Case Report

Anaesthetic Management of a Patient with Down Syndrome VSD, ASD & PDA in Pediatric Cardiac Surgery Department, Dhaka Shishu Hospital

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Abstract.

Down syndrome or trisomy 21 is a condition where extra genetic material causes mental and physical delays and deficits. It affects 1 in every 650 babies. Abnormalities of the cardiovascular system are common in down syndrome. Approximately half of all infants born with down syndrome have a heart defect. The most common heart defects in down syndrome are the following: atrioventricular septal defect (45%), ventricular septal defect (35%), atrial septal defects (8%) and patent ductus arteriosus (7%), tetralogy of Fallots (4%).

A Two years one-month-old baby was admitted in cardiac surgery department of Dhaka Shishu Hospital, with the diagnosis of Down syndrome with VSD,ASD& PDA with moderate pulmonary arterial hypertension. Clinical examination revealed diastolic murmur over mitral area. The child was treated with face musk oxygen, diuretics and digoxin and was stabilized medically and then was selected for surgery. We used balanced anesthetic technique using oxygen, air, fentanyl, midazolam and vecuronium. Patient was operated under cardiopulmonary bypass (CPB) with moderate hypothermia. Patient tolerated the whole procedure well and was ventilated electively for 4hrs in the intensive care unit. He was discharged on the 8thpostoperative day.

Keywords: *Down syndrome, Ventricular septal defect (VSD), Atrial septal defect (ASD), Patent ductus arteriosus (PDA). Cardiopulmonary bypass, Congenital heart disease, Pulmonary arterial hypertension.*

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Introduction

Down syndrome is a condition characterized by trisomy of chromosome 21.[1,2] Among all cases, 95% are primary trisomy and 5% are translocation and mosaic forms (3% and 2%, respectively).[3] The overall incidence of Down syndrome is one case in every 650 live births, although this rate varies according to the mother´s age. In mothers 45 years of age or older, the incidence reaches one in every 30 live births.[2,3] The risk of recurrence is 1% in the general population.[4]

The clinical manifestations of Down syndrome are numerous and can present in any body system. The most significant include intellectual impairment, short stature, heart disease, digestive disorders and orthopedic abnormalities. Heart disease is, without a doubt, the main factor contributing to a favorable or unfavorable course in these patients. Among all cases of congenital heart disease, 4%-10% are associated with Down syndrome, and 40%-60% of Down syndrome patients present congenital heart disease. Cardiac malformation is the principal cause of mortality in the first two years of life. The most frequent cardiac abnormalities in Down syndrome patients are patent ductus arteriosus (PDA), ventricular septal defect (VSD), and atrial septal defect (ASD).

Case Report

A two years one -month-old male child weighing 6.3 kg baby was admitted in Dhaka Shishu Hospital,

Cardiac Surgery Department with the diagnosis of Down syndrome with VSD,ASD &PDA. On clinical examination, pulse was 120/min and regular, respiratory rate 30/min and blood pressure 90/60 mm Hg. On auscultation a diastolic murmur was heard at the mitral area. The chest X-ray showed cardiomegaly with increased bronchovascular markings in both the lung fields. The electrocardiogram (ECG) showed both atrial and ventricular hypertrophy. Transthoracic 2D echo demonstrated moderate VSD with ASD with PDA. The child was treated with oxygen, face mask diuretics digoxin and was stabilized medically. Cardiac catheterization was performed to know the size and site of VSD, ASD & PDA, pulmonary artery pressure and to rule out any other associated lesions. It revealed VSD (1.5cm x 1.3cm), moderate PDA, pulmonary arterial hypertension (47/9 mmHg with a mean of 25 mmHg). Child was scheduled for surgical correction of VSD and under general anesthesia.

On the day of surgery, the child received intravascular ketamine (15 mg) & shift to operation theatre. Then have been given midazolam (0.5 mg) and glycopyrrolate (30 mcg) as premedication and oxygen 5 L/min by face mask. Peripheral venous access was obtained using 22-gauge cannula. Induction of anaesthesia was achieved with fentanyl (20 mcg) and additional titrated doses of midazolam. Tracheal intubation was facilitated by vecuronium (1 mg). Anaesthesia was maintained with oxygen, air, isoflurane, fentanyl, midazolam and vecuronium. A 20 gauge arterial catheter was inserted in the Rt femoral artery for continuous monitoring of blood pressure, blood gas and serum electrolytes analysis. A 4.5-F triple-lumen catheter was advanced into the right internal jugular vein for monitoring central venous pressure, infusions of inotropes and vasodilators. Pulmonary artery (PA) pressures were recorded before going on cardiopulmonary bypass (CPB) and were 50% of systemic pressure i.e. 38/24 (mean 28) mmHg.

After median sternotomy and thymus dissected out then 25mg of heparin was administered for anticoagulation. Ascending aorta and bi-caval cannulation was done. CPB was instituted with a maximum flow of 1.56 L/min. A moderate size PDA was seen .Multiple ligation of PDA was. Patient was cooled to 32°C (nasopharyngeal temperature). Blood cardioplegia was used Two times. The total CPB time was 94 min and aortic cross-clamp time was 60 mins. Right atriotomy revealed a moderate-sized VSD. PTFE patch closure was done by 5 –O pledgetted stitch. ASD was notfound. Hemofiltration was carried out during CPB to remove excessive

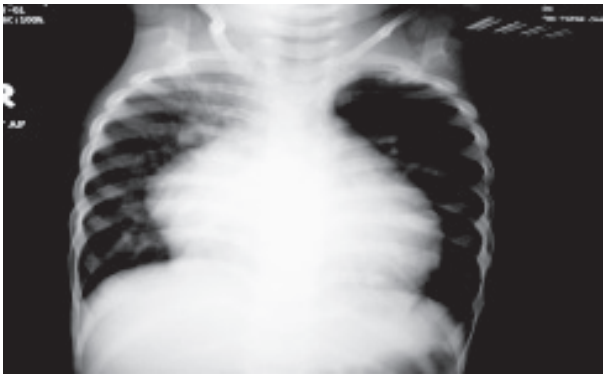

Fig 1 *Preoperative X -Ray*

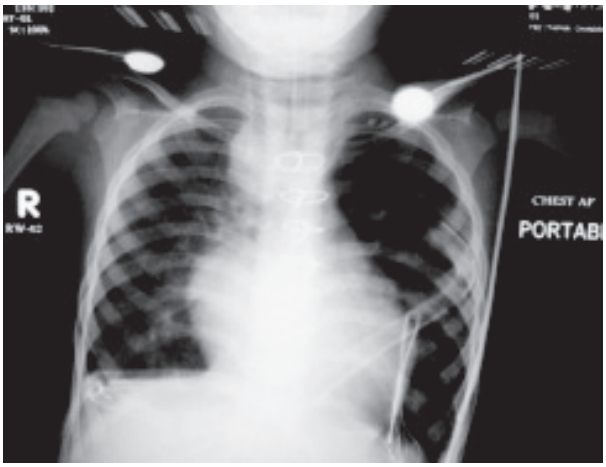

Fig 2 *Post operative X -ray*

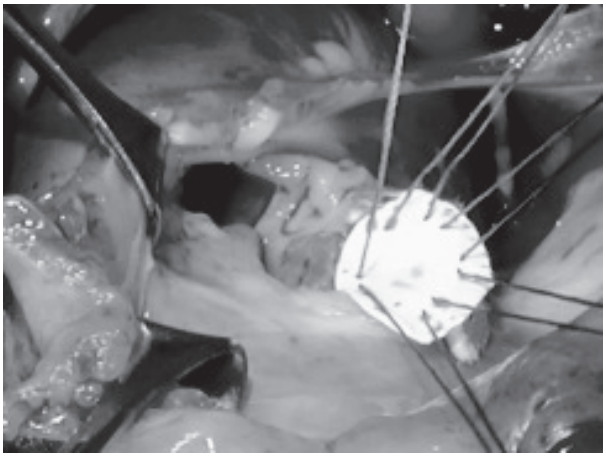

Fig 3 *PTFE patch closure of VSD*

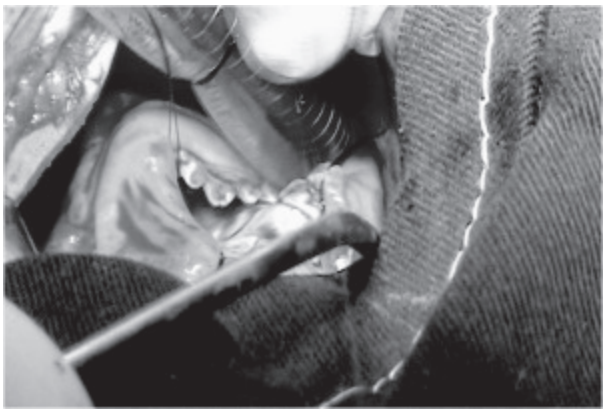

Fig 4 *Atrium closing*

fluid and to raise the hematocrit. Leukocyte depleter (PAL) was made use of during blood transfusion intraoperatively.

Adequacy of perfusion during CPB was assessed by arterial pressure, urine output, arterial blood gases and mixed venous oxygen tension. A 20-G single-lumen 10-cm catheter was inserted into the PA through the right ventricle to monitor PA pressure throughout the intra-operative period. After the completion of repair the patient was rewarmed to 37°C. He was weaned from CPB. Dobutamine, Adrenalin, and nitroglycerin were used during the weaning process. Hyperkalaemia was corrected. Blood glucose was monitored every hour. After weaning from CPB residual heparin was neutralized with 25 mg of protamine sulfate. The patient was electively ventilated for 4 hrs using pressure-regulated volume control mode of ventilation in the intensive therapy unit. Then the patient was extubated uneventfully. He was discharged from the hospital on the 8th postoperative day.

Discussion

Down syndrome is a condition characterized by trisomy of chromosome 21.[1,2] Among all cases, 95% are primary trisomy and 5% are translocation and mosaic forms (3% and 2%, respectively).[3] The overall incidence of Down syndrome is 1 case in every 650 live births, although this rate varies according to the mother´s age. In mothers 45 years of age or older, the incidence reaches one in every 30 live births.[2,3] The risk of recurrence is 1% in the general population.[4]

The clinical manifestations of Down syndrome are numerous and can present in any system of the body. The most significant manifestation are intellectual impairment, short stature, heart disease, digestive disorders and orthopedic abnormalities. Heart disease is, without a doubt, the main factor contributing to a favorable or unfavorable course in these patients. Among all cases of congenital heart disease, 4%-10% are associated with Down syndrome, and 40%-60% of Down syndrome patients present congenital heart disease. Cardiac malformation is the principal cause of mortality in the first two years of life. The most frequent cardiac abnormalities in Down syndrome patients are patent ductus arteriosus (PDA), ventricular septal defect (VSD), and atrial septal defect (ASD), where the predominant cardiac malformation (40%-70%) is atrioventricular septal defect (AVSD), with partial atrioventricular canal defect (ostium primum ASD with mitral cleft) being the most.

The only appropriate treatment is surgery. Medical treatment is indicated initially for those patients who are in heart failure. Surgery should have every chance of success with excellent prognosis.

The anaesthetic goal for such cases is to prevent exacerbation of pulmonary artery hypertension (PAH) and hemodynamic instability. PAH is managed by adequate ventilator support, inodilators (milrinone and dobutamine) and pulmonary dilators (nitroglycerin). Pain and anxiety are known to cause catecholamine release which in turn can adversely.

Influence pulmonary vascular tone .It has been reported that ketamine does not adversely affect pulmonary artery pressure.[8] We used intravenous ketamine for its sedative and analgesic effects. Thiopentone, propofol, inhalational agents are myocardial depressants. Fentanyl, midazolam and muscle relaxant-based anaesthesia has proved to be suitable in PAH. Inhaled nitric oxide and prostaglandin are beneficial in reducing the PAH.[9] PAH crisis is the main concern in the postoperative period and is treated with 100% oxygen with moderate hyperventilation; treatment of both respiratory and metabolic acidosis, removal or attenuation of precipitating factors should be undertaken. Inodilators, vasodilators and sildenafil citrate are also advocated.

To conclude, we report here the anesthetic management of a male child who presented with a

Down syndrome ,with VSD and ASD is quiet difficult. Understanding the pathophysiology and hemodynamics of combined or complex cardiac disease of this nature is important in the management of anesthesia. Fentanyl and midazolam-based anesthetic technique and appropriate management of PAH helped us in the perioperative management of this patient.

References:

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