduit capable of growth, which can be constructed early in infancy. (J Thorac Cardiovasc Surg 1997;114:552-9)**

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Since the original description of total venous diversion directly to the lungs by Fontan and Baudet[1] in 1971, numerous modifications have been proposed to Fontan's operation, many of which have been widely adapted and have proved successful in large series.[2-8] In the mid-1980s, de Leval and his colleagues[9] described the energy losses of cavopulmonary versus atriopulmonary connections and initiated the practice of constructing intraatrial lateral tunnels to connect the inferior vena cava (IVC) to the pulmonary artery (PA), coupled with a bidirectional Glenn shunt. Recently, Amodeo and associates[10] demonstrated the utility of constructing the lateral tunnel exterior to the heart, bridging the gap between the IVC and PA with Dacron, polytetrafluoroethylene,* or homograft tubes. In theory, this extracardiac lateral tunnel could be constructed without cardiac ischemia or cardiac incisions, thereby lessening the known morbid sequelae of Fontan's operation, namely low cardiac output and atrial arrhythmias. Unfortunately, extracardiac conduits have no potential for growth, so that their application in very young children may not be warranted.

The native pericardium, which is "sealed" posteriorly along the PA, atrium, and IVC, could be turned down onto the right atrium (RA) and connected to the PA above and the IVC below to form a viable extracardiac lateral tunnel, which we postulated would be a nonthrombogenic tube with growth potential. This report describes the technique of constructing this viable extracardiac lateral tunnel and the results of its application in 19 patients.

## Patients and methods

**Patient population.** Between December 1993 and June 1996, 19 patients between the ages of 9 months and 5 years underwent the modified Fontan operation with the use of a pedicled pericardial viable extracorporeal lateral tunnel at Loma Linda University Children's Hospital. All patients had previously undergone creation of bidirectional Glenn shunts, with three of these patients having had bilateral, bidirectional Glenn shunts for bilateral superior vena cavae (SVCs). Five patients had dextrocardia or mesocardia with a midline IVC. One patient had undergone a previous "standard" Fontan operation consisting of closure of the previous atrial septal defect and connection of the superior aspect of the RA to the PA. This child returned 6 months after the operation with uncontrolled atrial arrhythmias and severe congestive heart failure with low cardiac output and a "giant" RA.

* Gore-Tex conduit, registered trade name of W. L. Gore & Associates, Inc., Elkton, Md.

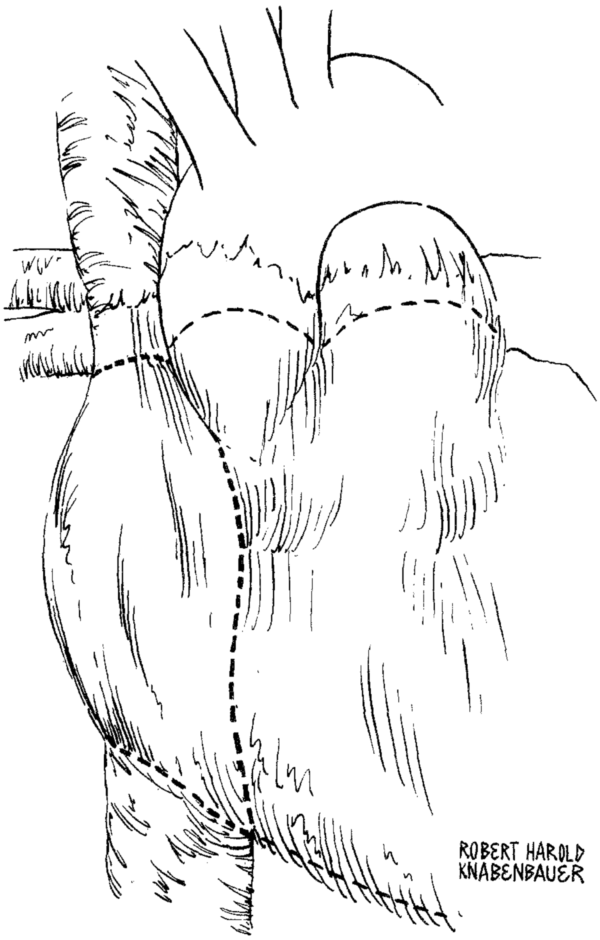


**Fig. 1.** Artist's rendering of proposed pericardial incisions in preparation for constructing pedicled pericardial lateral tunnel.

**Surgical technique.** After a second median sternotomy, the right-sided pericardial edge is identified and carefully dissected from surrounding structures (Fig. 1). Attention is paid to assure enough pericardium is present parallel to the PA above and the IVC-RA junction below. The pleura overlying the right lung is incised and the location of the phrenic nerve is identified. The viable pericardial flap is prepared by incising the pericardium perpendicularly down to the base of the right PA and to the diaphragmatic reflection of the pericardium inferiorly (Fig. 2). If the phrenic nerve overlies the proposed incision site(s), it is gently teased off the pericardium at that point, with the incision passing beneath it. The incisions are carried down to the pericardial-PA junction above and the pericardial-IVC junction below.

An aortic cannula is placed in the ascending aorta and a single venous cannula is placed in the RA appendage. Cardiopulmonary bypass is initiated and the patient cooled to 18° to 20° C. During cooling, the right PA is exposed, but care is taken not to dissect it circumferentially. Similarly, the atrium is further freed, down to its

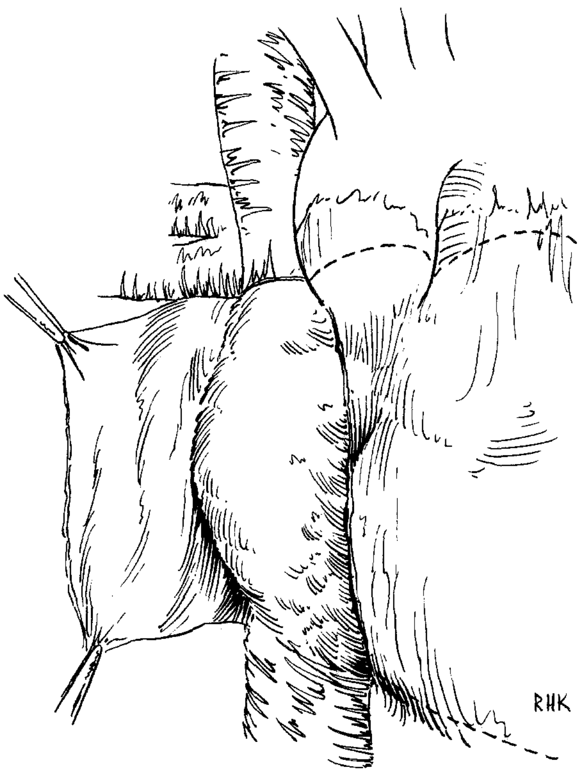


**Fig. 2.** Artist's rendering of perpendicular incisions of right-sided pericardial edge down to pericardial reflections on right PA superiorly and IVC inferiorly to create pedicled pericardial flap.

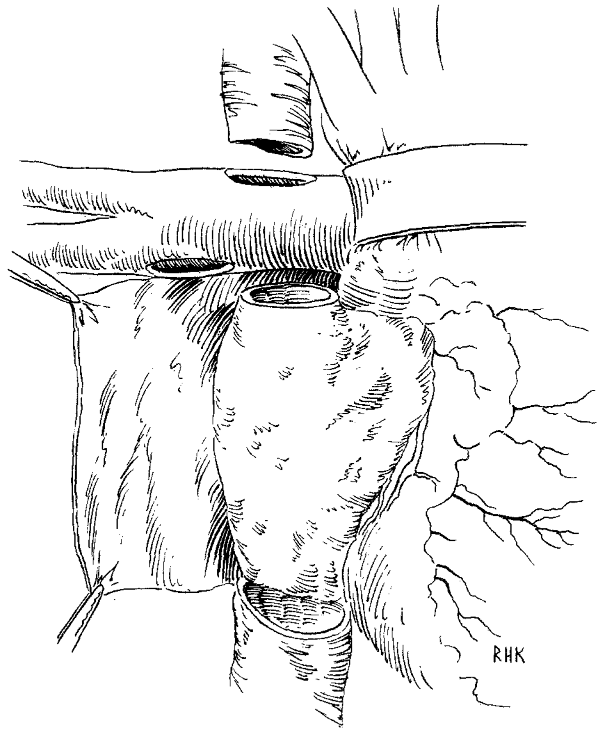


**Fig. 3.** Artist's rendering of incisions on anterior surface of IVC-RA junction and incisions above if Glenn shunt has not been previously performed. In current series, all patients had a prior Glenn shunt.

pericardial reflection posteriorly. Any severe "valleys" caused by the pulmonary veins are obliterated with fine running Prolene sutures (Ethicon, Inc., Somerville, N.J.). The anterior surface of the IVC-RA junction is also dissected, but the IVC is not encircled.

Low-flow bypass (25 to 50 ml/kg per minute) is instituted, the RA cannula is connected to a cardiotomy sucker, and the venous line is clamped. The IVC is opened transversely at its junction with the RA anteriorly, leaving the back wall in place (Fig. 3). A second cardiotomy sucker or a wire-weighted cardiotomy vent (DLP, Inc., Grand Rapids, Mich.) is placed into the IVC. The RA opening is sutured to its back wall with running 5-0 Prolene suture (Fig. 4). The pericardial flap is now measured for width and trimmed so that its leading edge reaches to the medial aspect of the opened IVC. Draping the pericardium over a dilator (the size of the IVC) placed temporarily into the IVC opening facilitates cutting the proper width of the pericardial flap. This flap is then sewn to the opened edge of the IVC, with care taken to ensure that the corner transition from pericardium to IVC is closed and carrying the suture line across the opened IVC, terminating at the still-connected IVC-RA junction medially.

Next, the underside of the exposed right PA is opened and the incision carried laterally to the pericardial reflection and medially until an opening slightly larger than the IVC is obtained. (This is usually located adjacent to the stump of the SVC-RA junction) (Fig. 4). A third cardiotomy sucker is placed in the PA. The posterior wall of the opened PA is then sewn to the posterior pericardial reflection, which at this location usually parallels the superior pulmonary vein beneath it. Then the previously incised and trimmed pericardial flap is sewn across the superior edge of the opened PA, completing the upper flap connection.

Finally, the free edge of the pericardial flap, pedicled on its lateral blood supply, is sewn to the free wall of the RA in a line parallel to the IVC connection below and PA connection above (Fig. 5). The RA catheter is reconnected to the pump tubing, regular bypass is begun, and the patient is rewarmed. During rewarming, gentle ventilation is started to aid venous return.

In cases in which the IVC was midline, the lateral tunnel was constructed to either side of the ventricle, four times coming under the systemic ventricle on the right and once coming left to the left PA. Deficient pericardium can be augmented with a piece of pericardium from the opposite side of the heart (one patient).

## Results

All patients were weaned from bypass without the use of inotropic drugs or with low-dose dopamine

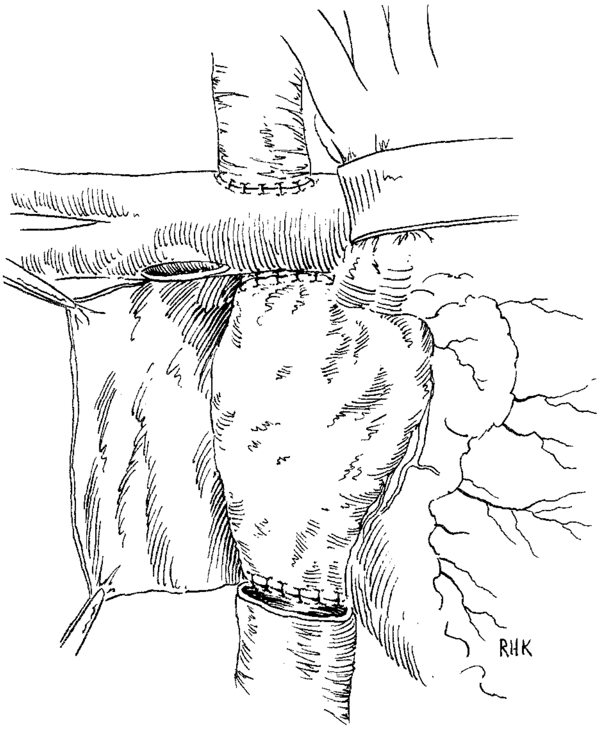


**Fig. 4.** Artist's rendering of RA sewn to its back wall, completion of Glenn shunt, and opening of underside of right PA.

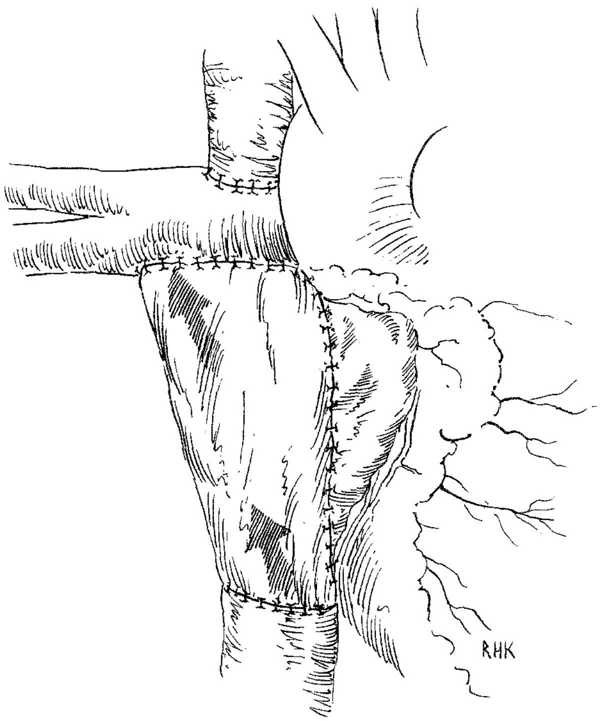


**Fig. 5.** Artist's rendering of pedicled pericardial flap brought across openings in IVC and PA and then sewn to edge of RA to complete extracardiac lateral tunnel.

and tolazoline (Priscoline), depending on surgeon preference. All but one patient was extubated in the perioperative period. No early deaths occurred. Prolonged chest tube drainage (greater than 7 days) was required in three patients (15%). Median hospital stay was 5 days (range 3 to 66 days). Two of these patients required subsequent pleurocentesis or reinsertion of a chest tube, but the drainage resolved. No patient had malabsorption syndrome or ascites. No injuries to the phrenic nerve occurred. No early or late anticoagulation has been used. Cardiac pulsations and respiratory variations were recorded in the PAs of patients after the operation (Fig. 6). Serial echocardiograms show continued growth of the tube in length and appropriate width (Figs. 7 and 8). The only catheterization, done 1½ years after the operation, showed a smooth-walled tube equal in size to the IVC (Fig. 9). No child has rhythm disturbances according to history or examination. No late Holter monitoring has been required. Three late deaths occurred, two from noncardiac causes. The child whose initial unsuccessful Fontan procedure was revised to a viable extracardiac lateral tunnel did well initially and was discharged uneventfully. The child was readmitted 2 weeks later to another service, was treated for sepsis and low cardiac output, and died. Postmortem examination revealed an unsuspected pericardial tamponade as the cause of death. A second 4-year-old child was declared brain dead 5 months after the operation after an arteriovenous malformation of the brain ruptured. The child's autopsy showed a well-formed tunnel with a smooth, endothelium-lined channel from the IVC to the PA (Fig. 8). The third child died 3 months after the operation of a severe respiratory syncytial viral infection that progressed to viral cardiomyopathy.

## Discussion

The introduction of the lateral tunnel concept by de Leval and coworkers[9] and the report of its successful application by Jonas and Castaneda[11] spawned a near universal acceptance of this technique to complete Fontan's operation. However, because this technique still requires ischemia and intraatrial manipulations, other alternatives have been sought, such as the construction of much of this connection during the previous Glenn shunt (the so-called hemi-Fontan)[12] or fenestration of the lateral tunnel at the time of its construction.[13] None

**Fig. 6.** Tracing from PA line showing respirations and cardiac pulsations in the PA after construction of a pericardial extracardiac lateral tunnel.

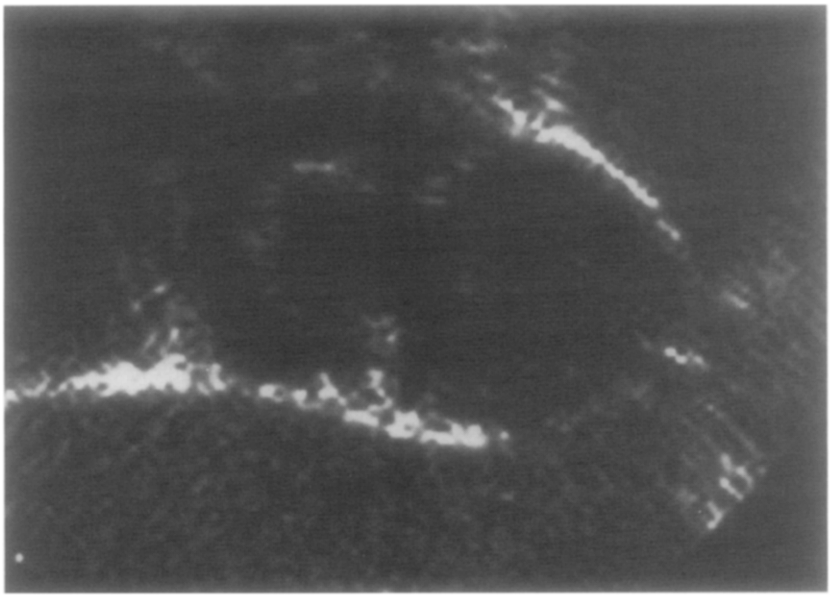

**Fig. 7.** Echocardiogram of extracardiac lateral tunnel 2 years after the operation showing smooth tube adjacent to RA connecting to right and left PAs.

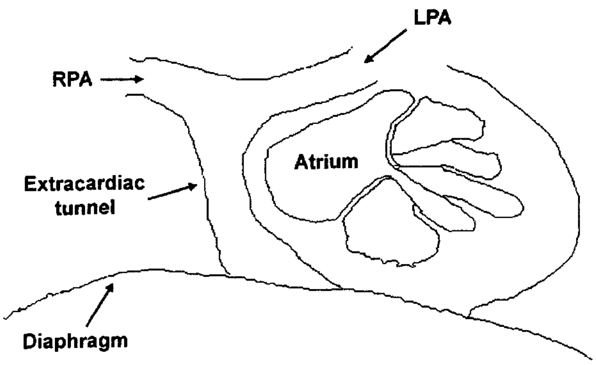


**Fig. 8.** Diagrammatic illustration of echocardiogram shown in Fig. 7. *RPA,* Right pulmonary artery; *LPA,* left pulmonary artery.

of these features has eliminated myocardial ischemia. Moreover, the use of synthetic tubes intraatrially has resulted in pulmonary and systemic emboli from clotting within or external to the tube within the atrium. Finally, some concern exists that the SVC orifice that serves as the lateral tunnel's exit to the pulmonary artery is smaller in caliber than the IVC and thus becomes a built-in resistance circuit.

The extracardiac lateral tunnel, as described by Amodeo and colleagues,[10] would appear to answer many of these failings. A tube placed between the IVC and PA is easy to construct, and although Amodeo and coworkers[10] have used aortic cross-clamping and circulatory arrest, as we have shown in this report and as has been described by others,

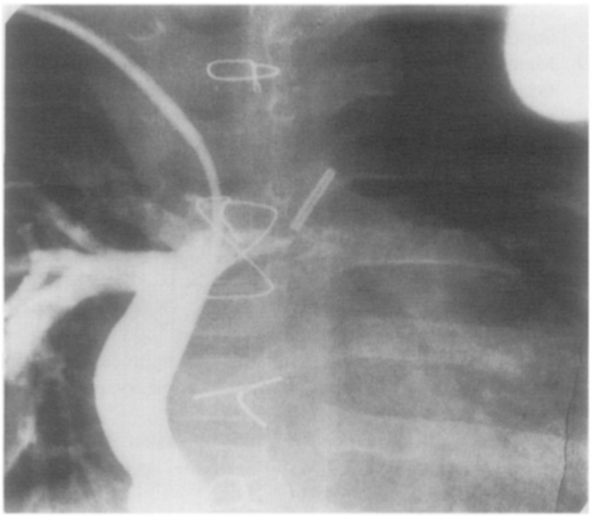

**Fig. 9.** Angiogram, 1½ years after the operation, of a pedicled pericardial lateral tunnel showing smooth-walled chamber leading from IVC to right PA.

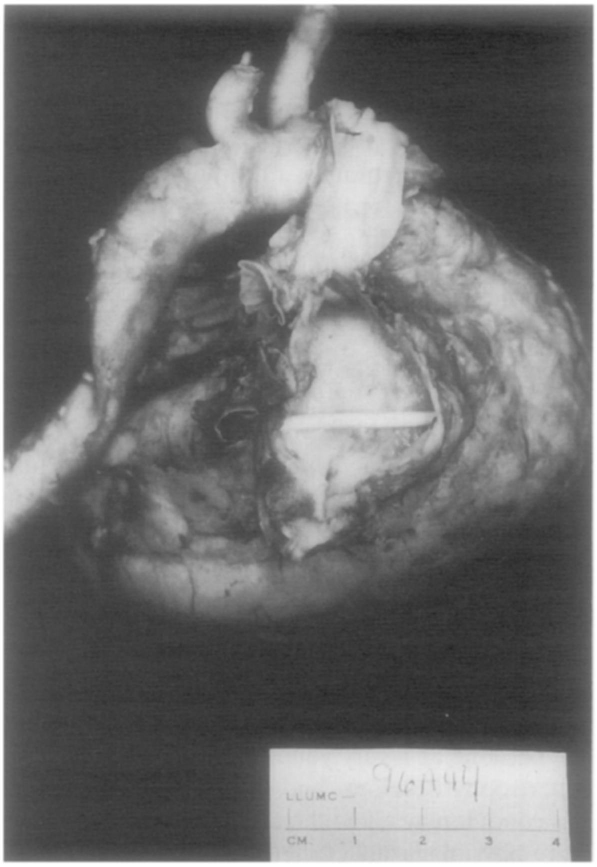

**Fig. 10.** Autopsy specimen, 6 months after the operation, of an opened pedicled pericardial lateral tunnel showing a smooth, endothelial cell–lined surface and open connections to the IVC *(below)* and PA *(above)*.

there is no reason to use ischemia to construct these anastomoses outside the heart. Because no suture lines are around the SVC-RA junction, atrial arrhythmias should be less prevalent. Left-sided emboli (unless the Fontan shunt is fenestrated) should be eliminated because no foreign material exists within the heart.

Unfortunately, extracardiac conduits with homografts are expensive and use a limited resource. Extracardiac conduits of Dacron or polytetrafluoroethylene are cheaper but show trends toward luminal size loss. At six months' follow-up of 80 patients with polytetrafluoroethylene or Dacron extracardiac lateral tunnels, Amodeo and associates[10] found an 18% mean decrease in luminal area and a 32% maximal decrease in area. Perhaps most worrisome is the lack of growth potential in width or length, limiting the application of these tubes in very young children. Young children are perhaps best served by early intervention to avoid ventricular hypertrophy or dilatation (or both) from prolonged volume overload.[14]

The pericardium represents a vascular graft substitute that has been used in the heart and great vessels from the earliest days of cardiac surgery. As such, its parietal surface is nonthrombogenic and it does not dilate, unless exposed to exceptional distal obstruction. The pericardial reflection along the lateral and posterior borders of the atria and PA provides a natural, smooth-walled passageway from IVC to PA. When the pericardium is pedicled along its lateral blood supply and turned down along the right atrium, the natural tunnel formed from IVC to PA is striking in its simplicity; because it is vascularized, the conduit should grow in length and width as physiologic needs change (see Figs. 6 to 8).

Because the pericardial tube is viable, it is also pliable, thus being capable of absorbing cardiac pulsations, as well as being affected by the respiratory bellows mechanism. Although difficult to quantify and prove, this bellows mechanism would affect only a pericardial viable extracardiac lateral tunnel, having no effect on a rigid extracardiac lateral tunnel or on an intracardiac lateral tunnel. This may account for the minimal difficulty to date with pleural effusions and excellent cardiac output, despite no fenestrations.

We have been struck with the lack of perioperative low cardiac output in this group of patients, despite the use of no fenestrations, but only the accrual of more patients will allow this observation to be tested more thoroughly. However, inasmuch as the right atrium forms the medial wall of the viable extracardiac lateral tunnel, fenestration of this connection could be easily accomplished in the catheterization laboratory after the operation, eliminating the need for adjustable snares or for automatically fenestrating all Fontan shunts. Although long-term follow-up is relatively short, the dramatic growth of the 9-month-old child's viable extracardiac lateral tunnel in the 2½ years since the operation (see Fig. 6) suggests that growth potential is real.

## Conclusion

Construction of a viable extracardiac lateral tunnel with autologous in situ pericardium pedicled on its lateral blood supply has been accomplished in 19 young patients with promising initial results. Viable pericardial extracardiac lateral tunnels may represent the optimal Fontan connection, because they are easy to construct without ischemia, use no foreign material, and have the potential to grow in width and length. Completion Fontan procedures can be done at a very young age before ventricular hypertrophy develops, and the unique elasticity of this conduit may allow the respiratory bellows to be more physiologically efficient.

## Addendum

Since submission of this manuscript, an additional 10 patients have been treated with this technique. The conclusions based on the 19 original patients remain valid after the addition of 10 more patients. No patients required fenestration, median postoperative stay was 4 ± 2 days, and no patient required inotropic support.

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## Discussion

**Dr. V. Mohan Reddy** *(San Francisco, Calif.).* Dr. Gundry and associates describe yet another modification of the modified Fontan procedure. It is an innovative and elegant technique and appears to be promising. In addition to all the advantages of the extracardiac conduit Fontan, the potential for growth is the most important. I have three questions for Dr. Gundry.

Inasmuch as the pericardium is not clean in reoperations, do you give anticoagulants or antiplatelet drugs for a short period of time?

**Dr. Gundry.** No, we have not used any anticoagulation in these patients. It is true that this area sometimes is a little bit rawer than others, and we take some time to clean off any debris that may be present, but we have not required anticoagulation.

**Dr. Reddy.** Have you had any episodes of pulmonary thromboembolism episodes in these patients?

**Dr. Gundry.** No, and we have not had any systemic emboli, obviously, because there is no fenestration. We do get echocardiograms of these patients and the tubes appear to stay smooth walled.

**Dr. Reddy.** Have you had any cases in which the pericardium was not usable or was inadequate?

**Dr. Gundry.** We have not encountered that yet. In one of Dr. Razzouk's patients we had to add a small piece of autologous pericardium from the other side to make an adequate tube. However, because the rest of the pericardium is pedicled, it is possible to add from the other side if need be.

**Dr. Reddy.** Although your experience suggests that no arrhythmias have occurred on short-term follow-up, the longitudinal atrial suture line may be of some concern. Do you routinely perform any Holter studies or other electrophysiologic studies to monitor rhythm at regular intervals?

**Dr. Gundry.** We have not done that as yet. Although the artist's drawing shows this atrial suture line primarily on the RA, in fact in most constructions this suture line exists more in the atrioventricular groove, rather than within the RA tissue itself. I think the worry about atrial arrhythmias is going to be small.

**Dr. Reddy.** I think the long-term follow-up of these patients will be especially useful for all the physicians who participate in the management of patients having the Fontan operation.

**Dr. Davis C. Drinkwater** *(Los Angeles, Calif.).* This technique is innovative. However, inasmuch as the pericardium is capable of growing, is there a risk of pulmonary venous obstruction developing, similar to what we have seen with the earlier lateral tunnels constructed with pericardium?

Second, would you consider fenestration a contraindication with this technique?

**Dr. Gundry.** No, in fact, I think the nice thing about this technique is that if you need a fenestration you can construct it in the catheterization laboratory after the operation. The cardiologist can take the patient to the catheterization laboratory and poke a hole in the RA wall, which makes it a lot easier and eliminates having snares on fenestrations.

**Dr. Drinkwater.** We have seen earlier pericardial lateral tunnels come back with obstructed pulmonary veins on the right side, and you are right over that area with this technique. Are you concerned?

**Dr. Gundry.** No. I think as long as pericardium is viable, and as long as the surgeon follows Laplace's law, the pericardium follows Laplace's law fairly well. Pericardium will not dilate unless it has a distal obstruction somewhere. We have known this for years in right ventricular outflow tract reconstruction and also aortic reconstruction. As long as we make that initial tube small and match the diameter of the intended flow, I do not think it is going to be a problem. We have not seen that problem in 2¾ years' follow-up.

**Dr. John Lamberti** *(San Diego, Calif.).* The work that Marc de Leval has done in London with engineers looking at flow in rigid tubes has supported the trend toward the lateral baffle type of Fontan operation. The goal of that baffle Fontan operation is to create a relatively rigid tube, if possible. When you introduce the concept of respiratory variation augmenting cardiac output, you may be challenging the "rigid tube" concept. Are you troubled by the fact that this runs counter to the teachings of other leaders in the field?

**Dr. Gundry.** Not really. Of course de Leval trained me and I became a disciple of his technique. We were struck actually from the first case, looking at PA tracings, by the wide swing in the PA with respiration, and we have been tracking that. The amount of respiratory pulsation that is available through this technique is striking. I think it actually may account for why we did not need fenestrations and why low cardiac output did not occur in this group of patients. I think it is going to be beneficial.

## Commentary

Dr. Gundry and his colleagues have described an interesting technical modification to achieve a Fontan circulation. In this technique they create an extracardiac tunnel from the inferior vena cava to the right pulmonary artery by suturing a pedicled pericardial flap to the free wall of the right atrium. The results in the first 19 patients appear to have been favorable, although the longest follow-up is only 2.5 years.

This technique has several attractive features. First, the pedicled nature of the pericardial flap should allow growth potential, and the authors provide limited data to support this potential advantage. Second, the extracardiac position of the Fontan pathway should preclude the acute creation of pulmonary venous obstruction by an intracardiac Fontan pathway, an important technical consideration in cases of aberrant systemic or pulmonary venous connection or in cases of left atrioventricular valvular stenosis or atresia. The extracardiac position of the Fontan pathway also would be expected to reduce the systemic thromboembolic risk by eliminating intracardiac patch/baffle material, although there are other intracardiac sources for emboli such as atrial appendages and the cardiac end of the original pulmonary artery.

However, it is my impression that there are two potential drawbacks to the extracardiac lateral tunnel technique performed with pedicled pericardium. First, as I understand the technique, the "floor" of the extracardiac lateral tunnel contains the right pulmonary veins. We have now seen three cases in which anterior compression of the right pulmonary veins by a markedly enlarged right atrium has occurred late after a "classic" Fontan procedure in which the atrial septal defect was closed and an atriopulmonary connection was created. Although the authors' technique should avoid creating a massively dilated right atrium, the location of the right pulmonary veins in the floor of the Fontan pathway would appear to put them at risk for compression by the chronically higher pressures within the Fontan pathway. The authors have not reported detailed echocardiographic angiographic follow-up to address this question, nor have they carried out lung scans to determine the relative distribution of blood flow between the right and left lungs. These data will be very important in the longer term evaluation of the technique. Finally, although the authors have reported no difficulties with sinus node dysfunction or arrhythmias, it is my impression that the pericardial flap to the atrial suture line must cross in some proximity to the sinus node; therefore long-term rhythm issues may still be a concern with this technique. Holter monitor data will also be important in the long-term evaluation of this technique.

In summary, Dr. Gundry and colleagues have described an interesting modification to the Fontan technique, which has significant potential advantages but some important potential for late problems. Longer follow-up information will be essential to more complete evaluation of this technique.

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