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## UNIQUE ANAESTHESIA PROBLEMS ENCOUNTERED IN CONGENITAL RUBELLA SYNDROME

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| <p><b>Article Info</b><br/>Received 15/03/2015<br/>Revised 27/04/2015<br/>Accepted 02/05/2015</p> <p><b>Key words:</b> Patent Ductus Arteriosus (PDA), Rubella cataract, Difficult airway.</p> | <p><b>ABSTRACT</b><br/>Congenital cataract is most common treatable cause of blindness in infants of which around 25% are associated with congenital rubella syndrome (CRS). CRS is a constellation of multi-system abnormalities and each problem has significant anaesthetic implications. These patients are a challenge to anesthesiologists at various fronts like unanticipated airway abnormalities, uncorrected cardiac lesions, low birth weight etc. Here we report the anaesthetic management of two neonates of CRS posted for congenital cataract extraction presented with unique problems of uncontrolled severe pulmonary hypertension with PDA and unanticipated difficult airway. Anticipation, extreme vigilance and multidisciplinary approach for management are essential for successful outcome.</p> |
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### INTRODUCTION

Congenital cataract is responsible for nearly 10% of all vision loss in children worldwide. It is one of the most common treatable causes of visual impairment and blindness during infancy with an estimated prevalence of 1-6 cases per 10,000 live births [1]. 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 [2].

CRS affects almost all structures with rubella cataract being the most common [1]. Anaesthesiologists are confronted repeatedly with such patients for variety of surgical interventions including eye procedures. This has to be dealt intensively at various fronts like severe pulmonary hypertension (PHT) associated with PDA / Pulmonary Stenosis (PS) [1], developmental defects leading to unrecognized airway abnormality, low birth weight (LBW) neonates and thus imposes a great challenge in anaesthetic management [3].

We describe 2 cases of CRS posted for cataract surgery with unique problems of hemodynamic instability and unanticipated difficult airway.

### Case 1

A month old male baby born of non consanguineous marriage, LBW weighing 1.6 kg posted for cataract aspiration surgery. Clinical examination revealed high arched palate with low set ears, continuous murmur of grade 3. 2D-ECHO showed a large PDA with predominant L-R shunt, small PFO with L-R shunt, right ventricular dilatation and severe PHT with PSAP of 62 mm of Hg. Antibody titers for TORCH showed Rubella IgM, IgG positive. Standard balanced general anaesthesia was administered with IV Glycopyrollate 0.06mg, IV Fentanyl 3µg, IV thiopentone sodium 8 mg and IV Atracurium 1mg. After intubation with No. 3 ETT anaesthesia was maintained with Desflurane (5%-6%), O<sub>2</sub> and air with intermittent atracurium. All emergency cardiac drugs and vasopressors were kept ready. Sudden hypotension



occurred intraoperatively which was successfully managed with titrated dose of phenylephrine (5 -10 mcg/kg/dose every 10-15 minutes as needed). IV fluids were carefully titrated to avoid cardiac overload. Total duration of surgery was one hour and baby was reversed with IV Glycopyrrolate 0.04mg and IV Neostigmine 0.08mg and extubated and shifted to post anaesthesia care unit for monitoring.

## CASE 2

A month old female baby, confirmed case of CRS, full term, LBW of 1.2 kg was posted for cataract aspiration surgery. Infant had almost all the features of CRS including PDA with left to right shunt with PASP of 35 mm of Hg. On airway examination high arched palate was noted.

Patient was induced with standard balanced general anaesthesia with assisted mask ventilation and adequate depth was achieved. On direct laryngoscopy large over hanging epiglottis was seen and vocal cords could not be visualized (Cormach Lehane grade IV). Endotracheal intubation was unsuccessful in the first attempt with endotracheal tube (ETT) no. 3.0 however proper mask ventilation was ensured. With external laryngeal pressure, and with the aid of styliet, intubation was successful with No. 2.5 ETT in 3<sup>rd</sup> attempt and ETT placement was confirmed. This unanticipated difficult intubation was kept in mind for next 2 surgeries in the same patient and intubated with similar technique.

## DISCUSSION AND CONCLUSION

CRS develops in an infant due to maternal infection in first trimester and following fetal infection with rubella virus (German measles). The classic triad for CRS among infants is sensorineural deafness (60%), eye abnormalities especially cataract (25%), microphthalmia, congenital glaucoma, and congenital heart disease (45%) mainly PDA. Also associated with LBW (23%), microcephaly (27%), hepatosplenomegaly (19%) and other multisystem abnormalities [4].

Patients of CRS are repeatedly posted for variety of eye procedures under anaesthesia. Cataract extraction is an urgent vision saving procedure so complete optimization (correction of cardiac defects, adequate weight gain) of neonate may not be possible [1,5]. The main considerations in above cases were LBW neonates with PDA and severe PHT and unanticipated difficult intubation.

Incidence of cardiac defects in CRS with eye involvement is as high as 95% and commonest is PDA [1,2]. 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 [5]. Factors like hypoxia, hypercarbia, hypothermia and acidosis which lead to increased PVR should be avoided. Fall in SVR increases right to left

shunt, decreases pulmonary perfusion and induces hypoxaemia [5]. Nitrous Oxide was avoided as it is known to increase PHT. High doses of opioids were avoided to prevent respiratory depression resulting altered PVR [1]. Smooth induction and intubation was done to prevent pressor response and adequate depth of anaesthesia was maintained. Low tidal volume without PEEP was used for assisted ventilation to decrease PVR. IV fluids were carefully titrated to avoid cardiac overload and air bubble precaution was observed. However, there was an episode of sudden hypotension which was managed by titrated doses of phenylephrine [5]. In spite of adequate precautions we should anticipate the variability in hemodynamics and be prepared to manage them. A very fine balance between adequate depth of anaesthesia to reduce PVR and simultaneously to avoid fall in SVR has to be maintained. LBW neonate 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. Both the neonates were LBW and they are very sensitive to normal doses of anaesthetic agents which might lead to sudden hypotension. Also exogenous sodium, water and glucose should be provided perioperatively as they have low GFR and are more prone for hypoglycemia [6].

Airway abnormality in CRS may remain undetected in the presence of other multiple congenital defects. Wells et al have reported various upper airway anomalies like subglottic stenosis, shortened trachea, and short glottis carinal length associated with many congenital syndromes [3]. Anaesthetic management is a challenge if it remains unrecognized until induction and may lead to disastrous complications. In case 2 airway examination was inconclusive of airway anomaly however on laryngoscopy, large overhanging epiglottis was seen and no vocal cords (Cormach Lehane grade IV). We could successfully intubate the neonate only in 3<sup>rd</sup> attempt with a smaller size ETT using aid. There are reports of successful usage of LMA to avoid intubation with its subsequent complications and PHT crisis in such congenital syndromes [3]. Hence anticipation and preparedness is the key for successful management.

These neonates are more prone for hypothermia. This problem was solved by increasing ambient temperature of OT, using warm blankets, warm IV fluids and active warming in the post anaesthesia care unit [6]. Rubella is contagious disease which spreads in droplets. The respiratory secretion, cataractous lens is one of the most infectious materials hence warrants universal precaution.

CRS is a constellation of multi-system abnormalities with each posing unique anaesthetic problem and anticipation, preparedness, extreme vigilance and multidisciplinary approach for management is essential for successful outcome.



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