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

Key words: Danon disease, Morrow procedure, hypertrophic cardiomyopathy, systolic anterior motion, cardiac magnetic resonance, case report.

Author information:
Dongdong Wu¹, Kai Yang², Hongyue Wang³, Jing Zhang¹, Shoujun Li¹.

1. Pediatric Cardiac Surgery Center, Fuwai Hospital, National Center for Cardiovascular Diseases, State Key Laboratory of Cardiovascular Disease, Chinese Academy of Medical Sciences, Peking Union Medical College, Beijing, 100037, China.

2. Department of Radiology, Fuwai Hospital, National Center for Cardiovascular Diseases, State Key Laboratory of Cardiovascular Disease, Chinese Academy of Medical Sciences, Peking Union Medical College, Beijing, 100037, China.

3. Department of Pathology, Fuwai Hospital, National Center for Cardiovascular Diseases, State Key Laboratory of Cardiovascular Disease, Chinese Academy of Medical Sciences, Peking Union Medical College, Beijing, 100037, China.

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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 hypertrophic cardiomyopathy 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 left ventricular outflow tract patency. CMR imaging demonstrated left ventricular 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 eight-month 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.
Patient consent statement

The patient signed the informed consent form before surgery.

Data availability statement

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

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There is not patient involvement.

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