JACC: CASE REPORTS VOL. 14, 2023

CASE REPORT

ADVANCED

CLINICAL CASE

Sternal-Sparing Transapical, Beating Heart Mitral Valve Repair in an Adult With Repaired Tetralogy of Fallot

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ABSTRACT

We present the case of a 60-year-old male patient who underwent tetralogy of Fallot repair at 7 years of age and then developed severe degenerative mitral regurgitation during adulthood. Given the increased surgical risk (obesity, obstructive sleep apnea syndrome, and reoperation), the patient underwent a successful microinvasive mitral valve repair with neochordae implantation. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2023;14:101818)

HISTORY OF PRESENTATION

A 60-year-old male patient with a history of repaired tetralogy of Fallot (ToF) was admitted to our hospital with reports of de novo worsening dyspnea and bilateral peripheral edema. He was then transferred to our Adult Congenital Heart Disease (ACHD) section for further investigation.

PAST MEDICAL HISTORY

The patient had a history of surgically repaired ToF in 1969 at the age of 7 years (no records were available regarding the surgical technique). However, he did not undergo regular cardiological follow-up until 1 month before hospitalization. In addition, his medical history was remarkable for arterial hypertension, smoke abuse, obesity (body mass index 41 kg/m^2), and obstructive sleep apnea syndrome (OSAS).

LEARNING OBJECTIVES

- New techniques for mitral valve repair in adult patients with congenital heart disease.

DIFFERENTIAL DIAGNOSIS

Given the clinical history of the patient, the differential diagnosis list included acute heart decompensation due to left ventricular dysfunction or left valve dysfunction, and acute right heart failure secondary to pulmonary or tricuspid regurgitation.

INVESTIGATIONS

A transthoracic echocardiogram showed severe mitral regurgitation (with pulmonary venous flow reversal)

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The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the Author Center.

Manuscript received November 16, 2022; revised manuscript received February 27, 2023, accepted March 2, 2023.

ISSN 2666-0849 https://doi.org/10.1016/j.jaccas.2023.101818

ABBREVIATIONS AND ACRONYMS

ACHD = adult congenital heart disease

OSAS = obstructive sleep apnea syndrome

ToF = Tetralogy of Fallot

caused by P2 flail, a dilated left ventricle (90 mL/m^2) with normal ejection fraction (69%), and a mildly dilated left atrium. No residual interventricular shunt was detected. The right ventricle showed normal function and dimensions, with a mild subpulmonary gradient (maximum/medium 29/13 mm Hg) and no pulmonary regurgitation. No electrocardiographic abnormalities were found (**Figures 1 and 2**).

MANAGEMENT

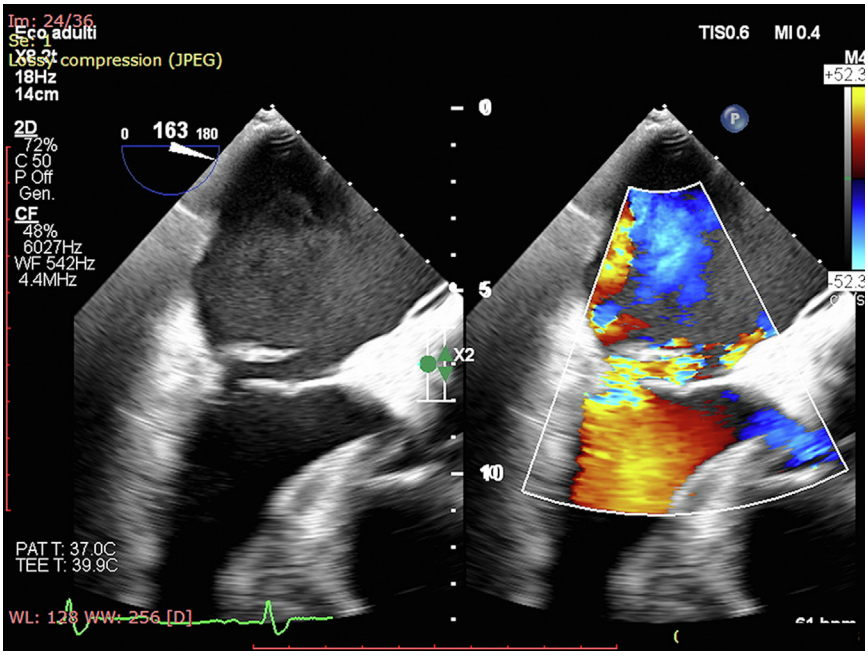
The case was discussed in our ACHD multidisciplinary round meeting and, given the higher risk of a conventional surgical procedure (re-sternotomy and anesthesia risks related to OSAS) and the favorable anatomy for a microinvasive mitral repair, the surgical team decided to perform a transapical off-pump, beating heart neochordae implantation. This medical decision was shared and discussed with the patient, who agreed to this microinvasive plan. The procedure was performed in a standardized fashion under general anesthesia and mechanical ventilation. Briefly, the cardiac apex was identified and exposed through an echocardiogram-guided fourth left space mini-thoracotomy, and two 2-0 Prolene concentric purse-string sutures (Ethicon Inc) were placed on it. After heparinization, the NeoChord DS1000 device (NeoChord, Inc) was inserted through an incision within the 2 purse-string sutures and, under real-time three-dimensional transesophageal echo guidance, 3 pairs of neochordae were implanted on the posterior leaflet at the level of P2 without any complications (Video 1). The total procedure time was 1 hour and 55 minutes. Extubation occurred 1 hour after surgery, and the patient did not require any intensive care unit stay. A transthoracic echocardiogram at discharge showed trivial residual mitral regurgitation (**Figure 3**).

DISCUSSION

Due to advances in the medical and surgical treatment of congenital heart disease (CHD), up to 90% of children born with CHD now reach adulthood.[1] Unfortunately, many ACHD who underwent previous repair, still experience residual intracardiac lesions or, less frequently, develop acquired disease (eg, coronary, valvular, or aortic disease).

Repaired ToF is more frequently associated with the development of pulmonary regurgitation during

FIGURE 1 **Preoperative Mitral Regurgitation**



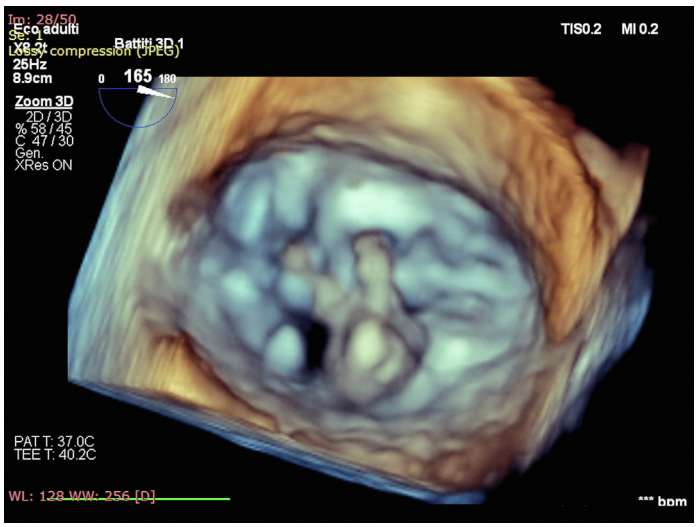
Preoperative bidimensional transesophageal echocardiographic image showing posterior leaflet prolapse **(left)** causing severe mitral regurgitation **(right)**.

follow-up. However, given the high risk of a redo surgery associated with the increasing expertise with the use of transcatheter valves, pulmonary regurgitation is currently preferentially managed by means of endoprosthesis implantation. Acquired degenerative diseases in this special population are rarely reported,[2,3] but their management might be a challenge when surgery is recommended.[4] Microinvasive surgery includes procedures that mimic conventional operations but are performed off-pump on a beating heart (eg, transapical neochordae implantation).[5,6] Thus far, as with transcatheter aortic valve replacement and edge-to-edge mitral valve repair, microinvasive procedures are still not widely performed in ACHD.

To the best of our knowledge, this case is the first of transapical neochordae implantation in an ACHD patient. Redo surgeries in this subset of patients are associated with a high risk of intraoperative and postoperative complications (sternal re-entry, bleedings, unexpected cardiac and pulmonary lesions, long cardiopulmonary bypass and cardioplegic arrest, postoperative cardiogenic shock, neurologic problems, arrhythmias, infections, and wound issues) and, consequently, lower overall postoperative survival.[7] To prevent these problems, congenital interventional cardiologists have already prioritized and standardized a few procedures to avoid (unless mandatory) conventional surgical reoperation (eg, pulmonary endoprosthesis or stent implantation). Unfortunately, current knowledge of acquired disease in ACHD remains limited, but it is going to become a serious issue in the very near future.[7]

Degenerative mitral regurgitation is not a frequent finding in ToF; however, it might require surgical treatment (according to current guidelines) when symptomatic and severe. In patients such as the current individual (with multiple risk factors such as OSAS, obesity, previous major cardiac intervention, and recent acute heart decompensation), a traditional surgical strategy might have been high risk. The absence of standardized surgical scores in this special population to predict postoperative mortality emphasizes the importance of an informed discussion with patients to offer them the best care. In fact, the more frequently used EuroSCORE (European System for Cardiac Operative Risk Evaluation) II for predicting in-hospital mortality after major cardiac surgery is not appliable to ACHD patients. In addition, patients with ToF who underwent late surgical repair (as in this case) frequently exhibit abnormal hypertrophy and biventricular fibrosis, which might negatively affect heart recovery after a redo cardiopulmonary bypass and cardioplegic arrest.[8] Even if no studies are available to compare transapical neochordae implantation vs transcatheter mitral valve repair (eg, MitraClip, Abbott), we were more confident with the first procedure, and we believe that neochordae implantation might be more physiological in restoring mitral competence at long-term follow-up.

FIGURE 2 **Intraoperative 3D Mitral Anatomy**

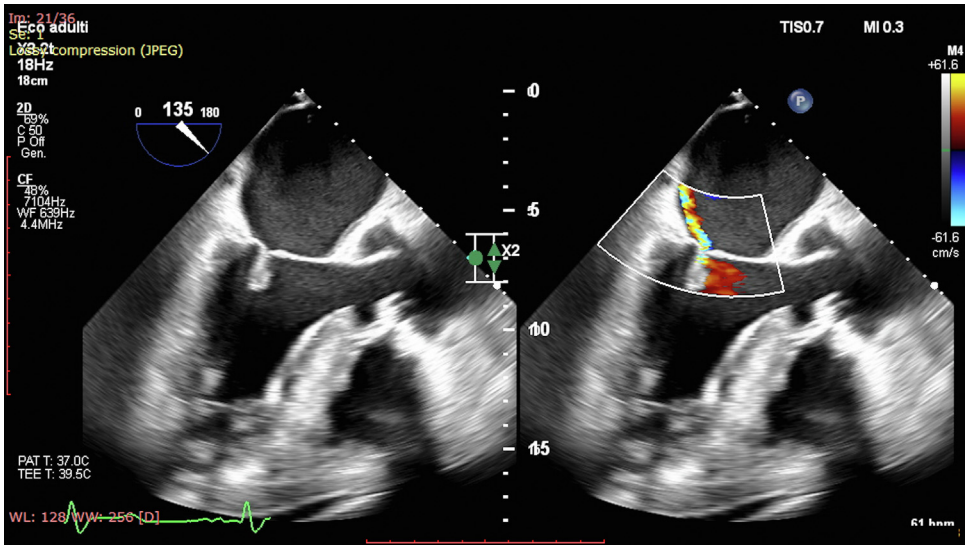


Preoperative tridimensional transesophageal echocardiographic images showing P2 mitral prolapse.

Given our experience with indications, technical aspects, and results of the transapical neochordae implantation,[9] we decided to direct the patient toward a microinvasive surgical strategy. In fact, according to our experience, in favorable mitral anatomies (isolated P2 prolapse), a transapical neochordae repair yields 5-year results similar to those of traditional open surgery in terms of freedom from death and reoperation.[10] Indeed, at 5 years of follow-up, incidence of severe mitral regurgitation in favorable anatomies after neochordae repair remains low (<15%).[11] Due to the novelty of this microinvasive procedure, the patient will require a strict follow-up. However, should long-term failure of this procedure occur, conventional redo mitral valve repair will be feasible.

With this case we want to highlight that microinvasive surgery can be successfully applied to complex cases, with the advantage of lowering procedural times, anesthesia risks, and, finally, hospital complications and length of stay.

FIGURE 3 **Postoperative Mitral Regurgitation**



Postoperative transesophageal echocardiographic images on mitral valve regurgitation.

FOLLOW-UP

The patient did not experience any major or minor postoperative complications according to MVARC (Mitral Valve Academic Research Consortium) criteria[12] and was discharged home 9 days after the procedure in good clinical condition. The patient will undergo strict clinical and echocardiographic follow-up during the first year (with outpatient visits every 3 months). These visits will then be reduced to one clinical and echocardiographic check-up every 6 months.

CONCLUSIONS

Transapical neochordae implantation off-pump on a beating heart in high-risk ACHD might be safe and feasible. This case highlights the importance of a multidisciplinary preoperative evaluation and proper planning to tailor the procedure according to each patient's characteristics. Indeed, microinvasive surgery might be considered as a good therapeutic alternative in high-risk ACHD patients with acquired cardiac disease. Furthermore, a microinvasive mitral valve repair will enable a more physiological approach to restoring normal mitral anatomy and function.

FUNDING SUPPORT AND AUTHOR DISCLOSURES

The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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KEY WORDS **adult congenital heart disease, cardiac surgery, microsurgery, mitral valve disease**

APPENDIX **For a supplemental video, please see the online version of this paper.**