ESC European Society of Cardiology

European Heart Journal - Case Reports (2024) **8**, ytae186
https://doi.org/10.1093/ehjcr/ytae186

CASE REPORT
Cardiovascular surgery

The elephant's Achilles' heel: a case report of acute obstruction of frozen elephant trunk after proximal aortic dissection repair

Stephan Nienaber [1], Xun Yuan [2], Susanna Price [2], Ulrich Rosendahl[2,3], and Christoph A. Nienaber [2]*

[1]University of Cologne, Faculty of Medicine and University Hospital Cologne, Clinic III for Internal Medicine, Cologne, Germany; [2]Department of Cardiology and Aortic Centre, Guy's and St. Thomas' Hospitals NHS Trust, Royal Brompton and Harefield Hospitals, London SW3 6NP, UK; and [3]Department of Cardiac Surgery, Guy's and St. Thomas' Hospitals NHS Trust, Royal Brompton and Harefield Hospitals, London, UK

Received 21 December 2023; revised 27 March 2024; accepted 8 April 2024; online publish-ahead-of-print 15 April 2024

Background The frozen elephant trunk (FET) technique as a hybrid combining surgical and endovascular repair is an emerging concept to treat complex aortic dissection. Early experience showed technical feasibility and promising clinical outcomes. However, unsuspected complications still arise.

Case summary A 25-year-old male presented to the emergency department with a 2-day history of chest pain. After exclusion of acute coronary syndrome, a computed tomography angiography (CTA) revealed Type A (DeBakey Type I) aortic dissection. The patient underwent median stenotomy for complete replacement of the ascending aorta, the aortic arch, and FET. Early after rewarming, the patient became unstable due to severe left ventricular dysfunction. Soon veno-arterial extracorporal membrane oxygenation (VA-ECMO) was required for circulatory support. The cause of deterioration remained unclear until repeated CTA showed acute obstruction of the FET. Invasive exploration confirmed a trans-FET gradient of 100 mmHg, successfully managed by repeated balloon inflation with resolution of both obstruction and gradient. The patient recovered completely without any sequela.

Discussion While the mechanism of acute obstruction after FET remains subject to speculation, the rescue intervention of ballooning the obliteration on VA-ECMO was life-saving. Intraoperative ultrasound and videoscopic inspection may be instrumental before chest closure to avoid such critical events.

Keywords Frozen elephant trunk • Pseudo-coarctation • Balloon dilatation • Type A aortic dissection • Thoraflex Hybrid prosthesis • Case report

ESC curriculum 9.1 Aortic disease • 7.1 Haemodynamic instability • 7.5 Cardiac surgery • 7.4 Percutaneous cardiovascular post-procedure • 6.4 Acute heart failure

* Corresponding author. Tel: +44 (0) 207 352 8121, Email: C.Nienaber@rbht.nhs.uk
Handling Editor: Can Gollmann-Tepeköylü
Peer-reviewers: Dror Ben Leviner; Alicja Zientara
Compliance Editor: Tom Wardill

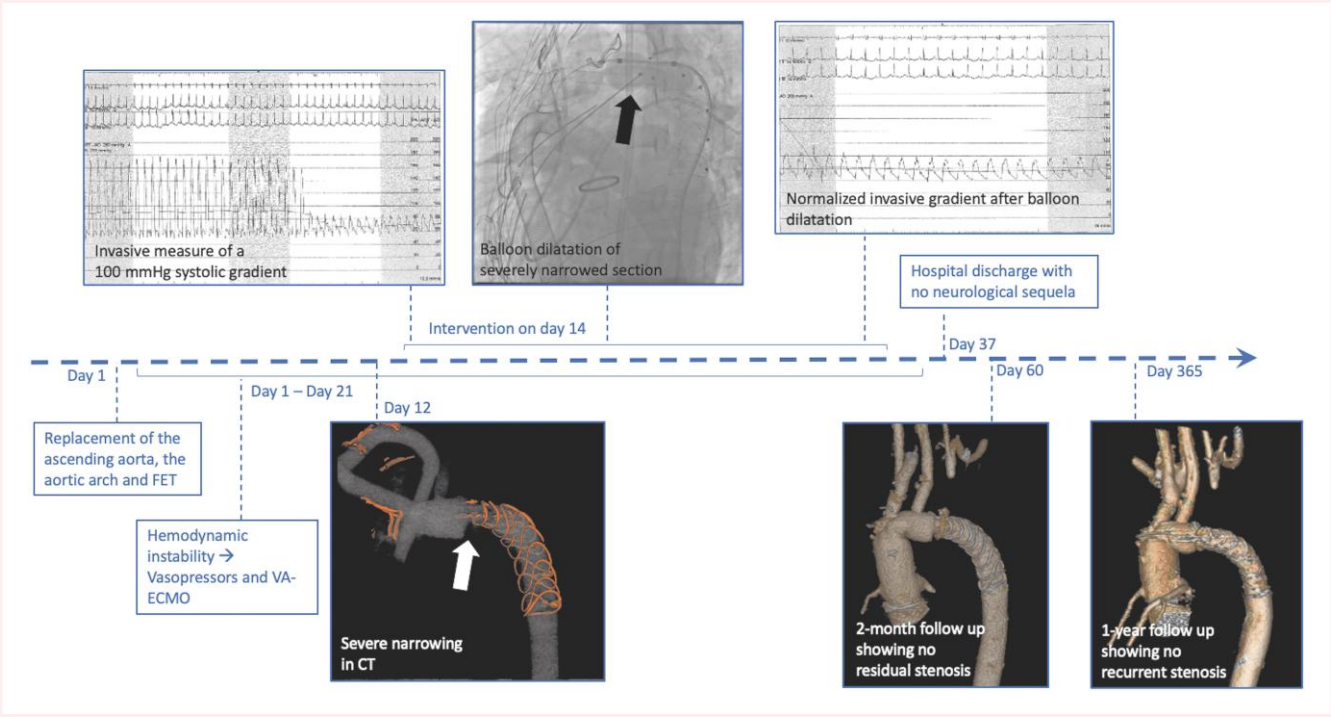
Learning points

- Complex aortic arch reconstruction and frozen elephant trunk (FET) placement may benefit from intraoperative ultrasound or videoscopic inspection.
- In case of post-cardiotomy heart failure, pseudo-coarctation needs to be ruled out by computed tomography angiography and managed immediately.
- Interventional ballooning or stenting has potential to abolish a 100 mmHg trans-FET gradient and remove the underlying obstruction.

Introduction

Combined aortic arch replacement with frozen elephant trunk (FET) insertion is an emergent surgical concept to manage patients with proximal aortic dissection restoring flow to the true lumen by exclusion of the false lumen and prophylactic protection against potential expansion of the descending thoracic aorta.[1–3] As experience with this complex surgery is growing, unexpected complications are observed and need to be managed. This case represents a unique presentation of a complication with a strategy to resolve it.[4]

Summary figure



Case presentation

A previously fit 25-year-old gentleman without any pre-existing medical conditions presented to the emergency department 2 days after onset of severe chest pain. Physical examination neither revealed phenotypic abnormalities nor other medical findings. Due to normal electrocardiography and troponin levels, acute coronary syndrome (ACS) appeared unlikely to be the cause of symptoms. Therefore a computed tomography angiography (CTA) was conducted and revealed Type A aortic dissection originating from the aortic root (DeBakey Type 1, *Figure 1*).

Transoesophageal echocardiography (TOE) showed the dissection membrane and massive aortic regurgitation (see Supplementary material online, *Video S1*).

Transfer to a specialist hospital for surgical care was arranged with arrival in theatre 3 h later (*Figure 2*). The patient underwent median sternotomy providing access to complete replacement of the ascending aorta, the aortic arch, and a FET (Thoraflex 22 mm × 24 mm × 100 mm, *Figure 3*). The cannulation strategy comprised femoral artery and grafted bilateral carotid artery cannulation for antegrade cerebral perfusion as well as venous access to the right atrium.[5]

Under antegrade cerebral perfusion, a mechanical composite graft (St. Jude Medical, Saint Paul, MN, USA) with a 28 mm mechanical valve was used with reimplantation of coronaries and total arch replacement with FET at 28°C. After rewarming, the patient was separated from cardiopulmonary bypass and stable for only 20 min until suddenly it was difficult to oxygenate the patient. Transoesophageal echocardiography showed severe left ventricular (LV) dysfunction and worsening mitral regurgitation (MR) (see Supplementary material online, *Video S2*) prompting re-initiation of cardiopulmonary bypass and conversion to central veno-arterial extracorporal membrane oxygenation (VA-ECMO) for ~7 days with no signs of high pressure via the sidearm of the graft; central ECMO is preferred in acute heart failure and suspected post-cardiotomy syndrome because peripheral ECMO may

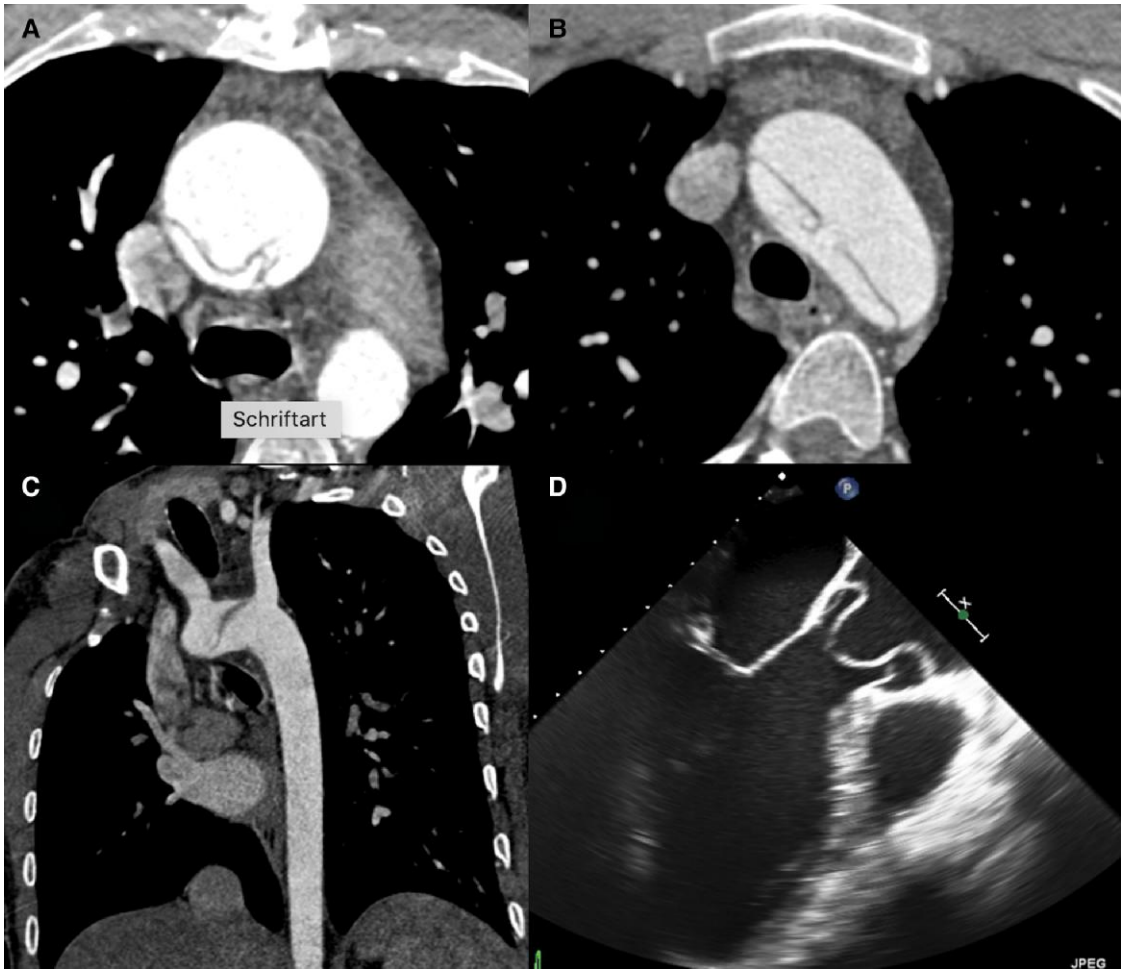


Figure 1 Type A aortic dissection in CTA (*A–C*) and TOE (*D*) (resolution: 300 dpi).

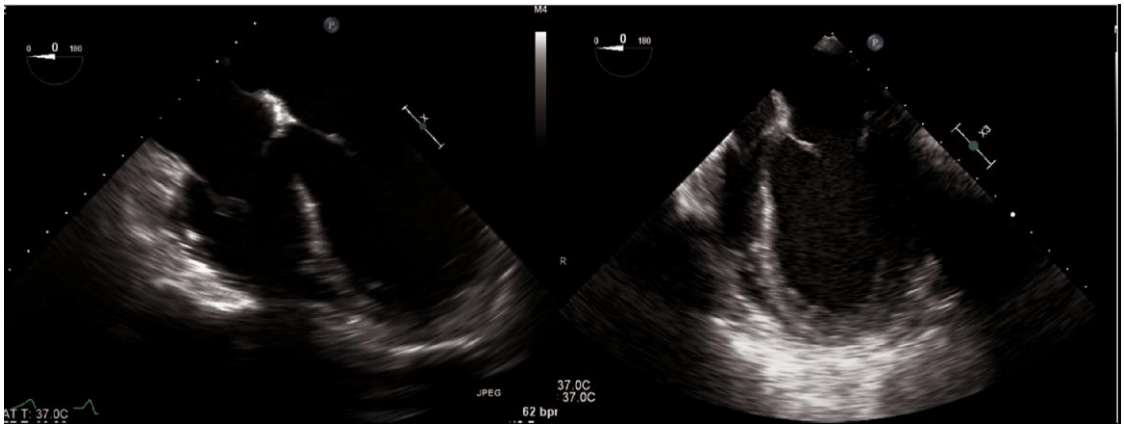


Figure 2 Transoesophageal four-chamber view and transoesophageal two-chamber view showing severely depressed left ventricular function (resolution: 300 dpi).

miss central oxygenation desaturation. When returning to theatre for ECMO decannulation, pericardial washout and an attempt to close the chest, LV function had apparently slightly improved. Soon after chest closure, he required noradrenaline (NA) while TOE demonstrated severely depressed LV function, no collection of pericardial blood, and severe MR. In the following 3 h, the patient became progressively

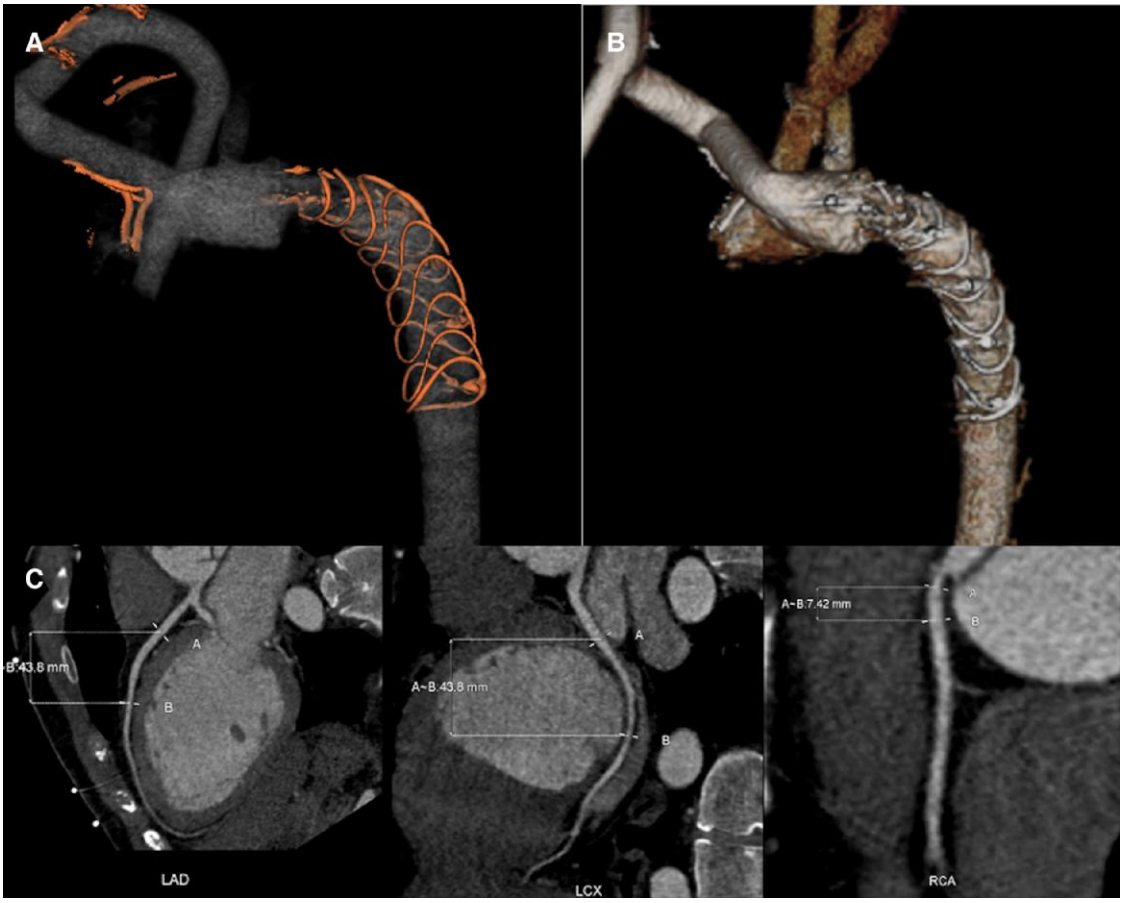


Figure 3 CTA showing severe narrowing at the transition of the grafted arch to the FET (*A* and *B*). CTA showing normal coronary arteries (*C*) (resolution: 300 dpi).

unstable revealing a cardiac index (CI) of 2.6 L/min/m^2 on continuous CI measurement, a systemic vascular resistance of 7.5 Wood units, and pulmonary capillary wedge pressure of 30 mmHg. Dobutamine was commenced but stopped due to excessive tachycardia.

The patient's chest was reopened while haemodynamic instability worsened, inotropes and pressors were escalated to 0.4 mcg/kg/min NA and 0.04 mcg/kg/min of vasopressin (VP) and nitric oxide was started at a pulmonary artery pressure of 66/32 mmHg. Regardless, the patient became increasingly unresponsive to VP and volume substitution and sustained cardiac arrest. Asystole was managed by internal cardiac massage prompting central VA-ECMO to be re-established. The patient was then supported on central VA-ECMO (flow with 5.3 L/min) with reduced doses of pressors now requiring haemofiltration. Broad spectrum antibiotics and antifungal agents were given in suspicion of sepsis, with ST depression and sinus tachycardia of 130 b.p.m., while pupils stayed reactive to light.

On Day 12 post-surgery and still on VA-ECMO for severe LV dysfunction, the patient was considered for long-term mechanical circulatory support, despite unknown neurological status, potential sepsis, and renal dysfunction. A repeat coronary CTA excluded any coronary obstruction (*Figure 3C*) to explain LV dysfunction, but raised suspicion of an obstructed or critically narrowed distal anastomosis of FET.

On post-operative Day 14, the patient was moved on central VA-ECMO to the catheterization suite for invasive exploration, confirming unobstructed coronary circulation via radial access. Retrograde arterial access from the left femoral artery via the true lumen revealed a trans-FET gradient of 100 mmHg with a pressure of 180 mmHg proximal and 80 mmHg distal to the FET on various ECMO flow conditions (*Figure 4A*). Subsequently, a pigtail catheter armed with a hydrophilic 0.018 inch guide wire in order to navigate the subtotal obstruction of the aortic arch was advanced into the ascending aorta (*Figure 4B*).

A contrast aortogram showed severe narrowing at the arch graft and transition to the FET (*Figure 4B*). The narrowing was documented, and repeated pressure measurements were taken for confirmation. With unequivocal documentation of gradient and narrowing, the pigtail catheter was used to guide a Lunderquist superstiff wire (Boston Scientific, Marlborough, MA, USA) across the narrowing to advance a Reliant balloon (Medtronic, Dublin, Ireland) into the narrow segment of the distal aortic arch/proximal FET. After repeated balloon inflations using 50 mL syringes, the gradient was entirely abolished with blood pressure of 100/60 mmHg both proximal and distal to the aortic arch (*Figure 4C and D*). At this time, a decision was made not to advance a Cheatham-Platinum stent into the previous narrowing, but rather watch and wait for potential recovery of LV function.

Within 7 days of the procedure and no intermittent signs of organ malperfusion, the patient was weaned off VA-ECMO with prior recovery of LV function and disappearance of MR allowing the chest to be closed on Day 8 post-intervention; 13 days post-intervention,

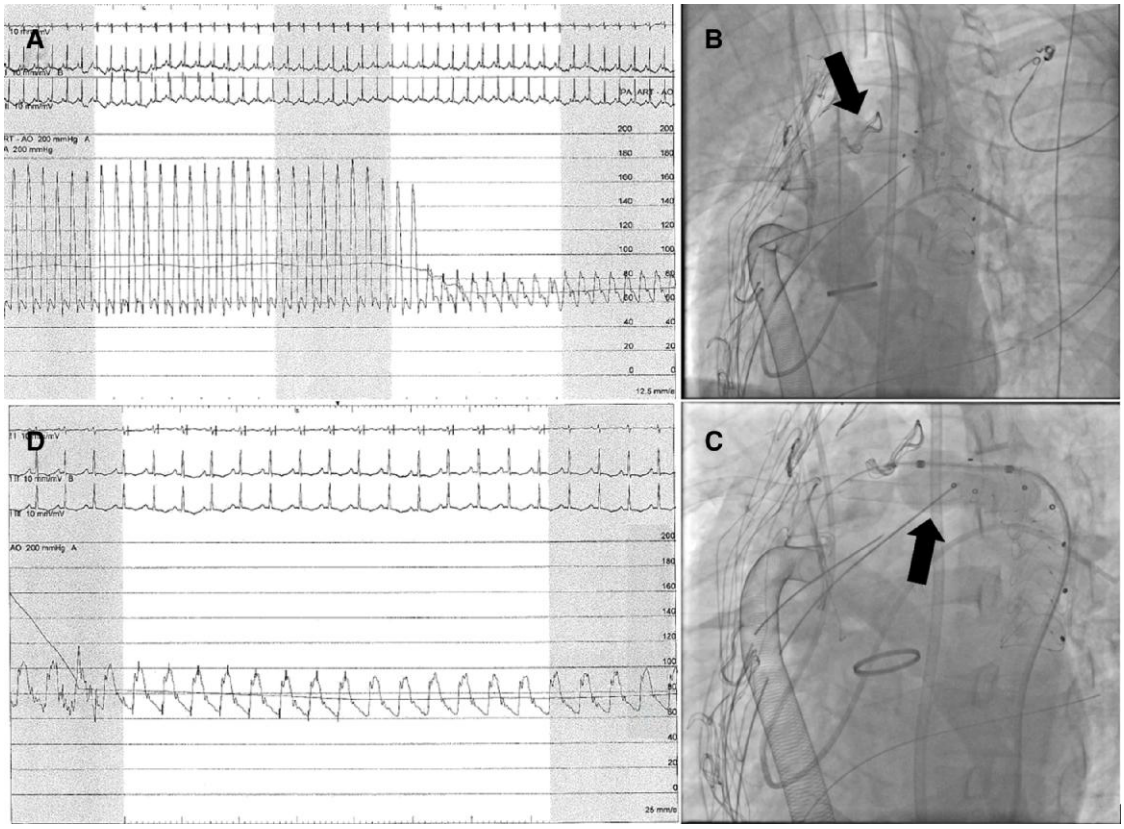


Figure 4 (*A*) Invasive exploration using catheterization of a systolic gradient of ~100 mmHg across a narrowed FET. (*B*) Contrast aortogram showing interrupted contrast in the arch due to severe stenosis at the transition between graft and FET. (*C*) Balloon inflation at the level of the severe stenosis. (*D*) Normalized systolic gradient across FET after successful intervention. (resolution: 300 dpi).

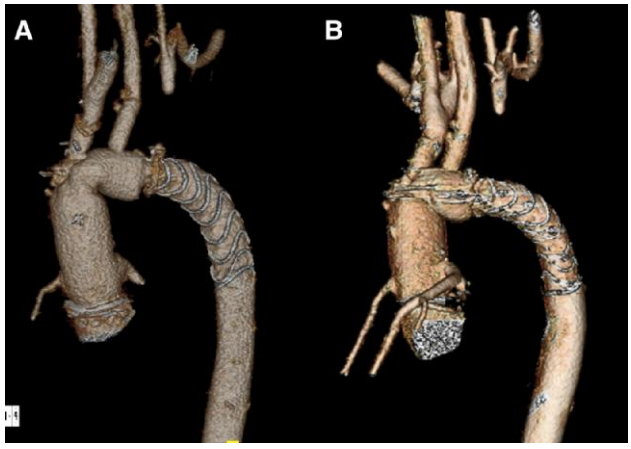


Figure 5 (*A*) Cardiac CTA at 2 months follow-up with no residual stenosis. (*B*) Cardiac CTA at 1 year follow-up with no recurrent stenosis (resolution: 300 dpi).

the patient was extubated with no neurological sequela and discharged 10 days later. At 2 months and 1-year follow-up, complete clinical recovery with no residual signs or symptoms of heart failure (HF) and a wide-open FET on a cardiac CTA were confirmed (*Figure 5A* and *B*).

Discussion

This case demonstrates a rare complication of surgical aortic arch repair with a FET. While the use of FET in the context of replacement of the

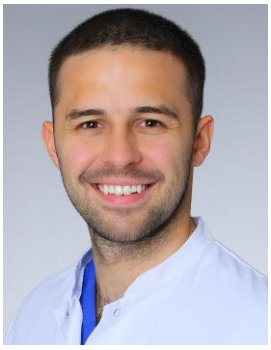
ascending aorta and arch is becoming popular in younger patients, unexpected complications are being encountered. Whereas the mechanism of potential spinal ischaemia after FET is understood, the cause of life-threatening acute obstruction is still subject of speculation.[6,7] It could be either related to twisting of the FET during deployment or external compression causing a sudden massive afterload to the left ventricle leading to acute HF, severe MR, and the need for prolonged VA-ECMO support. The pathophysiologic model of this scenario is a sudden onset pseudo-coarctation with massive increase in afterload to a naïve left ventricle.

The ease to remove the obstruction and abolish a 100 mmHg gradient with balloon inflations with sustained 1 year results may suggest twisting or kinking as the cause of obstruction rather than any external compressing force. Additional stent support after ballooning was not necessary as the result was lasting (*Figure 5*). The fact that the patient turned out to harbour a heterozygous ACTA2 mutation [considered a missense variant of unknown significance c.739>A p (Gly247Arg)] is unlikely to have played a role although the aortic tissue has been described as fragile and soft; ACTA2 mutation is known to dissect at normal aortic dimension.

The case should focus on both, design issues of the FET with better support and on implant technique to avoid any twisting or kinking within the aortic arch or descending aorta. While firmer support would require modified FET design and time to implement, implantation techniques are easier to optimize by use of intravascular ultrasound or even videoscopic guidance during implantation and inspection before closing the anastomosis.[8,9]

On aggregate, this dramatic case provides useful learning points: (1) complex aortic arch reconstruction and FET placement may benefit from intraoperative ultrasound or videoscopic inspection and haemodynamic assessment of transaortic arch gradient. (2) In case of post-cardiotomy heart failure, pseudo-coarctation needs to be ruled out by CTA and managed immediately. In case of an obstructed FET, catheterization and subsequent ballooning have potential to normalize LV afterload and avoid down-spiralling LV decompensation and the need for VA-ECMO. (3) Interdisciplinary cooperation with interventional cardiology may solve unexpected life-threatening problems caused by FET-related aortic obstruction.

Lead author biography



Mr Nienaber graduated from high school in 2013. He completed medical studies at the Medical Faculty of the University of Buenos Aires in Argentina and the Medical Faculty of the University of Cologne in Germany in 2020. Since September 2020, he is employed at the Clinic III for Internal Medicine of the University Hospital Cologne.

Supplementary material

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

Acknowledgements

Not mentioned in this document in order to not reveal the author's names.

Consent: The patient gave his consent in accordance with the COPE guidelines for the material presented to appear in the publication above and related publications. It was explained to him that the material has educational and scientific value and that the publication may help to improve the care that others will receive in the future; however, the patient did and will not receive any financial benefit. He has had the opportunity to see and read the material submitted for publication, and it was explained that the final publication may differ in style, grammar, consistency, and length.

Conflict of interest: None declared.

Funding: None declared.

Data availability

The data underlying this article are available in the article and in its online Supplementary material.

References

1. Shrestha M, Bachet J, Bavaria J, Carrel T, De Paulis R, Di Bartolomeo R, et al. Current status and recommendations for use of the frozen elephant trunk technique: a position paper by the Vascular Domain of EACTS. *Eur J Cardiothorac Surg* 2015;**47**:759–769.
2. Leone A, Beckmann E, Martens A, Di Marco L, Pantaleo A, Reggiani LB, et al. Total aortic arch replacement with frozen elephant trunk technique: results from two European institutes. *J Thorac Cardiovasc Surg* 2020;**159**:1201–1211.
3. Carrel T, Sundt TM, von Kodolitsch Y, Czerny M. Acute aortic dissection. *Lancet* 2023; **401**:773–788.
4. Kayali F, Qutaishat S, Jubouri M, Chikhal R, Tan SZCP, Bashir M. Kinking of frozen elephant trunk hybrid prostheses: incidence, mechanism, and management. *Front Cardiovasc Med* 2022;**9**:912071.
5. Capoccia M, Nienaber CA, Mireskandari M, Sabetai M, Young C, Cheshire NJ, et al. Alternative approach for cerebral protection during complex aortic arch and redo surgery. *J Cardiovasc Dev Dis* 2021;**8**:86.
6. Takagi D, Yamamoto H, Kadohama T, Kiryu K, Wada T, Igarashi I. Optimal stent length and distal positioning of frozen elephant trunks deployed from the aortic zone 0 for type A acute aortic dissection. *J Thorac Cardiovasc Surg* 2024;**167**:15–25.e2.
7. Preventza O, Liao JL, Olive JK, Simpson K, Critsinelis AC, Price MD, et al. Neurologic complications after the frozen elephant trunk procedure: a meta-analysis of more than 3000 patients. *J Thorac Cardiovasc Surg* 2020;**160**:20–33.e4.
8. Roselli EE. Trade in the hammer for a power driver—perspectives on the frozen elephant trunk repair for aortic arch disease. *Ann Cardiothorac Surg* 2013;**2**:633–639.
9. Ouzounian M, Hage A, Chung J, Stevens LM, El-Hamamsy I, Chauvette V, et al. Hybrid arch frozen elephant trunk repair: evidence from the Canadian Thoracic Aortic Collaborative. *Ann Cardiothorac Surg* 2020;**9**:189–196.