Perceval S aortic valve implantation in an achondroplastic Dwarf

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ABSTRACT

Despite cardiovascular disease in patients with dwarfism is not rare; there is a lack of reports referring to cardiac interventions in such patients. Dwarfism may be due to achondroplasia or hormonal growth disorders. We present a 58-year-old woman with episodes of dyspnea for several months. She underwent an transthoracic echocardiography, and she diagnosed with severe aortic valve stenosis. She referred to our department for surgical treatment of this finding. In accordance of her anthropometric characteristics and her very small aortic annulus, we had the dilemma of prosthesis selection. We decided to implant a stentless valve to optimize her effective orifice area. Our aim is to present the successful Perceval S valve implantation and the descriptions of the problems coming across in operating on these special patients. To our knowledge, this is the first case patient in which a Perceval S valve is implanted according to the international bibliography.

Key words: Achondroplasia; Aortic valve stenosis; Aortic valve surgery; Cardiac surgery in dwarf; Dwarfism; Perceval S valve

INTRODUCTION

Dwarfism is a term referring to people of short stature, and can be due to several musculoskeletal and hormonal growth anomalies.[1] Achondroplasia (ACH) is the most common dwarfing condition.[1,2] Hypopituitarism is the second cause of dwarfism and requires special management during heart surgery.[3,4]

In this paper, we would like to present a case of aortic valve replacement with the Perceval S (Sorin Group, Italy) valve implantation [Figure 1a], performed on an achondroplastic dwarf with severe calcified aortic stenosis. There are no data about the incidence of valve diseases in achondroplastic patients. To our knowledge, this is the first Perceval S valve is implanted to a dwarf. The aim of this report is to show that ACH and dwarfism itself is not a contraindication to aortic valve replacement or coronary artery bypass grafting.[3,5]

CASE REPORT

We present a 58-year-old achondroplastic woman, admitted to our department with a diagnosis of severe and symptomatic aortic stenosis. Neither her parents nor her brother was achondroplastic. Her habitus bore characteristic achondroplastic features: Although the truncal length was proportioned, her limbs were very short, and she had the typical facies of ACH (a large head, small face, and saddle nose). She weighed 28 kg and her height was 105 cm [Figure 1b]. Chest radiography revealed great thoracic skeleton anomalies with pectus carinatum [Figure 2a and b]. The echocardiographic examination showed a hypertrophic...
interventricular septum (13 mm) but well-functioning left ventricle (ejection fraction: 0.55) and severe aortic stenosis, with a maximum trans-valvular gradient of 85 mmHg, mean pressure gradient 45 mmHg and an aortic valve area of 0.5 cm². The pulmonary artery pressure was 45 mmHg. She was in sinus rhythm.

Cardiac catheterization confirmed the severe and calcific aortic stenosis, without coronary artery disease. During the preoperative preparation, we performed a respiratory test that proved a chronic obstructive pulmonary disease (COPD). Due to anomalies of the chest and its dimensions, it has been suggested the necessity of a specific pediatric tube for the intubation. The unusual skeletal structure of such patients makes percutaneous arterial and venous access difficult. Catheterization of jugular vein was also impossible due to the very short neck. For these reasons, we prepared surgically the femoral artery and vein; in the open surgical way for arterial and central venous access.

Because of her anthropometric numbers and her very small aortic annulus we had in our mind to implant the Perceval S aortic valve as the best solution for the patient.

Intra-operatively we found a tricuspid, fibrous, and heavily calcified aortic valve in a very small aortic root. After calcified valve removal, the aortic annulus was very small, about 11 mm.

We decided the Perceval valve implantation. This valve is available as small, medium, large, and extra-large. With the help of the sizer, a small valve is decided to be implanted. Aortic annulus dilatation (not enlargement), was necessary to implant this valve into a very small annulus.

Due to very small aorta, we were obliged to put the aortic cannula very distally in the aortic arch. We had to use pediatric cannula both for the aorta (20F) and for vena cava (28F). Anesthetic management was carried out with no complication: Both hemodynamic and hematologic values were normal. The operative procedure required 83 min of cardiopulmonary bypass (CPB) and 44 min of ascending aortic cross-clamp time under normothermia. For her chest closure, we initially used two wires and then some ethibond sutures due to very fragile and osteoporotic sternum.

The patient was in the intensive care unit for 36 h. The early postoperative period of the patient was completely uneventful. Postoperative chest-ray was normal without effusion [Figure 2c and d]. The patient was discharged on the 6th postoperative day without any special medication.

**DISCUSSION**

Peculiar difficulties arise in the anesthesiologic management of patients with ACH. First, the unusual skeletal structure of such patients makes percutaneous arterial and venous access difficult.[1,3,5] Tracheal intubation can be complicated by the abnormal anatomy of the upper respiratory tract and the skeletal abnormalities. The main problem in this patient with aortic stenosis was the very small size of the ascending aorta and aortic root. It has been described the necessity of the aortic root enlargement to implant the valve.[5] We implanted the Perceval S valve; a stentless and self-expanding valve to provide her a satisfied effective orifice area. It is a bioprosthesis loaded on a device-stent for its self-anchoring in the aortic root. This valve is especially indicated in patients with small aortic root and in patients with comorbidities. Is a rapid intervention because this valve is a sutureless and self-expanding. Using this valve, it is not necessary any aortic root enlargement or any other manipulation in the aortic annulus. We believe that this valve, the Perceval S, is the choice for patient with small aortic root. It provides them the biggest possible effective orifice area and simultaneously, is especially indicated in patients with comorbidities (COPD) which require a rapid operation without long cross clamp time and CPB time. In conclusion, aortic valve replacement

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**Figure 1:** The Perceval S aortic valve. This valve (Perceval S) has not got any ring. It is a bioprosthetic valve sutured on a special device for its anchoring in the aortic root (a). Our patient (105 cm) with her brother (174 cm) (b)
with Perceval S valve implantation is safely indicated in patients with dwarfism.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

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