ISSN 2515-8260 Volume 10, Issue 1, Winter 2023

Anaesthetic management in a paediatric patient posted for bilateral herniotomy with CHARGE syndrome.

1. **Dr. Sonal Sagar Khatavkar** (Professor Anaesthesiology Dr D.Y. Patil Medical College, Hospital & Research Centre, Dr. D Y Patil Vidyapeeth, Pimpri, Pune)
2. **Dr. Ujjwal Chandra**(Department of Anaesthesiology Dr D.Y. Patil Medical College, Hospital & Research Centre, Dr. D Y Patil Vidyapeeth , Pimpri, Pune)

Corresponding Author:
Dr. Ujjwal chandra
Address: Department of Anaesthesiology and Critical Care, Dr. D.Y. Patil Medical College, Hospital and Research Centre, Dr. D.Y. Patil Vidyapeeth, Pimpri, Pune 411018, Maharashtra.

ABSTRACT

Background: CHARGE syndrome is a genetic disorder with autosomal dominance, involved in the mutated gene CHD7 on chromosome 8. The abbreviation "CHARGE" characterizes the association of coloboma, heart anomalies, choanal atresia, and retardation of growth, genital and ear anomalies. This is a case report of 1.6 year old male baby who underwent bilateral herniotomy. The surgical procedure called for mild support of manual ventilation until the thorax was opened and then regulated ventilation with judicious fluid management was done.

Intra-operative care of hypercarbia and hypoxia with meticulous post-operative surveillance was done. In this case satisfactory respiratory function and bilateral herniotomy was successfully done.

Case report : We report a 1.6 year old male child with case of CHARGE syndrome . There are several features common in this disorder like coloboma, heart defects, choanal atresia, growth retardation, genital abnormalities and ear abnormalities. Here we are discussing the anaesthetic management for the same .

Conclusion: The anaesthetic care of a child with CHARGE Syndrome can be difficult. Although there are many problems associated to children with CHARGE Syndrome, and anaesthesia management is challenging and require expert skills. Early recognition and intervention can definitely improve the survival and outcome benefits. Assessing the risks of children with CHARGE syndrome should be monitored longer after surgery than the general paediatric population.

Key words: CHARGE syndrome, coloboma, heart anomalies, choanal atresia, growth retardation.

INTRODUCTION:

CHARGE is an abbreviation for several of the features common in the disorder like coloboma, heart defects, choanal atresia, growth retardation, genital abnormalities and ear abnormalities. The pattern of malformations varies among individuals with this disorder, and the multiple health problems can be life-threatening in infancy.

CHARGE syndrome occurs in approximately 1 in 8,500 to 10,000 new-borns.

ISSN 2515-8260 Volume 10, Issue 1, Winter 2023

CASE HISTORY AND MANAGEMENT:

A 1.6 year old, weighing 6 kgs male patient with swelling in the bilateral inguinal region since 1 month of life. Birth was at full term via LSCS in view of meconium stained liquor with absent cry, baby got admitted to NICU on ventilatory support for 7 days. Further investigations revealed that he had choanal atresia which presented as noisy breathing since birth, left facial nerve palsy, bilateral hearing loss and right kidney was absent.

In PAC, heart rate was 110 bpm with Sp02 98% on room air. It was observed that he was facing difficulty in breathing with nose. Respiratory rate was 30-35 breaths/minute, with no apparent signs of respiratory distress. Air entry was bilaterally equal with noisy breathing. Patient was reviewed after 2D Echo which came to be a normal heart study, ENT and Paediatrician advises were also taken. Finally, written informed consents for high risk, NICU and ventilator were taken and surgery was planned with GA with Caudal Block under ASA 2 risk.

Pre-op nebulization was given with Normal Saline as per ENTs advise. NBM for 6 hours was confirmed and pre-medication with Inj. Glycopyrrolate 0.004mg/kg was given.

Inside OT, monitors such as ECG, NIBP, Pulse Oxy meter and end tidal CO2 were attached. Induction was done with Inj. Propofol 2mg/kg, sevoflurane 2% with oxygen. After assessing his mask ventilation, confirmation by end tidal CO2 and rising of chest, Inj. Fentanyl 1µg/kg was given. After negative eye lash response, airway was secured with size 1.5 I-Gel and spontaneous ventilation was achieved. Thereafter, caudal block was given under all aseptic precautions with Bupivacaine 0.25% 1ml/kg.

Post-Surgery, I-gel was removed in deep plane of anaesthesia chest indrawing was observed. Patient was shifted to NICU for 6 hours in view of observation of any respiratory complications.

CASE DISCUSSION:

Patients with choanal atresia are considered to be at high risk as the anaesthetic management becomes challenging for them.

Both complete and partial nasopharyngeal obstruction exacerbates airway obstruction with relaxation of upper airway tone after sedation or induction of general anaesthesia. Inhaled anaesthetics are preferred when the airway is difficult to intubate.

Preliminary preparation in patients with choanal atresia who are anticipated to have a difficult airway, makes the intubation procedure easier and reduces complications that might arise. All airway conduits are to be kept ready.

Anaesthesiologists must be concerned about anticipated post operative airway difficulties which are common in children with CHARGE syndrome such as laryngospasm, bronchospasm, desaturation, and re-intubation are expected complications.

Assuming these risks, children with CHARGE syndrome should be monitored longer after surgery than the general paediatric population.

CONCLUSION

Children with CHARGE syndrome need intensive care, suitable to their particular features. Although there are many problems associated with CHARGE Syndrome, and anaesthetic management is challenging which requires expert skills. However, early recognition and intervention can definitely improve the survival and outcome benefits.

References:

1. Morgan DW, Bailey CM, Phelps P, Bellman S, Grace A, Wyse R. Ear - Nose - Throat abnormalities in the CHARGE Association. *Arch Otolaryngol Head Neck Surg*. 1993;119:49;54. doi:10.1001/archotol.1993.01880130051006. [PubMed] [Google Scholar]
2. Brooks P, Ree R, Rosen D, Ansermino M. Canadian pediatric anesthesiologists prefer inhalational anesthesia to manage difficult airways:a survey. *Can J Anaesth*. 2005;52:285–290. doi:10.1007/BF03016065. [PubMed] [Google Scholar]
3. Blake KD, Prasad C. CHARGE syndrome. *Orphanet J Rare Dis*. 2006;1:34. doi:10.1186/1750-1172-1-34. [PMC free article] [PubMed] [Google Scholar]
4. Naito Y, Higuchi M, Koinuma G, Aramaki M, Takahashi T, Kosaki K. Upper airway obstruction in neonates and infants with CHARGE syndrome. *Am J Med Genet A*. 2007;15:1815–1820. doi:10.1002/ajmg.a.31851. [PubMed] [Google Scholar]

5. Hara Y, Hirota K, Fukuda K. Successful airway management with use of a laryngeal mask airway in a patient with CHARGE syndrome. *J Anesth*. 2009;23:630–632. doi:10.1007/s00540-009-0791-y. [PubMed] [Google Scholar]