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Case Reports

Congenital Rubella Syndrome and anesthetic considerations

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Abstract:

Blindness in infants due to congenital cataract is up to 5-20% of all the vision loss of which 25% is due to Congenital Rubella Syndrome (CRS). CRS is a constellation of multi-system abnormalities with each posing its own variant of anesthetic dilemmas. CRS affects all the structures with rubella cataract being the commonest in the early age. Anesthetic management becomes a challenge in these rubella infants in view of Low Birth Weight (LBW), small infants, PDA/PS (pulmonary stenosis), ASD, PHT (pulmonary hypertension). Here we discuss the anesthetic management of three CRS infants coming for cataract surgery presenting with unique problems of the syndrome such as congenital cardiac septal defects, PDA, pulmonary hypertension and unanticipated difficult airway. Knowledge of the disease, anticipation preparedness, extreme caution and multidisciplinary approach for management is required for a successful outcome.

Key words: congenital Rubella Syndrome, anomalies, cataract, anesthesia

Introduction:

Congenital cataract is one of the most common treatable causes of visual impairment and blindness during infancy. [1, 2] In India, it is estimated that about 50,000 children are born blind from congenital cataract every year, of which at least 25% are due to maternal rubella. [3] In the developing countries like India 7.4-15.3% of childhood blindness is due to cataract.

Congenital rubella syndrome (CRS) is known to be associated with congenital cataract and congenital cardiac diseases. CRS develops in an infant as a result of maternal infections in the first trimester and subsequent fetal infection with rubella virus (German measles). The main abnormalities which occur are sensorineural deafness, eye defects cardiovascular defects (Patent Ductus Arteriosus, pulmonary artery stenosis or its branches, and septal defects), brain damage (microcephaly, mental retardation, and meningoencephalitis), hepato-splenomegaly, thrombocytopenia and neonatal jaundice. [4, 5] So such babies may be posted for cardiac and/or non-cardiac surgery. Optimization of cardiac status may be required prior to non-cardiac surgery pharmacologically or surgical correction. Infants with CRS require surgery for cataract in the early age. The timing of surgery is very critical for visual development. Most investigators recommend surgery within the first two months of life. [6] Anesthetic management becomes a challenge in these rubella infants in view of LBW, small infants, PDA/PS (pulmonary stenosis), ASD, and PHT (pulmonary hypertension). [3, 7]

We describe here 3 cases of CRS posted for cataract surgery with unique problems of congenital cardiac anomalies and unanticipated difficult airway.

Case Report:

Case 1:

Two months old female child with congenital cataract was posted for Right Lens aspiration. Parents gave history of preterm normal delivery, cried immediately after birth, developed physiologic jaundice for which phototherapy was given for 2 days. On examination, infant weighed 2.5 kg, lungs bilaterally clear and has continuous murmur with Hb of 9 gm%, RBC 4 millions/cumm, WBC 11 millions/cumm, Platelet count of 4.27

L/cumm, BT/CT of 1.5 and 4.45 minutes, TORCH – IgM positive suggesting Rubella Syndrome. Mother gave history of repeated URTI infections during first trimester. Child received immunization till date. 2D Echo showed ASD, PDA with L to R shunt. Patient was on syrup frusemide, aldactone, and phenobarbitone, with no other congenital anomalies. Cardiologist opinion was taken which suggested cardiac surgery to defer till the age of 1 yr and to go ahead with non-cardiac surgery under high risk. The same was discussed with parents and also informed regarding the requirement of post op ICU stay.

With 4 hours of NBM to breast milk child was taken to the OR connected to the monitors and induced with o2+Sevo2-6%, IV line secured given atracurium 1.5 mg, fentanyl 2+1 g and intubated with size 3 RAE tube and maintained on IPPV-PCV to achieve $ETCO_2$ between 35 to 45 mmHg with O_2, air and sevo1% and incremental doses of atracurium. Paracetamol rectal suppository was inserted to facilitate post operative analgesia. At the end of the surgery neuromuscular block was reversed with neostigmine and atropine and extubated uneventfully. Oral feeding was resumed after 4 hrs.

Case 2:

A two months old female child weighed 4 kg with congenital cataract, PDA with L to R shunt and mild Pulmonary Valvular Stenosis was posted for cataract surgery. With 4 hrs of NBM (breast feed) child was induced with O2+sevo8% given fentanyl 4+4 g and atracurium 2 mg, intubated with 3.5 mm size RAE tube and ventilated with PCV. ECG, SpO2, $ETCO_2$, temperature and NIBP monitoring was done. Anesthesia was maintained on 50% of O_2 and air, sevo1% and atracurium. Child behaved well during the intraoperative period. Extubated after reversing and post-op period was uneventful.

Case 3:

Child of three months with 3 kg was having congenital cataract, ASD, L to R shunt, dilated chamber with h/o IUGR and seizures. Antiepileptic was continued till on the day of surgery. After gas (o_2 & sevo) induction IV line was secured and intubated with atracurium 1.5 mg, fentanyl 3+2 g and midazolam 0.2 mg with 3 mm plain RAE tube. Here intubation could be done with the help of stylet on third attempt due to the anteriorly placed larynx and overhanging long epiglottis. Maintained on O_2/air, sevo1-2% and atracurium with pressure controlled ventilation with appropriate TV and rate to maintain $ETCO_2$. Vitals were maintained well throughout and reversed at the end of surgery. The recovery was satisfactory and the child was observed for any recurrence of seizures postoperatively.

Discussion:

CRS is an important cause of severe birth defects when a woman is infected with Rubella virus early in the pregnancy. The classic triad for CRS among infants is sensorineural deafness (58%), eye abnormalities especially retinopathy, cataract and micropthalmia (43%), microphthalmia, congenital glaucoma, and congenital heart disease (50%) mainly pulmonary stenosis PDA. This is also associated with LBW (23%), microcephaly (27%), hepato splenomegaly (19%) and other multisystem abnormalities [8] each of which pose a significant anesthetic challenge. These children seek surgical intervention for cataract extraction, cleft-lip/palate repair and congenital cardiac septal defect correction. [8, 9]

The main considerations in above cases were LBW infants with PDA, ASD, PS, PHT and unanticipated difficult intubation. Other congenital abnormalities were not present or could not be assessed as our cases were presented at their early age.

Incidence of cardiac defects in CRS with eye involvement is as high as 95% and commonest is PDA. [3, 10] In case 1 our concern was PDA with PHT. Main aim during non-cardiac surgeries is to maintain systemic vascular resistance (SVR) to pulmonary vascular resistance (PVR) ratio. [9] Factors which increase PVR like, hypercarbia, hypoxia, hypothermia and acidosis were avoided. Decrease in the SVR may lead to increase in R to L shunt, reduce pulmonary perfusion and hypoxemia [9] was also avoided. Nitrous oxide was not used as it is known to increase PHT. Inhalational induction was used as IV induction may cause hypotension in our cases. Case reports of Divekar et al [3] and Pallavi Gour et al [11] had fall in the blood pressure with thiopentone IV induction.

Anesthetic 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. Anesthetics goals in patients with PS, ASD & PDA are contrary to each other but when both these defects are present together a careful balanced technique needs to be maintained to ensure hemodynamic stability as well as to ensure tissue needs adequately.

NBM was kept minimal to avoid dehydration. Graded doses of opioids were used to prevent respiratory depression associated with high doses affecting PVR. [3] Smooth induction and intubation was done to prevent pressor response and maintained adequate depth of anesthesia. Lungs were ventilated with low tidal volumes without PEEP using Pressure controlled ventilation and the rate was adjusted to maintain $ETCO_2$ between 35-40 mmHg maintaining PVR. IV fluids were titrated cautiously to avoid cardiac overload and precautionary measures were taken to prevent air bubbles. Despite adequate precautions the variability in hemodynamic should be anticipated and one must be prepared to manage them. A very fine balance between adequate depth of anesthesia to reduce PVR and simultaneously to avoid fall in SVR has to be maintained.

LBW infants poses many challenges as they are highly sensitive to opioids, barbiturates and volatile anesthetic agents because of immature blood brain barrier and decreased ability to metabolize drugs. All the neonates in our cases were LBW and they are responded well to inhalational (O_2 + Sevo) induction and titrated doses of fentanyl. But cases have been reported having sudden hypotension to IV anesthetic induction. [3, 11] Also exogenous sodium, water and glucose should be provided peri operatively as they have low GFR and are more prone for hypoglycemia [12] if NBM period is prolonged.

CRS neonates may have undetected airway abnormality in the presence of other multiple congenital defects. Various upper airway anomalies like sub-glottic stenosis, shortened trachea, and short glottis carinal length associated with many congenital syndromes have reported by Wells et al. [7] Anesthetic management is a challenge if it remains unrecognized until induction and may lead to disastrous complications. In case 3 airway anomalies was made only on laryngoscopy showing large overhanging epiglottis and anterior vocal cords. The infant could be successfully intubated only with 3rd attempt with size 3 RAE tube with the help of stylet. LMA could be used successfully in syndromic babies to avoid intubation with its subsequent complications and PHT crisis. [7] Hence, one needs to anticipate and prepare for difficult intubation for successful management in CRS babies.

Infants are more prone for hypothermia. This was taken care by increasing ambient temperature of OT, using warm blankets, warm IV fluids and use active warming devices in the post anesthesia care unit. [12] Skin temperature monitoring was used both intra-op and post-operative period. All the infants were fed after 2-4hrs NBM post-operatively to prevent hypoglycemia.

Rubella is contagious disease which spreads in droplets. The respiratory secretion, cataractous lens is one of the most infectious materials hence warrants universal precaution.

Conclusion:

CRS is a constellation of multi-system abnormalities with each posing its own variant of anesthetic dilemmas. Knowledge of the disease, anticipation preparedness, extreme caution and multidisciplinary approach for management is required for a successful outcome.

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