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CASE REPORT

INTERMEDIATE

CLINICAL CASE

Resolution of Hypoxia and Ascites With Percutaneous Intervention of Mustard Baffle Obstruction and Leak

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ABSTRACT

A 45-year-old man with history of Mustard repair for transposition of the great arteries, cirrhosis, and chronic hypoxemic respiratory failure presented for subacute worsening of his chronic symptoms, which were found to be secondary to a previously unrecognized baffle stenosis and leak. Percutaneous intervention resolved his ascites and hypoxia. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2020;2:1079–83) © 2020 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

A 45-year-old man with complex medical history, including Mustard repair for simple transposition of the great arteries (D-TGA) presented for dyspnea. His D-TGA was palliated by balloon atrial septostomy and Blalock-Hanlon atrial septectomy as a neonate, followed by a Mustard repair at 2 years of age with a complicated post-operative course requiring revision with Dacron (Maquet, Rastatt, Germany) grafts 1 day after the initial procedure (**Figure 1**). Since then, he developed multiple arrhythmias including sinus node dysfunction necessitating pacemaker implantation at age 9 years, ventricular tachycardia requiring ablation at age 35 years, atrial tachycardia, and atrial fibrillation status post-ablation 1 year ago. Additionally, he had chronic resting hypoxia (saturations of 88% to 92% on room air) for which he had been receiving 2 l of oxygen for multiple years. He had cirrhosis (Child Pugh class B complicated by hepatic encephalopathy and ascites) that was first diagnosed 20 years ago after he underwent exploratory laparotomy for a ruptured colon. Since age 40 years, he had received monthly therapeutic paracenteses.

The patient presented with 3 to 4 months of worsening of his chronic exertional dyspnea, alongside increased lower extremity swelling and ascites requiring more frequent therapeutic paracenteses. He was referred to our congenital heart specialty center by his general cardiologist because of medical complexity.

LEARNING OBJECTIVES

- To review Mustard repair anatomy and physiology.
- To understand the clinical presentation and management of baffle complications.

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ABBREVIATIONS AND ACRONYMS

D-TGA = simple transposition of the great arteries

IVC = inferior vena cava

RV = right ventricle

SVC = superior vena cava

DIFFERENTIAL DIAGNOSIS

We were concerned about a long-term complication of the patient's Mustard repair. Systemic baffle stenosis may lead to elevated systemic pressures and explain his liver dysfunction, edema, and ascites. A baffle leak with right-to-left shunt would explain his hypoxia. Right ventricle (RV) failure may develop as a result of chronic exposure to systemic pressures with D-TGA anatomy. Tricuspid regurgitation may develop secondary to RV dysfunction or because of altered geometry post-surgery. Finally, atrial arrhythmias, including both bradyarrhythmias and tachyarrhythmias, are especially common in patients with a history of Mustard repairs.

INVESTIGATIONS

Pacemaker interrogation showed rate-controlled atrial fibrillation with minimal pacing requirements. A transthoracic echocardiography revealed moderate RV dysfunction without significant valvular regurgitation. Cardiac computed tomography demonstrated moderate to severe narrowing of the systemic baffle superior limb (Figure 2A) and moderate to severe narrowing of the inferior limb with dilation of the inferior vena cava (IVC) hepatic segment (Figure 3A).

MANAGEMENT

The patient was taken to the catheterization laboratory for evaluation and possible intervention of a baffle stenosis. Angiography demonstrated superior vena cava (SVC) and IVC baffle obstructions with mean gradients of 5 and 4 mm Hg, respectively (Figures 2B and 3B). In addition, there was a baffle leak across the systemic baffle superior limb with right-to-left shunting (Figure 2B). Cardiac index was low (1.8 l/min/m^2), pulmonary vascular resistance was normal (<2 WU), and pulmonary blood flow to systemic blood flow ratio was 0.9:1.

The pacemaker leads traversing the superior limb were first extracted by using a combination of a 14-F Guidelight laser sheath and an 11-F TightRail (Spectranetics, Colorado Springs, Colorado). Then, a Palmaz XL 4010 stent (Cordis, Santa Clara, California) followed by an 8-zig 3.4-cm Cheatham Platinum covered stent (Braun Interventional Systems, Hopkinton, New York) was deployed to relieve the SVC baffle obstruction and leak (Figure 4). The IVC limb

FIGURE 1 Mullen Diagram of Mustard Repair

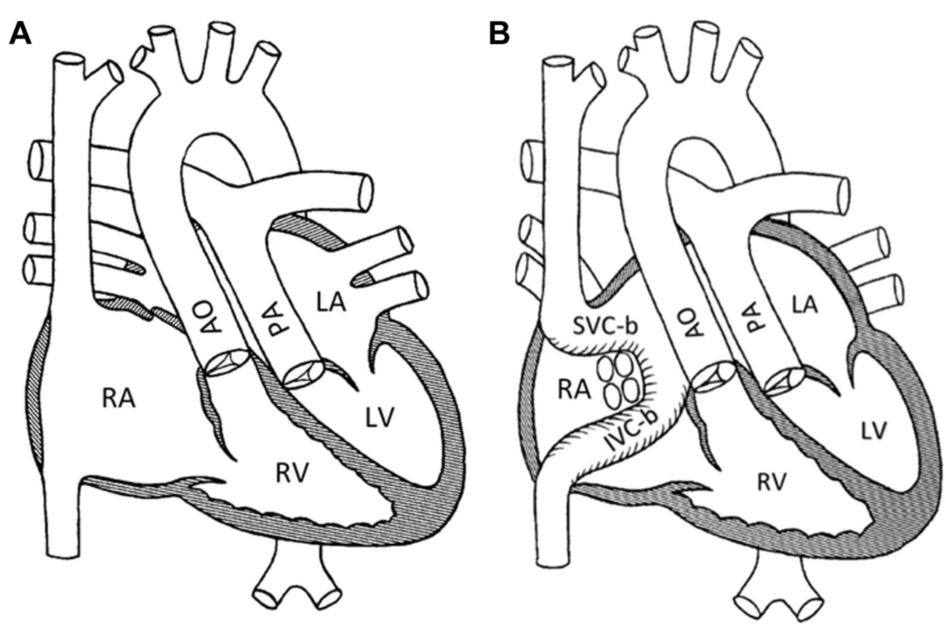
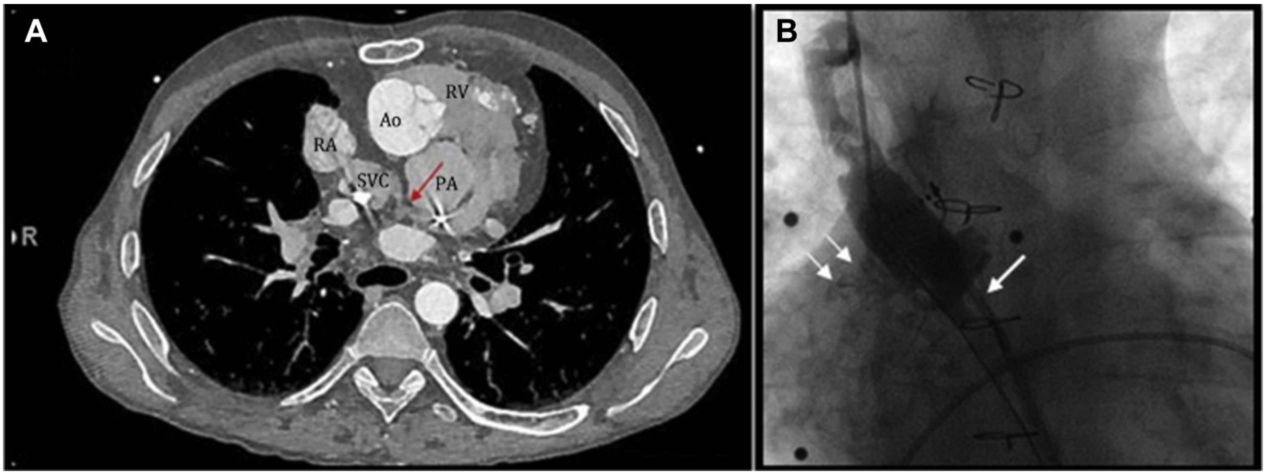


Diagram depicting our patient's cardiac anatomy **(A)** before and **(B)** after Mustard repair. Systemic blood is redirected to the LA via the SVC-b and IVC-b. Meanwhile, pulmonary blood is redirected to the RA via a pulmonary baffle (not pictured). Our patient had stenosis in the IVC-b segment in addition to combined stenosis and leak in the SVC-b segment. AO = aorta; IVC-b = systemic baffle inferior limb; LA = left atrium; LV = left ventricle; PA = pulmonary artery; RA = right atrium; RV = right ventricle; SVC-b = systemic baffle superior limb.

FIGURE 2 Systemic Baffle Superior Limb Stenosis



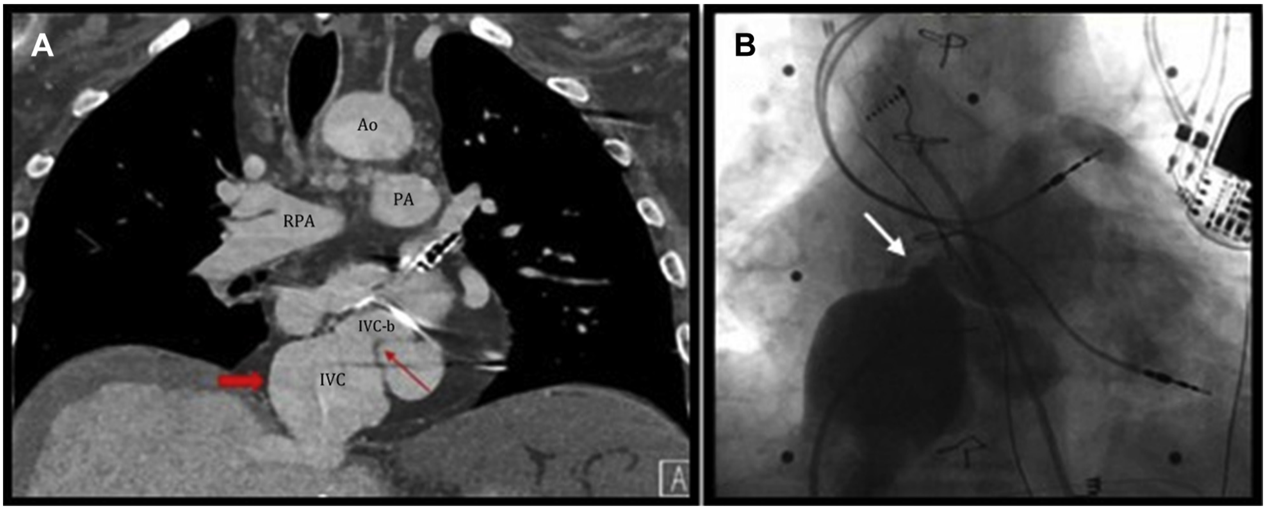
(A) Axial view computed tomography scan showing moderate to severe narrowing of the systemic baffle superior limb **(red arrow)**. **(B)** Angiogram of the systemic baffle superior limb demonstrating severe stenosis **(solid white arrow)** and baffle leak with right-to-left shunting **(small white arrows)**. Abbreviations as in Figure 1.

stenosis was relieved by placing a 36-mm Max Large Diameter stent (Boston Scientific, Marlborough, Massachusetts) (Figure 5). Following stent placement, pulmonary wedge angiography demonstrated patency of the pulmonary venous baffle. As the patient's pacemaker interrogation showed a low pacing requirement, a new pacemaker was not implanted.

DISCUSSION

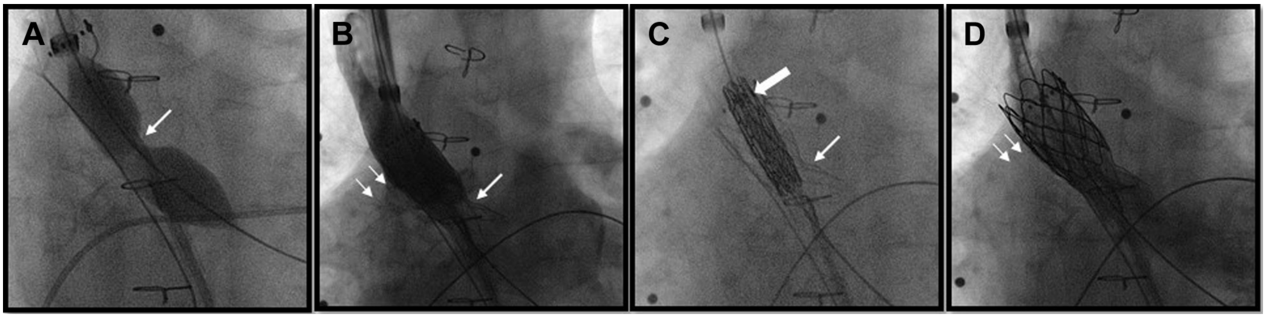
Baffles are surgical conduits composed of either biological or synthetic material commonly utilized for the redirection of blood flow in congenital heart surgery. Mustard and Senning repairs, commonly used in the 1960s to 1980s to correct

FIGURE 3 Systemic Baffle Inferior Limb Stenosis



(A) Coronal view computed tomography scan demonstrating narrowing of the systemic baffle inferior limb **(thin red arrow)** with upstream dilation of the IVC **(thick red arrow)**. **(B)** Angiogram of the systemic baffle inferior limb showing focal severe stenosis **(arrow)** with aneurysmal dilation proximal to stenosis. IVC = inferior vena cava; RPA = right pulmonary artery; other abbreviations as in Figure 1.

FIGURE 4 Systemic Baffle Superior Limb Intervention

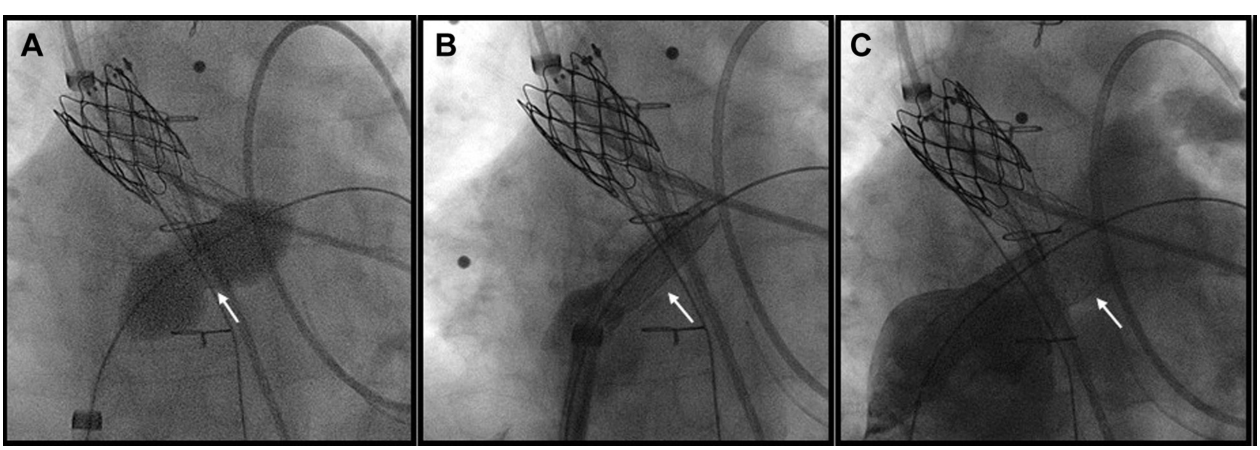


(A) Balloon angioplasty of the systemic baffle superior limb demonstrating tight waist **(arrow)**. **(B)** Angiogram showing Palmaz 4010 XL stent implantation with mild residual stenosis **(larger arrow)** and a baffle leak **(small arrows)**. **(C)** An 8-zig 3.4-cm CP covered stent **(larger arrow)** was positioned proximally within the Palmaz XL stent **(small arrow)**. **(D)** Angiogram demonstrates resolution of the baffle leak after covered stent deployment **(arrows)**.

previously fatal TGA, utilized baffles to reroute systemic and pulmonary blood flow such that the RV becomes the physiological systemic pump and the left ventricle becomes the physiological pulmonary pump (**Figure 1**) (1). Other surgeries that use baffles include Fontan procedures and the double-switch operation for congenitally corrected TGA (1).

Unfortunately, baffles are prone to deterioration. Among patients with Mustard repair, obstructions and leaks have a lifetime incidence of up to 30% (2-4). Their clinical presentation can be variable, including pulmonary hypertension (with pulmonary venous baffle obstruction), dyspnea, SVC syndrome, volume overload, chylothorax, and protein-losing enteropathy (1,3). Our patient had chronic hypoxia related to right-to-left shunting from an SVC baffle leak and liver cirrhosis secondary to IVC baffle stenosis and hepatic congestion. Although it is difficult to pinpoint when his baffle complications began, they likely went unrecognized for multiple years, as inferred by his symptom duration. Early referral to a specialized

FIGURE 5 Systemic Baffle Inferior Limb Intervention



(A) Balloon angioplasty of the systemic baffle inferior limb demonstrating moderate waist **(arrow)**. **(B)** Angiogram showing positioning of a 36 IntraStent Max (Boston Scientific) across the stenotic site **(arrow)**. **(C)** Angiogram of the inferior limb demonstrates no residual stenosis after stent deployment **(arrow)**.

clinical center of excellence could have prevented delays in care.

The largest available studies for transcatheter intervention in baffle stenosis report favorable outcomes with low rates of restenosis over midterm follow-up (5). Repairing defects may be performed by using a variety of approaches, with plain balloon angioplasty, bare-metal stents, covered stents, and endovascular grafts all being reasonable choices. We used a covered stent over the SVC baffle to exclude a concomitant leak. Because angioplasty alone has high restenosis rates, stents were placed to ensure longer-term patency and facilitate access for future electrophysiological procedures.

Finally, a notable complicating factor is the presence of pacemaker leads traversing the systemic baffle, which many patients with Mustard repair will have because of the high rates of sinus node dysfunction in this population. Joint procedures with lead extraction are often necessary. Previous reports show that combined transvenous lead extraction and stenting for baffle complications is safe (6).

FOLLOW-UP

The patient underwent an uncomplicated hospital course with dramatic symptom improvement. His dyspnea resolved, and he was successfully weaned off supplemental oxygen. At the 9-month follow-up, he required no more paracenteses. There has been no cause for reintervention.

CONCLUSIONS

Baffle obstruction and leak are common complications in patients with Mustard repair that may have subtle and variable presentations. Referral to a clinical center of excellence specialized in the care of patients with congenital heart defects may avoid delays in care.

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KEY WORDS baffle obstruction, baffle leak, congenital transposition of the great arteries, Mustard procedure