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CASE REPORT
Congenital heart disease

Case report of a Gore-Tex TCPC conduit dissection causing severe stenosis

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Background The patient is a 15-year-old male with situs inversus, dextrocardia, bilateral superior caval veins, atrioventricular discordance with a single outlet, large perimembranous ventricular septal defect, aortic override, pulmonary atresia, and right aortic arch. The complex anatomy with a Ventricular Septal Defect (VSD) distant from the aorta (unsuitable for baffling to the aorta) meant he was unsuitable for biventricular repair and proceeded down a univentricular palliation pathway.

Case summary Post-total cavopulmonary connection his clinical course was uneventful until the age of 5 when he developed fatigability with desaturation. An accessory hepatic vein was surgically banded with improved saturations and exercise tolerance. At the age of 15, cardiovascular magnetic resonance (CMR) was performed to investigate borderline saturations and as work-up for transition to adult services. Cardiovascular magnetic resonance and cardiac computed tomography (CT) imaging demonstrated an eccentric thrombus causing stenosis of the extracardiac conduit and a thrombus outside of the lumen contained by the thin outer membrane of the Gore-Tex conduit. Collateralization suggested this was longstanding. Cardiac catheterization demonstrated a 4 mm × 6 mm stenosis at the junction of the conduit with the pulmonary arteries. The region was successfully balloon dilated and stented with a 34 mm-long Cheatham Platinum stent, with no complications.

Discussion To date, this is the first documented case of a dissecting thrombus of a Gore-Tex graft in the literature. This case emphasizes the need for anticoagulation and serial cross-sectional imaging (CT or CMR) in Fontan patients with prosthetic grafts throughout a patients' lifetime.

Keywords TCPC • Fontan • Dissection • Thrombus • Gore-Tex • Case report

Learning points

- Gore-Tex grafts are also at risk of thrombus formation and dissection.
- Significant thrombosis and dissection can occur in asymptomatic patients.
- The risk of dissection may increase with age, somatic growth, and stretching of the conduit.
- There is a need for ongoing cross-sectional screening of prosthetic grafts in throughout a patients' lifetime.

Introduction

It is suggested that the total cavopulmonary connection (TCPC) with an extracardiac conduit (ECC) provides optimal flow dynamics, whilst minimizing the complications associated with other surgical techniques.[1]

The routine use of Dacron conduits has been abandoned due to the relatively high incidence of stenosis secondary to neointimal formation.[2] In the current era, Gore-Tex (polytetrafluoroethylene, W. L. Gore and Assoc, Flagstaff, AZ, USA) is a preferred material. The disadvantages of Gore-Tex are thrombogenicity and a lack of somatic

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growth potential. The advantages are minimal intimal formation, resistance to calcification and easy manipulation.[3]

All implanted prosthetic material will be subjected to shear stress, stretching, and some degree of neointimal formation. In the literature there is a lack of long-term data regarding the performance of the ECC[4] and the role of the conduit in Fontan failure.

Whilst stenosis of Gore-Tex conduits (which have been successfully stented) is described,[4] to the best of our knowledge, there are no previous reports of a dissection between the Gore-Tex layers of the tube causing stenosis of the ECC. Such a finding is important due to the increasing numbers of patients palliated with a Gore-Tex ECC Fontan who will require lifetime surveillance.

Timeline

Age	Event
Neonate	Modified right and left Blalock-Taussig (BT) shunt
Infant	Bilateral bidirectional cavopulmonary anastomosis with augmentation of central pulmonary arteries
2 years	Division of previous shunts and Atrial Septal Defect (ASD) creation
4 years	Total cavopulmonary connection (TCPC) completion with an extracardiac non-fenestrated 18 mm Gore-Tex conduit and a pulmonary artery plasty
5 years	Desaturating, fatigability, and mild exercise intolerance
5 years	Cardiac computed tomography (CT) demonstrated an unobstructed TCPC conduit, with accessory hepatic vein suspected as the reason for desaturation
6 years	Banding of accessory right hepatic vein
15 years	Cardiovascular magnetic resonance demonstrated an incidental conduit dissection with thrombus causing severe stenosis with venous collateralization
15 years and 10 months	Cardiac CT confirmed luminal thrombus and dissection contained within the outer layer of Gore-Tex membrane
15 years and 10 months	Discussed in cardiothoracic meeting with consensus for covered stent of stenotic region ± occlusion of collaterals
15 years and 11 months	Interventional cardiac catheter, angioplasty of stenosis, placement of 34 mm Cheatham Platinum stent, veno-venous collateral occlusion with Amplatzer Vascular Plug II device. and M Reye coils
16 years	No complications at clinical follow-up 2 months post-procedure

Case presentation

The case describes a 15-year-old male with situs inversus, dextrocardia, bilateral superior caval veins, atrioventricular discordance with a single outlet, large perimembranous ventricular septal defect, aortic override, pulmonary atresia, and right aortic arch. The complex anatomy with a VSD distant from the aorta (unsuitable for intracardiac baffling) meant biventricular repair was not possible and he proceeded down a univentricular palliation pathway.

Eleven years after a TCPC, cardiovascular magnetic resonance (CMR) demonstrated incidental, severe stenosis of the Gore-Tex ECC, secondary to thrombus formation and subsequent dissection.

The patient previously underwent modified right and left Blalock-Taussig (BT) shunt in the neonatal period, bilateral bidirectional cavopulmonary anastomosis, augmentation of central pulmonary arteries and a TCPC with an extracardiac, non-fenestrated, 18 mm Gore-Tex conduit, and a pulmonary artery plasty at the level of the left Glenn. The cardiac anatomy required the anastomosis of the Gore-Tex conduit obliquely at the pulmonary artery end. An accessory right hepatic vein draining directly to the atrium was left (to function as a small fenestration). He was maintained on aspirin.

At the age of 5, he developed fatigability with desaturation. Cardiac computed tomography (CT) suggested veno-venous shunting was causative. The accessory hepatic vein was banded. This improved exercise tolerance and saturation.

At the age of 15, the patient was generally asymptomatic, with mild effort intolerance and lower but acceptable saturations (91%). His clinical examination, electrocardiogram, chest X-ray, and echocardiogram were unremarkable. His liver function was normal (see *Table 1*). Cardiovascular magnetic resonance was performed to investigate this and as part of his workup for transition to adult services. Cardiovascular magnetic resonance demonstrated a dissection of the extracardiac tunnel starting ∼17 mm above the liver reaching above the level of the pulmonary arteries (see *Figure 1*). The narrowest dimension of the tunnel measured 7 mm × 9 mm just before joining the pulmonary branches and was additionally compressed by suspected thrombus. There was a large tortuous vein noted to the left of the extracardiac tunnel. This was thought to decompress the inferior vena cava (IVC) and draining into left pulmonary veins. Although the exact entry could not be identified, the suggestion that it has arisen was due to a calculated increased flow in the left pulmonary veins when compared to left pulmonary artery (net forward flow 38 vs. 22 mL/beat). The patient refused cannulation therefore contrast angiography showing the anatomy in more detail was not possible.

An urgent CT (see *Figure 2*) confirmed eccentric thrombus narrowing the conduit, but more significantly, organized thrombus outside of the conduit lumen, confined to the shape of the conduit as demonstrated on the previous CT. This was presumed to represent prior rupture of the Gore-Tex conduit lining with containment via its thin outer membrane with subsequent thrombosis. The lumen of the TCPC conduit was reduced to 4 mm × 3 mm and an enlarged phrenic vein demonstrated in keeping with collateralization. There was no enhancement of the 'false lumen' to suggest active leak and the appearance was considered long standing (with collateralization noted).

He was discussed in our cardiothoracic meeting with consensus to proceed to cardiac catheter and stenting of the stenotic region.

At cardiac catheter, haemodynamic assessment showed acceptable Fontan pressures (see *Table 2*), likely due to the large decompressing collateral. The angiogram (see *Figure 3*) demonstrated a tight distal stenosis (4 mm × 6 mm) at the junction of the conduit with

Table 1 Normal liver function tests before and after interventional catheterization

Liver function tests	20 October 2015 (pre-catheter)	5 January 2021 (post-catheter)
Total bilirubin (µmol/L) (0–20)	5	19
ALP (U/L) (60–425)	272	218
ALT (IU/L) (8–40)	28	22
AST (IU/L) (<50)	34	28
Total protein (g/L) (60–80)	77	76
Albumin (g/L) (30–50)	44	41

Units and ranges included.
ALP, Alkaline Phosphatase; AST, Aspartate Aminotransferase.

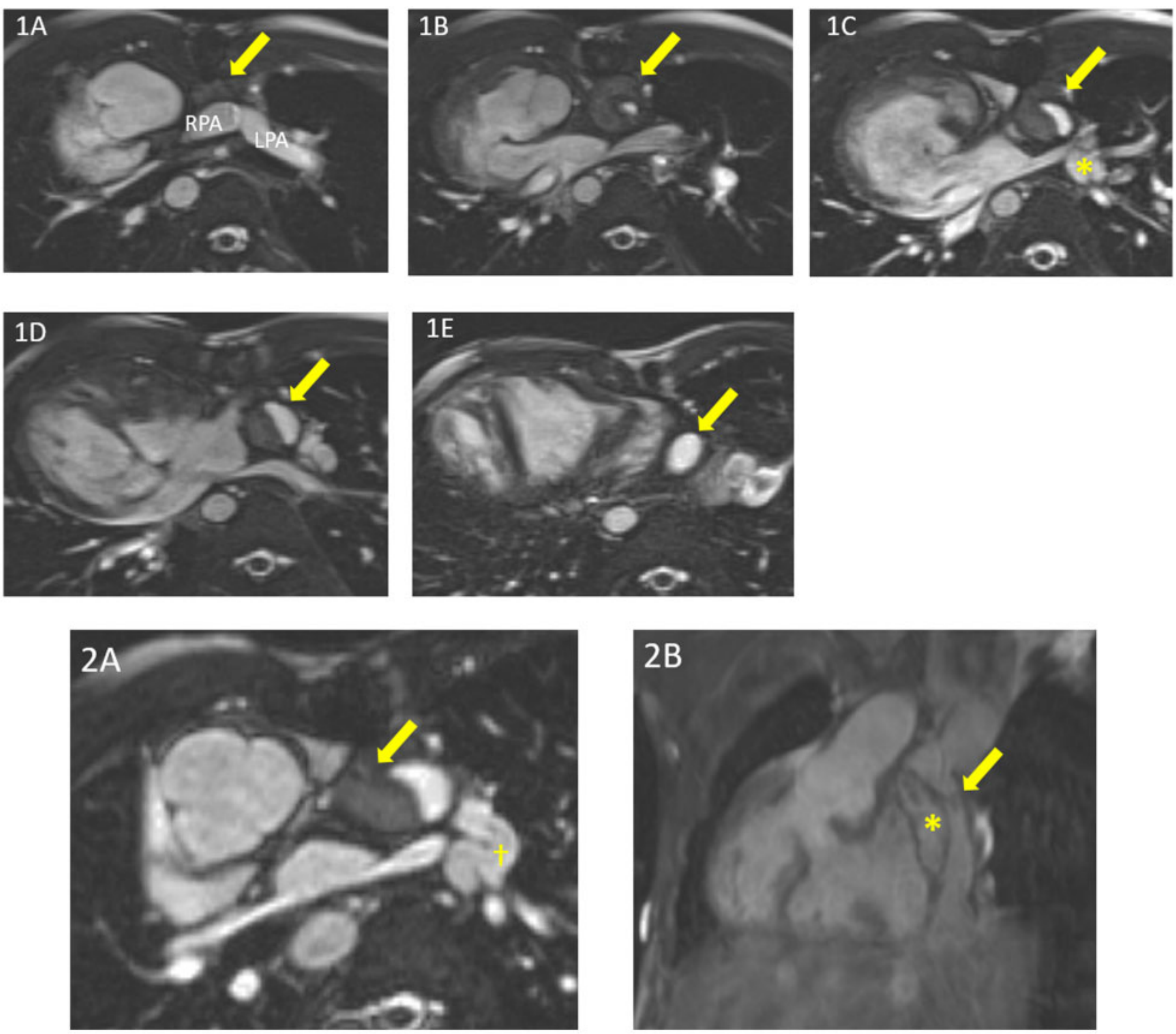

Figure 1 Transaxial cardiovascular magnetic resonance cines showing the extracardiac conduit (ECC, yellow arrow) in superior to inferior direction. (1*A*) A mass above the level of the pulmonary arteries; (1*B*) severely stenotic conduit compressed by a rounded mass; (1*C* and 1*D*) the lumen of the ECC is larger; and (1*E*) inferior vena cava of normal appearance. Asterisk: veno-venous collateral to the left and posterior to the ECC. LPA, left pulmonary artery; RPA, right pulmonary artery. (2*A*) Mass at the level of the pulmonary arteries compressing the conduit. †Veno-venous collateral to the left and posterior to the ECC. (2*B*) Coronal oblique slice demonstrating narrowing of the total cavopulmonary connection lumen with thrombus both inside (arrow) and outside (*) the internal conduit layer.

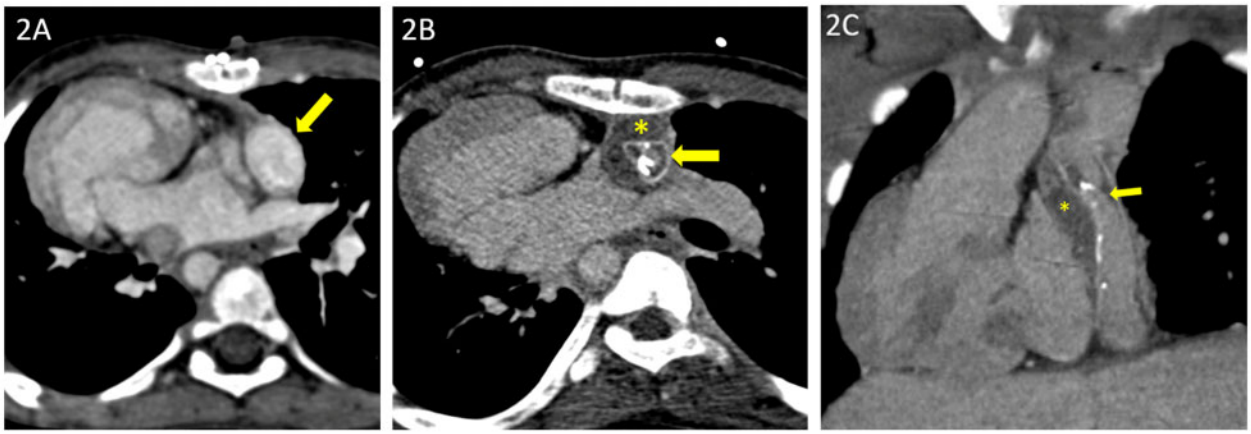

Figure 2 Cardiac computed tomography. Delayed post-contrast computed tomography sections following total cavopulmonary connection demonstrating (*A*) homogeneous opacification of the extracardiac total cavopulmonary connection conduit (arrow). (*B*) On follow-up there is low attenuation thrombus surrounding the conduit (*) with separation of the layers of conduit material and compression of the lumen. Further intraluminal thrombus is also demonstrated (arrow). (*C*) Coronal oblique computed tomography reconstruction demonstrating narrowing of the total cavopulmonary connection lumen with thrombus both inside (arrow) and outside (*) the internal conduit layer.

Table 2 Total cavopulmonary connection haemodynamic and oximetry data

Site	Pressure S/D/M (mmHg)	Oximetry
RSVC	14 mean	77.9
LSVC	17 mean	72
IVC	20 mean	
RPA	12/13/2012	71.7
LPA	15/14/14	74.4
Asc Ao		91
LVEDP	11	

Asc Ao, ascending aorta; IVC, inferior vena cava; LPA, left pulmonary artery; LSVC, left superior vena cava; LVEDP, left ventricular end-diastolic pressure; RPA, right pulmonary artery; RSVC, right superior vena cava; S/D/M, systolic/diastolic/mean.

pulmonary arteries. In anterior–posterior and lateral projections, the conduit measured 12 and 19 mm, respectively. There was significant dilation of the hepatic veins. There was one right hepatic vein draining into the right-sided atrium. There was a large haemodynamically significant collateral emerging from the IVC that divided into a paravertebral collateral and a large, long, and tortuous collateral draining into the left pulmonary veins. The distal Fontan conduit stenosis was progressively dilated with 8 and 10 mm × 20 mm Conquest Balloon and 12 mm × 40 mm Atlas Gold Balloons. The conduit was stented with a 34 mm-long Cheatham Platinum stent over a 14 mm × 40 mm BIB balloon. The stent was then further dilated with a 16 mm × 40 mm Atlas Gold balloon. The subsequent angiogram showed no gradient between the IVC and pulmonary arteries. The large haemodynamically significant collateral from the IVC was successfully occluded with a 12 mm Amplatzer Vascular Plug 2 and two 15 mm × 15 mm MReye coils.

The patient was discharged the next day on aspirin and clopidogrel. At clinic follow-up, 2 months later, the patient was well with improved exercise tolerance and saturations of 92%.

Discussion

Conduits can be made from pericardial tissue, human or bovine vessels, or synthetic materials such as Dacron or Gore-Tex. There are variable data on the optimal choice of conduit material and its durability, especially in the paediatric setting.[5]

Dissection has been described with Dacron conduits.[6,7] The use of Dacron conduits in the Fontan circulation has now been abandoned due to the higher incidence of stenosis and thick neointimal peel formation.[4]

The main problem with Contegra conduits is stenosis, secondary to inadequate graft preparation, kinking/distortion, calcification, or surgical technique.[5] Dissection in Contegra conduits used in the right ventricular outflow tract position has been reported.[5,6,8,9] The hypothesized mechanism is secondary to a chronic inflammatory process causing platelet-fibrin deposits, neointimal proliferation, calcification, and distortion. This in turn increases shear stress and endothelial disruption, separating the inner peel/neointima from the conduit wall by a dissecting haematoma/thrombus.[3,5]

Gore-Tex grafts are used for ECCs as the material is easy to manipulate, does not readily form an intimal peel, and is resistant to calcification.[3] However, there are concerns regarding the lack of somatic growth and the longevity of the material.[3] A case series of 33 patients (Ochiai, 2009) suggested Gore-Tex conduits are safe in the medium term with no structural or haemodynamic problems.[3] Hagler *et al.*[4] demonstrated that Gore-Tex ECC stenosis is rare, with only 20/233 patients needing intervention over a 16-year period. The study suggested that a major mechanism of the stenosis is the stretch

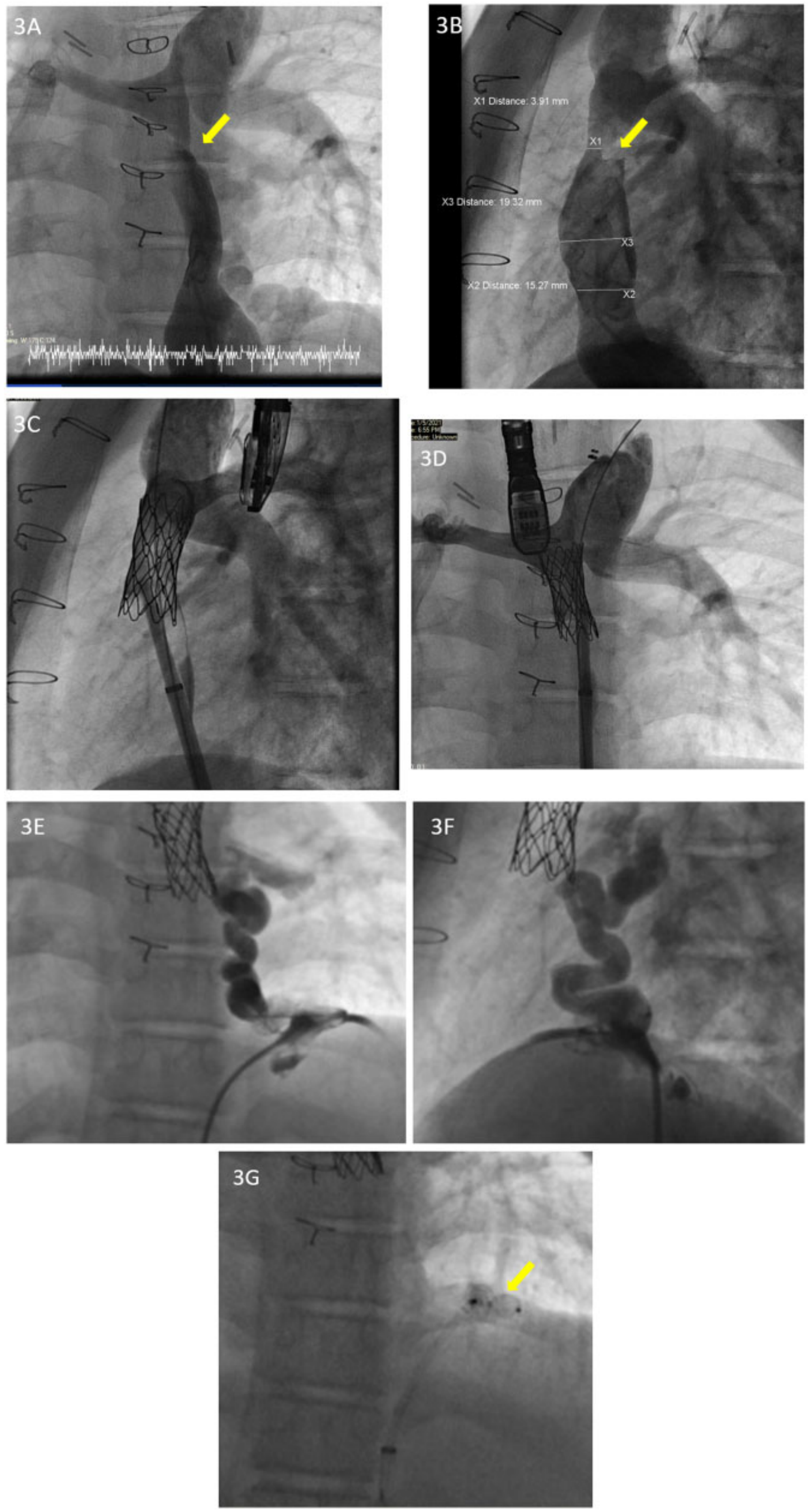

Figure 3 Angiography in anterior–posterior and lateral views. (*A and B*) Demonstrates a proximal filling defect in the total cavopulmonary connection conduit, with a lumen of 3.9 mm at the narrowest point. (*C and D*) Angiogram post-stent deployment demonstrating good opacification of the branch pulmonary arteries. (*E and F*) Demonstrates the large tortuous venous collateral. (*G*) Closed with a 12 mm Amplatzer Vascular Plug (AVP II) (arrow), and two 15 mm × 15 mm M Reye coils.

associated with somatic growth. Gore-Tex stretchable conduits have been developed to address this problem.[4]

There is little data on the long-term follow-up of the ECC used in smaller Fontan patients. Concerns remain regarding the distortion of the pulmonary artery during the TCPC, causing flow distortions in the ECC. In our case, the left pulmonary artery was stenotic and required extensive reconstruction. Furthermore, the Gore-Tex conduit was anastomosed obliquely to this reconstructed region. Over time and with somatic growth these factors may have altered the shear stresses and haemodynamics of this region promoting neointimal formation, stenosis, and a nidus for dissection.

The diameter of the conduit has important implications for kinetic energy loss and thereby thrombus formation. Extracardiac conduits >20% of the IVC diameter should not be used[3] and in our case an 18 mm conduit was an appropriate choice.

The data highlighted above emphasize the need for anticoagulation and serial cross-sectional imaging (CT or CMR) in Fontan patients. It has been recommended that Gore-Tex ECCs should be assessed at 10 years after implantation and then periodically after that.[4] In our case, the stenosis and dissection were detected 11 years after Fontan completion.

Fortunately, in this case, the stenosis and dissection had no severe adverse clinical consequences at follow-up. However, this important finding highlights the need for ongoing vigilance with prosthetic grafts throughout a patients' lifetime.

Lead author biography

Oliver Bates is a trainee in paediatric cardiology, London, UK.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

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Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The authors confirm that written consent for the submission and publication of this case, including images, has been obtained from the patient and his parents in line with COPE guidance.

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