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PERSPECTIVE

# Anesthetic challenges in patients with multicompartmental lymphatic failure after Fontan palliation undergoing transcatheter thoracic duct decompression

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## Abstract

Lymphatic flow abnormalities are central to the development of protein losing enteropathy, plastic bronchitis, ascites and pleural effusions in patients palliated to the Fontan circulation. These complications can occur in isolation or multicompartmental (two or more). The treatment of multicompartmental lymphatic failure aims at improving thoracic duct drainage. Re-routing the innominate vein to the pulmonary venous atrium decompresses the thoracic duct, as atrial pressure is lower than systemic venous pressure in Fontan circulation. Transcatheter thoracic duct decompression is a new minimally invasive procedure that involves placing covered stents from the innominate vein to the atrium. Patients undergoing this procedure require multiple general anesthetics, presenting challenges in managing the sequelae of disordered lymphatic flow superimposed on Fontan physiology. We reviewed the first 20 patients at the Center for Lymphatic Imaging and Intervention at a tertiary care children's hospital presenting for transcatheter thoracic duct decompression between March 2018 and February 2023. The patients ranged in age from 3 to 26 years. The majority had failed prior catheter-based lymphatic intervention, including selective embolization of abnormal lympho-intestinal and lympho-bronchial connections to treat lymphatic failure in a single compartment. Fourteen had failure in three lymphatic compartments. Patients were functionally impaired (ASA 3–5) with significant comorbidities. Concurrent with thoracic duct decompression, three patients required fenestration closure for the resultant decrease in oxygen saturation. Ten patients had improvement in symptoms, seven had no changes and three have limited follow up. Five (25%) of these patients were deceased as of January 2024 due to non-lymphatic complications from Fontan failure.

**KEYWORDS**
Fontan failure, lymphatic flow abnormalities, thoracic duct decompression

## 1 | INTRODUCTION

The Fontan circulation is the result of surgical palliation of complex congenital heart defects where bi-ventricular separation and function cannot be safely achieved. The non-pulsatile pulmonary blood flow (PBF) is driven by the pressure gradient from the higher-pressure vena cavae to the lower-pressure pulmonary venous atrium. Adequate PBF is feasible when the pulmonary vascular bed resistance is low. The absence of a ventricle providing PBF results in increased dependency on intravascular volume and sensitivity to hypovolemia or impaired

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systemic venous return. Some patients have a fenestration, a surgically created communication between the systemic venous return and the atrium. The purpose of the fenestration is to augment ventricular preload when conditions that limit PBF exist, at the expense of causing a right to left shunt.[1] Incremental improvements in patient selection, surgical technique and long-term medical management have led to greater long-term survival in this population. While the exact prevalence is unknown, it is estimated that 50000–70000 patients live with Fontan Circulation worldwide, with this population expected to double in the next 20 years.[1] The need for anesthesia services for the patient with Fontan circulation outpaces the rate of growth of anesthesiologists with sufficient expertise (and perhaps familiarity) to manage them for non-cardiac surgery.[2]

While survival after Fontan palliation has improved, long-term complications are increasingly recognized. Fontan failure is characterized by one or more of the following: death, transplant, functional impairment (New York Heart Association class III or IV), protein-losing enteropathy (PLE) or plastic bronchitis (PB).[3] Investigators have identified disordered lymphatic flow in the genesis of Fontan failure in many patients. Lymphatic flow abnormalities are central to the development of PLE, PB, ascites, and often pleural effusions. These complications can occur in isolation or in combination of two or more (multicompartmental).[4] Selective lymphatic embolization alone has been an effective treatment for isolated PB and PLE. It is not effective for multicompartmental lymphatic failure.[4] The lymphatic system drains into the venous system via the thoracic duct (TD) at the junction of the left internal jugular vein and the innominate vein. Because systemic venous pressure is elevated in the Fontan circulation, lymphatic drainage via the TD is impaired. Rerouting the innominate vein to the lower-pressure atrium address the problem by decreasing TD afterload, thereby allowing more lymphatic drainage.

Hraska described a surgical approach where the innominate vein is detached from the superior vena cava and is anastomosed, either directly or via an interposition graft, to the left atrial appendage.[5] Although this procedure, requiring cardiology pulmonary bypass and in most at least a 3rd sternotomy, has been successful in four of the six highly selected original patients, many are poor surgical candidates due to anatomic considerations, medical fragility, or both.

Smith et al. described transcatheter TD decompression (TTDD) which involved placing covered stents via either an intra or extravascular course from the innominate vein to the atrium.[6] This procedure has demonstrated promising results, but patients require multiple general anesthetics and reintervention is not uncommon.

Patients were identified in a registry of the Center for Lymphatic Imaging and Intervention at the Children's Hospital of Philadelphia. Permission for prospective data collection stored in the registry was obtained from parent/patient at the time of hospital admission. The institutional review board for the protection of human subjects approved access to the data for this study with waiver of informed consent. Over the period from March 2018 to February 2023, TTDD was performed on 20 patients. Additional data, including post procedure course and follow up was obtained from the hospital electronic medical record.

Before the scheduled procedure, multiple imaging modalities are necessary. They include cardiac magnet resonance imaging evaluating anatomy (patency and location of the TD outlet) and physiology with flow velocity mapping. Lymphatic imaging using T2 weighted MRI as well as dynamic contrast MRI lymphangiography (intranodal, intrahepatic, and mesenteric) lymphangiography are performed. Half of patients also underwent computed tomography imaging to aid in developing a 3-dimentional model for plotting the extravascular course of the covered stent pathway. General anesthesia is necessary for one or more of the required diagnostic imaging.

## 2 | TRANSCATHETER THORACIC DUCT DECOMPRESSION PROCEDURE

Initial procedural strategy involved placing covered stents via an intravascular route from the innominate vein, through the floor of the pulmonary artery and into the atrium. Due to concerns for protrusion of the stents leading to potential obstruction or thrombosis of the Fontan pathway, the technique was modified to create an extravascular course from the innominate vein to the ipsilateral atrium. Left internal jugular access is obtained to access the innominate vein and a transeptal needle is advanced via the extravascular course toward the atrial appendage, carefully avoiding the aorta, left pulmonary artery and the bronchi. The needle is then snared in the atrium, the tract balloon-dilated, and a stent placed across the pathway (Figure 1). Catheters are positioned in the aortic root and left pulmonary artery to identify these structures using manual contrast dye injections.

## 3 | ANESTHETIC AND INTRAOPERATIVE MANAGEMENT

Compartments of lymphatic failure and characteristics of the Fontan circulation are depicted in Table 1. Seventy percent of patients had lymphatic failure in three or more compartments and 90% had a failed prior embolic lymphatic intervention.

Four patients were admitted from home for the procedure. Sixteen patients required in-hospital care for their condition, five of whom were in intensive care. Pre-procedure management included some or all of the following: pleural and/or peritoneal drain, PICC line placement, correction of electrolyte derangements, albumin replacement, and initiation or continuation of outpatient medication, including diuretics and pulmonary vasodilators. Three of the five ICU patients received dopamine for inotropic support.

Table 2 contains data from each anesthetic. Pre-anesthetic assessment determined a high degree of impaired physical status (ASA 3, 4 or 5). Pre-anesthetic sedation, either midazolam or midazolam and ketamine, was administered to 85% of patients. Induction of anesthesia was most often achieved with ketamine, alone or in combination with propofol, followed by vecuronium, endotracheal intubation, and mechanical ventilation. Two patients underwent rapid

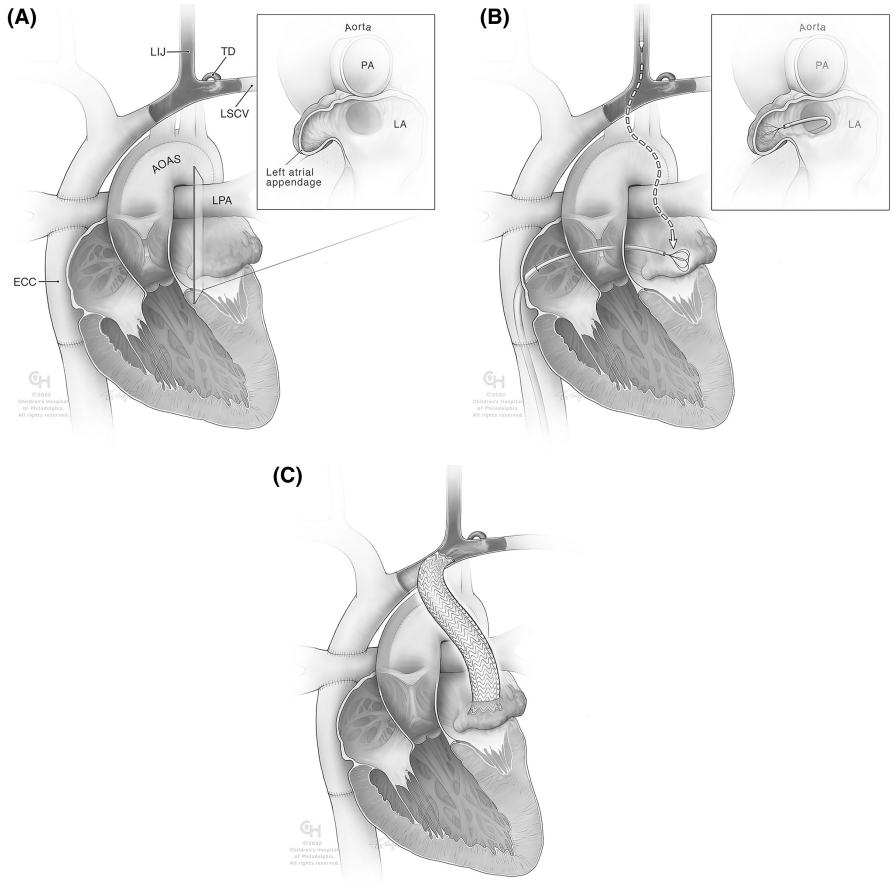



**FIGURE 1** Transcatheter thoracic duct decompression procedure. (A) The anatomy of an extracardiac Fontan. (B) Femoral venous access is obtained and a transeptal approach utilized to access the atrium, and a snare is placed near the left atrial appendage. Left internal jugular access is obtained and a transeptal needed is advanced along an extravascular course (dotted line) and enter the atrium where it is snared. (C) Completed stented pathway of the innominate vein to left atrium, allowing thoracic duct decompression. *Figure reprinted with permission from Wolters Kluwer.*

sequence induction. Anesthesia was maintained with sevoflurane. In addition to the three patients on inotropic support pre-procedure, three additional patients were started on dopamine during the procedure because of persistent hypotension.

Two patients developed supraventricular tachycardia, one terminated spontaneously while the other required adenosine. One patient had arterial oxygen desaturation to 40% due to a bronchial cast which was effectively removed with a suction catheter. One patient developed a small pericardial effusion after transeptal needle puncture but did not require intervention. Three patients required transcatheter fenestration closure for decreased oxygen saturation after TTDD. Eighty five percent of the patients were extubated at the end of the procedure. Anesthetic duration ranged from 4.3 to 9.3 h (median 6.7 h).

All but two patients after TTDD were admitted to the ICU for observation/management. The length of the ICU stay ranged from 0 to 59 days (median 2) and length of hospital stay ranged from 2 to 109 days (median 28) before being discharged.

## 4 | DISCUSSION

### 4.1 | Anesthetic considerations

#### 4.1.1 | Pre-procedure

Maintaining an optimal transpulmonary gradient to achieve adequate PBF and hence cardiac output is fundamental in the anesthetic management of Fontan patients. An adequate transpulmonary gradient is dependent on adequate systemic venous volume and pressure (preload), low pulmonary vascular resistance (PVR), atrio ventricular valve competency, sinus rhythm and a well-functioning single ventricle. Assessment of the intravascular volume status in Fontan patients with serous effusions, often treated with diuretic therapy and continuous catheter drainage, is critical. If a hypovolemic state is not corrected prior to, at the time of induction, and continuously throughout the procedure, decreased venous tone induced by anesthetics and increased resistance to PBF secondary to mechanical ventilation will result in hypotension.

Long term pulmonary vasodilator therapy needs to be continued.

Patients with ongoing drainage of ascites and pleural effusions lose a significant amount of protein resulting in hypoalbuminemia. Low level of plasma albumin results in increased free drugs which partially bind to proteins that in turn can result in greater potency of these drugs.[3]

#### 4.1.2 | Conduct of anesthesia

An essential component of anesthetic management is the prevention of patient movement that could dislodge precisely placed needles or catheters during imaging and intervention(s). Rapid sequence induction may be considered in the setting of a large, distended abdomen secondary to ascites.

Maintenance of low PVR is an important intraoperative goal. The pulmonary vasculature is reactive, and the PVR can change

**TABLE 1** Characteristic of Fontan circulation and lymphatic failure compartments.

| Number of patients = 20 | Count | Frequency (%) |
|---|---|---|
| Lymphatic failure characteristics | | |
| Compartment combinations | | |
| PLE + ascites | 5 | 25 |
| PLE + pleural effusion | 1 | 5 |
| PLE + pleural effusion + ascites | 10 | 50 |
| PLE + PB + pleural effusion | 1 | 5 |
| PLE + PB + pleural effusion + ascites | 3 | 15 |
| Previous lymphatic intervention | 18 | 90 |
| Fontan circulation | | |
| Fontan technique | | |
| Lateral tunnel | 6 | 30 |
| Extra-cardiac | 14 | 70 |
| Pre-procedure fenestration | 10 | 50 |
| Ventricular function | | |
| Normal | 11 | 55 |
| Mildly diminished | 3 | 30 |
| Moderately diminished | 3 | 15 |
| Atrioventricular valve regurgitation | | |
| Trivial | 9 | 45 |
| Mild | 9 | 45 |
| Moderate | 1 | 5 |
| Severe | 1 | 5 |
| Pre-procedure room air saturation | Median | Range |
| Fenestration present | 91 | 83–95 |
| No fenestration | 94 | 90–100 |

**TABLE 2** Conduct of anesthesia.

| Number of patients = 20 | Count | Frequency (%) |
|---|---|---|
| ASA status | | |
| 3 | 7 | 35 |
| 4 | 12 | 60 |
| 5 | 1 | 5 |
| Pre anesthetic sedation | | |
| None | 3 | 15 |
| Midazolam | 15 | 75 |
| Ketamine/Midazolam | 2 | 10 |
| Anesthesia induction drugs | | |
| Propofol | 1 | 5 |
| Ketamine | 6 | 30 |
| Ketamine/Propofol | 11 | 55 |
| Ketamine/Fentanyl/Dexmedetomidine | 1 | 5 |
| Sevoflurane | 1 | 5 |
| Access–arterial | | |
| Pre-procedure arterial | 5 | 25 |
| Arterial line placed for procedure | 1 | 5 |
| Inotropic support | | |
| Pre-procedure | 3 | 15 |
| Intra-procedure | 6 | 30 |
| Extubation at end of procedure | | |
| Yes | 17 | 85 |
| No | 3 | 15 |

abruptly. Factors which increase PVR include hypoxemia, hypercarbia, acidosis, hypothermia, and increased intrathoracic pressure. Overdistention of alveoli with excessive mean airway pressure and reduced lung volumes can increase PVR. Maintaining a low PVR with periods of apnea (sometimes required to facilitate the procedure) and suboptimal ventilation and/or oxygenation is challenging. PB is characterized by the formation of rubbery plugs or "casts" in the tracheobronchial tree that can acutely occlude the airway leading to rapid desaturation. Endotracheal suction and bronchoscopy need to be readily available to remove casts.

Frequent assessment of intravascular volume and acid/base status is crucial with prompt correction. Antiarrhythmic therapy should be available to treat an arrhythmia induced by wire or catheter manipulation as arrhythmias are poorly tolerated with the Fontan circulation.

### 4.1.3 | Post procedure

A decrease in arterial oxygen saturation is expected given the newly created right to left shunt. In some patients with an existing fenestration, the creation of an additional right to left shunt resulted in desaturation that was significant enough to warrant transcatheter fenestration device closure. In most patients, there was no need to continue mechanical ventilation on completion of the procedure and most were successfully extubated in the catheterization laboratory. Those who remained intubated had hemodynamic instability due to decreased function of the single ventricle with escalation of inotropic support.

Pain from the procedure is mild and managed with acetaminophen. All patients were started on anticoagulation, initially with enoxaparin and converted to warfarin. Length of stay was highly variable. In some, this was due to persistent lymphatic failure requiring monitoring and re-intervention. The most common reasons for return to the catheterization laboratory was to treat endoleaks (retrograde flow through the covered stent pathway into the innominate vein) or narrowing of the stented pathway. A total of nine patients required reintervention.

Not all patients in this small cohort were responders to the intervention. Reasons for the lack of improvement included narrowing of the stented pathway, occlusion of the TD and one patient had a second TD that had gone unrecognized during the initial procedure. Further investigation is needed in the five patients who failed to respond despite a patent pathway.

A total of five patients in this 20 patient cohort died from complications related to Fontan failure since undergoing TTDD, highlighting the fragility of this population. In many patients with Fontan and other end organ failure, transplant is not an option. Re-routing the TD drainage is the only option to improve physiology and quality of life. TTDD is currently the lowest risk option. While the procedure has shown promising results in some patients, we need to understand why others fail to improve. The need for repeat diagnostic imaging and re-intervention with general anesthesia in this high-risk patient population should be recognized.

## 5 | FUTURE CONSIDERATIONS

Despite improved survivability for single ventricle patients over the last five decades, patients with Fontan circulation present unique long-term comorbidities. While mechanisms for potential etiologies of lymphatic failure have been identified, management and treatment of long-term sequelae is ongoing. Given the high surgical risk in this population, advances with catheter-based interventions of complex cardiovascular problems is desirable. It is imperative that the anesthesiologist understand the procedural implications, intraoperative management of failing-Fontan physiology and recognize the need for repeat intervention. Care by or consultation with a pediatric cardiac anesthesiologist could be beneficial, as they are familiar with these patients.

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### DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available from the corresponding author upon reasonable request.

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