

# Robotic mitral valve repair and resection of a pericardial cyst in Maffucci syndrome with sternal manifestations: A case report



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Multiple enchondromas in the hand, characteristic for Maffucci syndrome.

## CENTRAL MESSAGE

We present a case report with video of robot-assisted mitral valve repair in a patient with Maffucci syndrome with sternal manifestations, contraindicating a conventional sternotomy approach.

▶ Video clip is available online.

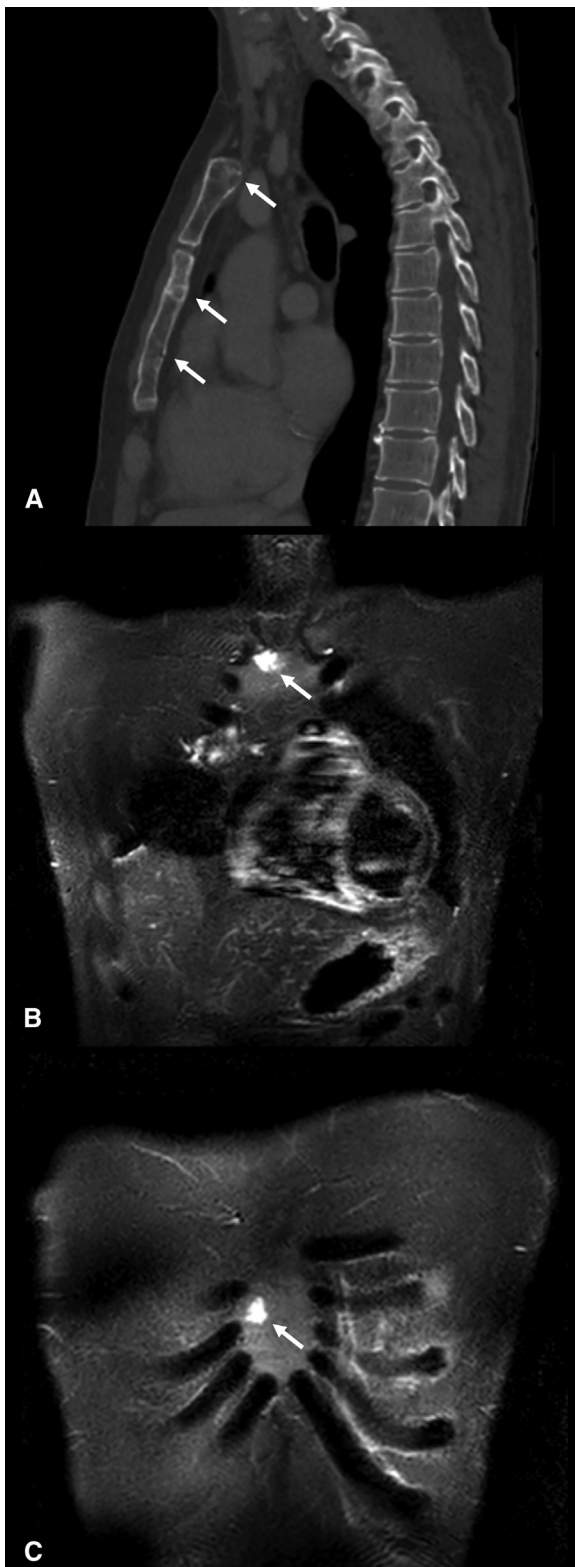
Maffucci syndrome is a very rare skeletal disorder characterized by multiple enchondromas (benign cartilaginous malformations) and soft-tissue angiomas.<sup>1,2</sup> Currently, there is no treatment available. Surgery is performed for complications such as deformities around the joints, scoliosis, and pathologic fractures. Patients with Maffucci syndrome have an increased risk of malignant transformation of an enchondroma to a low-grade chondrosarcoma, as well as a high risk of developing a fatal visceral malignancy.<sup>1</sup> Transformation of benign enchondromas to chondrosarcomas, their malignant counterparts,<sup>3</sup> has been reported, with rates varying between 52% and 57%.<sup>1</sup> Hence, when disease manifestations are present in the sternum, a median sternotomy is contraindicated for cardiac surgery, and alternative approaches should be considered. We present a case report with video on robotic mitral valve repair and resection of a pericardial cyst through a right minithoracotomy in a patient with Maffucci syndrome and sternal manifestations. The patient provided written informed consent for publication of all study data included in this case report with video.

## CASE REPORT

A 41-year-old male patient was referred to our clinic with dyspnea complaints in the presence of severe primary mitral

regurgitation. The patient had Maffucci syndrome with manifestations at multiple locations, including multiple chondroid lesions in the sternum (Figure 1). Throughout his life, he had undergone multiple reconstructive orthopedic procedures. On echocardiography, a bileaflet mitral valve prolapse with predominant prolapse of the P2 segment and excess tissue of both leaflets was observed, resulting from advanced Barlow disease. The effective regurgitant orifice area of the eccentric jet was 60 mm<sup>2</sup> and resulted in backflow in the pulmonary veins. The left ventricle was slightly dilated, with an end-diastolic diameter of 56 mm, but had a good function. Computed tomography revealed a right-sided paramediastinal mass of 3.9 by 5.7 cm with a calcified wall (Figure 2).

The presence of multiple enchondromas in the sternum was a contraindication for a conventional sternotomy due to potential of tumor spread in the mediastinum and the risk of malignant transformation. Hence, robot-assisted mitral valve repair (using the Da Vinci X, Intuitive Surgical



**FIGURE 1.** Preoperative computed tomography (A) and magnetic resonance imaging (B and C) demonstrating sternal manifestations of Maffucci syndrome.

robotic platform) was performed through a right minithoracotomy. First, the pericardial mass was resected en bloc, after which extracorporeal circulation with peripheral cannulation was installed and aortic crossclamping with a Chitwood clamp was performed. The mitral valve was exposed using a left atrial approach. Valve analysis demonstrated a Barlow-like valve with excessive leaflet tissue, a dilated annulus, and a prolapse of the P2 segment of the posterior leaflet with multiple ruptured primary chords. The valve was repaired by implanting 4 pairs of neochords and a ring annuloplasty (size 40, Memo-4D; LivaNova Group). The postoperative echocardiogram depicted no residual mitral regurgitation, sufficient leaflet coaptation, and an inflow gradient of 1.5 mm Hg. The patient was discharged on postoperative day 5 after an uneventful clinical course, although he experienced a transient ischemic attack at home, for which warfarin was continued after the initial first 3 months. Histologic examination revealed that the pericardial mass was a calcified cyst. Details on pre- and postoperative imaging, and the surgical procedure itself are summarized in [Video 1](#).

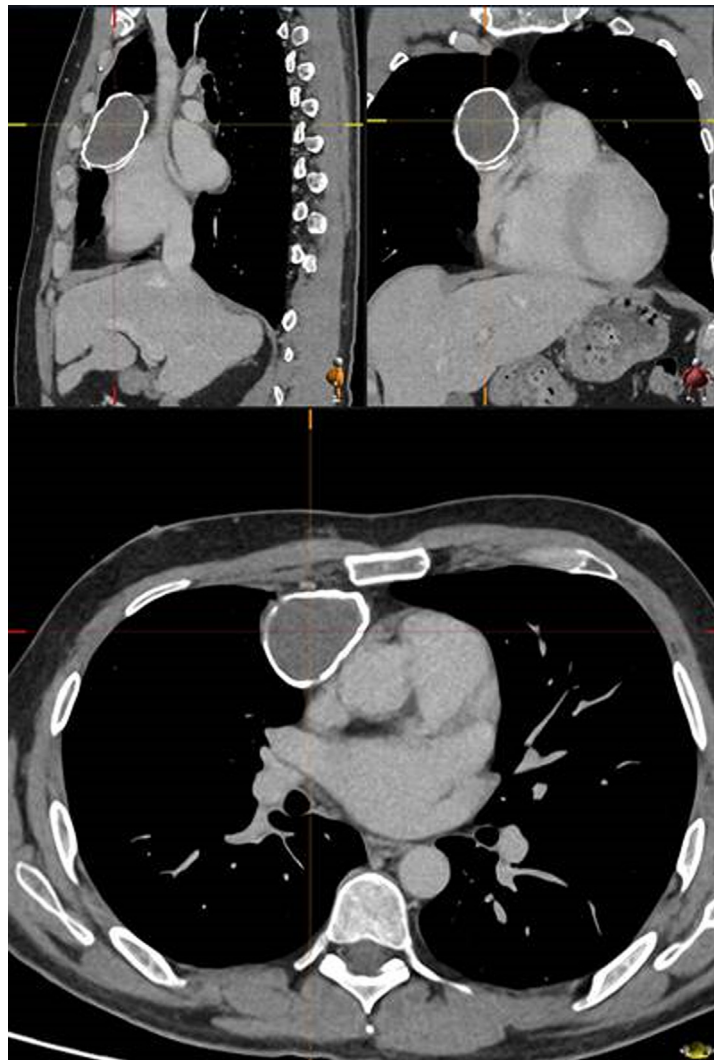
## DISCUSSION

The surgical approach to the heart should be carefully determined when dealing with patients with Maffucci syndrome and documented sternal manifestations. If enchondromas are present in the sternum that cannot be avoided when performing a sternotomy, then sternotomy is contraindicated because of the potential of dispersion of cells and the risk of malignant transformation. In this case, we chose a robot-assisted right minithoracotomy approach because it offered versatility of motion and 3-dimensional vision, which are beneficial to obtain good mitral valve repair and resection of the pericardial cyst<sup>4</sup> and because of our experience with this technique. However, alternative endoscopic approaches, like port-access mitral repair, could also be considered. Therefore, the choice between these 2 minimally invasive techniques should be based on the experience of the surgical team. To conclude, robotic mitral valve repair is a safe and attractive treatment option for patients with Maffucci syndrome with multiple sternal chondroid lesions.

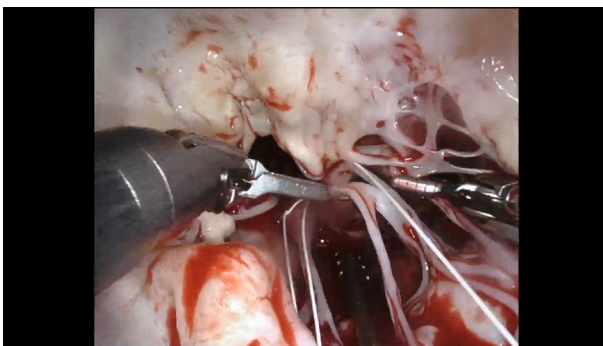
## Conflict of Interest Statement

B.J.J.V reported institutional research grant and speaker fees paid to his department by Medtronic. R.J.M.K. reported research support, consultation fees, and European Principal Investigator of the PERIGON Pivotal Trial for Medtronic. All other authors reported no conflicts of interest.

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**FIGURE 2.** Preoperative computed tomography demonstrating a large right-sided paramediastinal mass with a calcified wall.



**VIDEO 1.** Robotic mitral valve repair and resection of a pericardial cyst in Maffucci syndrome with sternal manifestations. The case is explained in a step-by-step approach. The video includes details on pre- and postoperative imaging, as well as the surgical procedure itself. Video available at: [https://www.jtcvs.org/article/S2666-2507\(24\)00045-2/fulltext](https://www.jtcvs.org/article/S2666-2507(24)00045-2/fulltext).

of interest. The editors and reviewers of this article have no conflicts of interest.

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