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VALVULAR HEART DISEASE

CLINICAL CASE

The First Pediatric LAMPOON Procedure for TMVR (Valve-in-Ring)

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ABSTRACT

BACKGROUND The LAMPOON (laceration of the anterior mitral leaflet to prevent outflow obstruction) procedure is an innovative transcatheter technique designed to prevent left ventricular outflow tract obstruction during transcatheter mitral valve interventions.

CASE SUMMARY A 14-year-old male patient with a history of infective endocarditis, previously treated with surgical aortic valve replacement and mitral annuloplasty, presented with severe mitral regurgitation and decompensated heart failure. Owing to prohibitive surgical risk, a transcatheter mitral valve-in-ring replacement (TMViR) was performed with adjunctive LAMPOON to prevent left ventricular outflow tract obstruction. This is to our knowledge the first reported pediatric case of TMViR facilitated by LAMPOON. The procedure was technically successful, and the patient recovered without major complications.

DISCUSSION This case highlights a novel application of LAMPOON in the pediatric population. Given the absence of surgical options, this approach offers a life-saving, minimally invasive alternative and expands the role of structural interventions in children with complex valvular disease.

TAKE-HOME MESSAGES LAMPOON can enable safe TMViR in high-risk pediatric patients. Successful outcomes require careful planning, imaging, and multidisciplinary coordination. (JACC Case Rep. 2025;30:105094)

HISTORY OF PRESENTATION

A 14-year-old male adolescent presented with worsening dyspnea, fatigue, and reduced exercise tolerance. He was limited to NYHA functional class III. On physical examination, he appeared cachectic, with a body weight of 40 kg, a height of 1.57 m, and a body

TAKE-HOME MESSAGES

- LAMPOON can enable safe TMViR in high-risk pediatric patients.
- Successful outcomes require careful planning, imaging, and multidisciplinary coordination.

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ABBREVIATIONS AND ACRONYMS

LAMPOON = laceration of the anterior mitral leaflet to prevent outflow obstruction

LVOT = left ventricular outflow tract

TMViR = transcatheter mitral valve-in-ring replacement

TTE = transthoracic echocardiography

VA-ECMO = venoarterial extracorporeal membrane oxygenation

mass index of 16.2 kg/m^2. Laboratory work-up revealed elevated N-terminal pro-B-type natriuretic peptide (2,456 pg/mL), anemia (hemoglobin: 88 g/L), elevated lactate dehydrogenase (1,504 U/L), and increased free hemoglobin (1.00 g/L).

PAST MEDICAL HISTORY

Three years prior, the patient was treated for infectious endocarditis and subsequently underwent surgical mitral valve annuloplasty with an incomplete flexible ring (Carbomedics Annuloflex #28, Sorin Group), and he underwent surgical aortic valve replacement with a mechanical prosthesis (St Jude Regent #19, Abbott) in 2019. His noncardiac history was notable for liver cirrhosis, with laboratory findings of elevated direct bilirubin (2.21 mg/dL), elevated aspartate aminotransferase (107 U/L), and liver elastography showing F3 stage fibrosis.

DIFFERENTIAL DIAGNOSIS

The differential diagnoses included recurrent severe mitral regurgitation due to mitral valve annuloplasty failure, pulmonary hypertension, and worsening right heart function.

INVESTIGATIONS

Baseline transthoracic echocardiography (TTE) demonstrated preserved left ventricular ejection fraction (51%) with a dilated left ventricle and severe mitral regurgitation, characterized by a restricted posterior mitral leaflet and a vena contracta of 0.74 cm (Figure 1). The mean mitral valve gradient was 12 mm Hg, and the mean left ventricular outflow tract (LVOT) gradient measured 15 mm Hg. Severe

FIGURE 1 Baseline Transesophageal Echocardiography Findings

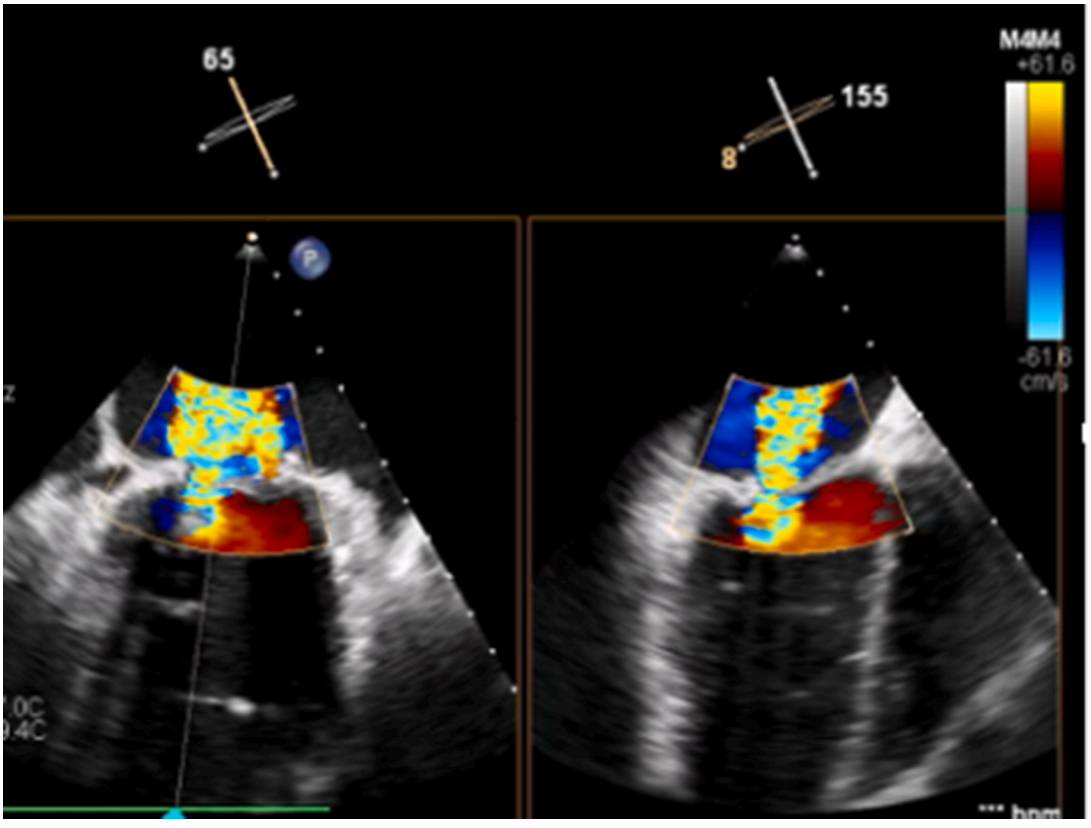
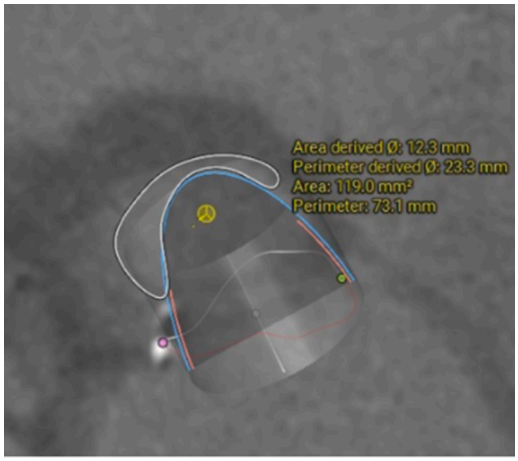


FIGURE 2 **Predicted Neo-LVOT Area on Multislice Computed Tomography**



LVOT = left ventricular outflow tract.

tricuspid regurgitation was also present (vena contracta: 0.68 cm), with right ventricular dysfunction (TAPSE: 11 mm).

Right heart catheterization revealed a systemic blood pressure of 101/65 mm Hg (mean: 77 mm Hg), mean right atrial pressure of 14 mm Hg, pulmonary artery pressure of 75/32 mm Hg (mean: 46 mm Hg), and a pulmonary wedge pressure of 26 mm Hg with a prominent v-wave. Pulmonary vascular resistance was elevated at 7.6 indexed WU; systemic vascular resistance was 23.9 indexed WU. The cardiac index was 2.6 L/min/m^2. Multislice computed tomography was performed for procedural planning. Three-dimensional reconstruction (3-Mensio software) revealed a critically small predicted neo-LVOT area of 119.8 mm^2 (**Figure 2**) and a narrow aortomitral angle of 112° (**Figure 3**).

MANAGEMENT

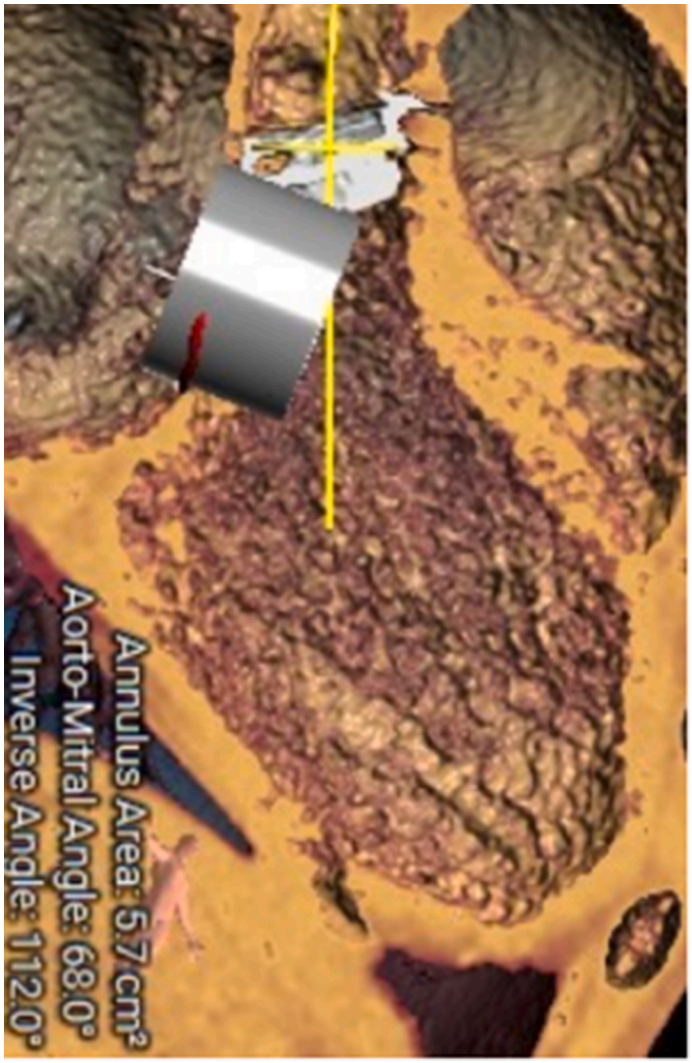
Given the severity of the patient's clinical condition, history of prior cardiac surgery, and elevated pulmonary vascular resistance, the heart team determined that he was at extreme surgical risk and that further intervention would be futile. As a result, the focus shifted to palliative and symptomatic management. Percutaneous options for treatment were investigated, however this was also complicated by a combination of an incomplete flexible annuloplasty ring and critically small predicted neo-LVOT, thus the decision was made to perform a base-to-tip LAMPOON (laceration of the anterior mitral leaflet to prevent outflow obstruction) procedure to mitigate the risk of LVOT obstruction.

Bespoke valve sizing was performed via multislice computed tomography measurements to allow for prediction of how the incomplete flexible annuloplasty ring would expand during balloon-expandable valve implantation (**Figure 4**). A 27.5-mm Myval transcatheter heart valve (Meril Life Sciences) was selected, with oversizing of 19%. It was felt that with the improved effective orifice area, hemodynamics would be favorable with this valve, with an expected mean transmitral gradient of <5 mm Hg.

The procedure was performed under general anesthesia with transesophageal echocardiographic and fluoroscopic guidance. Owing to high procedural risk, prophylactic venoarterial extracorporeal membrane oxygenation (VA-ECMO) was initiated via right axillary artery and right internal jugular vein cannulation to maintain hemodynamic stability (**Figure 5**). A transseptal approach was used for transcatheter mitral valve-in-ring replacement (TMViR). After atrial septal puncture and septostomy, dual steerable catheters were positioned in the left atrium to perform a base-to-tip LAMPOON.

To ensure coaxial valve deployment and reduce the risk of severe paravalvular leak and embolization, a percutaneous transapical access was established to facilitate guidewire rail creation. In our patient, this risk was further amplified by a severely angulated Emory angle of 35° (**Figure 6**), which significantly limited coaxial alignment. Additionally, the first-generation Myval features limited outer skirt coverage (~25% of frame height), providing less margin for positional error. Given these factors, we determined that implantation without an apical rail would carry an unacceptably high risk of transcatheter heart valve malposition and severe paravalvular leak. Coronary angiography was performed to prevent coronary injury during left ventricular apex puncture. After anterior mitral leaflet laceration, the Myval was implanted via conventional venous access using the transseptal technique, with deployment under rapid right ventricular pacing at 150 beats/min. The left ventricular apical access site was closed using an Amplatzer Duct Occluder II (Abbott). Final imaging confirmed stable valve

FIGURE 3 Aorto-Mitral Angle With 3-Dimensional Transcatheter Heart Valve Simulation



position, no paravalvular leak, and successful apical access closure. VA-ECMO was weaned and decannulated at the end of the procedure. A step-by-step demonstration of the procedural technique is provided in the supplemental videos (Videos 1 to 14).

OUTCOME AND FOLLOW-UP

The total procedure duration was 360 minutes (VA-ECMO time: 210 min), with a fluoroscopy time of 133 minutes and a dose-area product of 2,776 cGy × cm^2. A successful outcome resulting in an LVOT mean gradient of 13 mm Hg was confirmed by TTE (Figure 7). The patient remained hemodynamically stable throughout the procedure, followed by uneventful recovery, and he was discharged on postoperative day 4. The patient's condition improved rapidly, with a reduction in pulmonary artery pressure down to 40 mm Hg peak. At the time of discharge (postoperative day 5), the patient had a mean mitral valve gradient of 3 mm Hg with normal left ventricular function, and his NYHA functional class had improved from III to I.

At the 6-month follow-up, the patient had gained 9 kg lean mass and remained well. TTE revealed a mean LVOT gradient of 22 mm Hg and a well-functioning mitral valve, with a mean gradient of 4 mm Hg.

DISCUSSION

Currently there are no specific measurement criteria to predict potential LVOT obstruction in pediatric patients. Therefore, our decision to perform LAMPOON was based on findings from studies conducted in the adult population. To the best of our knowledge, this case represents the first documented pediatric LAMPOON procedure combined with TMViR in a 14-year-old patient with complex cardiac history. The decision to perform transseptal TMViR with LAMPOON under temporary mechanical circulatory support was made after a multidisciplinary structural heart team consensus, with the goal of optimizing the patient's hemodynamic and clinical status as a potential bridge to future surgical intervention when required.

Preprocedural imaging plays a crucial role in predicting the risk of LVOT obstruction and guiding procedural planning. Multislice computed tomography with three-dimensional reconstruction is a well-established method in adult LAMPOON cases for assessing the aortomitral angle and anticipated neo-LVOT area, enabling procedural modification to mitigate obstruction risks.[1,2] In our case, preprocedural multislice computed tomography analysis confirmed a high risk of LVOT obstruction after TMViR, reinforcing the necessity of anterior mitral leaflet laceration to prevent this complication.

Alternative strategies to allay LVOT obstruction, such as transcoronary alcohol septal ablation, have been described.[3] Although technically simple to perform, the results can be suboptimal in terms of effect on neo-LVOT, and there is a considerable

permanent pacemaker risk. For those reasons, the heart team opted against alcohol septal ablation in this patient in favor of LAMPOON, which offers an immediate and targeted solution.

The antegrade base-to-tip LAMPOON technique was specifically chosen because of the incomplete mitral annuloplasty ring; therefore, tip to base was contraindicated. These approaches are well documented in adult cases, with a step-by-step procedural description provided by Lisko et al.[4]

VA-ECMO support was implemented to secure hemodynamic stability in view of pulmonary hypertension and right ventricular dysfunction; the patient had no real surgical alternative in the event of complication.

Unlike valve-in-valve procedures, valve-in-ring implantation carries a higher risk of malalignment, as the transcatheter valve may sit "canted" within the incomplete ring, leading to incomplete apposition; if any portion of the outer skirt falls below the annular plane, the risk of significant paravalvular leak and embolization increases. To mitigate this, apical access was employed to create a guidewire rail, providing enhanced coaxial alignment and precise depth control during valve deployment in this anatomically challenging case.

Passage of the drainage ECMO cannula via right internal jugular vein was controlled under fluoroscopy, placing the tip at the center of the right atrium. This position ensures blood drainage from the right atrium for adequate ECMO performance, not interfering with upcoming intracardiac and inferior vena cava interventions. Having gained experience with VA-ECMO in transcatheter valve interventions,[5] we initiated VA-ECMO before any intracardiac intervention to ensure hemodynamic stability, considering the high pulmonary artery pressure and reduced right ventricular function.

CONCLUSIONS

This case demonstrates the feasibility of LAMPOON and percutaneous TMViR in carefully selected, critically ill pediatric patients who are unsuitable for surgery owing to prohibitive perioperative risk. Successful outcomes in such complex cases require meticulous procedural planning, comprehensive imaging, multidisciplinary collaboration, and thorough contingency preparation.

FIGURE 4 In-Situ Surgical Ring Internal Diameter Measurements

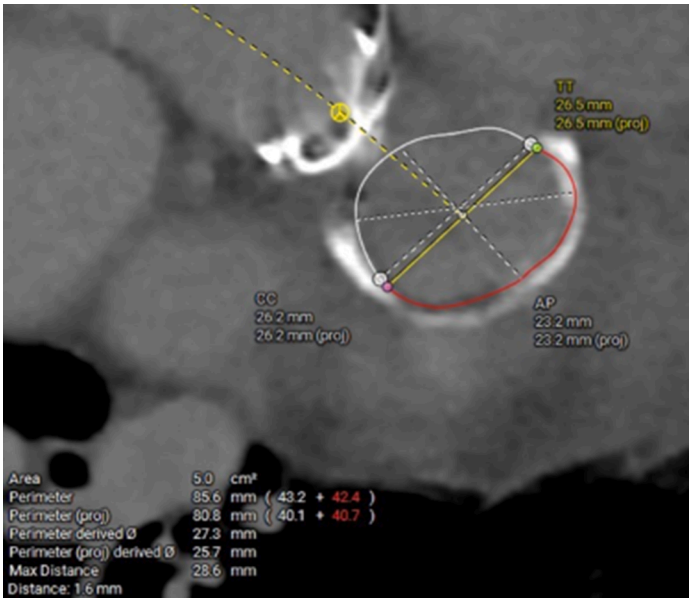


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FIGURE 5 VA-ECMO Cannulation

VA-ECMO = venoarterial extracorporeal membrane oxygenation.

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FIGURE 6 Emory Angle

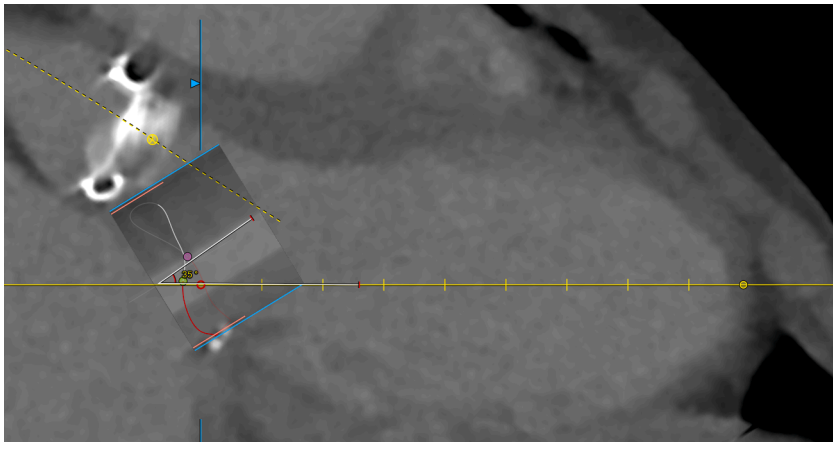

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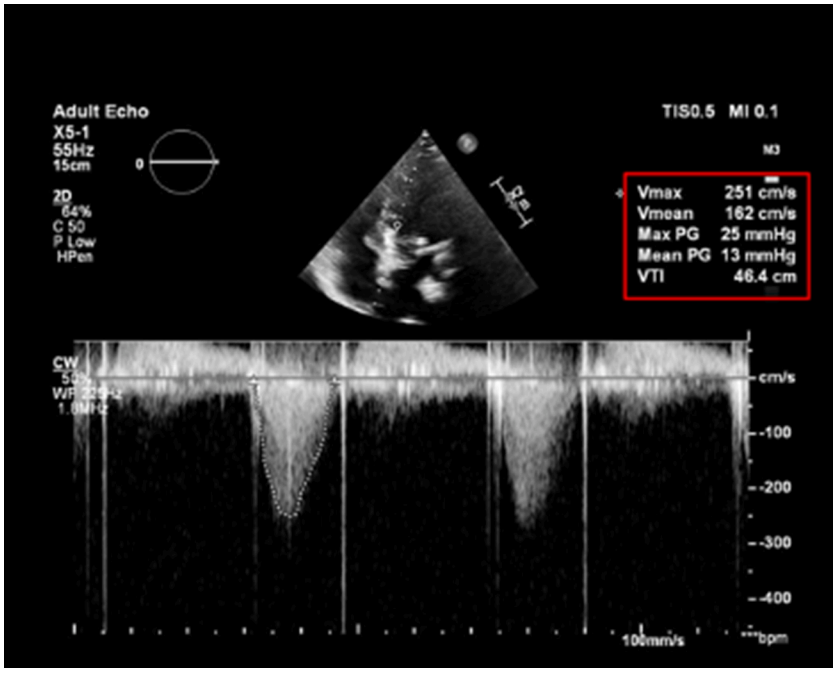
FIGURE 7 Postprocedural Transthoracic Echocardiography



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KEY WORDS computed tomography, echocardiography, hemodynamics, imaging, insufficiency, left ventricle, mitral valve, pediatric surgery, pulmonary hypertension, valve repair

APPENDIX For supplemental videos and figures, please see the online version of this paper.