

Anaesthetic management of a child with severe valvular aortic stenosis and mild pulmonary stenosis for ophthalmic surgery

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Abstract

Congenital rubella syndrome (CRS) has been associated with cataract and congenital cardiac disease. Incidence of cardiac defects with eye involvement could be as high as 95%. Anaesthesia for ophthalmic surgeries in children with severe valvular aortic stenosis (VAS) and pulmonary stenosis (PS) have been seldom reported. Anaesthetic management in a child with AS should be based on avoidance of systemic hypotension, maintenance of sinus rhythm and an adequate intra vascular volume, with awareness of the potential for myocardial ischemia. Here we describe the anaesthetic management of a 2 year old child with severe VAS and mild PS presented for intraocular lens implantation.

INTRODUCTION

Congenital rubella syndrome (CRS) has been associated with ocular and systemic anomalies including cataract and congenital cardiac disease (CHD) ¹. Complication rates of cardiac catheterization are higher in pediatric patients than in adults ²⁻⁴. Anaesthetic management for ophthalmic surgeries in children with severe valvular aortic stenosis (VAS) and pulmonary stenosis (PS) have been seldom reported. Severe complications including death after induction of anaesthesia in an infant with supra valvular aortic stenosis (SVAS) have been reported ³. Here we report and describe the anaesthetic management of a child with severe uncorrected VAS and mild PS presented for intraocular lens (IOL) implantation.

CASE REPORT

A 2 year old male child, weighing 10kg was admitted for bilateral IOL implantation. He was term baby. At 7 month of age, he was diagnosed as CRS with CHD and bilateral congenital cataract. Echocardiography revealed PS with peak systolic gradient (PSG) of 60mmHg and VAS with thickened, dysplastic aortic valve (AV) with aortic annulus of 6.5mm, PSG of 100mmHg and mean systolic gradient (MSG) of 60mmHg. Aortic valve and pulmonary valve balloon dilatation (AVBD/PVBD) were performed. Post procedure, due to absent lower limb pulses, streptokinase infusion was started which caused intracerebral hemorrhage

and status epilepticus. Subsequently, he was started on antiepileptics. Residual PSG were 50mmHg at the pulmonary valve (PV) and 50mmHg at the AV. After one month, child underwent bilateral lens aspiration under general anaesthesia which was uneventful.

Presently, the child had mental retardation and was on antiepileptics. There was no history of cyanosis, breathing difficulty, recurrent respiratory tract infections, dependent edema or limitation of activity. On examination, the child was active, pink, heart rate (HR) varied between 95/min during sleep to 120-130/min during crying. Non invasive blood pressure (NIBP) was 85/35 mmHg. The child's hemoglobin was 12.8g. Electrocardiogram (ECG) showed right ventricular hypertrophy (RVH), RS in V1-3 and no ST-T changes. X-ray chest showed cardiomegaly and prominent pulmonary artery. Review echo revealed PSG of 20mmHg at pulmonary valve, PSG of 90mmHg and MSG of 51mmHg at aortic valve with aortic annulus of 9-10mm. Ejection fraction was 60% with normal biventricular function.

Fasting guidelines were followed. Intravenous (IV) access was obtained after application of eutectic mixture of local anaesthetics (EMLA) cream. Child was premedicated with midazolam and infective endocarditis (IE) prophylaxis was administered in the preoperative room.

In the operative room, defibrillator and emergency cardiac drugs were kept ready. Oxygen saturation (SpO₂), ECG,

NIBP monitors were attached and anaesthesia was induced with fentanyl 20µg, incremental doses of thiopentone and with oxygen (O₂) /Air/isoflurane. Airway was secured with Proseal laryngeal mask airway (PLMA) after muscle relaxation with vecuronium. Pressure controlled ventilation was initiated to maintain peak airway pressure ±15cmH₂O and EtCO₂ of 35-40cmH₂O. Anaesthesia was maintained with vecuronium, O₂, air and isoflurane (MAC of 0.5). Intra-operatively, HR was maintained between 90-98/min, NIBP between 75-85mmHg systolic/35-45mmHg diastolic. SpO₂ was between 99-100%. Analgesia was administered with fentanyl, topical anaesthesia and paracetamol suppository. The surgery lasted for 60mins. Adequate fluid replacement was done with lactated ringer solution. After resumption of spontaneous efforts, neuromuscular blockade was reversed. PLMA was removed once child was having adequate spontaneous ventilation. Post-operatively, recovery was uneventful. The child was discharged 2days after the surgery with the advice to follow up in cardiology department.

DISCUSSION

Congenital rubella syndrome is associated with ocular manifestations, cardiac abnormalities, sensorineural deficit, psychomotor and mental retardation¹. Incidence of cardiac defects in CRS with eye involvement could be as high as 95%⁴. Commonest cardiac anomaly in CRS is patent ductus arteriosus (PDA)⁵. Although AS accounts for nearly 5-10% of CHD with predominance in males, there is only single case report of CSR with severe subaortic stenosis⁶⁷.

Aortic valve area is normally 2cm²/m² BSA. Two thirds of children with congenital AS present with bicuspid aortic valve, the remaining one third have tricuspid valves that may have fused, thickened or dysplastic cusps⁶. According to classification of AS in children based on valve area and valvular PSEG (peak systolic ejection gradient), our child had severe VAS with PSEG of 90mmHg (≥75mmHg) with bicuspid aortic valve⁶.

In our child, valvular PS is characterized by the fusion of the leaflets producing dome shaped valve with a central opening.

Patients are Children undergoing balloon valvuloplasty of the aortic valve are especially at risk for adverse events during cardiac catheterization as happened in our case². First eye surgery was uneventful as the child had undergone balloon dilatation. However for present surgery, PSEG at aortic valve was high due to restenosis of aortic valve. The

indication for valvuloplasty of the aortic valve is a PSG greater than 50mmHg². A definitive surgery or cardiac intervention would be ideal before the eye surgery, but child's parents were not ready for any cardiac intervention due to complications during previous balloon dilatation.

Usually in children less than 6 years of age, spectacles are prescribed following lens aspiration. Secondary IOL implantation is performed once the globe is fully developed (after 6 years of age). In our mentally retarded child, it was difficult for parents to encourage him to wear spectacles all the time to prevent ametropic amblyopia. So, early IOL implantation was necessary to improve vision.

Presently, the child was asymptomatic and surgery was minor without anticipated fluid shift, we accepted this child for IOL implantation under general anaesthesia with explained risks to his parents.

Review of medical literature to the best of our knowledge, does not reveal much information about anaesthetic management of congenital severe VAS for non cardiac surgery in pediatric population. Only one case of non cardiac surgery has been reported in a 9 year girl for liver transplantation and two cases of cardiac surgery i.e. PDA ligation and percutaneous transcatheter device occlusion have been described in children with VAS⁷⁸⁹. Myocardial ischemia during sevoflurane mask induction has been reported in an infant with undiagnosed supravulvular AS³. Adachi T et al used opioid with benzodiazepines successfully for induction and maintenance of anaesthesia for liver transplantation⁸. The authors monitored pulmonary arterial pressure and inserted intra aortic balloon pump for protection against myocardial ischemia.

Anaesthetic management in a child with AS should be based on avoidance of systemic hypotension, maintenance of sinus rhythm and an adequate intra vascular volume, with awareness of the potential for myocardial ischaemia¹⁰. Anaesthetic goal for PS include maintenance of a normal or slightly high heart rate, augmentation of preload and avoidance of factors that increase pulmonary vascular resistance (PVR). Defibrillator and emergency drugs were kept ready and arrangements were made for emergency cardiac surgery in view of difficult resuscitation in these children⁶.

IE prophylaxis was administered as the turbulent flow produced by the high velocity systolic jet in AS increases the

potential for development of endocarditis⁶.

The child was adequately premedicated to alleviate separation anxiety and crying which can lead to tachycardia and hypertension. EMLA cream was applied prior to IV access to abolish pain. Intravenous induction with titrated doses of thiopentone was planned as exposure to higher concentrations of sevoflurane may depress the myocardium. High dose of narcotics was not used to avoid postoperative respiratory depression as the procedure was short. LMA was preferred over endotracheal tube, to avoid the stress response to laryngoscopy, intubation and to facilitate smooth recovery. Nitrous oxide was avoided due to its adverse hemodynamic effects and EtCO₂ was maintained in range of 35-40mmHg with normal lung volumes and adequate depth of anaesthesia to decrease PVR¹¹. IV fluids were carefully titrated to avoid cardiac overload. Multimodal analgesia was administered to avoid side effects of pain during perioperative period. LMA was removed when the child was awake as any obstruction or hypoventilation might lead to acute arterial and pulmonary hypertension.

Invasive monitoring was not considered essential in view of a minor surgery without major fluid shift and acute cardiac failure may be reflected by ischemic changes in the ECG¹². Central venous catheter may not reflect exact ventricular status due to presence of PS. Transesophageal echocardiography could have been used as it is more useful than CVP monitoring for evaluation of ventricular filling and function¹³. Our patient had an uneventful perioperative period and child was pain free in postoperative room with stable hemodynamic.

CONCLUSION

No case of AS with PS in such a small child has been previously described. Though a prior definitive cardiac surgery would have been the best option before ophthalmic surgery, a proper understanding of the hemodynamic effects of severe AS with mild PS and vigilance regarding their anaesthetic implications enables us to manage this child successfully.

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