Surgical ventricular restoration and mitral valve replacement in a pediatric patient with complex congenital heart disease and malignant ventricular arrhythmias

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CASE REPORT

We report the case of female child with neonatal (2006) diagnosis of aortic coarctation and partial atrioventricular septal defect, including cleft and hypoplasia of the left atrioventricular valve (LA VV) and primum atrial septal defect. After coarctation repair as a neonate, at 3 years of age she underwent partial closure of LAVV cleft and atrial septal defect closure. During separation from cardiopulmonary bypass, a large hematoma of the LV lateral wall was noticed along with ST abnormalities on the electrocardiogram. Injury to the circumflex coronary artery was suspected, but no revision of the intracardiac repair or attempt to revascularize the myocardium was performed. Postoperative extracorporeal membrane oxygenation support was necessary for approximately 12 hours to obtain a gradual LV recovery; clinical improvement allowed discharge to home on postoperative day 10. Discharge echocardiogram showed mild residual LAVV regurgitation and mild LV ejection fraction (EF) impairment. ST abnormalities persisted in the inferolateral and septal leads along with new pathologic Q waves in leads II and V5-6.

The patient was readmitted at 6 years of age for worsening LAVV regurgitation and increasing LV dimensions. Cardiac catheterization showed normal coronary arteries, and the patient underwent surgical closure of the residual LAVV cleft. Predischarge echocardiography showed moderate residual LAVV stenosis and regurgitation in association with moderate LV dilation and dysfunction. At 9 years of age, the patient was readmitted to the intensive care unit for loss of consciousness due to ventricular tachycardia. After defibrillation and restoration of sinus rhythm, an electrophysiology study was performed: Sequential mapping of LV identified the lower septal and apical regions as the focus for the arrhythmias, but transcatheter ablation was unsuccessful. The arrhythmias resolved with antiarrhythmic therapy (amiodarone and beta-blockers), and the child was discharged after 2 weeks. Shortly thereafter, clinical conditions worsened: Echocardiogram and cardiac magnetic resonance showed severe LAVV regurgitation and moderate stenosis, LV dilation and dysfunction (end-diastolic volume index, 127.2 mL/
m² end systolic volume index, 84.5 mL/m², EF 35%), and an extensive LV posterior scar, presumed to be a result of evolution of the ischemic injury and exacerbated by the arrhythmias (Figure 1, A-C).

After a discussion with the Heart Team, the patient underwent redo surgery consisting of posterior SVR and LAVV replacement with the mechanical prosthesis Sorin Bicarbon number 23 mm (Sorin, Saluggia, Italy). The LV was opened at the level of the inferior scarred wall, parallel to posterior descending artery. After digital identification of the border between normal and scarred tissue, termed the “transitional zone,” the incision was extended along the entire length of myocardial scar (Figure 1, D). The opening was closed by 2 direct continuous 2/0 Prolene sutures aiming to reapproximate the myocardium beyond the “transitional zone,” thus excluding the whole scar.

The child has been observed over 2 years; the last echocardiography showed a well-functioning prosthesis, with

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**FIGURE 1.** Preoperative cardiac magnetic resonance imaging showing an extensive LV posterior scar in late gadolinium enhancement (A, B), and the dilated LV (C) chamber due to an iatrogenic ischemic lesion in the circumflex artery during a previous surgical correction for a partial AVSD, possible ventricular arrhythmia trigger in a 9-year-old patient who underwent posterior SVR. Schematic surgical procedure of the posterior SVR showing the elevated and incised LV from the LAVV annulus (in yellow) to the apex and the “transitional zone” (black dashed line), representing the border between normal and scarred tissue. Blue arrows represent the direct continuous 2/0 Prolene sutures aiming to reapproximate the myocardium beyond the “transitional zone,” thus excluding the whole scar (D).

**VIDEO 1.** The LV is lifted up and opened at the level of the inferior scarred wall, parallel to the posterior descending artery. After initial opening and exposure, a digital exploration and identification of papillary muscles and the “transitional zone,” the border between normal and scarred tissues, are performed and the incision is extended along the entire length of myocardial scar. After careful examination of the left ventricular chamber, a first running suture from the annulus of LAVV toward the apex is performed to get the rapprochement of myocardium beyond the “transitional zone” and the exclusion of the scarred tissue. A second suture, started from the distal opening down to the basal segments is conducted, thus excluding whole scar. The remnant-excluded tissue is finally overlapped on the first suture to ensure a better hemostasis. Video available at: https://www.jtcvs.org/article/S0022-5223(19)31034-7/fulltext.
restored LV contractility and dimensions (end-diastolic volume index 82 mL/m², end-systolic volume index 40.6 mL/m², EF 50%) (Figure 2, A and B). No major arrhythmias or adverse clinical events have been recorded.

DISCUSSION
To the best of our knowledge, this is the first case of SVR in a pediatric patient who had undergone multiple major surgical procedures. Our 9-year-old patient had a large posterior scar due to LV remodeling and LAVV disease, and presented with dyspnea and malignant ventricular arrhythmias. Our surgical solution aimed to restore LV volume and shape and to exclude the myocardial scar, a potential trigger for electrical instability.

CONCLUSIONS
In ischemic heart failure, mitral valve repair and SVR have shown controversial results. In our patient, considering her young age, the higher risk of the need for a postoperative permanent pacemaker, and the potential restriction of annular growth, valve repair could have been considered over replacement. A mechanical prosthesis was chosen given the presence of mixed LAVV disease, the history of multiple valve repair attempts, and the need for a definitive surgical solution. The antiarrhythmic benefits of SVR has been demonstrated by several groups and may play an important role in cardiac resynchronization. In our patient, LV function and volumes improved after SVR and have remained stable during follow-up. The absence of adverse clinical events and arrhythmias speak favorably regarding the feasibility and efficacy of SVR.

References