Original Research Paper

Volume - 13 | Issue - 04 | April - 2023 | PRINT ISSN No. 2249 - 555X | DOI : 10.36106/ijar

Anaesthesiology

ANAESTHETIC MANAGEMENT OF EDWARD'S SYNDROME FOR CLEFT PALATE REPAIR

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ABSTRACT Trisomy 18 is the 2nd most common autosomal trisomy. It presents with a multisystem involvement including cardiovascular, respiratory, musculoskeletal and developmental anomalies. We hereby have a 1 year 8 month old child, diagnosed with Edward's syndrome who presented to our hospital for a cleft palate repair. Dysmorphic features such as low set ears, micrognathia, and reduced neck extension were noted. No cardiac abnormalities were found. Investigations were within normal limits. Our anaesthetic concerns mainly involved difficult airway, delayed recovery and shared airway. General anaesthesia with IPPV was planned keeping in mind the difficult airway. Intraoperative and postoperative management was uneventful.

KEYWORDS : Edward's syndrome, airway management, cleft palate

INTRODUCTION:

Trisomy 18, also known as Edward's syndrome is a chromosomal abnormality[1]. It is the 2nd most common autosomal trisomy after trisomy 21 (Down's syndrome). The incidence of Edward's syndrome is 1 in 5000 births, with a female preponderance[2]. Many babies with trisomy 18 don't survive past the second or third trimester of pregnancy. Its survival rate is 5-10% beyond the first year[3]. Multisystem involvement is common in these babies that include cleft palate, cardio respiratory system defects, defects of kidneys, stomach and intestines; deformed feet, low set ears, developmental delay, microcephaly and micrognathia[4,5]. The common surgical interventions include cardiac surgeries, palliative surgeries, gastrointestinal anomalies correction, cleft lip/ palate repair. Due to an elevated risk of mortality in the first month of life and presence of significant developmental disability in those who survive, ethical concerns exist regarding the necessity of surgical intervention[6]. Anaesthetic management of a child with trisomy 18 poses several challenges and involves a multidisciplinary approach to ensure a smooth recovery.

CASE REPORT:

A 1 year 8 month old male child, weighing 11 kgs, presented for a cleft palate repair to our institute. The child was diagnosed with Edward's syndrome at a private hospital and was referred to our centre for further management. The child presented with complaints of cleft palate and noisy mouth breathing with mild developmental delay. On clinical examination, low set ears, micrognathia, reduced neck extension and anemia was observed. No cardiac defects were noted. Investigations including CBC, coagulation profile, ECG and echocardiogram were within normal limits. MRI brain revealed CNS ventriculomegaly. General anaesthesia with IPPV was planned for cleft palate repair. After taking parental consent and confirming nil per oral status, the child was taken into the theatre.

A 22 G cannula was secured in the ward. Airway preparation with lignocaine nebulisation was done. OT preparation included warming blankets, warm fluids and ambient temperature of 28°C to prevent hypothermia. A difficult airway cart was kept ready. Premedication with injection Midazolam 20 mcg/kg IV was given. Routine noninvasive monitors were connected. Induction was done with titrated doses of sevoflurane 6-8% and injection Propofol 2 mg/kg, fentanyl 2 mcg.kg and dexamethasone 0.04mg/kg. Spontaneous respiration was maintained and confirmed with EtCO2 waveform. Check laryngoscopy was done with a MAC 2 blade, CL grade was 3b. With maintenance of spontaneous respiration, intubation was done with a reinforced OETT 4 mm with a MAC 2 blade on the 2 nd attempt over a bougie with OELM. General anaesthesia was maintained with air and oxygen mixture and sevoflurane. Atracurium was the muscle relaxant of choice. IV paracetamol and fentanyl boluses were used for analgesia. IVF was maintained according to the Holliday- Segar formula. The child was reversed and extubated once awake. Difficult airway cart was kept ready for emergency reintubation. Intraoperative and postoperative course in the hospital was uneventful.

DISCUSSION:

The trisomy 18 syndrome is a common chromosomal disorder due to presence of an extra chromosome 18, either full, mosaic trisomy or partial trisomy 18q[7]. Currently most cases of trisomy 18 are prenatally diagnosed, based on screening by maternal age, maternal serum marker screening, detection of sonographic abnormality (increased nuchal translucency thickness, growth retardation, choroid plexus cyst, overlapping of fingers, CHD)[8,9].

Anaesthetic concerns while managing a surgical patient with Edward's syndrome includes anticipated difficult airway, concerns regarding structural heart defects, growth defects, global developmental delay, delayed recovery and shared airway. Airway management considerations include dolicocephaly, micrognathia, and small mouth and reduced neck extension[10]. It is essential to be prepared with a difficult airway cart. In our case, intubation was relatively easy and was managed with a Macintosh size 2 blade. Masuda et al reported a boy with trisomy 18, undergoing testicular fixation, who developed muscular rigidity following suxamethonium administration, making endotracheal intubation impossible[11]. Intraoperatively, he developed a temperature of 38.4°C and elevated serum creatinine phosphokinase. However, no clear link exists between trisomy 18 and malignant hyperthermia. We monitored core and peripheral temperature with interthreshold range and no changes were encountered.

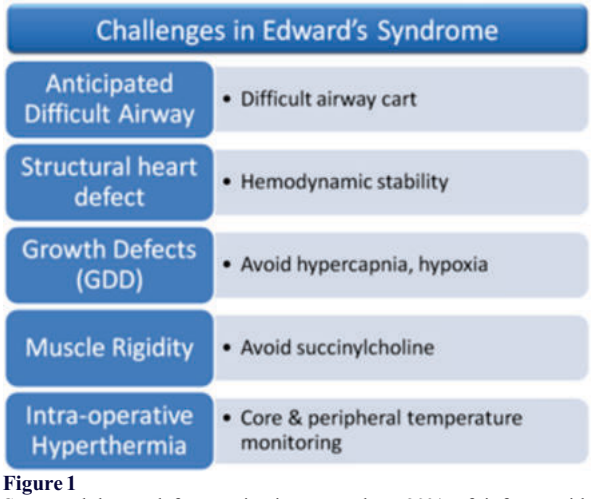

Figure 1

Structural heart defects exist in more than 90% of infants with Edward's syndrome. The common cardiac lesions encountered are ASD, VSD, PDA and polyvalvular diseases. Courreges et al suggested that these patients be considered as cardiac patients[12]. Hence it is imperative to maintain homeostasis between systemic and pulmonary circulation. Additional goals involve maintaining hemodynamic stability, avoiding hypoxia and hypercapnia.

Inhalational induction using agent with less cardiac depressant effects has been done successfully. Use of fentanyl as analgesic can be considered in children with pulmonary hypertension[13].

CONCLUSION:

Anaesthetic management of Edward's syndrome is uncommon, yet t

poses numerous challenges. No definitive anaesthetic protocol exists. Due to a short life expectancy, ethical concerns regarding surgical intervention also need to be addressed.

CONFLICTS

None

REFERENCES:

1. Edwards, J.H, Harnden D.G, Cameron A, et al. 1960. A new trisomic syndrome. Lancet 1:787-790
2. Dan Bernan, MD, 19 July, 2019; What is trisomy 18? 30 Sept 2020; webmd.com/baby/what is trisomy 18
3. Arun O, Oc B, Oc M and Duman A. 2014. Anaesthetic management of newborn with trisomy 18 undergoing closure of patent ductus arteriosus and pulmonary artery banding. Cardiovasc.J.Afr. 25: e 10-e12.
4. M Alghamdi, R Diogo, R Izquierdo et al. Detailed musculoskeletal study of a fetus with trisomy 18 (Edward's syndrome) with Langer's axillary arch, and comparison with other cases of human congenital malformations. J. Anat .Sci. Res 1, 1-8, 2018
5. Katasi C. Edward's syndrome: A case study. IJCN.2019, Vol 20; 18-21.
6. Arun Oguzhan, June 2017; Anaesthesia recommendation for patients suffering from trisomy 18 syndrome; 30 Sept 2020, www.orpha.net
7. A Cereda, Jc Carey- Orphanet journal of rare diseases, 2012- Springer
8. Irwing C, Richmond S, Wren C et al. Changes in fetal premature and outcome for trisomies 13 and 18: a population based study over 23 years. J Matern Fetal Neonatal Med 2011;24:137-141
9. Yawanaka M, Setoyama T, Igarashi Y, et al. Pregnancy outcome of fetuses with trisomy 18 identified by prenatal sonography and chromosomal analysis in a perinatal centre. Am J Med Genet. 2006; 140:1177-1182
10. Mifsud S, Bezzina M, Paris S. Anaesthetic management of a patient with Edward's syndrome. Clin.Case Rep 2016 Aug; 4(8):740-742.
11. Matsuda H, Kaseno S, Gotoh Y et al. 1983. Muscle rigidity caused by succinylcholine in Edward's syndrome. Masui 32:125-128.
12. Courreges P, Nievviarts R and Lecoutre D. 2003. Anaesthetic management for Edward's syndrome. Pediatr Anaesth. 13:267-269
13. Shew, S.B., Keshen J.H, Glass N.L, et al. 2000. Ligation of a patent ductus arteriousus under fentanyl anaesthesia improves protein metabolism in premature neonates. J.Pediatr.Surg.35:1277-1281.