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Mitral Valve Replacement and Hunter Syndrome: Case Report

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ABSTRACT

We report a rare case of mitral valve stenosis secondary to Hunter syndrome, mucopolysaccharoidosis (MPS) type II in a 33-year-old man. Anatomical abnormalities in patients with MPS present anesthetic and surgical challenges during cardiac surgery. Management of this particular patient was complicated by excessive oral secretions and atrial fibrillation. With a detailed preoperative assessment and planning for airway management, this patient successfully underwent mitral valve replacement and had an uncomplicated hospital course. After 6 months of follow-up, the patient was still in stable condition.

INTRODUCTION

Hunter syndrome is an X-linked recessive mucopolysaccharoidosis (MPS) [Hunter 1917] resulting in lysosomal storage disease due to deficiency of the enzyme iduronate-2-sulfatase. This deficiency results in an inability to metabolize glycosaminoglycans, which in turn leads to accumulation of connective tissue in the extracellular matrix of various organs, causing nonreversible dysfunction of the whole or part of the target end-organ. Valvular heart disease (most commonly mitral, but aortic disease also occurs) [Zimmermann1988], obstructive pulmonary disease, sleep apnea, respiratory tract infections [Semenza 1988], liver and spleen enlargement, decreased cervical spine range of motion, and central nervous system involvement are manifestations of the syndrome [Schwartz 2007].

Hunter syndrome type II (MPS II) is a rare condition affecting 1 in 100,000 live births. Males are most often manifest the disease, whereas females are usually carriers. Hurler (MPS type I H), Scheie (MPS type I S), Sanfilippo (MPS type III A, B, C, D), Morquio (MPS type IV A, B), Maroteaux-Lamy group (MPS VI), and Sly (MPS VII) are different types of the same disease origin. These different types are related to abnormalities in different enzymes involved in mucopolysaccharide synthesis [Walker 1994].

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Characteristic physical features of MPS patients include short stature, a large scaphoid skull caused by premature closure of the metopic and sagittal sutures, a depressed nasal bridge with broad nasal tips, and enlarged lips. The neck is short and vertebral subluxation with cord compression can occur. Joint stiffness is a common feature of all MPS and probably results from a combination of metaphyseal deformities secondary to glucosaminoglycan deposition and fibrosis.

There are 2 recognized entities of MPS II. The early-onset type (severe form) develops shortly after age 2 years and presents with a large skull, coarse facial features, profound mental retardation, spasticity, aggressive behavior, joint stiffness, and death before age 20. The late-onset type (mild form) presents later in life, with less severe symptoms.

We performed a literature review that yielded reports of 28 patients with different types of MPS who required surgical treatment for various problems. They are summarized as follows: 40% ear, nose, throat, and airway procedures; 19% neurological investigation; 8% neurosurgery; 11% orthopedic surgery; 7% hernia repair; 4% ophthalmologic procedure; and 1% open-heart surgery [Moores 1996].

CASE REPORT

A 33-year old man was admitted to our hospital with diagnosed mitral stenosis secondary to Hunter syndrome. The patient had late-onset type MPS (mild MPS II), with an enlarged head, broad nose, thickened lips, short extremities, limited joint mobility with flexion deformity in both elbows, and a short neck. Both liver and spleen were palpable below the costal margin. The patient's IQ was normal. His height was 140 cm and his weight was 40 kg.

The diagnosis of the syndrome occurred when the patient was 5 years old and urine screening tests were positive for MPS type II. The diagnosis of mitral valve stenosis was made 3 years prior to admission and was confirmed by echocardiography. The patient had been in atrial fibrillation in the last 12 months. His medications consisted of warfarin, digoxin, and furosemide. On admission the patient's condition was American Society of Anesthesiologists class IV and New York Heart Association class III.

The echocardiographic examination revealed thickened leaflets, a mitral valve area of 0.75cm^2, a transvalvular pressure gradient of 10 mmHg, a left atrium of 69 mm, and satisfactory function of the left and right ventricles.

Pulmonary function test results were as follows: forced vital capacity (FVC) 0.73 (2.87) 25%, forced expiratory volume (FEV_1) 0.52 (2.57) 20%, and FEV_1/FVC 86%, consistent with restrictive lung disease.

During the preanesthetic examination, the patient's airway was grade IV according to the Mallampati classification. We decided not to premedicate the patient, after a long and detailed discussion with him during which we were assured of his complete understanding of the potential danger from the compromised airway in the setting of mitral stenosis and pulmonary hypertension.

For induction of anesthesia, propofol 100 mg and midazolam 2 mg were used while spontaneous ventilation was maintained. To avoid pulmonary hypertension episodes during intubation, we gave diltiazem in incremental doses (total dose of 15 mg). The patient was intubated, on the third attempt, with the "stylet in tube" method (Figure 1).

We elected not to use nasopharygeal intubation, because of the risk of hemorrhage. The patient had excessive oral secretions, which contributed to difficulty in managing the airway. Because he was in atrial fibrillation with a relatively fast ventricular response, we decided to not use atropine.

The patient was intubated with a 6.5 cuffed endotracheal tube, and anesthesia was supplemented with another 2 mg of midazolam and 200 μg fentanyl. Mechanical ventilation was adjusted to maintain arterial carbon dioxide ($PaCO_2$) at 32-38 mmHg and *cis*-atracurium 2 μg/kg per min was used for maintenance of muscle relaxation. Sevoflurane 2%-3% and fentanyl were titrated to provide analgesia throughout surgery (total dose 500 μg). Tranexamic acid 1.5 g was also administered.

Intaoperative transesophageal echocardiography confirmed the preoperative findings. Echocardiographic monitoring throughout the operation allowed for continuous monitoring of the right and left ventricular function and for indirect measurement of the systolic pulmonary artery pressure, which did not increase.

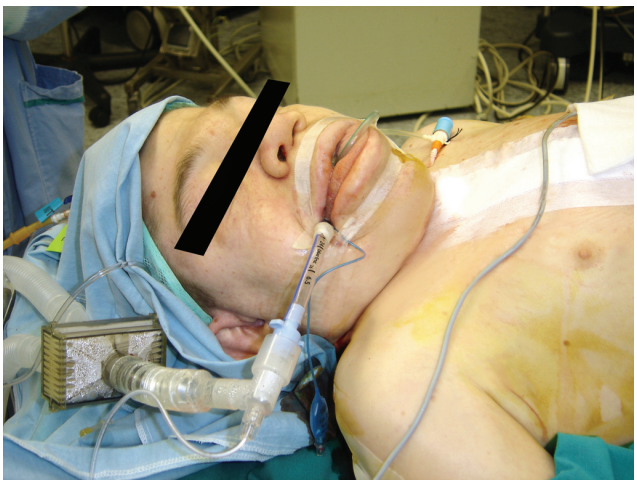

Figure 1. The patient in the operating room after application of airway instrumentation. An enlarged head with broad nose and short neck can be noted.

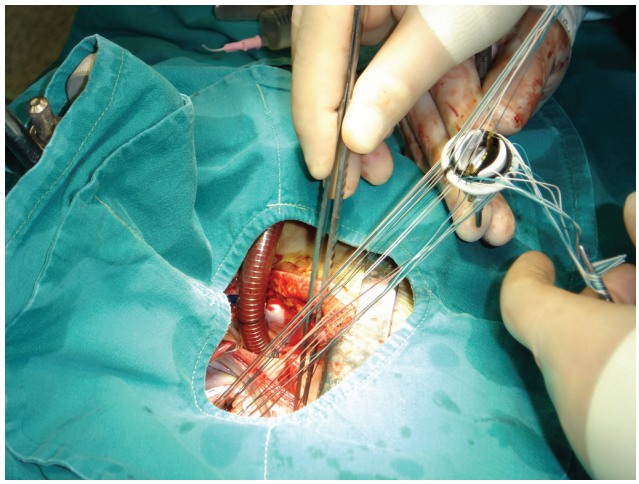

Figure 2. The aortic prosthesis placed reversed in the mitral position.

The patient was placed on aortobicaval cardiopulmonary bypass through a median sternotomy, and the mitral valve was replaced with a mechanical aortic valve 21S, placed reversed, in the mitral position (Figure 2). The mitral valve specimen had manifestations of MPS disease, including marked thickening of the mitral leaflets and chordae tendinae (Figure 3).

The patient remained hemodynamically stable throughout the operation, with a central venous pressure of 18 mmHg. The cardiac index was 3.6 L/min per m^2 at the prebypass period and 5.9 L/min per m^2 at the postbypass period, as measured by a new semiinvasive device, the FlO Track/Vigileo™ (Edwards Lifesciences, Irvine, CA, USA), using arterial pressure waveform analysis for cardiac output measurement [Mayer 2007]. The patient was weaned from bypass on moderate inotropic support with dobutamine 7 μg/kg per min.

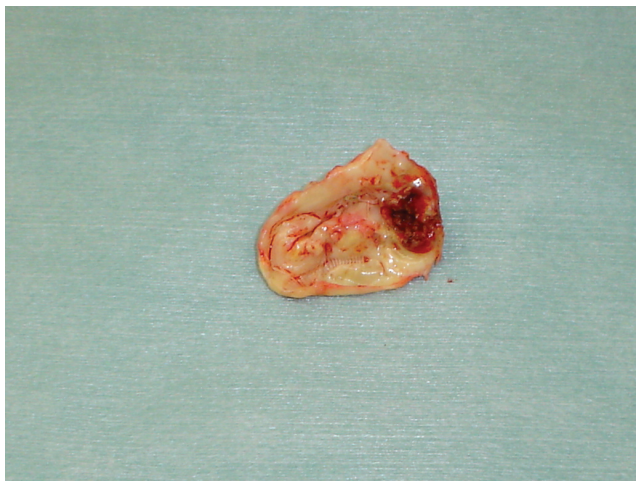

Figure 3. The mitral valve specimen, manifesting gross thickening of the mitral leaflets and the chordae tendinae.

Postbypass transesophageal echocardiography demonstrated a well-functioning prosthesis. The patient was extubated on the second postoperative day in the intensive care unit and was weaned off the inotropic support on the same day. He was discharged from the hospital on postoperative day 8, after an uneventful recovery.

DISCUSSION

Cardiac disease is common in Hunter syndrome, presenting most frequently with valvular involvement, coronary artery narrowing, and endocardial fibroelastosis. The intracellular accumulation of glycosaminoglycans causes cellular enlargement with resulting disruption of the structure and function of the tissue involved.

Echocardiographic examination reveals striking changes in the mitral valve, which is dense and multilayered with a decreased diastolic-descent rate on M-mode, and thick leaflets with diminished opening visible on 2-dimensional echocardiogram [Johnson 1981; Gross 1988]. Also, the aortic valve shows thickening along with diffuse hypertrophy of the intraventricular septum and of the posterior wall of the left ventricle. The right-sided valves (tricuspid and pulmonary) are usually unaffected.

Upper and lower respiratory tract infections are common. Multiple factors are present in MPS patients and make airway management and tracheal intubation potentially hazardous. The glucosaminoglycan deposition in the connective tissues is accompanied by an enlarged and thickened epiglottis, tonsillar hypertrophy, tongue enlargement, supraglottic narrowing, and a narrow and flattened trachea.

There have been reports of combined aortic and mitral valve replacement in a patient with Shei syndrome [Fischer 1999], but in our literature search we could find only 1 report of mitral valve replacement in a patient with Hunter syndrome [Bhattacharya 2005].

According to several published reports, anesthetic problems in this set of patients are mostly related to airway management difficulties attributable to anatomical abnormalities [Kempthorne 1983; King 1984]. The usually existing short neck in these particular patients may make tracheostomy difficult [Baines 1983].

According to a report by Brown and colleagues [1984], the use of a nasopharygeal tube improved the airway in 6 of 10 patients; however, its insertion may be difficult because of the mucopolysaccharide deposits. A laryngeal airway mask was used in only 2 patients, and its usefulness in this setting requires further investigation.

Bhattacharya et al [2005] report a patient with Hunter syndrome scheduled for mitral valve replacement who was intubated with a laryngeal-mask airway and an endotracheal tube inserted with the aid of a rigid bronchoscope. This particular patient required major inotropic support and intraortic balloon pump due to myocardial stunning.

The increased mortality of patients with MPS is often due to a combination of restrictive lung disease, upper and lower respiratory infections, and valvular heart disease.

Our patient did not present any airway complications that could have had deleterious effects on the already elevated pulmonary vascular resistance on induction, during intubation, and later, after extubation, in the intensive care unit. We attribute this success to a detailed preoperative assessment and a well-devised plan for airway management that avoids compromising of the pulmonary circulation and the right heart, which are cornerstones of care in patients with Hunter syndrome and a stenotic mitral valve.

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