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Case Report

A Case Report: Successful Cardiac Catheterization Management of Post-Glenn complete Thrombosis of the Left Pulmonary Artery and Severe SVC to RPA Anastomosis Stenosis in a 1- Year-Old Infant

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Abstract

The Glenn palliation is a vital step in the staged surgical management of patients with single ventricle physiology. While it offers hemodynamic improvement, certain post-operative complications can be life-threatening.

Case Presentation: We report a case of a 1-year-old with single ventricle physiology, who underwent Glenn palliation and subsequently presented with severe complications, including complete thrombosis of the left pulmonary artery (LPA) and significant stenosis at the superior vena cava-right pulmonary artery (SVC-RPA) anastomotic site. The child presented clinically with profound cyanosis and features suggestive of superior vena cava syndrome. These complications were successfully managed via cardiac catheterization, with repermeabilization of the LPA and stenting of both the LPA and SVC-RPA junction.

Keywords: acute coronary syndrome; essential thrombocytemia; cardiovascular surgery

Introduction

The Glenn procedure, also known as the bidirectional Glenn or hemi-Fontan operation, represents an intermediate stage in the surgical palliation of patients with single ventricle congenital heart defects. By directing the superior vena cava (SVC) blood flow directly to the pulmonary arteries, the Glenn procedure aims to reduce the volume load on the single ventricle while providing passive pulmonary blood flow. This improves oxygen saturation levels and preconditions the pulmonary vascular bed for the subsequent Fontan procedure.

Despite its benefits, the Glenn operation is not devoid of potential complications. These can range from mild and self- limiting issues to severe life-threatening scenarios, the latter of which mandates prompt recognition and intervention. Thrombosis of the pulmonary arteries and stenosis at the SVC-pulmonary artery anastomotic site are among the more severe complications that can profoundly affect a patient's clinical status.

In this report, we delve into a particularly challenging case of a 1-year-old post-Glenn palliation, highlighting the complexities encountered and the therapeutic strategies employed for a favorable outcome.

Case Presentation:

A 1-year-old patient with a history of single ventricle physiology, post-Glenn palliation,

Clinical presentation:

Severe cyanosis and lethargy.

Oxygen Saturation: Significantly reduced on pulse oximetry, consistent with the patient's cyanotic presentation.

Head and Neck Examination:

Facial Edema: Swelling was evident, particularly around the eyes and cheeks.

Jugular Venous Distention (JVD): Prominent jugular veins were noticeable, consistent with superior vena cava syndrome.

Echocardiography findings:

Single Ventricle Physiology: Echocardiography confirmed the anatomy of a dominant Right ventricular chamber, consistent with the patient's known single ventricle physiology.

Systemic Venous Return:

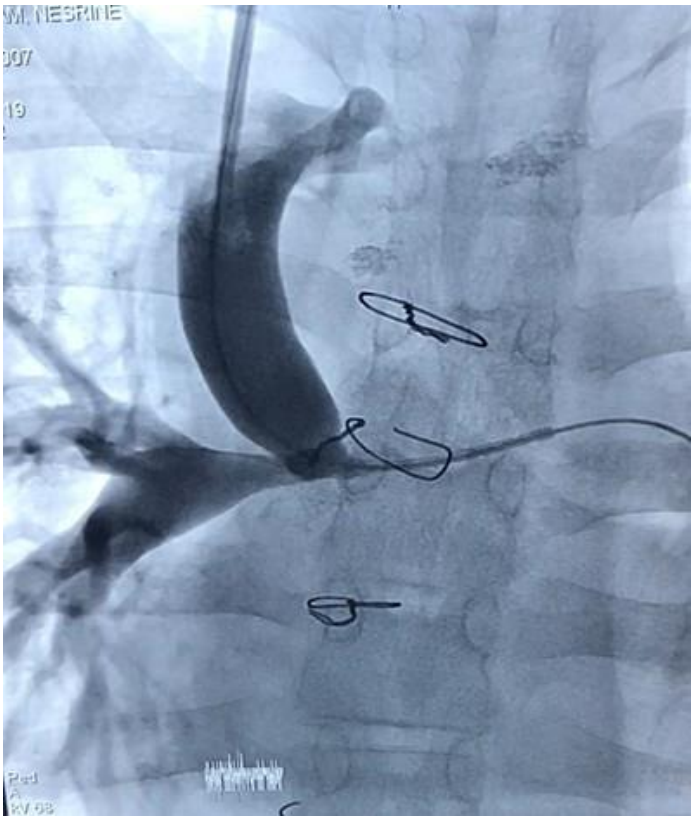
Superior Vena Cava (SVC) Flow: Turbulent flow was visualized in the SVC, suggesting significant obstruction or stenosis at the site of its anastomosis with the right pulmonary artery.

Pulmonary Artery (PA) Evaluation:

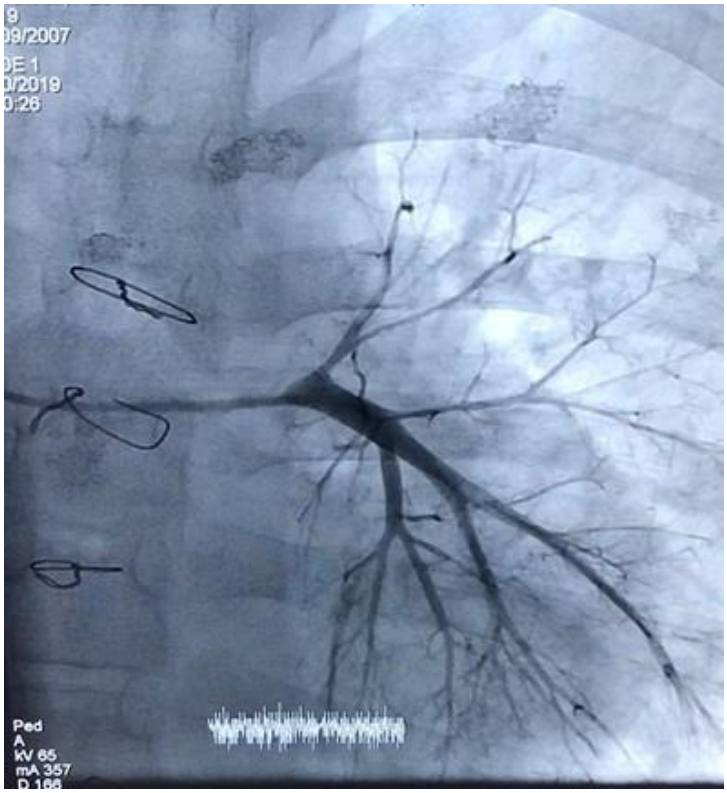
Right Pulmonary Artery (RPA): Normal flow and patency were observed.

Left Pulmonary Artery (LPA): Absence of color flow Doppler signal in the LPA, suggestive of complete occlusion or thrombosis.

Bidirectional Glenn Angiography showing a complete LPA thrombosis and severe SVC/RPA anastomosis stenosis. Note dilated SVC and Veno-venos fistulas



Hydrophilic Guide wire was gentely advanced throught the long LPA thrombus



NIH 4F catheter was than advanced on the guide wire, selective injection into the LPA.

Angiography:

1.Superior Vena Cava (SVC) to Right Pulmonary Artery (RPA) **Anastomosis:**

Stenosis:

- A localized narrowing was visualized at the SVC-RPA anastomotic site. The narrowing was consistent with stenosis and demonstrated turbulent flow during the contrast injection.

2. Left Pulmonary Artery (LPA):

Complete Occlusion:

The LPA showed a complete lack of contrast opacification, indicating full thrombosis or occlusion.

1.Preparation:

- Sedate the patient appropriately, ensuring airway patency and vital sign stability.
- Sterilize and drape the chosen catheterization site(s), commonly the femoral vein and/or artery.

1. Vascular Access:

- Utilizing a needle, puncture the jugulare internal vein.
- Introduce a sheath over the guidewire to maintain venous access.

Initial Assessment:

- Perform a baseline angiogram to visualize the anatomy and confirm suspected complications (see above).

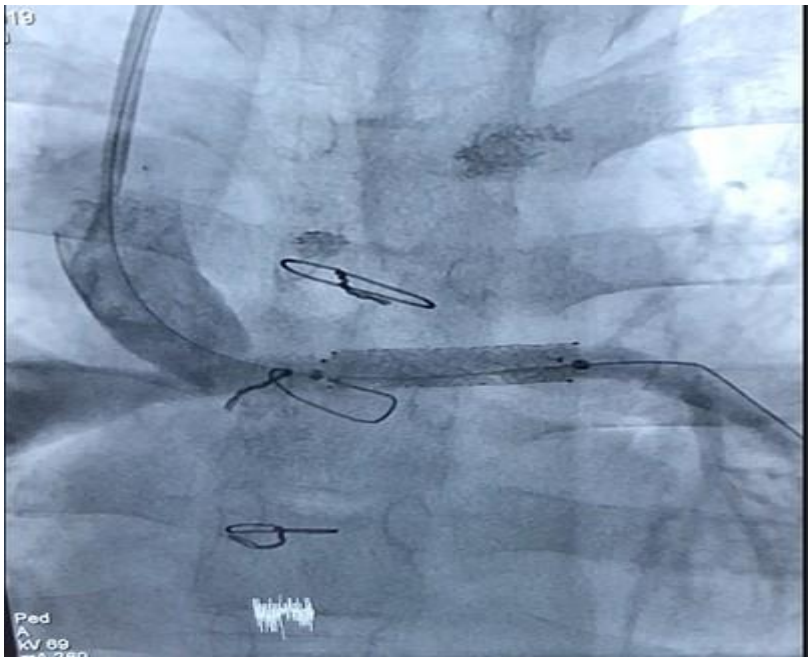
Recanalization of Thrombosed LPA:

- Under fluoroscopic guidance, navigate a hydrophilic guidewire across the occluded segment of the LPA.
- Use balloon Tayshak II 6 mm to open up the occluded segment by inflating the balloon at the site of occlusion.
- Deploy the stent, alleviating the stenosis.
- Again, validate stent placement and improved flow with an angiogram.

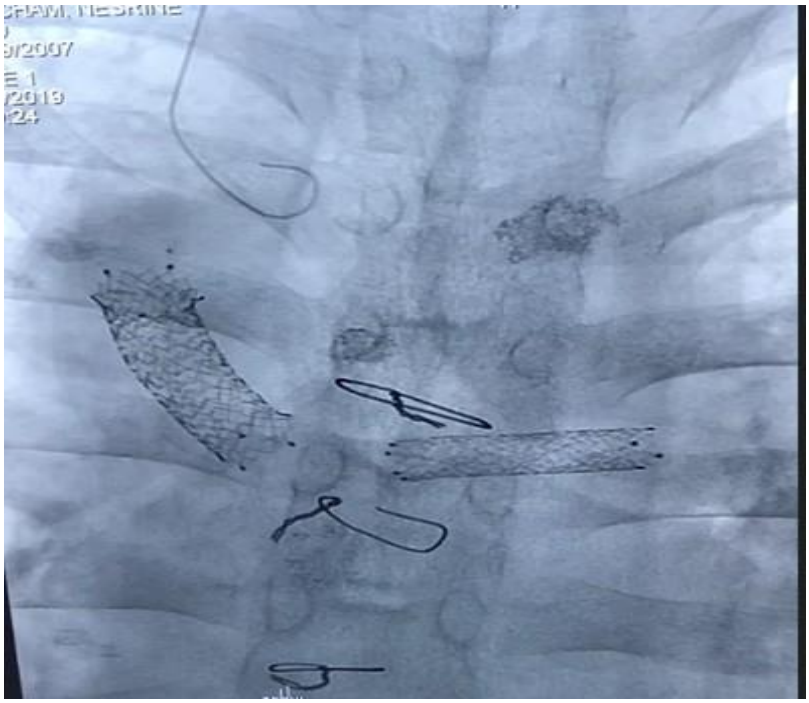
Stenting of the LPA:

- Position a Formula 6/16 stent over the guidewire, ensuring accurate placement across the previously occluded segment.
- Deploy the stent by inflating the balloon. The stent expands and scaffolds the vessel open.
- Confirm stent position and patency via angiography.
- Addressing SVC-RPA Anastomosis Stenosis:
- Direct the guidewire across the stenosed SVC-RPA junction.
- Position another stent Formula 10/20 mm over this guidewire, ensuring it spans the stenotic segment.

Left Pulmonary artery recanalisation



Stent position in the LPA



Second stent to treat SVC to RPA anastomosis thrombosis

Discussion:

In children with single ventricle physiology who have undergone Glenn palliation, the postoperative period can present with an array of complications. Two such severe complications are thrombosis of a major pulmonary artery and significant stenosis at the anastomotic site of the

superior vena cava (SVC) with the pulmonary artery. In the described scenario, the child manifested both these complications

Surgical Intervention:

- Surgical revision of the Glenn palliation or direct surgical correction of the noted complications is a possibility. However, reoperation carries inherent risks, especially if the child is hemodynamically unstable or if there are other contraindications for surgery.

1. Cardiac Catheterization:

- This allows for both diagnostic assessment and therapeutic interventions. Balloon angioplasty can open stenosed areas, and stents can maintain vessel patency. Thrombosed arteries can be recanalized using catheter-based techniques.

Why Choose Cardiac Catheterization?:

Minimally Invasive: Compared to surgical intervention, cardiac catheterization is less invasive, thus reducing recovery time and potential morbidity.

Immediate Relief: The interventions, like stenting or balloon angioplasty, can provide immediate hemodynamic benefit by addressing the obstruction or thrombosis.

Versatility: Catheterization offers the flexibility to manage multiple issues in one setting. In this case, both the SVC stenosis and LPA thrombosis could be addressed.

Reduced Risk: Avoiding reoperation minimizes risks associated with general anesthesia, surgical wound complications, and postoperative adhesions.

Diagnostic & Therapeutic: Cardiac catheterization allows for immediate angiographic assessment, verifying the success of the interventions and ensuring other areas are not compromised.

Trending Practice: Over the years, the success and safety of catheter-based interventions in congenital heart disease have burgeoned, making it a preferable option in select scenarios.

In conclusion, the decision to pursue cardiac catheterization in this child was based on the severity and complexity of the complications, the immediate therapeutic options available through catheterization, and the aim to avoid more invasive surgical re-interventions. As always, the choice of therapy should be individualized, considering the specific anatomy, the patient's clinical condition, and the expertise of the treating team.

Conclusion:

The management of children with single ventricle physiology, particularly following Glenn palliation, is complex and multifaceted. The emergence of severe complications, such as significant stenosis at the anastomotic site and complete thrombosis of major pulmonary arteries, emphasizes the critical need for meticulous post-operative monitoring and early intervention. These complications not only deteriorate hemodynamics but also jeopardize the patient's candidacy for future palliative stages.

In this reported case, the successful utilization of cardiac catheterization demonstrated its pivotal role in the contemporary management of post-Glenn complications. By allowing for real-time diagnostic evaluation and simultaneous therapeutic intervention, catheterization offers a minimally invasive yet efficacious avenue to restore pulmonary blood flow and rectify hemodynamic imbalances.

The choice of cardiac catheterization over other therapeutic options was underscored by its ability to offer immediate relief, minimize potential morbidity associated with reoperations, and provide comprehensive assessment of the circulatory system. Moreover, the intervention exemplifies the growing trend and trust in catheter-based procedures in pediatric cardiology, driven by technological advancements and growing expertise.

It is imperative for clinicians to be cognizant of the potential complications following Glenn palliation, to detect them early, and to have a strategic blueprint for management. Cardiac catheterization, as evidenced in this case, can be the linchpin in achieving optimal outcomes and setting the stage for successful future interventions.

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