ANESTHETIC MANAGEMENT OF PEDIATRIC PATIENTS WITH CONGENITAL RUBELLA SYNDROME: OUR EXPERIENCE AND REVIEW OF THE LITERATURE

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Introduction

- Congenital rubella syndrome (CRS) is a triad of congenital malformations (Greg’s triad) which includes hearing impairment, congenital cataract, heart disease.
- Rubella infection presents with is maculopapular rashes and is caused by a toga virus of the genus *Rubivirus*.
- Rubella infection causes multiple congenital malformations which are collectively called as CRS.
- Apart from Greg’s triad, CRS may present with congenital glaucoma, pigmentary retinopathy, purpura, hepatosplenomegaly, jaundice, microcephaly, developmental delay, meningoencephalitis or radiolucent bone disease.
- Infants and children with CRS need anaesthesia for surgeries (ophthalmic, cardiac and cochlear implant) as well as for imaging studies.
- Even after improvement of anesthetic techniques, these cases are still an anesthetic challenge due to scarce literature on perioperative management of such children.
- We performed a retrospective review of the perioperative course of children with CRS, who underwent ophthalmic surgery in our institute.

Materials and Methods

- Institutional ethical committee approval was obtained for this retrospective study.
- Medical records of children with CRS who have had ophthalmic surgeries in our institute were collected and reviewed. Children with CRS, less than 10 years of age who underwent ophthalmic surgery or examination under anesthesia (EUA) were included for analysis.
- Case records that were incomplete were excluded. Preoperative variables like demographic data, cardiac anomaly, extra cardiac manifestations, symptoms, drug therapy, mental retardation and associated anomalies were tabulated.
- Intraoperative variables including type of anesthesia, surgery, analgesia, airway, other cardio-respiratory events, recovery and anesthetic complications were recorded. Postoperative course including cardiac failure, respiratory distress & intensive care (ICU) stay were also noted.

Results

- Total 46 cases were eligible for analysis

Table 1: Preoperative Variables

Preoperative Variables	Number of children n (%)
0 – 6 months	18 (39.13)
7 – 12 months	10 (21.74)
> 1 year	18 (39.13)
Male	26 (56.52)
Female	20 (43.48)
Cataract	33 (71.74)
Glaucoma	4 (8.70)
Pseudophakia	1 (2.17)
Squint	3 (6.52)
Corneal perforation	2 (4.35)
Aphakia	3 (6.52)
PDA	14 (30.43)
DEVICE CLOSED PDA	1 (2.17)
LIGATED PDA	1 (2.17)
PDA & PS	5 (10.87)
PDA, PS & AS	2 (4.35)
PDA & PAH	3 (6.52)
PDA & ASD	2 (4.35)
PDA & PFO	2 (4.35)
PDA, AS & COA	2 (4.35)
PS	3 (6.52)
TOF	3 (6.52)
ASD	1 (2.17)
MS	1 (2.17)
MS & PAH	1 (2.17)
OPERATED TGA	1 (2.17)
Presence of Cardiac Symptoms	12 (26.09)
On Cardiac drugs	13 (28.26)
Extracardiac Manifestations	20 (43.48)
Mental retardation (Yes/No)	15 (32.61)

Table 2: Intraoperative variables

Intraoperative Variables	Frequency (%)
Inhalational	38 (82.61)
Intravenous	6 (13.04)
MAC	2 (4.35)
LMA	27 (58.70)
Endotracheal tube	15 (32.61)
Face mask	4 (8.70)
Ventilation	
Controlled / Assisted	42 (91.30)
Spontaneous	4 (8.70)
Fentanyl	22 (47.83)
Fentanyl & PCM	16 (34.78)
Proparacaine , gel	4 (8.70)
Fentanyl, PCM & Topical	4 (8.70)
Eventful	3 (6.52)
Uneventful	43 (93.48)

Table 3: Postoperative complications

Complication	Age / Sex	Weight	Diagnosis	Cardiac Lesion	Cardiac symptoms
Fever	2 month	2.5kg	Cataract	PS	Previous hospitalization
Respiratory Distress	Male			ASD	Forehead sweating
				PDA	
Rest distress	25 Days	1.3 kg	Glaucoma	PDA	Suck rest suck cycle
Cardiac failure symptoms	Female			PS	
Respiratory distress	21 Days	1.2 kg	Glaucoma	Large	Suck rest suck cycle
Cardiac failure symptoms	Female			PDA	

Table 4: Postoperative complications

Suspected CRS	Clinically Confirmed CRS	Laboratory Confirmed CRS	Congenital Rubella Infection (CRI)
Infant presenting with 1)Heart disease (±) 2)Suspected deafness (±) 3)≥ 1 of •Microphthalmos •Nystagmus •Leucocoria •Strabismus •Buphthalmos •Diminution of vision	Infant with ≥2 of following 1)Congenital glaucoma 2)Cataract 3)Pigmentary retinopathy 4)CHD 5)Hard of hearing OR One finding above & 1 below 1)Purpura 2)Splenomegaly 3)Microcephaly 4)Mental retardation 5)Meningoencephalitis 6)Radiolucent bone disease 7)Neonatal jaundice	Clinically confirmed CRS & rubella-specific IgM antibody in blood (100% positive - 0-5 months& 60% positive - 6-11 months) Rubella virus can be isolated from pharyngeal and urinary samples if facility available. (60% shed rubella virus at age of 1-4 months; 30% at 5-8 months; 10% at 9-11 months)	Infant with no clinical signs of CRS, but who has a positive rubella-specific IgM

Discussion

- In the present study, high arched palate; microcephaly & micrognathia were present in children resulting in difficult mask ventilation needing two hand techniques.
- We preferred SGA in CRS children with CHD as SGA intubation and extubation response could be avoided and does not increase IOP during its insertion and removal.
- In this present study we could not establish a significant association between age and incidence of intraoperative events (p = 0.787), postoperative complications (p = 0.222) and Post op ICU stay (p = 0.222).
- Sex of the child did not establish significance with intraoperative complications, postoperative complications (p =0.075 & 1.000 respectively) & ICU stay (p = 0.572).
- There was positive correlation between preoperative presence of cardiac symptoms and postoperative ICU stay.
- Rest of the preoperative and intraoperative variables did not correlate with postoperative complications

Conclusion

- To conclude CRS is a multiorgan dysfunction with varied cardiac anomalies, knowledge about varied manifestations and apt clinical suspicion is vital.
- Extreme care should be exerted in children with CRS and symptomatic heart disease to improve perioperative outcomes.

References

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