ESC European Society of Cardiology

European Heart Journal - Case Reports
doi:10.1093/ehjcr/ytab155

CASE REPORT
Congenital heart disease

Case report of recurrent haemoptysis in an older patient with repaired tetralogy of Fallot

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Received 19 November 2020; first decision 17 December 2020; accepted 9 April 2021

Background Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart defect. Although most infants nowadays undergo surgical repair approximately at the age of 6 months with excellent outcomes, older patients typically underwent a staged approach with an initial systemic-to-pulmonary Blalock–Taussig–Thomas (BTT) shunt reducing hypoxaemia, followed by surgical TOF repair at an older age (with takedown of the BTT shunt). Late complications related to the BTT shunt are rare.

Case summary We report a case of recurrent haemoptysis late after classic BTT shunt and subsequent surgical TOF repair. Axial imaging indicated a stellate nodule with isotope accumulation in the right upper lung lobe, whereas bronchoscopy showed a rope-like structure in the bronchus to the right upper lung lobe. The patient underwent a lobectomy of the superior right lobe, with identification of what appeared to be the old suture around the BTT shunt. Anatomopathology confirmed diffuse necrotizing inflammation with erosion into the bronchus which eventually caused haemoptysis.

Discussion Management of older patients with congenital heart disease requires a thorough knowledge of the anatomy and surgical history of the patient. Although late complications related to a BTT shunt are rare, a residual shunt, endocarditis, pseudoaneurysm, or chronic inflammation with haemoptysis may occur.

Keywords Congenital heart disease • Tetralogy of Fallot • Blalock–Taussig shunt • Blalock–Taussig–Thomas shunt • Haemoptysis • Case report

Learning points

- Even though palliative surgery with a Blalock–Taussig–Thomas (BTT) shunt for tetralogy of Fallot (TOF) is rarely performed these days, physicians may still encounter older TOF patients who previously had a BTT shunt. Therefore, physicians should be aware of its function, anatomy, and implications.
- In patients with surgically repaired congenital heart disease suffering from haemoptysis, cardiac catheterization should be considered before moving on to invasive surgery or obtaining tissue samples. The catheterization should ideally be performed by a cardiologist with experience of adult patients with congenital heart disease.

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Handling Editor: Anastasia Egorova
Peer-reviewers: A. Shaheer Ahmed and Luis Antonio Moreno-Ruiz
Compliance Editor: Daniel Tardo
Supplementary Material Editor: Mariame Chakir

Introduction

Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart defect.[1] Tetralogy of Fallot consists of obstruction of the right ventricular (RV) outflow at the infundibular level, a large non-restrictive ventricular septal defect (VSD) with the aorta overriding this defect and concentric RV remodelling completing the 'tetralogy' as described by Etienne-Louis Arthur Fallot. Only a few decades ago, intracardiac surgical repair in neonates was impossible. Tetralogy of Fallot, being one of the first and most commonly described cyanotic lesions, sparked a dramatic evolution in congenital cardiac surgery resulting in the development of the Blalock–Taussig–Thomas (BTT) shunt in 1945 (John Hopkins),[2] and a first intracardiac repair by Lillehei *et al.* (Minnesota) in 1954.[3] For most older patients with repaired TOF followed in the adult congenital heart disease (ACHD) clinic, this typically implies that they underwent two-staged surgical interventions; a first palliative surgery to increase pulmonary blood flow and alleviate cyanosis and a second surgical repair with closure of the VSD and relief of the right ventricular outflow tract (RVOT) at an older age. Nowadays, newborns with TOF will undergo intracardiac repair surgery relieving the RVOT obstruction and closing the VSD at ∼6 months old.[4] Nevertheless, for cardiologists in general and ACHD physicians specifically, it remains crucial to understand prior surgical interventions even if these are no longer (or rarely) performed. As these individuals grow older, they are at increased risk of developing cardiac complications and other non-cardiac co-morbidities. Consequently, cardiologists are occasionally faced with complex and challenging medical issues in adult TOF patients where the answer for the problem is hidden in the medical charts.

Timeline

1962	Classic Blalock–Taussig–Thomas shunt at the age of 12 years
1967	Surgical repair of tetralogy of Fallot with ventricular septal defect patch closure and relief of right ventricular outflow tract obstruction (infundibulectomy) at the age of 17 years
1992	Cerebrovascular accident with complete recovery
1993	First episode of haemoptysis, treated conservatively
2002	Stellate nodule in right upper pulmonary lobe found on computed tomography scan—watchful waiting
2004	Chest pain with normal coronary arteries on coronary angiogram
2007	Permanent atrial fibrillation, started on oral anticoagulation and betablocker therapy
2019	Recurrence of haemoptysis, bronchoscopy shows mucosal swelling in the right upper pulmonary lobe

Case presentation

A 69-year-old woman with repaired TOF suffering from recurrent haemoptysis was referred to our institution for further investigations. She underwent palliation in 1962 (age 12) with a classic right-sided BTT shunt and TOF repair in 1967 (age 17) with closure of the VSD, relief of the RV outflow tract obstruction, and closure of the BTT shunt. Over the course of the years, she developed atrial fibrillation for which she was started on oral anticoagulation. In 1993, she had a first episode of massive haemoptysis which was treated conservatively. In 2002, computed tomography (CT) indicated a stellate nodule in the right upper lung lobe. Over the next years the lesion remained stable on axial follow-up imaging. In 2019, after new episodes of haemoptysis, this time with progressively worsening dyspnoea, a positron emission tomography–CT was performed showing isotope accumulation in the lesion of the right upper lung lobe (*Figure 1A–C*). She underwent a bronchoscopy that showed mucosal swelling in the right upper lobe. Cytology indicated acute inflammation without evidence of malignancy.

On physical examination, the right-sided radial pulse was absent and blood pressure (BP) not measurable. Blood pressure and heart rate taken on the left arm were 112/45 mmHg and 78 b.p.m., respectively. Respiratory rate was 22 per minute. The second heart sound was pronounced with a grade 3/6 systolic and 2/4 diastolic murmur at the second intercostal space on the left. C-reactive protein was normal (2.9 mg/L) and N-terminal B-type natriuretic peptide slightly elevated (733 ng/L). The electrocardiogram showed atrial fibrillation with inferolateral ST depression and Q-waves in V1–V2–aVR (*Figure 2*). Transthoracic echocardiography indicated a moderately dilated RV with normal systolic function, a peak/mean gradient of 53/32 mmHg across the pulmonary valve with severe pulmonary valve regurgitation (*Figure 1D–F*).

A diagnostic cardiac catheterization was performed but no arteriovenous connections were seen and the BTT shunt was not patent. Right atrial pressure was 14 mmHg, RV systolic pressure 56 mmHg, systolic pulmonary artery pressure 33 mmHg, and mean pulmonary artery pressure 21 mmHg. The RV was dilated with severe pulmonary valve regurgitation. A bronchoscopy with EndoBronchial UltraSound (EBUS) in order to perform EBUS guided biopsy was performed. A rope-like structure was seen through the bronchoscope, but we were unable to take a biopsy (*Figure 3C*).

After multidisciplinary discussion, the findings appeared to be most consistent with chronic inflammation due to a foreign body with secondary chronic inflammation, rather than vasculitis or a granulomatous infection, and the patient underwent a thoracotomy with lobectomy of the superior lobe of the right lung. The foreign body was removed without any resistance nor bleeding. Pathology indicated diffuse necrotizing granulomatous inflammation around the rope-like structure (*Figure 3C* and *D*) which appeared to be the old suture around the BTT shunt that had caused a chronic inflammatory reaction invading the right upper bronchus and causing haemoptysis. The patient remained stable after the procedure. She left the hospital after a couple of days. The postoperative course was uncomplicated. After 3 months, she underwent percutaneous pulmonary valve replacement. After the surgical intervention, given a CHA2DS2 VASc score of 6/9, a HAS-BLED score of 5 and the fact that the most likely source of haemoptysis was removed, she was started on edoxaban 60 mg once daily.

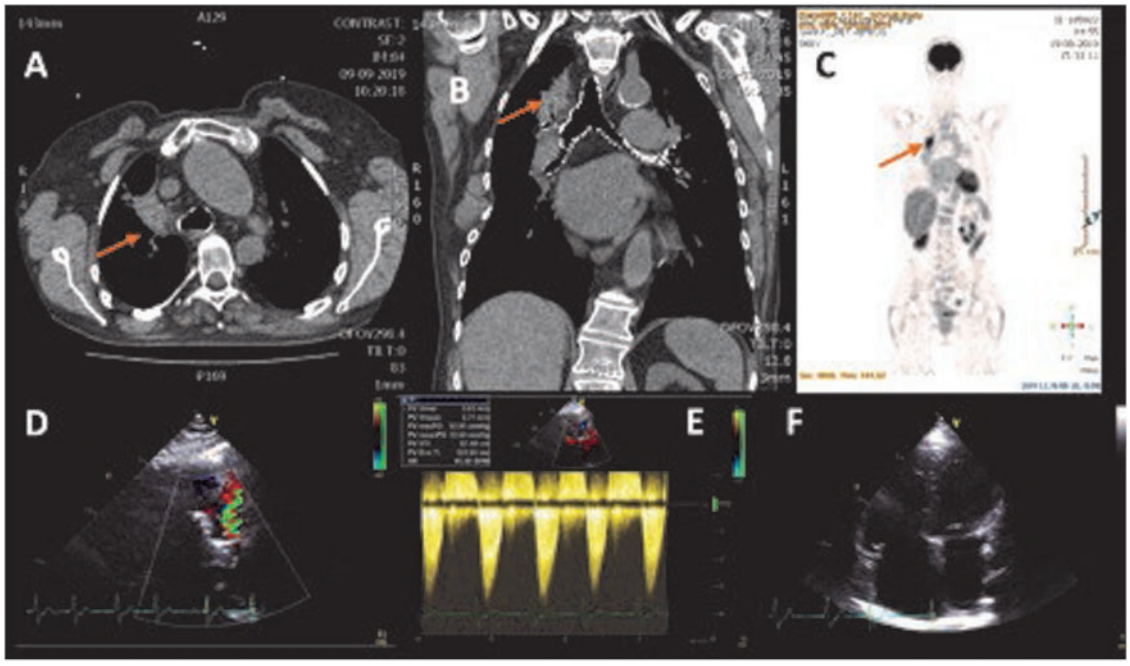

Figure 1 Positron emission tomography–computed tomography (*A–C*) showing axial and coronal planes. The orange arrow indicates the stellate nodule with isotope accumulation in the right upper lung lobe. Transthoracic echocardiography (*D–F*) with parasternal short-axis view (*D*), continuous wave velocity over the pulmonary valve (*E*), and apical four-chamber view (*F*). Colour Doppler flow indicates regurgitant flow in diastole, indicating pulmonary valve regurgitation. Continuous wave velocity indicates a peak/mean gradient in systole across the pulmonary of 53/32 mmHg indicating pulmonary valve stenosis.

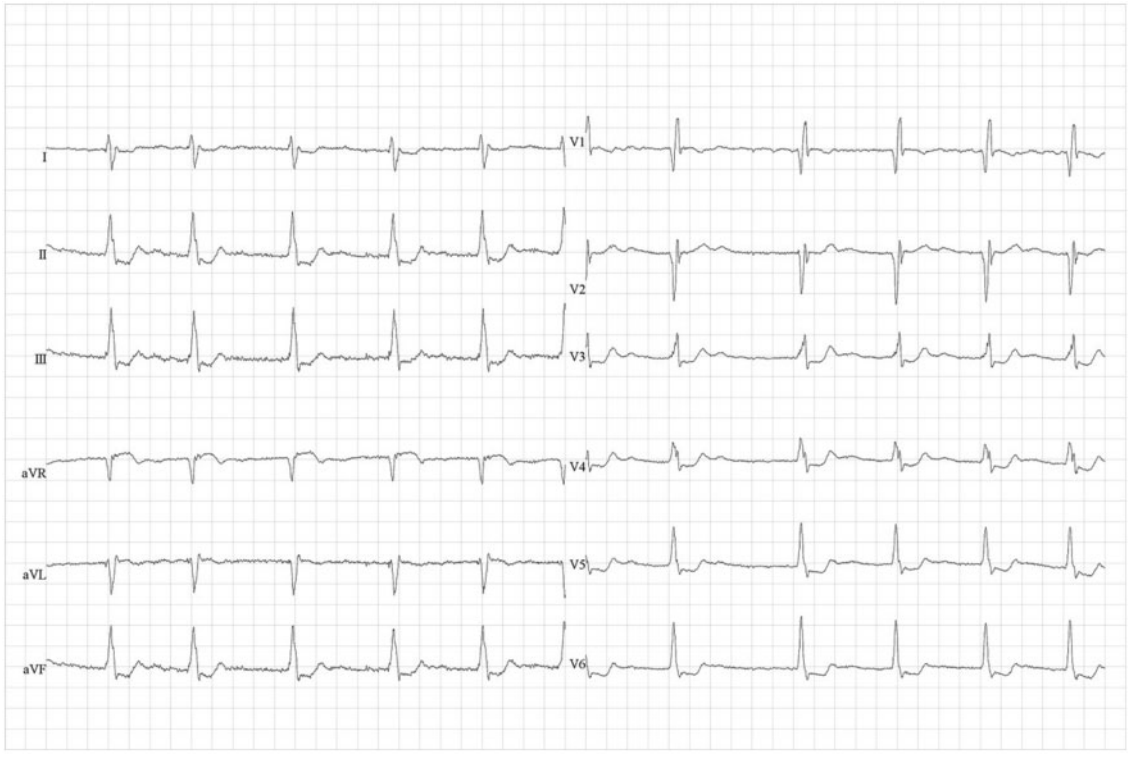

Figure 2 Electrocardiogram at first presentation.

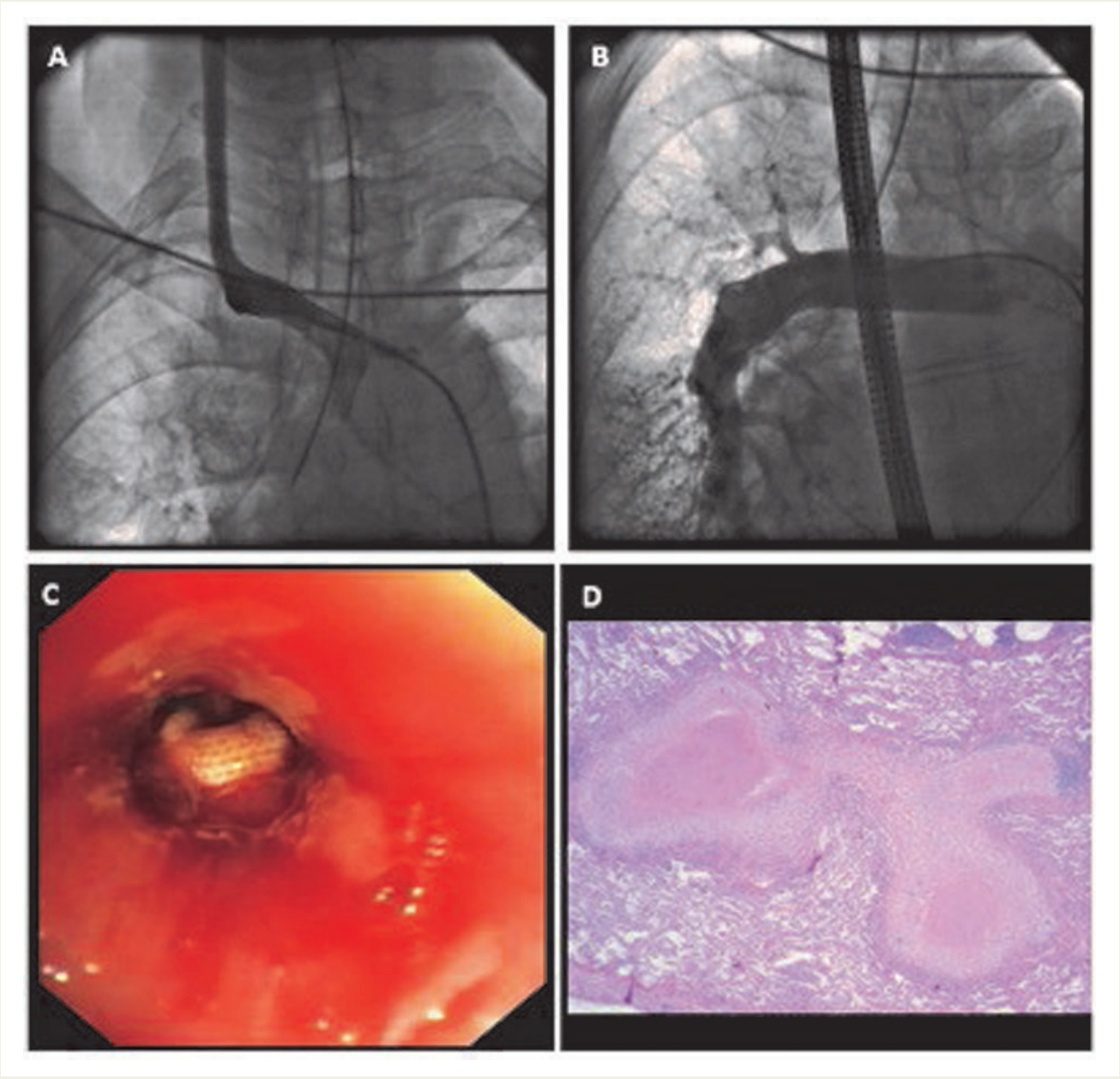

Figure 3 Cardiac catheterization with contrast injection in the right subclavian artery (*A*) and right pulmonary artery (*B*). Bronchoscopy with endoluminal image of the bronchus to the right upper lung lobe (*C*) and anatomopathological image after right upper lobectomy (*D*) showing diffuse necrotizing granulomatous inflammation (haematoxylin and eosin stain, magnification ×50).

Discussion

The classic BTT shunt was developed in the 1940s by Dr Helen Taussig, Dr Alfred Blalock, and Mr Vivien Thomas at John Hopkins University as palliative surgery to increase pulmonary blood flow and alleviate cyanosis by connecting the subclavian artery to the pulmonary artery.[2,5] In the early stages, the subclavian artery was disconnected from the aorta and the proximal part inserted to the pulmonary artery (classic BTT shunt), explaining the difficulties to measure BP and pulse in the right arm. The BTT shunt would bridge the time until the patient's age and size would allow surgical TOF repair. At that point, the BTT shunt was interrupted, initially using a suture, later using a clip or takedown of the shunt. Later, the classic BTT shunt was replaced by the modified BTT shunt, using an interposition tube graft.[6] Thanks to improvements in extracorporeal circulation methods in modern surgery, the staged approach for TOF repair is no longer necessary since neonates with TOF undergo curative surgical repair at the age of 3–6 months. However, late complications after surgical TOF repair still occur and typically encompass pulmonary valve regurgitation and/or

stenosis with secondary RV dilatation, which explains the systolic and diastolic murmur heard at the second intercostal space on the left side on physical examination.

There are few case reports of haemoptysis in TOF patients, mostly related to a vascular origin such as a pseudoaneurysm of the BTT shunt or rupture of major aorto-pulmonary collateral arteries (large abnormal arterial vessels connecting the aorta to the pulmonary circulation, providing extra pulmonary blood flow).[7] Moreover, there are reports of patients post-BTT shunt, with expectoration of the shunt years after the initial operation.[8] After excluding malignancy, granulomatous reactions, and vasculitis, cardiac catheterization plays a crucial role in excluding a vascular origin of haemoptysis.

In this case, we confirmed disconnection of the right subclavian artery (classic BTT shunt), without abnormal vascular connection and/or aneurysm from either subclavian artery or pulmonary artery.

Although indications have shifted, the modified BTT shunt is still used in the current era, mostly in the setting of palliation of patients with univentricular physiology.[9]

In conclusion, surgical treatment for congenital heart disease has evolved a great deal from where it started a century ago. Nevertheless, it is still important physicians are aware of—and understand these historical procedures as late complications may still arise later in life.

Lead author biography

Emma Vanderschueren is a third-year resident at the University Hospital of Leuven, Belgium. She has a broad range of interests within internal medicine and cardiology including congenital heart disease. She is thankful to have had the chance to work alongside specialist in different fields within cardiology who would train her on problem solving and guide her through complex cases.

Supplementary material

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

Acknowledgements

We thank Prof. Dr Dirk Van Raemdonck and Prof. Dr Christophe Dooms for their insights during the unfolding of this case.

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 submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: None declared.

Funding: none declared.

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