Annals of Case Reports
Wang H, et al. Ann Case Rep: 8: 1313
www.doi.org/10.29011/2574-7754.101313
www.gavinpublishers.com

Case Report

A Rare 10-Year Outcome Inverted Replacement of the Mitral Valve with a Mechanical Aortic Valve Prosthesis in an Achondroplastic Dwarfism

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Citation: Wang H, Xue Q, Wu J, Zhou G, Bai Y, et al (2023) A Rare 10-Year Outcome Inverted Replacement of the Mitral Valve with a Mechanical Aortic Valve Prosthesis in an Achondroplastic Dwarfism. Ann Case Report. 8: 1313. DOI:10.29011/2574-7754.101313

Received: 14 May 2023, **Accepted:** 19 May 2023, **Published:** 22 May 2023

Abstract

Achondroplastic dwarfism is a rare hereditary metabolic disorder associated with a higher incidence of cardiovascular disease. There are few epidemiological data and surgical experience of rheumatic valvular disease in this population. This report is the only one to date on mitral replacement in an achondroplastic dwarfism. In addition, our approach involves placing the mechanical aortic valve prosthesis upside down in the mitral position. The 10-year results show benefits, but intraoperative management, valve type, and anticoagulation regimen are real challenges.

Keywords: Achondroplastic Dwarfism; Valvular Disease; Valve Replacement Surgery

Introduction

Achondroplasia is a group of diseases in which abnormal bone and cartilage growth results in poor bone development. It is a systemic metabolic disease, with short limbs and a higher risk of heart disease. However, both adult and pediatric heart surgery centres lack experience in treating such patients with heart surgery. In this report, we summarize the experience of a patient with achondroplastic dwarfism undergoing valve replacement surgery, which is slightly different from that of other patients.

Presentation of Case

A 51-year-old woman came to our hospital for treatment of worsening chest tightness. The patient has been stunted since childhood, with a height of 94cm and a weight of 25Kg. She has normal intelligence, short limbs but a long torso, a flat face with compact features, consistent with achondroplastic dwarfism (Figure 1A). Chest radiographs revealed changes consistent with rheumatic heart disease, thickening of the metaphyseal, flattening of the bilateral humeral heads and multiple vertebral bodies (Figure 1B). Echocardiography demonstrated rheumatic valvular disease, including severe mitral stenosis (orifice area 0.5-0.6 cm2) and moderate tricuspid insufficiency (regurgitation of 6ml). The atrioventricular volume was generally small, with left atrium (LA): 42ml, left ventricle (LV): 21ml, right atrium (RA): 39ml, right ventricle (RV): 27ml. Systolic function was normal (left ventricular ejection fraction [LVEF]: 61%). Pulmonary function tests indicated severe impairment of ventilation and premature closure of small airways. The patient underwent mitral replacement and tricuspid valvuloplasty through the median sternum. The anaesthesiologist selected endotracheal intubation with an inner diameter of 5.5mm and a depth of 17cm. Cardiopulmonary bypass (CPB) was performed with 16F aortic intubation and 20F superior and inferior vena cava intubation. We found that the mitral orifice was changed like a fish's mouth, and the area was obviously small (Figure 2A). There was obvious fibrosis and thickening of the lobe,

and the chordae tendinae was shortened. The anterior lobe regions A1 and A2 were adhered to the posterior lobe regions P1 and P2 respectively. We resected the diseased valve and its subvalvular tissue, retained part of the posterior valve and folded it. A 21mm Carbomedics two-leaf mechanical aortic valve was used, and the prosthesis was reversely fixed and set with 12 stitches of 2-0 interstitial mattress suture ring with gasket. As for tricuspid, we used Devega method to complete valvuloplasty. No obvious reflux was found after intraoperative injection. After discharge, the patient had been given regular oral warfarin anticoagulation therapy. The patient's performance of activity tolerance has been good, with no apparent discomfort. Unfortunately, a cerebral infarction occurred in the sixth postoperative year, and now has left hemiplegia with stuttering ambiguity. Reexamination of echocardiography 10 years after surgery showed that the valve prosthesis function was still normal, orifice area was 1.93cm², the maximum cross-valve pressure difference was 12mmHg, and the average was 4mmHg (Figure 2B).

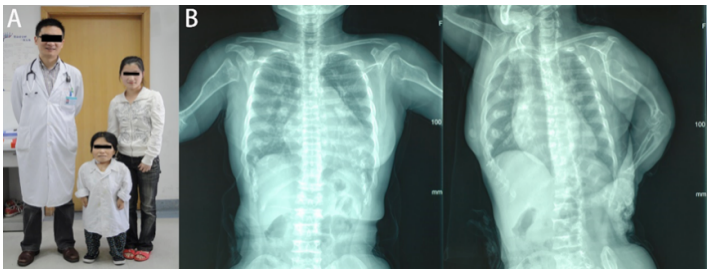

Figure 1: (A) From left, doctor, patient and her daughter. (B) Preoperative chest X‐ray.

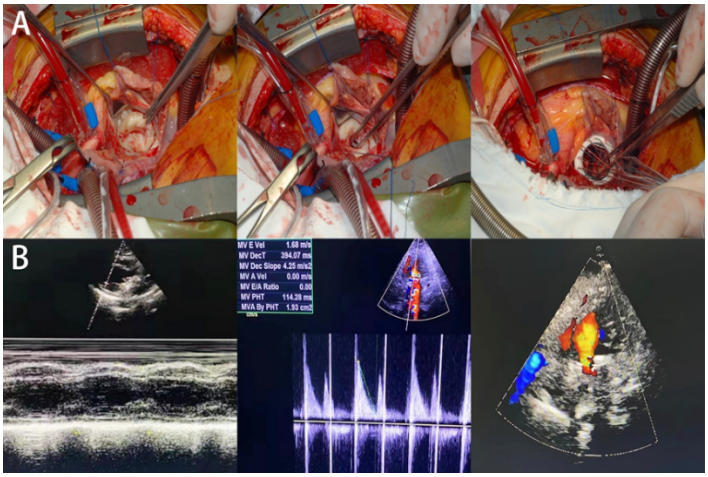

Figure 2: (A) Mitral morphology and mechanical valve prosthesis installation during operation. (B) Echocardiograms reviewed 10 years after surgery (2022). Left ventricular M-mode ultrasound and mitral valve function are shown from left to right. There was no significant regurgitation of the other valves, and LA: 30ml, LV: 38ml, RA: 21ml, RV: 14ml, EF: 54%.

Discussion

People with achondroplastic dwarfism have a higher incidence of cardiovascular disease than the general population [1]. Although achondroplastic dwarfism is not contraindicated in cardiac surgery, it has only been reported in aortic valve replacement and dissection [2,3]. This is the first report to date of mitral replacement in achondroplastic dwarfism with a rare 10-year follow-up [4]. Both anaesthesia and CPB in patients with achondroplastic dwarfism are a challenge, especially for physicians in centres that do not perform pediatric heart surgery. Age-related calculations are not appropriate for achondroplastic dwarfism, but endotracheal intubation with an internal diameter of about 5.5mm is recommended, which is consistent with the characteristics of children around 3 years old. Achondroplastic dwarfism patients can also prepare the size required for CPB intubation in advance with reference to body weight, usually 12-20F for 15-25Kg patients. Patients with small body weight are prone to high pump pressure, drainage and under perfusion even with thin tube wall intubation, so the position of the tube and negative pressure should be checked frequently during the operation. In addition, we also completed the inversion of the mechanical aortic valve prosthesis in the mitral valve position. Aortic valve prosthesis is similar in design to mitral, except that it needs to be installed upside down to match the direction of blood flow. Judging from the results, the practice is safe and reliable, so adult heart centres can perform similar procedures without obvious surgical confines and ethical issues. Another point of concern in this case report is that the patient had a stroke. In the general population, we usually keep the INR index around 2.0 after mechanical valve replacement surgery. The fact that we inverted the valve itself may have facilitated the application of higher anticoagulant targets. In addition, we also speculate that this may be related to the specificity of anticoagulation in patients with achondroplasia. Because achondroplasia is a metabolic disorder and vascular malformations develop. However, there are no relevant studies on anticoagulation in patients with achondroplasia, and only higher blood clotting has been found in animals with chondrodysplasia [5]. More research is needed to determine whether conventional anticoagulant targets are unsuitable for achondroplastic dwarfism, or whether they are simply the unfortunate individuals who have had a stroke.

Conclusion

Cardiac surgery for achondroplastic dwarfism should focus on intraoperative pipe management, valve type, and anticoagulation characteristics. Inverted aortic valve mechanical prosthesis is also safe and effective in the absence of suitable mitral models for achondroplastic dwarfism.

Acknowledgement: This study was supported by the Teaching Achievement Cultivation Program of the First Affiliated Hospital of Naval Medical University and the Deep Blue 123 Military Special Project (No. CHPY2021B23 and 2019SLZ006 to Guanxin Zhang). All authors participated in the development of this article and no conflicts of interest to report.

Ethical Guidelines: The Institutional Review Board (IRB) or equivalent ethics committee of the Changhai Hospital did not approve this study [There is no content in the article that obviously exposes patient information, and this study is a non-clinical human trial]. The subject provided informed written consent for the publication of the study data.

References

1. Sukhavasi A, O'Malley TJ, Maynes EJ, Choi JH, Gordon JS, Phan K, et al (2020) Cardiac interventions in patients with achondroplasia: a systematic review. Journal of thoracic disease. 12: 998-1006.
2. Al-Jughiman M, Yanagawa B, Rondi K, Dalamagas C, Peterson MD, et al (2014) Acute Type A Dissection Repair in an Achondroplastic Dwarf: Anesthetic, Perfusion, and Surgical Concerns. Aorta (Stamford, Conn). 2: 143-146.
3. Mikami T, Kainuma S, Toda K, Sawa Y (2022) Rare case of severe non-calcific aortic stenosis in an achondroplastic dwarf: surgical consideration. Interactive cardiovascular and thoracic surgery. 34: 495-497.
4. Khairallah S, Rahouma M, Gaudino M, Mick SL (2022) Mitral and tricuspid repair in an adult achondroplastic patient. J Card Surg. 37: 1425-1427.
5. Butty EM, McKinney CA, Prisk AJ (2021) Treatment of a flunixin meglumine overdose with intravenous administration of lipid emulsion and therapeutic plasma exchange in a Nigerian dwarf buck kid (Capra aegagrus hircus). Journal of veterinary internal medicine. 35: 1626-1630.