# Novel lateral tunnel Fontan operation in children promotes continued pathway growth into adulthood

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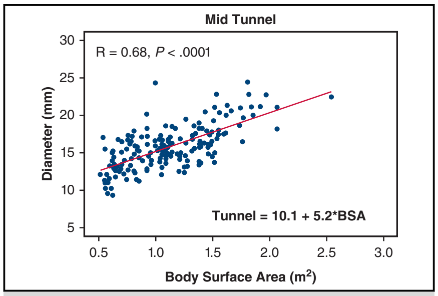
## ABSTRACT

**Objective:** Most Fontan operation techniques currently used in children are limited by a blood flow pathway with no growth potential. The external pericardial lateral tunnel Fontan is a novel technique using the patient's pedicled pericardium and atrial wall in situ to create a Fontan pathway. We hypothesize that viable, autologous tissue will allow growth potential of the pathway and adapt to increasing physiologic demands of somatic growth.

**Methods:** We performed a single-center retrospective review of serial echocardiographic measurements of the inferior vena cava junction and mid tunnel in patients with an external pericardial lateral tunnel Fontan. Linear mixed model and Pearson correlation tests were used for analysis of changes in pathway size with time and body surface area.

**Results:** A total of 1592 echocardiographic studies from 172 patients up to 23 years after the procedure were identified. Significant enlargement of the pathway at the inferior vena cava junction and mid tunnel was observed over time ($P < .0001$, $P < .0001$, respectively). There is a strong positive correlation between increasing size of the inferior vena cava junction and increasing body surface area (R = 0.81, $P < .0001$). There is also a strong positive correlation between increasing size of the mid tunnel with increasing body surface area (r = 0.67, $P < .0001$).

**Conclusions:** The external pericardial lateral tunnel Fontan technique creates a Fontan pathway that enlarges into adulthood. The correlation of increasing pathway size with increasing body surface area suggests pathway growth, responding to increasing physiologic demands of somatic growth into adulthood. Further studies are needed to investigate the impact of this novel technique on Fontan physiology and long-term patient outcomes. (JTCVS Techniques 2025;29:138-45)




**Linear relationship of Fontan pathway tunnel diameter and BSA.**

### CENTRAL MESSAGE

An extracardiac Fontan pathway constructed of in situ pericardium and atrial wall grows in response to increasing physiologic demands of somatic growth.

### PERSPECTIVE

Optimal Fontan hemodynamics require a Fontan pathway that adapts to individual patient physiology. The intra-atrial LTF pathway has growth potential, whereas fixed-size external conduits do not. Complications of conduit and patient size mismatch are increasingly being recognized. Dynamic growth of the EPLTF pathway may lead to improved Fontan physiology and outcomes.

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▶ Video clip is available online.

The Fontan operation is widely used for palliation of single-ventricle congenital heart disease. The procedure creates a direct blood flow pathway from the inferior vena cava (IVC) to the pulmonary arteries early in childhood, resulting in a total cavopulmonary connection: the Fontan circulation. The operative technique has continued to evolve since its introduction in 1971 in an ongoing effort to minimize complications and improve outcomes.[1,2]

**Abbreviations and Acronyms**

BSA = body surface area
ECF = extracardiac conduit Fontan
EPLTF = extracardiac pericardial lateral tunnel Fontan
IVC = inferior vena cava
LTF = lateral tunnel Fontan

The 2 most used techniques include the lateral tunnel Fontan (LTF), which creates an intra-atrial pathway, and the extracardiac conduit Fontan (ECF), which uses an extracardiac artificial tube, commonly made of polytetrafluoroethylene (Gore-Tex). Both techniques have their own limitations. The LTF is prone to atrial arrhythmias, places a thrombogenic artificial surface in the systemic circulation, and requires a period of myocardial ischemia to be completed. The ECF is limited by a fixed-size tube, which has been shown to have decreased cross-sectional area as early as 6 months after the procedure.[3] Implanting an adult-sized conduit in a small child can be technically challenging and hemodynamically undesirable.

The extracardiac pericardial lateral tunnel Fontan (EPLTF) technique, described by Gundry and colleagues[4] in 1997, uses a flap of autologous pericardium and atrial wall in situ to create an extracardiac tunnel from the IVC to the pulmonary arteries. This creates a pathway of native tissue that is vascularized and pliable, and may have growth potential. The EPLTF also eliminates the need for intra-atrial suture lines, thrombogenic foreign material, and the requirement for a period of myocardial ischemia to complete the operation. Favorable outcomes up to 15 years after EPLTF have been reported, as well as observed changes in pathway diameter correlating with changes in patient body surface area (BSA) in 10 patients.[5] An analysis of 38 patients who underwent the EPLTF reported pathway enlargement to accommodate somatic growth in childhood.[6] The objective of this study is to investigate the EPLTF pathway growth potential in a larger group of patients into adulthood.

## MATERIAL AND METHODS

### Surgical Technique

After the initiation of cardiopulmonary bypass with a single venous cannula in the atrial appendage, the patient is cooled to 22 °C. Direct cannulation of the superior and inferior venae cavae is avoided to minimize distortion and eliminate the need for circumferential dissection of the venae cavae. While cooling, the pericardial flap is prepared and the need for supplemental pericardium is assessed. The pericardium is dissected free from the lung, then the atrial wall, and posteriorly until the pulmonary veins are identified. The pericardial flap is detached from the diaphragm with electrocautery to the level of the IVC. Superiorly, the pericardial flap is separated laterally from the Glenn and branch pulmonary artery, leaving attachments along the inferior edge of the branch pulmonary artery. The location of the phrenic nerve is continuously noted, and the use of electrocautery near the phrenic nerve is minimized. The phrenic nerve is most vulnerable to injury near the hilum of the pulmonary artery. While temporarily restricting venous return and allowing the atrium to distend, a proposed pericardial suture line along the atrial wall is marked with ink.

Approximately 5% of patients require additional pericardium that is harvested from the contralateral pericardium or less frequently supplemented with bovine pericardium. Pericardial deficiency is most common at the pulmonary artery end of the tunnel. Surgical planning for a successful Fontan begins at stage 1 palliation with intentional preservation of as much viable pericardium as possible. The initial pericardial incision should be to the opposite side of the midline and away from the future Fontan pathway with minimal dissection near the venae cavae.

Myocardial fibrillation is induced by epicardial slush ice, eliminating the need for aortic crossclamping. The patient is maintained in the Trendelenburg position with venting of the aortic root. Hypothermic, low-flow perfusion (30-50 mL/kg/min) is maintained using atrial cardiotomy suckers that replace the venous cannula for venous return. The anterior wall of the IVC is incised, at least 60% of the circumference, and sewn to the posterior wall to close off the cardiac end. Careful suturing is required to avoid the inadvertent creation of a fenestration. The angled suture line directs blood entering the Fontan pathway laterally, away from atrial tissue. The IVC is sized with a Hegar dilator. The pericardial flap is sewn to the anterior edge of the IVC from lateral to medial, extending onto the atrial wall parallel to the atrioventricular groove. The IVC is again sized with a Hegar dilator to ensure the pathway is not constricted. The height of the pericardial flap is trimmed to match the marked atrial wall suture line, creating a cylindrical pathway without excessive bulging. Hemostasis of the highly vascularized pericardium with collateral vessels is best accomplished with electrocautery along the cut edge and hemoclips where collateral vessels are divided. The venous cannula is returned to the atrium, and slow rewarming of the patient is begun.

The branch pulmonary artery is dissected along the inferior edge. An arteriotomy is created opposite the Glenn anastomosis and extended toward the aorta, at least 2 mm longer than the IVC diameter. Blood from the superior vena cava and pulmonary arteries is captured with cardiotomy suckers. Multiple interrupted sutures are placed along the inferior edge of the pulmonary arteriotomy, securing it to the floor of the Fontan pathway. Any excess tissue or ridges from scar tissue in the Fontan pathway are removed. The pericardial flap is sewn to the anterior edge of the pulmonary arteriotomy from lateral to medial, transitioning away from the arteriotomy and onto the aortic wall. The running suture line is continued on the lateral aortic wall, toward the aortic root, and onto the atrial wall to create a broad connection of the pericardial tunnel to the pulmonary artery. The Fontan tunnel is completed by securing the pericardial flap along the previously marked atrial wall suture line. Patient rewarming is completed, and bypass is discontinued. The Fontan lateral tunnel setup is shown in Figure 1, thee completed Fontan lateral tunnel is shown in Figure 2, and a cross-section of the Fontan lateral tunnel is shown in Figure 3. The surgical technique (Video 1) and imaging of the completed tunnel by biplane cineangiography (Video 2) are included in the Supplemental material.

### Study Population

A retrospective review of serial echocardiographic studies and medical records for patients who received an EPLTF operation between January 1, 1994, and June 31, 2022, at Loma Linda University Children's Hospital was conducted. All patients with at least 1 echocardiographic study done by our cardiology group after hospital discharge for the EPLTF operation were included. Those with poor image quality that limited our ability to obtain accurate measurements were excluded from the study. The follow-up period was defined as the time from the EPLTF operation to the most recent echocardiogram. The Institutional Review Board of Loma Linda University Health approved the study protocol and publication of data (IRB #5220009. February 15, 2022). Patient written consent for publication of the study data was waived by the IRB.

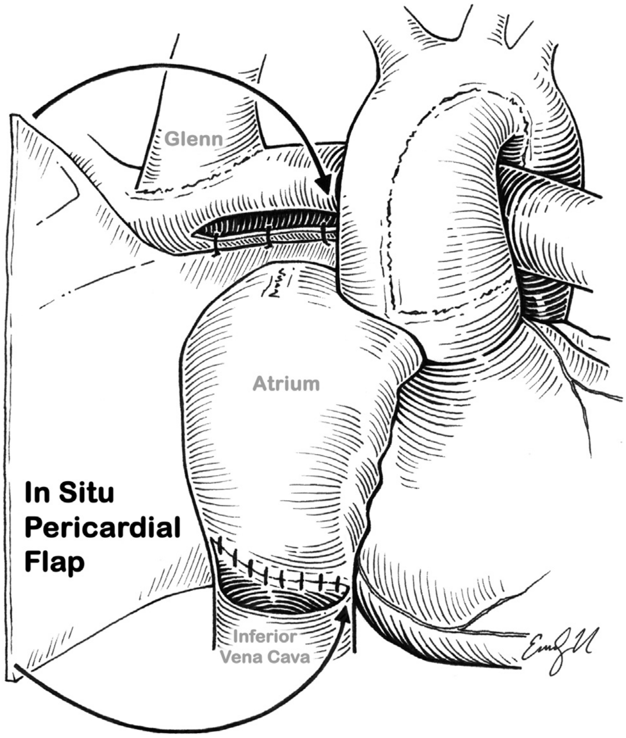



FIGURE 1. Extracardiac pericardial LTF setup. The in situ pericardial flap is prepared to roll onto the lateral atrial wall. The IVC and branch pulmonary arteries are widely opened to ensure an obstructed Fontan pathway.

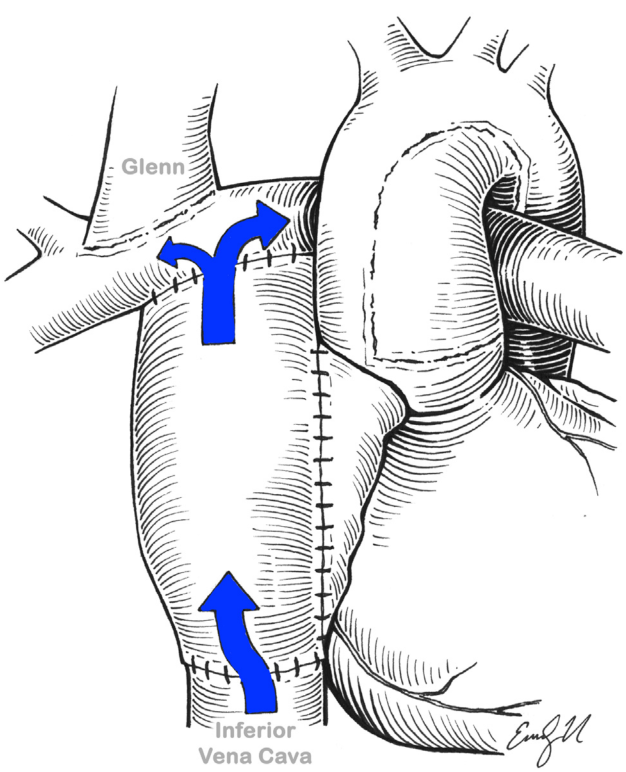



FIGURE 2. Completed EPLTF. Venous blood from the IVC is channeled outside the atrium to the branch pulmonary arteries.

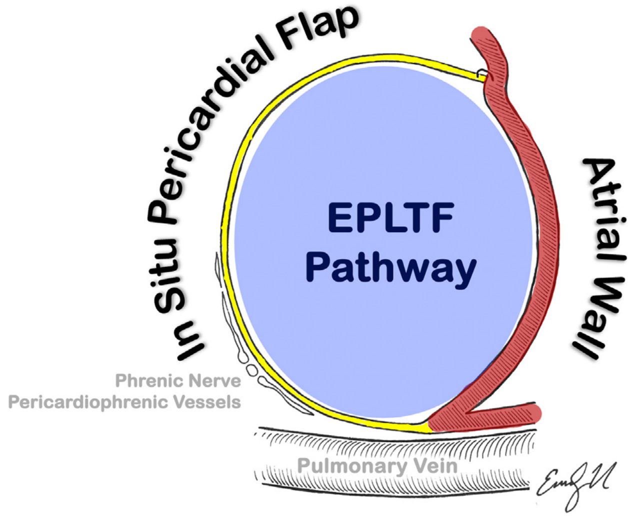



FIGURE 3. Cross-section of the completed EPLTF. The Fontan pathway is composed entirely of in situ pericardium and atrial wall with preservation of the phrenic nerve and pericardiophrenic vessels. *EPLTF*, Extracardiac pericardial lateral tunnel Fontan.

### Echocardiography

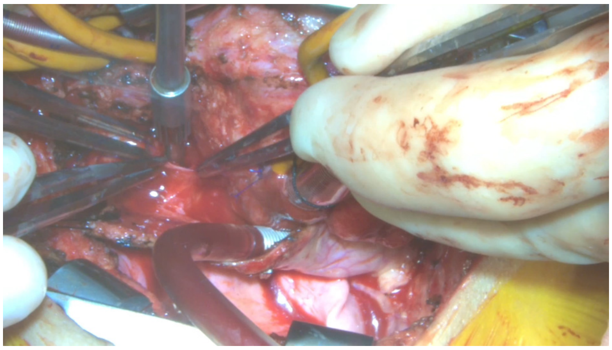
Echocardiographic images were reviewed and analyzed using Syngo Dynamics (Siemens Healthcare/Siemens Medical Solutions USA Inc). Two distinct measurements of the Fontan pathway diameter were obtained from a standard subcostal bicaval view: the IVC junction with the Fontan and mid tunnel. BSA was calculated at the time of each echocardiogram.

### Statistical Analysis

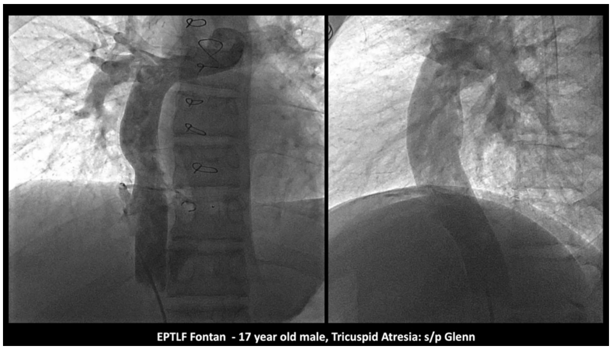
A linear mixed model was used to evaluate the association of increasing size of the IVC junction and mid tunnel measurements with increasing time from the Fontan operation. The Pearson correlation test was used to evaluate the association of IVC junction dimension and BSA, mid tunnel dimension and BSA, and dimension of the IVC junction and mid tunnel. All analyses were done using SAS 9.4 (SAS Institute, Inc) and R 4.2.2 (https://www.rproject.org/) with conclusions made at the 5% significance level. Variables are summarized as median (25th percentile, 75th percentile) or n (%).

## RESULTS

Of 263 patients who underwent EPLTF identified, 172 patients were included in the study. A total of 1592



VIDEO 1. Extracardiac pericardial LTF surgical technique. Video available at: https://www.jtcvs.org/article/S2666-2507(24)00440-1/fulltext.




**VIDEO 2.** Extracardiac pericardial LTF biplane cineangiography. Video available at: https://www.jtcvs.org/article/S2666-2507(24)00440-1/fulltext.

echocardiographic studies were reviewed, and 1580 images were used for pathway measurements. The median number of images per patient is 10 (6-12). The median follow-up period is 12 (6.5-14.6) years with maximum follow up 23.2 years. Patient characteristics at the time of Fontan are summarized in Table 1.

**TABLE 1. Patient demographics at the time of extracardiac pericardial lateral tunnel Fontan operation (n = 172)**

| | |
|---|---|
| Age (y) | 2.5 (2.1-3.2) |
| Height (cm) | 89 (84-94) |
| Weight (kg) | 12.7 (11.2-14.2) |
| BSA ($m^2$) | 0.54 (0.50-0.59) |
| Sex | |
| Female | 72 (42%) |
| Anatomic features | |
| AVSD | 41 (24%) |
| HLHS | 31 (18%) |
| TA | 29 (17%) |
| DILV | 20 (12%) |
| PA/IVS | 19 (11%) |
| DORV | 17 (10%) |
| MA | 5 (3%) |
| IAA | 5 (3%) |
| Ebstein | 3 (2%) |
| Other | 2 (1%) |
| +TAPVC | 8 (5%) |
| +Heterotaxy | 26 (15%) |
| Dominant ventricle | |
| Left | 81 (47%) |
| Right | 79 (46%) |
| Balanced | 12 (7%) |

Variables summarized as median (25th quartile, 75th quartile) or n (%). *BSA*, Body surface area; *AVSD*, atrioventricular septal defect; *HLHS*, hypoplastic left heart syndrome; *TA*, tricuspid atresia; *DILV*, double inlet left ventricle; *PA/IVS*, pulmonary atresia with intact ventricular septum; *DORV*, double outlet right ventricle; *MA*, mitral atresia; *IAA*, interrupted aortic arch; *TAPVC*, total anomalous pulmonary venous connection.

We observed significant enlargement of the EPLTF pathway at the IVC junction and mid tunnel over time ($P < .0001$, $P < .0001$, respectively) with a strong correlation between increasing size at the IVC junction and increasing size at the mid tunnel (R = 0.67, $P < .001$). The median IVC junction diameter increased from 10.2 mm at 1-year follow-up to 20.5 mm at 20-year follow-up, representing a 50% increase in size. The median mid tunnel diameter also increased from 12.5 mm to 22.4 mm during the same time period, a 56% increase in size (Figure 4).

Strong positive correlations between IVC junction size and BSA (R = 0.81, $P < .0001$) and mid tunnel size and BSA (R = 0.68, $P < .0001$) were also observed. The relationship between EPLTF pathway measurements and BSA appears to be linear (Figure 5).

## DISCUSSION

A previously published analysis from our institution showed that in a population of 38 patients, significant growth was measured in the EPLTF pathway through childhood, the first 10 years after Fontan completion. The relationship between pathway measurements and BSA is significant, suggesting the pathway is able to enlarge during this period of rapid somatic growth, adapting to the increasing physiologic demands placed on the Fontan circulation.[6]

In the current analysis, we have confirmed that the EPLTF pathway continues to enlarge with increasing BSA into adulthood, up to 23 years after Fontan completion. An approximately 60% increase in median mid tunnel size from year 1 to year 20 after Fontan completion suggests that constructing a Fontan blood pathway composed entirely of viable, autologous tissue preserves growth potential (Figure 6). The linear relationship between pathway measurements and BSA further suggests that increasing physiologic demands of somatic growth may be driving growth of the Fontan pathway into adulthood. Optimal hemodynamics of the Fontan circulation require matching Fontan pathway dimensions with physiologic demands of the individual patient. The relationship between the EPLTF pathway dimensions and the BSA may help to define the ideal pathway size.

The Fontan pathway created by the LTF is partially constructed of viable atrial wall and has shown growth potential in multiple studies.[7-9] Magnetic resonance imaging studies show increased volume and flow correlated with increasing BSA.[10] However, it has been noted that growth can be "inconsistent and heterogeneous" and may require stenting.[11,12]

In contrast to the EPLTF and LTF, the ECF requires insertion of a fixed-size external conduit in children that has no growth potential. Optimal-sized conduits for children are small for adults. The IVC to conduit size mismatch in

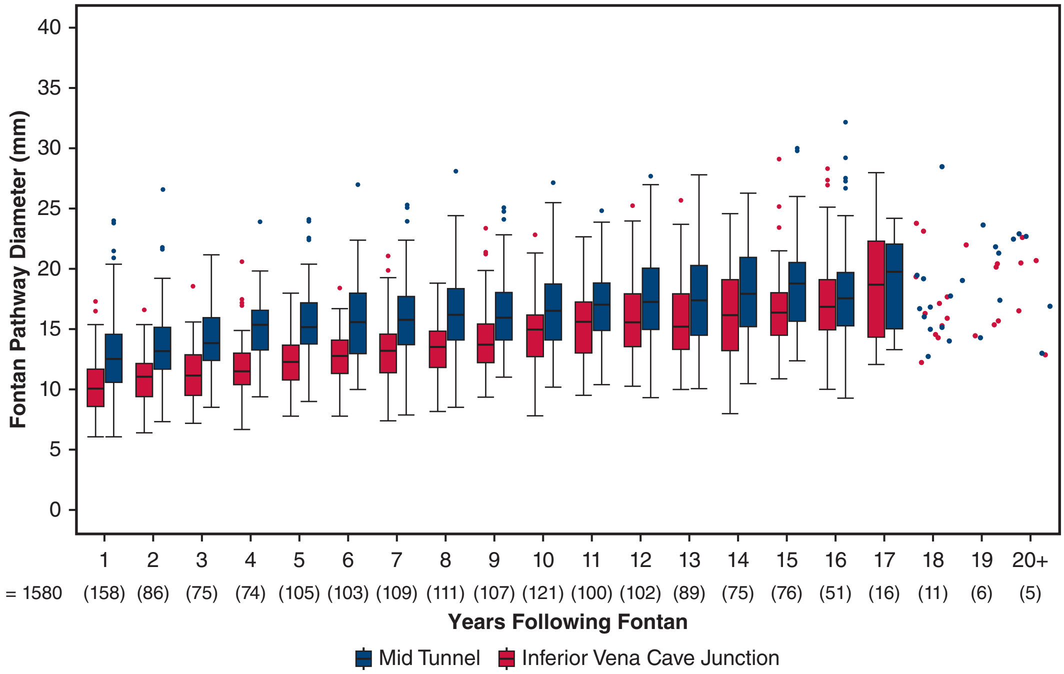



**FIGURE 4.** Fontan pathway measurements over time from Fontan operation. The box *horizontal line* represents median with the *lower* and *upper borders* representing the 25th and 75th percentiles. The *lower* and *upper whiskers* represent the minimum and maximum values of nonoutliers. Extra *dots* represent outliers. For year 17 and beyond, each measurement is plotted individually due to less than 14 measurements per year. The number of measurements each year are shown in parentheses.

adulthood results in increased flow velocity and unfavorable hemodynamics with high energy loss.[7,13] Decreased exercise capacity is a common finding.[7,14-16] Hepatic dysfunction, fibrosis, and cirrhosis are increasingly recognized beyond the first decade post-Fontan completion.[15,17,18] Many patients have reduced renal function.[18,19]

In addition to concerns about the lack of growth potential of the ECF, progressive stenosis of external conduits is common. Cardiac catheterization and magnetic resonance imaging have shown decreases in cross-sectional area by 33% as early as 6 months after Fontan completion.[3] Others have found 50% to 66% decreases in cross-sectional area in patients followed up to 17 years with time-dependentworsening of stenosis.[8,20,21] Pseudo-intimal hyperplasia, thrombus, calcification, and luminal irregularity have been implicated.[16] The ECF surgical technique for conduit insertion also may contribute to Fontan pathway stenosis.[22]

Adult-size external conduits often are placed in children in anticipation of future flow requirements in adulthood. Oversized conduits have poor flow hemodynamics with areas of turbulence and stagnation, pseudo-intimal hyperplasia, and mural thrombus with resultant narrowing of the proximal and distal segments.[16,23] Flow reversal with exercise and decreased exercise capacity have been identified.[23,24] Placing an oversized external conduit in a child

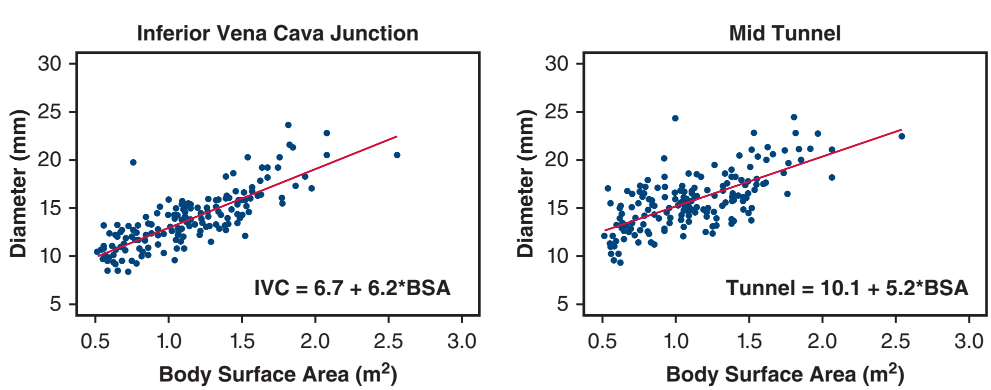



**FIGURE 5.** Fontan pathway measurements by BSA. *BSA*, Body surface area; *IVC*, inferior vena cava.

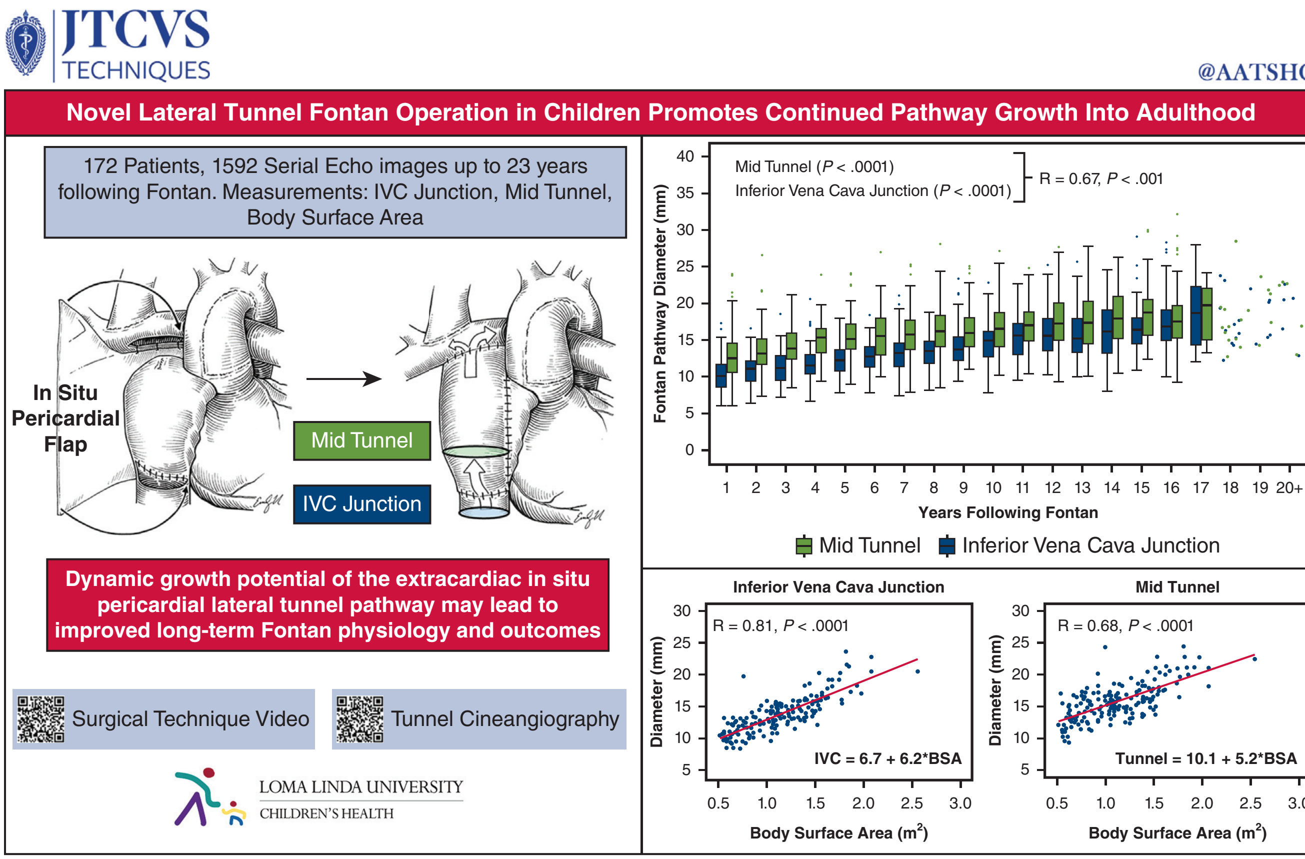



**FIGURE 6.** Extracardiac pericardial LTF pathway growth correlates with somatic growth into adulthood. *IVC*, Inferior vena cava.

will not result in optimal hemodynamics during childhood or once physiologic demands of growth have plateaued decades later. Other authors have noted that "matching Fontan pathway to somatic growth will require a different strategy than extracardiac conduit oversizing."[3] Concerns about the lack of growth and increasing recognition of external conduit stenosis and the associated complications have prompted some to abandon external conduits in favor of the lateral tunnel operation for Fontan completion.[25]

The management of undersized or stenotic external conduits has received considerable attention. Stenting can improve hemodynamics and functional capacity.[26] It has been shown that polytetrafluoroethylene tubes (Gore-Tex) can be expanded to a nominal size and larger.[20] New-generation expanded polytetrafluoroethylene grafts (exGraft, PECA Labs) that have the potential to be progressively dilated 200% may improve conduit longevity.[27] Patients with an external conduit are likely to need reinterventions in adulthood, including both stenting and surgical replacement of dysfunctional conduits.[25,27] Tissue-engineered vascular grafts may offer an alternative to address the issue of small conduits.[28]

The EPLTF has remained our chosen technique since the 1990s for creation of the Fontan circulation. Like the ECF, the operation can be completed on the beating heart without disruption of myocardial perfusion. In contrast to the LTF, the lack of intra-atrial suture lines should decrease the risk of atrial arrhythmias after EPLTF completion. Because the pathway is formed of viable, autologous tissue, the need for thrombogenic foreign material in the Fontan pathway is eliminated. Most of our patients do not have a fenestration and are maintained on aspirin only for anticoagulation. Patients with a fenestration receive low-dose warfarin and aspirin. Although this study only examined the changes in pathway size, our previously reported 15-year follow-up data showed low morbidity and mortality with relation to arrhythmias, stenosis, and thrombosis.[5] These findings suggest that the EPLTF technique is a favorable option for Fontan completion. Surgical programs with limited resources may further benefit from this novel Fontan completion technique that uses only autologous tissue rather than prosthetic materials. Although complications and outcomes of the Fontan circulation may be more a reflection of abnormal physiology than the Fontan operation per se, our challenge is to continually refine surgical technique in "the pursuit of excellence" (personal communication, Leonard L. Bailey, MD).

### Study Limitations

Because of the retrospective nature of this study, adequate echocardiographic images were available in only 172 of 263 patients, representing 65% of the population. The number of studies per patient and interval between imaging are variable. Echocardiographic images only allow visualization of the proximal portion of the EPLTF pathway in 2 dimensions, limiting our ability to evaluate the distal portion of the pathway and overall pathway geometry. We are also unable to evaluate flow dynamics of the pathway with this modality.

## CONCLUSIONS

The EPLTF technique creates a Fontan blood pathway that enlarges into adulthood. Correlation of increasing pathway size with increasing BSA suggests pathway growth, responding to increasing physiologic demands of somatic growth into adulthood. Further studies are needed to investigate the impact of this novel technique on Fontan physiology and long-term patient outcomes.

### Conflict of Interest Statement

The authors reported no conflicts of interest.

The *Journal* policy requires editors and reviewers to disclose conflicts of interest and to decline handling or reviewing manuscripts for which they may have a conflict of interest. The editors and reviewers of this article have no conflicts of interest.

Surgical technique illustrations were created by Emily Nagele, BS. Additional statistical assistance was provided by John Dibato, PhD.

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