ANAESTHETIC CHALLENGES FOR CONGENITAL RUBELLA SYNDROME

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Abstract

Congenital Rubella Syndrome (CRS) is known to be associated with congenital cataract and congenital cardiac disease. Incidence of cardiac defects with eye involvement could be as high as 95%. Anaesthesia for ophthalmic surgery in infant with uncorrected/ unpalliated severe valvar pulmonary stenosis (PS), right ventricular hypertension, atrial septal defect (ASD), patent ductus arteriosus (PDA) has seldom been reported. Anaesthetic management in an infant with PS should be based on decrease in pulmonary vascular resistance (PVR), avoidance of tachycardia, myocardial depression, systemic hypotension, maintenance of sinus rhythm and an adequate intra vascular volume, with awareness for potential myocardial ischemia. Here we describe the anaesthetic management of a 4 month female child with severe PS, hypoplastic branched pulmonary arteries (Pas), right ventricular hypertension, right atrial & right ventricular volume overload and moderate ASD, PDA, tricuspid regurgitation (TR) who presented for cataract extraction surgery.

Keywords: Congenital Rubella Syndrome, Ophthalmic surgery, atrial septal defect, patent ductus arteriosus, pulmonary stenosis

Introduction

Congenital rubella syndrome (CRS) develops in an infant as a result of maternal infection in first trimester and subsequent fetal infection with rubella virus (German measles).The overall burden of CRS is unknown but more than 100 000 cases occur each year in developing countries [1].

The main abnormalities which occur are sensorineural deafness, eye defects, cardiovascular defects [Patent Ductus Arteriosus (PDA), stenosis of the pulmonary artery or its branches, septal defects], brain damage (microcephaly, mental retardation, meningoencephalitis), hepatosplenomegaly, thrombocytopenia, and neonatal jaundice [2, 3]. Other manifestations are Type I diabetes mellitus, growth retardation, transient hemolytic anemia, metaphyseal 'celery stalking' changes in long bones, transient pneumonitis, transient generalized lymphadenopathy, cryptorchidism, inguinal hernia, and dermal erythropoiesis ('blueberry muffin syndrome'). Such patients may require cardiac and/or non cardiac surgery. Usually, the cardiac status is optimized prior to non cardiac surgery, which may involve performing cardiac surgery. We report a case of a 4month-old child, with Rubella syndrome in whom non cardiac surgery was performed. Very rarely such cases appear in the literature.

Case report

A 4 month-old, 4 kg female baby presented to the ophthalmology clinic with bilateral congenital cataract. She was born by Pre-term Caesarean section in a primi para. Birth weight - 2.24kg, with low APGAR score and managed in radiant warmer for 10 days. The child had bilateral congenital cataract & was suspected to have CRS. 2D echo done on second day of life showed atrial septal defect (ASD) 8mm having bidirectional shunt, patent ductus arteriosus(PDA) 2.8mm. Pulmonary stenosis gradient 18mm Hg, Estimated RVSP by TR jet=95 mm Hg and moderate tricuspid regurgitation.

Paediatric cardiologist advised early BPV (Balloon pulmonary Valvuloplasty) with follow up for ASD every 6 months & Cataract extraction surgery after cardiac interventions. Cardiac Surgeons when consulted by parents during second month opined that as the patient is having severe diffuse hypoplasia of pulmonary arteries, balloon pulmonary valvuloplasty is not possible and cataract surgery to be done immediately to save eyes with high risk consent. Intra cardiac repair to be done after cataract surgery at a higher centre. During pre anaesthetic check up it was found that infant was having tachycardia, tachypnoea, and delayed mile stones. Saturation in both upper & lower limbs varied between 84 - 88%. Pansystolic murmur was heard all over the chest and best heard over left lateral sternal border. Other systemic examinations were within normal limits. X ray showed globular cardiomegaly (biventricular type). Proximal 1/3 rd pulmonary vessels were more prominent suggestive of pulmonary hypertension. Apex was also lifted up due to cardiomegaly. ECG showed right ventricular hypertrophy with P pulmonale suggestive of pulmonary hypertension, without significant axis deviation. Fresh 2D echo (at 4months) findings were moderate ASD 6mm (Left to Right shunt), moderate TR, Estimated RVSP by TR jet=100 mm Hg. Severe RV Hypertension. Marked RAVO, RVVO. Tiny PDA 2mm (Left to Right

shunt). PSG – 53mm Hg. Severe Pulmonary Stenosis – Valvar subtype PSG- 85 mm Hg. Tethered septal leaflet of tricuspid valve to IVS (6mm).Pulmonary annulus = 5mm RPA = 3.5 mm, LPA = 3.5 mm & Hypoplastic branches of PAs. To improve her visual prognosis, cataract surgery was planned to prevent amblyopia. Fasting guidelines (as short as possible) were followed. Intravenous (IV) access was obtained day before surgery only. Parents were counselled for high risk consent. Infective endocarditis (IE) prophylaxis was administered (Injectable [Inj.] cefotaxim 50mg/kg and inj. amikacin 2mg/kg) one dose prior to surgery. In the operative room, defibrillator & emergency cardiac drugs were kept ready. Difficult airway cart was arranged. Induction with inj. thiopentone sodium pre-added by inj. glycopyrrolate 0.02 mg was done in mother's lap in response to avoid separation related adverse effects and later shifted to operation table covered by blanket & cotton roll below to avoid hypothermia. Monitors simultaneously attached & IV fluid (Ringer lactate [RL] + 25% dextrose 50ml) started according to pre-calculated requirements. Additional dose of inj. thiopentone sodium 15 mg followed by inj fentanyl 3mcg IV was given. Anaesthesia deepened with oxygen + air + halothane to facilitate oral intubation with uncuffed portex tube number 4. Utmost care was taken to suppress all presser response. Maintained with Oxygen + Air + halothane by Jakson Ree circuit on controlled ventilation. Inj Fentanyl and Inj Milrinone were given in incremental doses. Nitrous Oxide was avoided as it is known to increase pulmonary hypertension. High doses of opioids were avoided to prevent respiratory depression resulting altered PVR. During operation hemodynamic parameters were closely observed. Surgical steps were meticulously performed. Adequate hydration was ensured and heamodynamic stability was maintained. Duration of surgery was 1.5hrs and duration of anaesthesia was 2hrs.

Discussion

Congenital rubella syndrome is a constellation of multi-system abnormalities, each of which poses a significant anaesthetic challenge. These children seek surgical intervention for cleft lip/cleft palate repair, cataract extraction, and congenital cardiac septal defect correction [4, 5].

Incidence of cardiac defects in CRS with eye involvement could be as high as 95% [6]. Commonest cardiac anomaly in CRS is patent ductus arteriosus (PDA) [7]. The indication for valvuloplasty of the pulmonary valve is a PSG greater than 50mmHg [8]. But was not performed due to presence of hypoplasia of pulmonary arteries. A definitive surgery (intra-cardiac intervention) or at least BPV would be ideal before the eye surgery. Secondary IOL implantation is performed once the globe is fully developed (after 6 years of age). Considering the necessity of urgent cataract extraction in this patient, as otherwise minor procedure without much physiological effects, we proceeded to conduct the surgery after high risk consent from the parents. Review of medical literature to the best of our knowledge, does not reveal much information about anaesthetic management of congenital severe PS for non cardiac surgery in pediatric population. A 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 valvular aortic stenosis (VAS) [9,10,11]. Adachi T et al used opioid with benzodiazepines successfully for induction and maintenance of anaesthesia for liver transplantation [10]. The authors monitored pulmonary arterial pressure and inserted intra aortic balloon pump for protection against myocardial ischemia. Anaesthetic goal for PS include maintenance of a normal or slightly low heart rate, augmentation of preload and avoidance of factors that increase pulmonary vascular resistance (PVR). While for ASD & PDA maintenance of heart rate with increase in the preload &

pulmonary vascular resistance along with decrease systemic vascular resistance reduces the flow across the defect. Anaesthetic goals in patients with PS, ASD & PDA are contrary to each other but when both these defects are present together a careful balanced technique has to be maintained to ensure haemodynamic stability as well as to ensure tissue needs are adequate met. Defibrillator and emergency drugs are to be kept ready and arrangements to be made for emergency cardiac surgery in view of difficult resuscitation in these children [(12)] . IE prophylaxis is essential as the turbulent flow produced by the high velocity systolic jet in PS increases the potential for development of endocarditis [(12)]. The child was adequately pre medicated to allay separation anxiety and crying which can lead to tachycardia and hypertension. Intravenous induction with titrated doses of thiopentone was done to avoid exposure to higher concentrations of halothane which would depress the myocardium. High dose of narcotics was not used to avoid postoperative respiratory depression which may alter the PVR. Adequate amount of thiopentone sodium & fentanyl helped to achieve deeper planes of anesthesia so that all presser responses were suppressed and intubation was smooth. Nitrous oxide was avoided due to its adverse hemodynamic effects. Air was used so that dilution of gases occurred helped in maintaining $EtCO_2$ in range of 35-40 mmHg. Low tidal volume without PEEP was used for assisted ventilation maintaining the adequate depth of anaesthesia to decrease PVR [(13)]. Inj milrinone was used as it is potent pulmonary vasodilator without marked decrease in systemic vasodilation. IV fluids were carefully titrated to avoid cardiac overload. Extubation was done in deeper planes to avoid 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 ECG [(14)] . Central venous catheter may not reflect exact left 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 function [15]. Our patient had an uneventful perioperative period and child was pain free in postoperative room with stable hemodynamic. Post operative follow up after 2 weeks showed happy parents as the child had improved visual acuity.

Conclusion

No case of ASD, PDA with PS & hyplostic branched pulmonary arteries 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 above mentioned congenital heart diseases and vigilance regarding their anaesthetic implications enables us to manage such patient successfully.

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