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CASE REPORT
Congenital heart disease

Stenting of modified Blalock–Taussig shunt in adult with palliated pulmonary atresia and ventricular septal defect: a case report

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Background Adults with complex congenital heart disease palliated with systemic-to-pulmonary artery shunts have become rare and represent a particularly challenging patient group for the cardiologist. One of the complications and causes of severe clinical deterioration during long-term follow-up are progressive obstruction or total occlusion of the shunt. The risk for surgical intervention is frequently high and catheter intervention may be complicated by complex anatomy and shunt calcification.

Case summary We report the case of a 47-year-old man with uncorrected (palliated) pulmonary atresia and ventricular septal defect who presented with progressive cyanosis (oxygen saturation 69%) and decreasing exercise capacity. Computed tomography revealed a totally occluded modified left Blalock–Taussig (BT) shunt and a severely stenosed central shunt (Waterston–Cooley) in a patient with confluent but hypoplastic pulmonary arteries and multiple major aortic pulmonary collaterals. Due to a high operative risk, an interventional, percutaneous approach was preferred to re-do surgery. From a radial access the calcified BT shunt could be crossed with a hydrophilic guidewire. Then, a rotational thrombectomy, balloon dilatation, and bare-metal stenting at the proximal and distal anastomoses were performed. Post-interventionally, peripheral oxygen saturation increased from 69% to 82%. Clopidogrel was administered for 1 month after bare-metal stenting. At 1-year follow-up, the BT shunt was still patent on echocardiography and exercise tolerance markedly improved.

Discussion This case highlights the benefit of percutaneous rotational thrombectomy followed by stenting of chronically occluded systemic-to-pulmonary artery shunts for further palliation in adult patients with complex congenital heart disease not suitable for surgical repair.

Keywords Bare-metal stent • Case report • Modified Blalock–Taussig shunt • Rotational thrombectomy • Pulmonary atresia with VSD

Learning point

- Percutaneous rotational thrombectomy and stenting of pulmonary-systemic shunts, such as the modified Blalock–Taussig shunt, may offer further palliation in selected adult patients with complex congenital heart disease.

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Introduction

Unlike today, complex congenital heart diseases such as pulmonary atresia with ventricular septal defect and tetralogy of Fallot were not corrected in infants in earlier times. Instead, pulmonary-systemic shunts like the (modified) Blalock–Taussig (BT) shunt or the Waterston–Cooley (WC) anastomosis were performed for palliation. Some of those patients had to undergo surgical re-do operations, others survived with the palliation shunts until adulthood. Since the invention of interventional cardiac catheterization techniques in the 1980s and 90s, acute stent-thrombosis of pulmonary-systemic shunts in infants and stent stenosis in adults were treated via percutaneous catheter procedures.

In neonates and infants, cases of acute thrombotic occlusions of BT shunts were reported. When surgical re-do was not an option, they were treated with recombinant tissue plasminogen activator for systemic thrombolysis[1] or needed emergency cardiac catheterization.[2–5] To date, coronary artery stents were used in infant BT shunt catheterization procedures for multiple purposes: for re-opening fresh thrombotic occlusions, for down-sizing a BT shunt lumen,[6] and for covering pseudoaneurysms of BT shunts.[7] Kokov *et al.* and Wehman *et al.* reported the use of the Angio Jet rheolytic catheter system for thrombus removal in infant patients.[8,9]

In adults, very few reported patients underwent angioplasty and stenting of a severe BT shunt stenosis.[10,11] However, to the best of our knowledge, the use of a rotational thrombectomy system for re-opening a completely occluded BT shunt in an adult has not been described so far.

Timeline

Year (month)	Event
1970	Patient born with pulmonary atresia and ventricular septal defect
1977	Modified Blalock–Taussig shunt for palliation
1997	Waterston–Cooley anastomosis for palliation
2017 (August to November)	Clinical deterioration (progressive fatigue, limited walking distance, hypoxaemia pO_2 69%)
2017 (November)	Rotational thrombectomy and bare-metal stenting of modified Blalock–Taussig shunt
2018 (April)	Five-month follow-up with alleviated peripheral cyanosis and improved oxygen saturation (pO_2 79% under room air)
2018 (November)	One-year follow-up with significantly improved exercise tolerance and improved oxygen saturation (pO_2 85% under room air)

Case presentation

A 47-year-old male patient with palliated pulmonary atresia with ventricular septal defect (VSD) presented with progressive fatigue and hypoxaemia to our outpatient clinic. He was no more able to perform his daily activities, such as washing himself and his walking distance was limited to 25 m. His peripheral oxygen saturation was only 69% (haematocrit 71%). As to his history, a modified BT shunt was performed between the left subclavian artery and the left pulmonary artery at the age of 7 years. At the age of 27 years, a WC anastomosis was performed to increase blood flow through the pulmonary circuit, because the still patent BT shunt was not sufficient for pulmonary blood supply. Surgical correction was not a preferred option because of the technical difficulties.

Since 1997, the patient did not need any further cardiac surgery or intervention and was able to carry out his daily routine activities until 3 months prior to hospital admission. In the preceding months, the patient received treatment for left basal pneumonia and pneumosepsis at another hospital. Presenting with atrial fibrillation and atrial flutter, the patient needed cardioversion twice to convert his heart rhythm to sinus rhythm. There was no history of dizziness or syncope at any time. In the presence of intermittent left thoracic pain, coronary angiography was performed to rule out any relevant coronary artery stenosis. Oxygen therapy was initiated 2 weeks prior to admission to our hospital, but did not significantly ameliorate the progressive desaturation.

On physical examination, distinctive cyanosis was notable (SpO_2 69%). There were neither peripheral oedema nor heart murmurs or pulmonary crackles on auscultation present. Blood pressure was 140/100 mmHg, and heart rate was 90 b.p.m.

On admission, electrocardiogram showed a sinus rhythm and complete right bundle branch block. Echocardiography revealed the ventricular septal defect, occluded right ventricular outflow tract with hypertrophic right ventricle and dilation of both atria. Systolic function of the ventricles was preserved, and the aortic root was known to be ectatic. Computed tomography scans showed the occluded BT shunt and a reduced perfusion of the WC anastomosis (*Figure 1*); besides that, many major aortopulmonary collateral arteries (MAPCAs) were detected.

Because of the limited pulmonary blood flow oxygen therapy did not elevate saturation. Therefore, possible therapeutic approaches such as an interventional re-opening of the BT shunt or the WC anastomosis as well as a new surgical aortopulmonary shunt or correction were discussed in our interdisciplinary Heart Team meeting. Surgery was ruled out due to the excessive risk. The WC anastomosis showed a small lumen and unfavourable angle for access and intervention. In this respect, the occluded BT shunt was addressed as an interventional target. Prior to intervention, we carried out a pulmonary function test. A restrictive lung function with a reduced forced vital capacity (FVC) of 2.75 L (= 51%) was documented. Lowest oxygen level of pO_2 40.9 mmHg under room air and pO_2 48 mmHg under 4 L oxygen were measured.

For re-opening the BT shunt, the left radial artery was punctured. The occluded BT shunt could be wired with a Radifocus®-wire with a flexible tapered tip and a Glide-Cath® (both Terumo). Then, we performed a rotational thrombectomy using a 6 Fr-Rotarex®-system (AB Medica, Duesseldorf, Germany). Afterwards, the shunt was dilated with a standard balloon. Due to significant recoil of both the proximal and the distal anastomosis, then two Genesis® Stents (6 × 25 and 7 × 25 mm) were deployed in the proximal and distal anastomoses of the shunt (*Figure 2*).

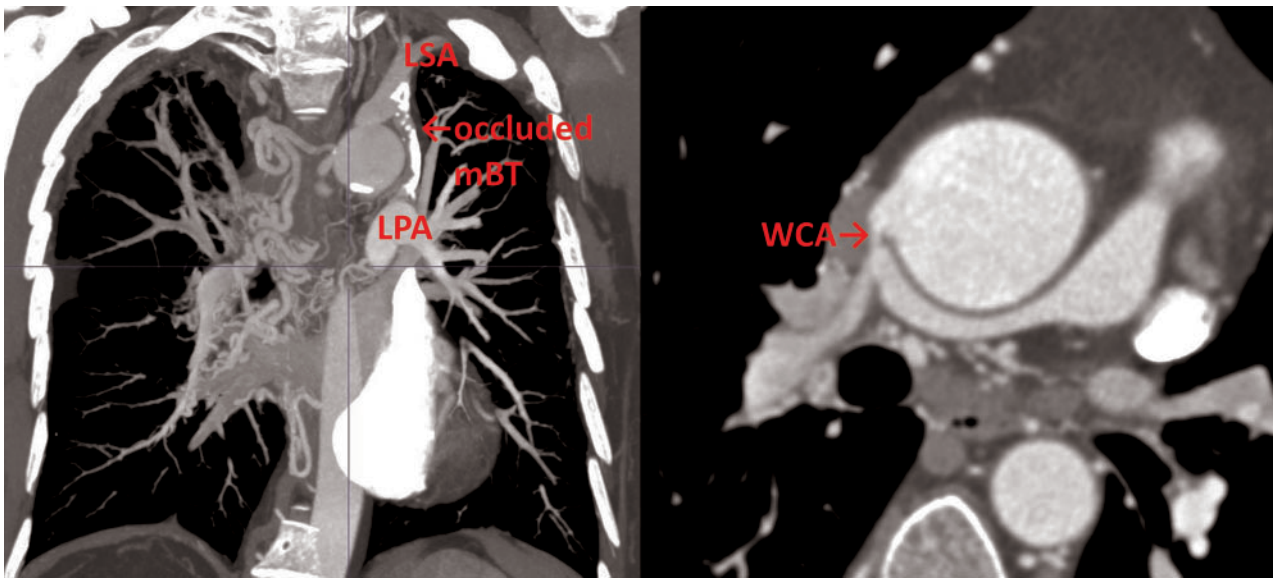

Figure 1 Computed tomography scans of the occluded modified Blalock–Taussig shunt before intervention (left) and the highly stenosed Waterston–Cooley anastomosis (right). LSA, left subclavian artery; LPA, left pulmonary artery; mBT, modified Blalock–Taussig shunt; WCA, Waterston–Cooley anastomosis.

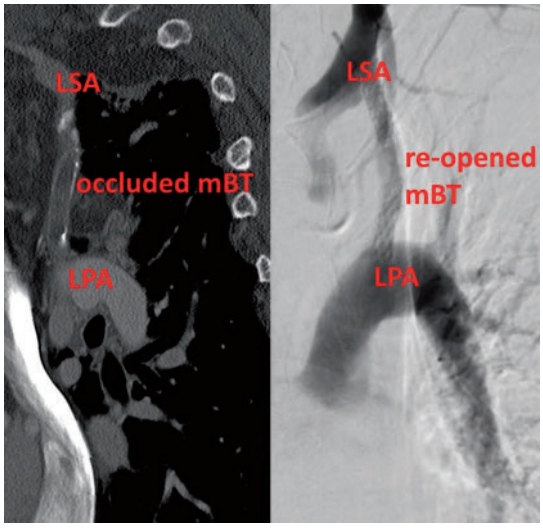

Figure 2 Computed tomography scan of the occluded modified Blalock–Taussig shunt (left) and angiographic result after rotational thrombectomy and stenting of both anastomoses (right). LSA, left subclavian artery; LPA, left pulmonary artery; mBT, modified Blalock–Taussig shunt.

Finally, the shunt was patent with a rapid perfusion of the left lung. Immediately, the peripheral oxygen saturation increased from 69% to 82% under room air.

The day after, the patient noticed significant improvement of his physical abilities. Vascular access site showed no complications. The patient was discharged in a stable general condition.

At his 5-month and 1-year follow-up appointments in our outpatient clinic, the patient reported on an improved exercise tolerance. There was a reduction in peripheral cyanosis with improvement in oxygenation (79%) and haematocrit (66%).

Echocardiography demonstrated patent BT shunt. Clopidogrel treatment was stopped 1 month after bare-metal stent implantation. Anticoagulant treatment with rivaroxaban was continued for thrombo-embolic prophylaxis due to atrial fibrillation.

Discussion

In summary, we report the successful rotational thrombectomy with consecutive bare-metal stenting of an obstructed BT shunt in an adult patient with an uncorrected, but palliated pulmonary atresia with VSD.

Transcatheter recanalization of an obstructed BT shunt is an appreciable alternative to surgical re-do shunt operations due to significantly lower risk. It may increase oxygen saturation and improve patients' exercise tolerance and quality of life. Advantages of catheter interventions in comparison with surgical repair are based on reduced bleeding risk, less post-interventional wound pain, reduced wound infection risk and shorter duration of hospital stay. Possible short and long-term risks of BM-stenting may be the formation of an in-stent-thrombosis with an acute deterioration of oxygen saturation and haemodynamics despite anti-platelet therapy and the development of an in-stent-re-stenosis due to intimal growth over time. Other possible targets of interventional therapies to improve lung perfusion are stenosed MAPCAs, but vessel morphology and wall

characteristics carry the risk of complications such as dissection and acute closure. The combined technique of thrombectomy and stenting of pulmonary-systemic shunts, such as the modified BT shunt, may offer further palliation in selected adult patients with complex congenital heart disease.

Lead author biography

Dr Julia Illner graduated from the University of Luebeck in 2016, and she started her specialist training in cardiology at the University Hospital Muenster. She gained her doctoral degree in 2018.

Supplementary material

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

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 image(s) and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: none declared.

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