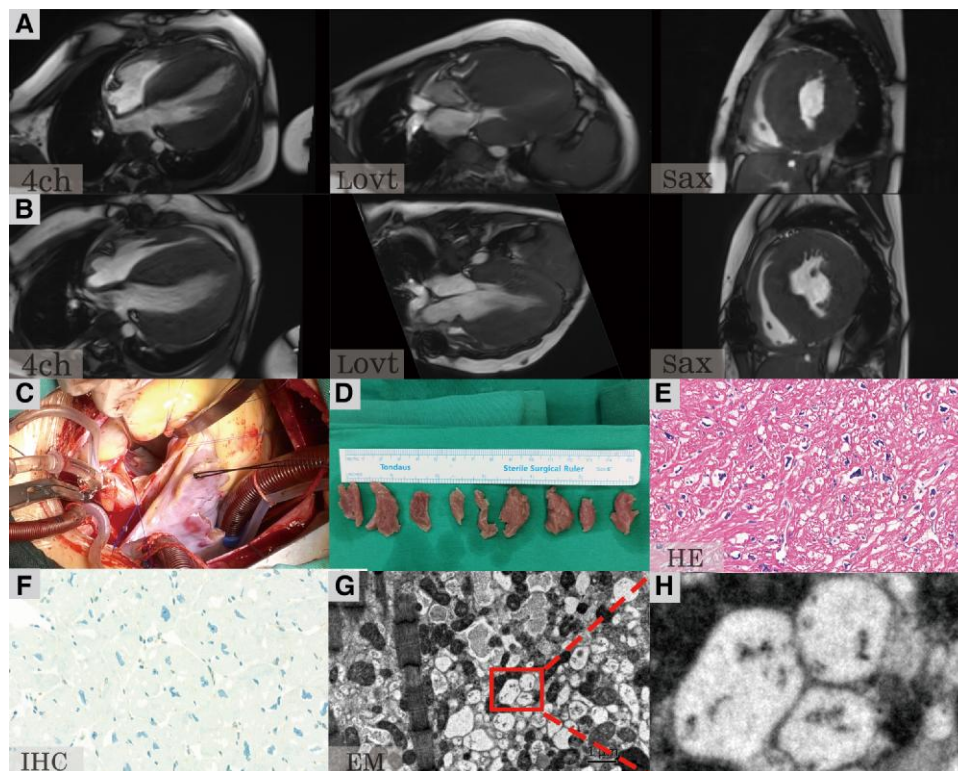


A modified extended Morrow procedure to relieve symptoms of the patient with Danon disease: a case report

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Received 8 June 2023; first decision 12 July 2023; accepted 21 July 2023; online publish-ahead-of-print 26 July 2023



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Handling Editor: Asad Shabbir

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A 12-year-old boy was diagnosed with hypertrophic cardiomyopathy (HCM) 3 years ago due to a preoperative examination of patent ductus arteriosus (PDA), and he didn't receive treatment. He was admitted to the hospital due to syncope and frequent convulsions. He also had decreased muscle strength, elevated serum creatine kinase levels, abnormal liver function (e.g. elevated transaminase activity), and intellectual decline. Echocardiography showed obstructive HCM and systolic anterior motion (SAM). Cardiac magnetic resonance (CMR) imaging demonstrated left ventricular (LV) hypertrophy (especially ventricular septum), biventricular outflow tract obstruction, and a SAM sign (*Panel A*; [Supplementary material online, Videos S1](#)). Genetic analysis identified a maternal hemizygous frameshift *LAMP2* variant (NM_002294: c.973dupC, p. L325fs*). A modified extended Morrow procedure was performed under cardiopulmonary bypass to relieve his symptoms (*Panels C and D*). Postoperative echocardiography showed LV outflow tract patency. CMR imaging demonstrated LV outflow tract obstruction was relieved without a SAM sign (*Panel B*, [Supplementary material online, Video S2](#)). Pathological examination revealed marked myocyte hypertrophy, disarray (*Panel E*), and *LAMP2* protein deletion (*Panel F*). Electron microscopy shows intracytoplasmic vacuoles containing autophagic material and glycogen (*Panels G and H*). The patient had an improvement in exercise capacity at follow-up 8 months post-discharge.

Danon disease (DD) is a rare, X-linked dominant, multisystem autophagic disease. Hemizygous male patients are typically affected earlier and more severely than women. Because of the pathophysiology of DD, the operation's objective is to improve the patient's quality of

life before his condition worsens to the point where heart transplantation is needed.

Supplementary material

[Supplementary material](#) is available at *European Heart Journal – Case Reports*.

Acknowledgements

I would like to express my gratitude to Mr. Zheng Wenhan for his contribution to the layout of the images.

Consent: The patient signed the informed consent form before surgery. There is no patient involvement.

Conflict of interest: All authors declare no conflict of interest for this contribution.

Funding: This study was supported by the Beijing Science and Technology Planning Project, China (Z191100006619003), the Capital Health Research and Development of Special Fund (2022-1-4032) and the Shenzhen Fundamental Research Program (JCYJ20220531091605012).

Data availability

The data underlying this article will be shared on reasonable request to the corresponding author.