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CASE REPORT

ADVANCED

CLINICAL CASE SERIES

Percutaneous Venous Reconstruction for Central Thrombosis-Associated Chylothorax

A Safe and Efficacious Option

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ABSTRACT

Central thrombosis–associated chylothorax is underrecognized in children and frequently refractory to conservative management. Central venous catheterizations are the predominate cause. We present 3 cases highlighting endovascular techniques used to treat persistent chylous effusions. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2021;3:1569–1575) © 2021 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

INTRODUCTION

Chylous effusions manifest as a "milky" exudate with high triglyceride content and are associated with respiratory, nutritional, hematologic, or immunologic dysfunction (1,2). Postoperative chylothorax is associated with longer hospital stay and increased risk of in-hospital mortality (3).

Chylothorax after cardiac surgery in children has an incidence of 4% to 9% (2,4). Causes include iatrogenic trauma of the thoracic duct and its tributaries, increased central venous pressures after cavopulmonary anastomosis, or impaired drainage of the duct secondary to central venous thrombosis (CVT) (3). Many reports have suggested a relationship between CVT and chylothorax, although this is still largely underrecognized (5). Children with central thrombosis-associated chylothorax (CTaC) tend to have worse outcomes (3).

The role of interventional recanalization of thrombosed central veins for treating CTaC is underreported (6), despite recommendations for aggressive treatment of CVT in such patients (4). We present a series of 3 pediatric cases of refractory CTaC

LEARNING OBJECTIVES

- To be able to make the diagnosis of CTaC by performing high-quality cross-sectional imaging, especially CT venography.
- To understand the role of endovascular revascularization techniques to re-establish flow in the left subclavian vein, brachiocephalic vein, and SVC that can lead to relief of chylothorax by reducing back-pressure on the thoracic duct.

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ABBREVIATIONS AND ACRONYMS

CT = computed tomography

CTaC = central thrombosis–associated chylothorax

CVT = central vein thrombosis

IVC = inferior vena cava

MCT = medium-chain triglyceride

MPA = main pulmonary artery

MRI = magnetic resonance imaging

RPA = right pulmonary artery

SVC = superior vena cava

TPN = total parenteral nutrition

successfully treated by interventional recanalization of thrombosed central veins.

The Institutional Review Board of Evelina London Children's Hospital deemed the anonymized retrospective case series review to be exempt from requiring formal approval.

CASE SERIES

PATIENT 1. An 8-year-old girl with end-stage kidney disease (unclassified multisystemic inflammatory disease) had a 4-year history of peritoneal dialysis and hemodialysis, complicated by infection and catheter malfunction. After the sixth hemodialysis catheter insertion, she presented with superior vena cava (SVC) obstruction syndrome and a left pleural effusion (**Figure 1**), and she subsequently developed bilateral chylous effusions. After bilateral chest drains, she switched to a medium-chain triglyceride (MCT) diet and parenteral nutrition supplementation. Later, she had a pulseless electrical activity cardiac arrest resulting from chylous pericardial tamponade and became ventilator dependent for respiratory support. While ventilated, she developed a dialysis catheter-associated right atrial thrombus. The catheter was removed, she underwent anticoagulation, and she was switched to peritoneal dialysis. At thoracoscopy, the thoracic duct could not be seen, and she underwent pleurodesis, but this failed to resolve the chylous effusions. Computed tomography (CT) venography demonstrated thrombosis of both brachiocephalic veins and the SVC. Magnetic resonance imaging (MRI) lymphangiography did not identify lymphatic leakage. After 2 months of respiratory ventilatory support, complicated by a plastic bronchitis syndrome, she underwent interventional recanalization of the left subclavian and brachiocephalic veins, SVC venoplasty and stenting (**Figures 2A, 2B, and 3A to 3G**), and catheter-directed thrombolysis (Videos 1, 2, 3, 4, 5, and 6). The chest drains were removed 3 days after the procedure. Warfarin administration and peritoneal dialysis were continued. A low-fat diet and parenteral nutrition were continued for 6 weeks, after which the patient resumed regular oral feeding.

The child underwent live donor kidney transplantation 6 months after recanalization and is doing well at 24-month follow-up.

PATIENT 2. A female child was born following antenatal diagnosis of a severely hypoplastic transverse aortic arch and coarctation of the aorta, with postnatal confirmation of an abnormal origin of the right pulmonary artery (RPA) from the ascending aorta and a large persistent arterial duct. Extended aortic arch repair and RPA reimplantation to the main pulmonary artery (MPA) were undertaken at 5 days of age. Eight days later, she developed both a right-sided chylous pleural effusion and a pericardial effusion. Total parenteral nutrition (TPN) was started; however, the pleural and pericardial fluid persisted. CT scan demonstrated thrombotic occlusion of the left subclavian and innominate veins and SVC and stenosis of the reimplanted RPA at its anastomosis with the MPA. She was heparinized, a surgical pleuropericardial window was created, and a right-pleural drain was placed, draining chylous fluid.

At 42 days, the child underwent catheter-directed thrombolysis, followed by recanalization and balloon venoplasty of the thrombosed left axillary, subclavian, and innominate veins and the SVC, as well as stenting of the occluded reimplanted RPA (**Figures 4A and 4B**). Anticoagulation with heparin and subsequently dalteparin was continued. Pleural and pericardial effusions reduced dramatically, and on day 3 after the procedure, the drains were removed.

At follow-up 2 years later, the child was thriving. She needed repeat intervention for in-stent stenosis of the RPA stent, but none for the central veins, and no chylous pleural or pericardial effusions recurred.

PATIENT 3. A $5^{1}/_{2}$-year-old female child, born with a hypoplastic left heart and abnormal pulmonary vein anatomy, underwent the hybrid procedure in the neonatal period, consisting of surgical bilateral pulmonary artery bands and transcatheter stenting of the arterial duct. Before proceeding to the Norwood operation, she underwent atrial septectomy, surgical diaphragm plication, left pulmonary vein stenosis repair, adjustment of pulmonary artery bands and stent placement in the left pulmonary vein. Subsequently, she had a Norwood procedure, a hemi-Fontan operation, and a left pulmonary vein procedure, and, finally, fenestrated lateral tunnel Fontan completion at the age of 4 years.

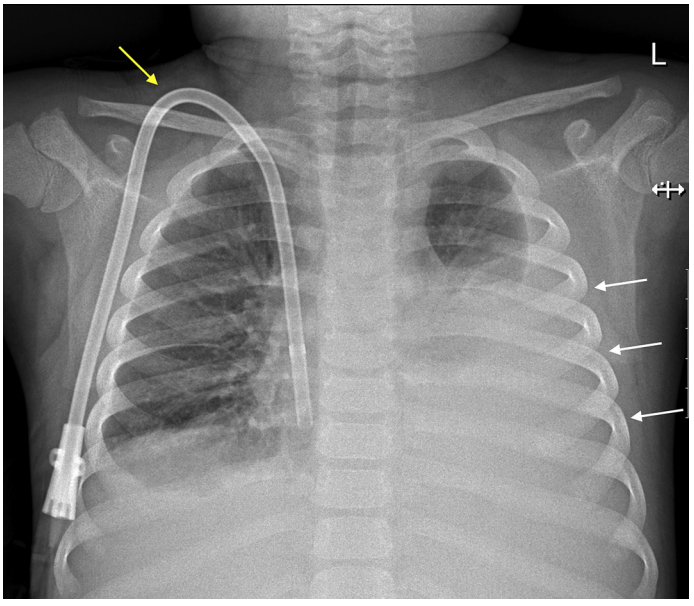
Following the Fontan operation, she had recurrent ascites, which was managed conservatively with angiotensin-converting enzyme inhibitors and sildenafil. A year later, she developed a large right chylous pleural effusion. CT demonstrated high-grade stenoses of the left brachiocephalic and subclavian veins and of the infrarenal inferior vena cava (IVC) (**Figures 5A to 5D**). After an MCT diet, TPN was commenced because of persistent chylothorax.

The child underwent recanalization and venoplasty of the left innominate and subclavian veins, with a high-pressure, scoring, and cutting balloon (**Figures 5A to 5D**) (Videos 7, 8, and 9). Balloon venoplasty of the IVC was also performed to assist with through-and-through access for the central venoplasty. The child was anticoagulated with unfractionated heparin and then warfarin and aspirin. She was placed again on an MCT diet.

The chylothorax resolved. However, there were slow reaccumulations that required intermittent drainage over the subsequent 3 months. MRI lymphangiography did not reveal any leak, but a large network of collateral lymphatic channels was demonstrated in the left supraclavicular fossa, with probable obstruction of the thoracic duct drainage into the left subclavian vein.

Percutaneous transvenous retrograde recanalization and balloon dilatation of the thoracic duct were performed, successfully establishing good flow from the thoracic duct into the left subclavian vein (**Figures 6A to 6G**). Thereafter, stable appearances of the mild right pleural effusion were noted, with no need for further chest drainage during 18 months of follow-up.

FIGURE 1 **Case 1: Presentation Chest Radiograph**

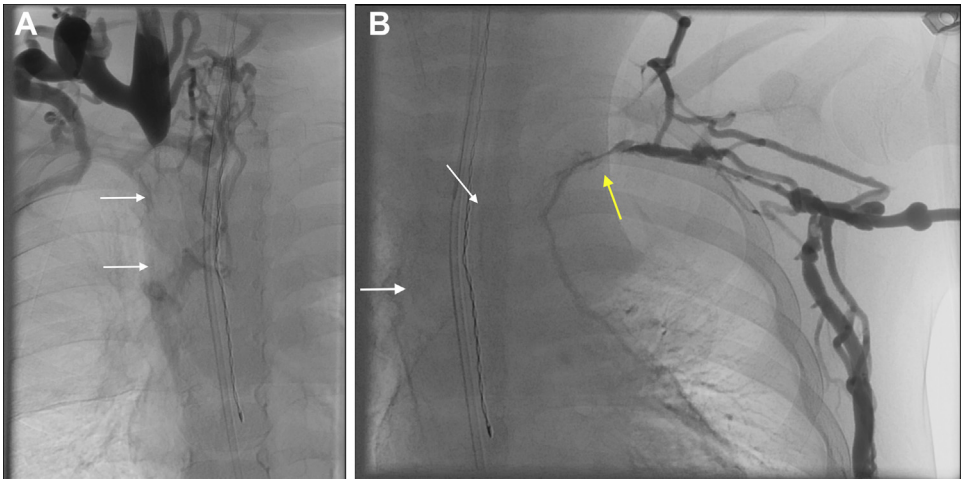


Left chylothorax **(white arrows)** and right Permcath (Medtronic) on presentation **(yellow arrow).**

DISCUSSION

Injury or obstruction of the thoracic duct may lead to chylothorax, which may be associated with significant morbidity, prolonged hospital stay, and increased mortality (1). CTaC is considered rare, and in children it often relates to use of central venous catheters and an underlying procoagulant state resulting from surgery, malignant disease, sepsis, or nephrotic syndrome (3).

FIGURE 2 **Case 1: Prereconstruction Venograms**



Superior vena cava and brachiocephalic vein occlusion **(white arrows)** and high-grade left subclavian vein stenosis **(yellow arrow)** on **(A)** right internal jugular and **(B)** left basilic venograms.

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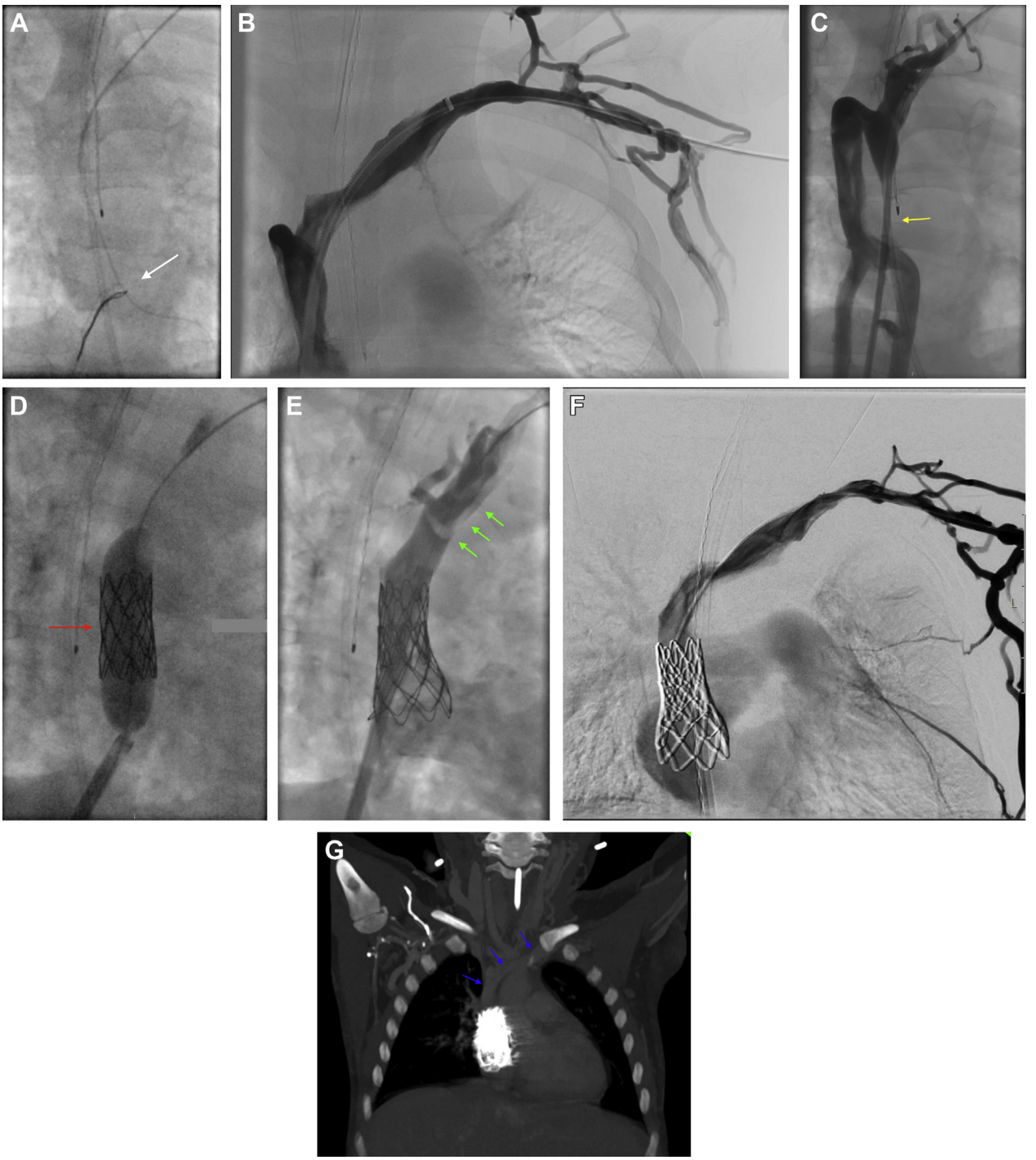
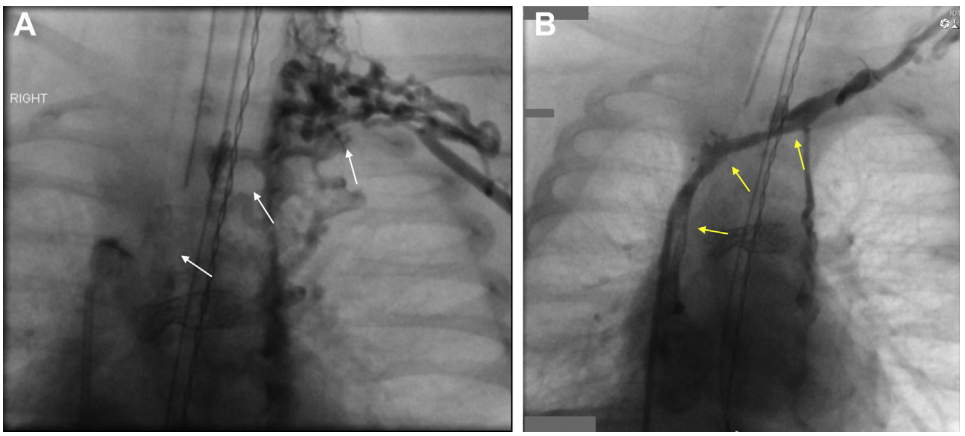
FIGURE 3 **Case 1: Venous Reconstruction**



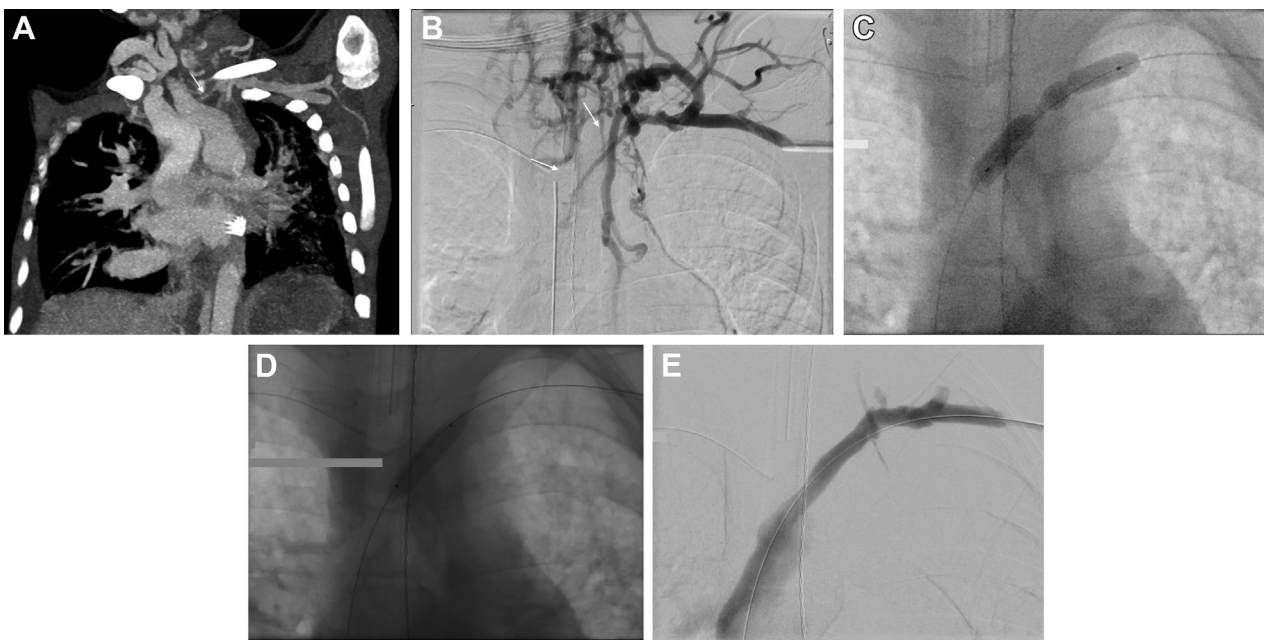
The occluded segment was crossed, **(A)** hydrophilic wire (Radifocus, Terumo) snared in the right atrium (Amplatz Goose Neck snare, Medtronic) **(white arrow),** and through-and-through access established. Superior vena cava, left brachiocephalic vein, and superior vena cava dilated with a scoring balloon (Enforcer, Cook Medical) resulting in **(B)** improved appearances. **(C)** Recoil of cavoatrial junction stenosis **(yellow arrow)**, **(D)** treated with a bare CP Stent (NuMED Inc; BVM) **(red arrow)**. **(E)** Non-flow-limiting thrombus in the left brachiocephalic vein **(green arrows),** cleared with catheter-directed thrombolysis (Cragg-McNamara, Medtronic) for 48 hours. **(F)** In-line flow after catheter-directed thrombolysis. **(G)** Patent left brachiocephalic vein and superior vena cava on 4-month computed tomography venography **(blue arrows).** Echocardiogram confirmed CP Stent patency.

FIGURE 4 **Case 2: Prevenous and Postvenous Reconstruction**



(A) Occluded left brachiocephalic vein and superior vena cava **(white arrows)** and extensive collateral veins. Crossed from below, compliant balloon (18LP, Cook Medical) dilatation and **(B)** in-line flow **(yellow arrows).**

FIGURE 5 **Case 3: Venous Reconstruction**



(A) Left brachiocephalic vein stenosis **(white arrows)** occluded at superior vena cava confluence (computed tomography venography) confirmed on **(B)** left arm. Crossed with a CTO wire (Asahi Gaia Second, Asahi Intecc USA, Inc) using a pigtail catheter in the superior vena cava as a target and guidewire snared from below. **(C)** Lesion resistant to a compliant balloon and **(D)** treated with a cutting balloon (Boston Scientific). **(E)** In-line flow established.

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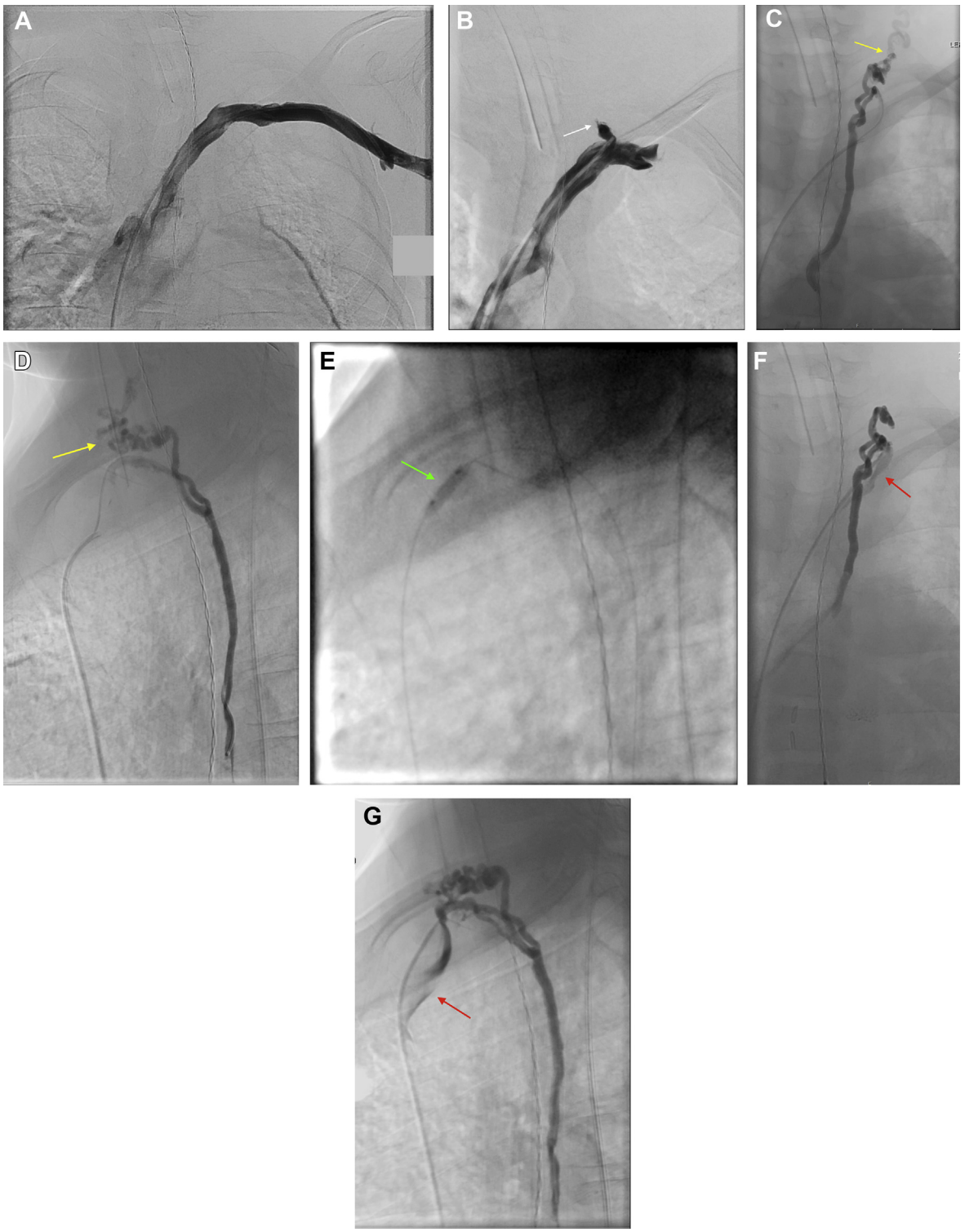
FIGURE 6 **Case 3: Thoracic Duct Recanalization**



(A) In-line flow maintained at 4 months and **(B)** thoracic duct occluded **(white arrow)**. Retrograde transvenous cannulation of the thoracic duct performed and a large collateral lymphatic network **(yellow arrows)** demonstrated in the supraclavicular fossa on **(C)** frontal and **(D)** lateral projections. **(E)** Thoracic duct confluence dilated with 2.5 × 12 mm 0.014-inch balloon **(green arrow)**. **(F and G)** Filling of the superior vena cava with some collateral lymphatic filling **(red arrows)**.

CVT causes venous hypertension with disruption of the thoracic duct anatomy and function. The causal relationship of CVT with the development of chylothorax was first documented in animal models in 1936. However, CVT does not always result in chylothorax because of collateral lymphatic channels (5,6).

Conservative treatment consists of an MCT diet with or without parenteral nutrition with or without octreotide. These measures aim to reduce lymphatic production and allow recovery of the thoracic duct system. However, CTaC is associated with prolonged and high-volume accumulation of chylous fluid, frequently requiring intervention (7). The optimal time for proceeding to intervention is not clearly defined. In our experience, approximately 4 to 6 weeks of conservative management is usually undertaken before contemplating intervention.

Thoracic duct ligation with video- assisted thoracoscopic surgery is considered the surgical treatment of choice for recurrent chylothorax (8). Following thoracic duct ligation, the lymphatic fluid is diverted, leading to resolution of the chylothorax. However, in CTaC, establishing central venous patency allows for natural drainage and flow of the thoracic duct. Percutaneous central venous reconstruction is minimally invasive and better tolerated in this patient cohort.

Anticoagulation is recommended and may occasionally be effective alone in treatment of CVT-related chylothorax (5). Kumar et al (9) proposed a technique involving open surgical thrombectomy and reconstruction of the SVC and innominate vein for treatment of CVT-associated chylothorax in infants.

In this series, all 3 patients failed to improve with anticoagulation and nutritional management alone. The subsequent radiologic treatment plans were tailored to the clinical condition and involved anticoagulation, catheter-directed thrombolysis, repeated venoplasty, stent insertion, and thoracic duct dilation. There were no associated complications. Case reports from other authors have described similar positive results with central venous recanalization for treatment of chylothorax (6).

CONCLUSIONS

CTaC is frequently refractory to conservative management. Anticoagulation and endovascular recanalization of the thrombosed central venous system offer minimally invasive, effective, and safe treatment and should be considered before surgical intervention.

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KEY WORDS central line, central vein obstruction, chylothorax, chylous effusion, endovenous reconstruction

APPENDIX For supplemental videos, please see the online version of this paper.