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Successful stenting of an obstructed Glenn anastomosis in a 20-month-old child

Oadi N. Shrateh [a, *], Afnan W.M. Jobran [a], Nizar Hijjeh [b], Nidal Haymouni [c], Iyad Sbeitan [c], Samer Abdelraziq [c], Ruba Rizik [c], Ahmed Darsalim [c], Mohammed Abutaqa [c]

[a] Al-Quds University-School of Medicine, Abu-Dis, Jerusalem, Palestine
[b] Department of Pediatric Thoracic Surgery, Al-Ahli Hospital, Hebron, Palestine
[c] Department of Pediatric Cardiology, Al-Makassed Islamic Charitable Hospital, Jerusalem, Palestine

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ABSTRACT

Children with univentricular heart disease may benefit from the Glenn operation (superior vena cava to right pulmonary artery anastomosis), one of the palliative treatment stages. Preoperative and postoperative variables like thrombosis, arrhythmias, and superior vena cava syndrome affect postoperative mortality. A 20-month-old child, known to have complex congenital heart disease with single ventricle physiology. She had Blalock–Taussig shunt in early infancy and then left Glenn anastomosis while she was recovering. Twenty-five days later, she developed acute thrombosis of her Glenn anastomosis. She was hemodynamically unstable with severe desaturation, and the decision was made for catheter-based intervention. Balloon dilatation of the thrombosis initially failed. However, relief of the blockade using 2 overlapping stents was successful and improved her pulmonary blood flow, avoiding urgent surgery for her.

1. Introduction

The management of patients with single-ventricle physiology is complex and challenging. Currently, a 3-staged procedure is performed for these patients in order to limit the ventricular output to the systemic circulation. These 3 stages are, in sequence, the Blalock-Taussig shunt, superior cavo-pulmonary anastomosis, and Fontan anastomosis. The bidirectional Glenn (BDG) is one of two ways used to achieve superior cavo-pulmonary anastomosis (SCPA) in which SVC is divided from the right atrium (RA) and sutured to the pulmonary artery, redirecting venous return from the upper body directly into the pulmonary circulation [1].

Most patients undergoing BDG do well after the procedure. However, one of the complications that may occur is thrombosis of the Glenn anastomosis, and it is a cause of significant morbidity and mortality [1]. We are reporting a 20-month-old child who developed acute thrombosis of the Glenn anastomosis twenty-five days after the procedure.

* Corresponding author.Al-Quds University-School of Medicine, Abu-Dis, Ramallah, Palestine.
E-mail addresses: oadi.shrateh@students.alquds.edu (O.N. Shrateh), afnanjobran26@gmail.com (A.W.M. Jobran), Hijjeh@hotmail.com.com (N. Hijjeh), novonidal@yahoo.com (N. Haymouni), iyad_sbeitan@yahoo.com (I. Sbeitan), Dr.samerar@gmail.com (S. Abdelraziq), Rubarizik265@gmail.com (R. Rizik), Dr.darsalim1@gmail.com (A. Darsalim), Moh.abutaqa@gmail.com (M. Abutaqa).

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2. Case report

Our female patient was diagnosed at birth with complex congenital heart disease; double outlet right ventricle with a large subarterial Ventricular Septal Defect (VSD) and malposed great arteries with pulmonary atresia. She underwent first stage palliation surgery, Blalock Taussig shunt insertion, at the age of two weeks.

She lost follow-up till the age of 20 months, when she underwent left bidirectional Glenn anastomosis with left pulmonary artery (LPA) plasty. After surgery, she developed a pulmonary hypertension crisis requiring nitric oxide, mechanical ventilation, a Milrinone infusion, and sildenafil. She stayed in the pediatric cardiac intensive care unit for 20 days post-surgery, then was transferred to the pediatric ward. While in the ward, she developed sudden hypoxia and bradycardia, requiring endotracheal intubation and cardiopulmonary resuscitation. On physical examination, she had congestion in the upper part of her body. She had persistent hypoxia despite 100% oxygen supply via mechanical ventilation. An X-ray of the chest revealed oligemic lung fields; a Doppler ultrasound of the neck revealed a thrombus in the internal jugular vein; and an echocardiogram revealed an obstructed and thrombosed Glenn anastomosis.

Measures taken to increase pulmonary blood flow included intravenous fluid expansions and heparin infusion, but these failed to improve oxygenation. She developed hypotension and required inotropic support, and vasopressors were used to maintain peripheral perfusion. After medical measures failed to improve her condition, percutaneous catheterization was performed. Via direct puncture to the left internal jugular vein through a 5 F introducer, angiography was done and showed complete obstruction of the flow to the lungs and the contrast seen in the hemiazygos vein, which was large and tortious (Fig. 1A). Trials to cross the clotted Glenn via a Terumo wire succeeded in introducing it to the RPA. Then a 5F RJ catheter was introduced and angiography was done, showing good flow in both lungs except for the right upper lobe (Fig. 1B). A filling defect of 15 mm length was found and thus a 12 × 20 mm balloon was inflated across the obstructed part with no improvement seen on angiography post angioplasty with no flow across that obstructed segment (Fig. 2A). The obstruction was crossed again and two stents were deployed along the obstructed segment 6 x 29 and 6 × 15 mm via 7 F sheath and then saturation improved and control angiography showed excellent flow in the Glenn to the pulmonary tree (Fig. 2B). At the start of the procedure, the patient was given 350 units of heparin. At the end of the procedure, all sheaths and catheters were removed. Hemostasis was achieved within 5 min and bleeding was less than 10 cc.

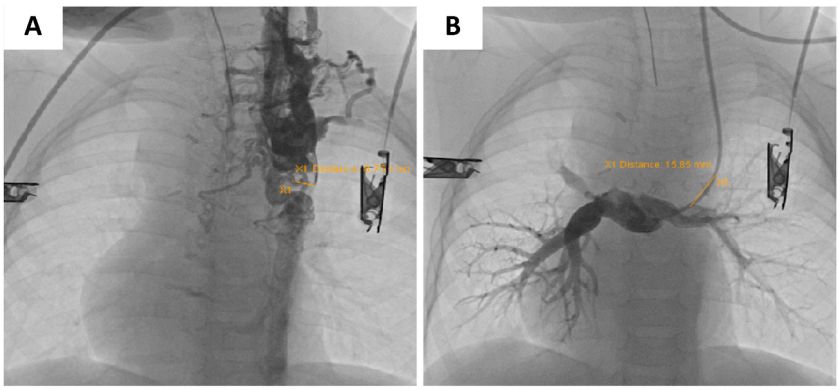

Fig. 1. (A): is an angiography in the left internal jugular access showing complete obstruction of the Glenn with no flow to the lungs and the contrast is seen in the hemiazygos vein. **(B):** is an angiography showing that the catheter is crossing the obstructed Glenn (the measured segment) with flow to the lungs.

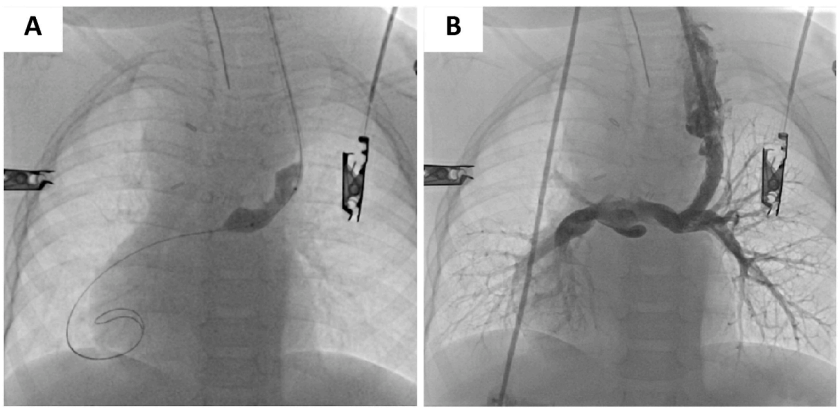

Fig. 2. (A): is an angiography showing that the balloon is inflated across the obstructed portion with residual waist that could not be opened. **(B):** is an angiography demonstrating stented distal Glenn with good flow to the both lungs.

Oxygen saturation immediately increased from 20% to 75% after placement of the two stents. She required mechanical ventilation for 6 days; inotropic support was stopped gradually. After three weeks, she went home with good pulmonary blood flow through the stented Glenn anastomosis.

3. Discussion

Compared to patients with biventricular hearts, individuals with univentricular hearts are more susceptible to morbidity and mortality [2]. Prior to performing palliative BDG and Fontan shunts, it is crucial to identify patients who are at risk. Significant morbidity and mortality are linked to identifiable risk factors in patients with univentricular hearts, including poor ventricular function, significant atrioventricular valve regurgitation (AVVR)/stenosis, aortic coarctation/obstruction, collaterals of hemodynamic significance, elevated pulmonary vascular resistance >3 Wood units m^2, and distortion/hypoplasia of pulmonary arteries [3,4]. The majority of the time, echocardiography can deliver the anatomical and functional information needed for BDG surgery. In addition to its reliance on acoustic windows that may not be ideal, echocardiography has difficulties in the evaluation of distal pulmonary arteries and APCs. This is especially true in patients who have chest deformities as a result of previous cardiac procedures and have hyperinflated lungs [5,6].

The morbidity and mortality of children for whom Fontan surgery is suggested is decreased when bidirectional cavopulmonary anastomosis (the Glenn technique) is performed as the initial stage of palliative care for univentricular patients. In various patient cohorts, the death rate associated with cavopulmonary anastomosis ranges from 1 to 13% [7]; risk factors for mortality and morbidity include age, the length of cardiopulmonary bypass, and the requirement for re-intubation. In patients with hypoxemia or low cardiac output syndrome, high pulmonary artery resistance, ventricular dysfunction, AV valve regurgitation, and anastomosis obstruction must be checked out [8].

Thrombotic events following BDG were reported in one systematic review to range from 0 to 28% [9]. Manlhiot et al. reported the incidence of thrombotic events following SCPA as high as 28% [10]. Thrombotic prophylaxis protocols vary between centers. Regimens include aspirin, enoxaparin, or warfarin as single agents or in combination [9]. At our institution, daily aspirin is the standard regimen used for achieving thrombotic prophylaxis after BDG. Even though our patient was maintained on aspirin since the surgery, she developed acute thrombosis of the BDG, causing severe hypoxia and hemodynamic compromise.

Early detection and rapid, timely intervention are of utmost importance in managing thrombosis and obstruction of the Glenn anastomosis. Options include medical measures, catheterization, and sometimes, surgical revision [9,10].

In our patient, medical measures using fluid expansions and heparin infusion failed to re-establish blood flow through the Glenn anastomosis. There was no improvement in oxygenation and she continued to have severe hypoxia despite receiving 100% oxygen supply and mechanical ventilation, leading to myocardial suppression, hypotension, and peripheral hypoperfusion requiring inotropic support. Non-surgical intervention was discussed and a decision was made for interventional catheterization as a last resort before surgery. Initially, attempts at balloon dilatation failed. However, a trial was made to insert two overlapping stents across the thrombosed segment of the distal Glenn anastomosis, which resulted in success in re-establishing pulmonary blood flow, avoiding surgical intervention for the patient.

Several studies have reported success in re-establishing pulmonary blood flow in thrombosed SCPA using catheter-based thrombolysis and stenting. Nomura K et al. reported successful use of catheter-directed thrombolysis in a patient with thromboembolism of the subsegmental branches of the right pulmonary artery after BDG anastomosis [11]. Mukherji A et al. reported success in thrombolysis and balloon dilatation of a thrombosed Glenn anastomosis in a 7-month-old infant [12]. Our study also reports the successful use of catheter-based techniques for the management of such cases.

4. Conclusion

Obstructed Glenn circulation, although very rare, can be lethal. Catheter-based endovascular stent implantation of Glenn anastomosis can be efficient, lifesaving, and a successful alternative to invasive surgical intervention. We reported a case in which using two overlapping stents to open a thrombosed Glenn anastomosis had been successful. This could be a viable option for certain patients to avoid surgical intervention. This report adds to the pool of previous studies demonstrating the efficiency of catheter-based interventions in patients with thrombosis after bidirectional Glenn (BDG).

Patient consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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Authorship

All authors attest that they meet the current ICMJE criteria for authorship.

Author contribution

Writing the manuscript: Oadi N. Shrateh, Afnan W.M. Jobran, **Imaging description:** Oadi N. Shrateh, Nizar Hijjeh, Nidal Haymouni, Iyad Sbeitan, **Reviewing & editing the manuscript:** Oadi N. Shrateh, Samer Abdelraziq, Ruba Rizik, Ahmed Darsalim, Mohammed Abutaqa.

Guarantor

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Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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