Airway management in a child with hydrocephalus and cleft lip scheduled for VP shunt procedure

First and presenting author - Dr Hima R Nambari, Senior Resident, Second author - Dr Shaila S Kamath, Professor and Head, Department of Anaesthesiology, Kasturba Medical College, Mangaluru.

Abstract

Hydrocephalus is commonly seen among paediatric patients in the neurosurgical and neuro-anaesthesia practice and it poses a difficulty in proper positioning of the patient during airway procedures. Cleft lip and cleft palate make laryngoscopy and ventilation a challenge in a majority of the patients. Individually these conditions indicate difficult airway management in patients. Together, the risk of difficulty in ventilation and securing an airway increases and adequate precautions need to be taken. It further necessitates a need for a proper plan in airway management and the need for careful execution of this plan with an experienced Anaesthesiologist at the core of this plan. A combination of hydrocephalus with midline structural anomalies is uncommon. We report a case of a 2-years-old child who presented with hydrocephalus, cleft lip and cleft palate. The clinical condition of the child posed a difficult airway management challenge.

Case report

A 2 years old female child presented with complaints of a large head and a midline defect in the upper lip and roof of the oral cavity since birth for a ventriculoperitoneal shunt procedure. She showed gross developmental delay. Her gross motor milestones corresponded to 6 to 9 months of age such as – lifting head more than 90 degrees without support and walking with support. Her fine motor milestones also corresponded to 9 months – pincer grasp. She spoke jargon and hence had a delay in language milestones as well. Her social milestones also corresponded to 6 – 9 months of age – stranger anxiety. Her mother gave no history suggestive of epilepsy. She had no difficulty in feeding the child. The child had no other known comorbid conditions. Her 5 years old elder sibling and twin did not have any similar problems. She was delivered via caesarean section in the 4th month of a twin pregnancy. She was the second twin in that pregnancy. Post-birth, she was observed in an intensive care unit due to signs of respiratory distress. However, she was not mechanically ventilated. She had no clear history of immunization. Before surgery, she showed no signs suggestive of respiratory tract infections or any other acute infections. The child was examined while she was comfortable in her mother’s presence. She was found to be cooperative and alert. Her vital signs were found to be within normal limits. Her chest was clear and there were no audible murmurs. Her oesophageal examination showed left-sided cleft lip and alveolus, high arched palate and micrognathia. There was no crowding of teeth. Other examination findings were - wide-spaced eyes, a flat nasal bridge, skin tags and coloboma of the eyelid. Her occipitofrontal circumference was 61 cm. She weighed 12.10 kilograms. Her laboratory investigations were grossly normal except haemoglobin of 9.9 g/dL and a total count of 1510/mm3. Her ECG showed sinus tachycardia with a rate of 175 to 180. Her subsequent echocardiogram showed – situs solitarius. Normal chamber sizes, AV and VA concordance with no evidence of ASD/VSD/PDA. The biventricular function was good.

MRA - a vascular study with causing proximal gross obstructive hydrocephalus with a significant reduction in white matter volume and focal destruction of the cortical mantle in the right superior frontal and parietal regions. She was to be taken up for a ventriculoperitoneal shunt procedure. The surgery, she was started on antiepileptic medication like levetiracetam 20 mg/kg/day and haloperidol intravenously a via a 24 gauge iv cannula on her left hand. She was advised 6 hours of fasting for solids and 2 hours for clear fluids. On the day of the procedure, she was shifted to the operating room and standard monitors like SpO2, ECG and NIBP were connected. The table was ramped using sheets and adequate padding for the bony prominences was ensured. With the patient sedated with fentanyl 2mcg/kg, ventilation was checked and deemed adequate. Sedation was then induced using sevoflurane and check direct laryngoscopy was done. Using a direct laryngoscope with a macintosh size 2 blade, size 4 micro cuffed ET tube was secured and fixed after confirming bilateral airway entry and e-tube trace. Neuromuscular blockade was achieved with atracurium 0.5mg/kg intravenously.

Postoperative period was uneventful. Intra operatively she was maintained on oxygen: air at 1:1 ratio and sevoflurane to maintain a minimum alveolar concentration of 0.7 – 0.8 and atracurium boluses of 0.1mg/kg every 20 minutes after the first 30 minutes. The procedure was uneventful and post procedure her residual neuromuscular blockade was reversed using neostigmine 0.05 mg/kg and atropine 0.02 mg/kg. A 160 mg rectal paracetamol suppository was inserted for analgesia. After a full recovery, she was extubated and shifted to the PICU for observation. Postoperative period was uneventful.

Discussion

Hydrocephalus is a clinical condition seen in pediatric neurological patients where an excess of cerebrospinal fluid (CSF) accumulates in the cisterns of the ventricular system of the brain causing an increase in intracranial tension (ICT) and neurological sequelae. Grossly put, there is an imbalance between the production and drainage of CSF leading to the pressure effect on the surrounding structures. It may be classified as non-communicating (obstructive) and communicating hydrocephalus. Congential hydrocephalus is seen in about 0.2–0.5/1000 live births. Its etiology remains unknown. An X-linked recessive form of aqueductal stenosis has been described in males. Choroid plexus papillomas often causes over production of CSF. Impaired absorption is seen in venous sinus occlusions, vein of Galen malformations, craniosenosis and diseases of the arachnoidal villi. The treatment options were excision of the choroid plexus, relieving any mass causing obstruction or use of devices to divert CSF from the intracranial compartment. Among the conduit devices, ventriculoperitoneal shunt is considered the best available form of treatment. Another upcoming and popular treatment modality is the endoscopic 3rd ventriculostomy. Cleft lip and palate is one of the most common congenital deformities with an average incidence of 1 in 700 worldwide. In the Asian population, its incidence is around 2.6 per 1000 live births or higher. As epidemiological data on a national scale is unavailable in India, several regional studies have reported a varied incidence of the cleft anomalies. Due to this, only rough estimates are available - approximately 35,000 per year. Very rarely do the above mentioned congenital anomalies occur in combination. A difficult airway as defined as ‘the clinical situation in which a conventionally trained anaesthesiologist experiences difficulty with facemask ventilation of the upper airway, difficult with tracheal intubation or both’ by the ASA Task Force on difficult airway management.

Intubation maybe difficult in the presence of dysmorphic features, reduced neck extension, restricted mouth opening, decreased temporomandibular joint mobility, macroglossia, mandibular anomalies reducing sub mandibular space, and any anatomical abnormalities involving the laryngo-tracheal passage. They require several counter measures to increase the chances of successful intubation. In our case, we issued strict nil per oral orders for six hours, planned for ramping and stacking, and inhalational induction and check laryngoscopy prior to intubation. The contents of the difficult airway cart were checked and kept ready nearby. Plans A, B and C were discussed well beforehand and the team was well prepared prior to the procedure. And this preparedness had led to the smooth execution of the intubation and extubation in this case. In conclusion, difficult airway situations in paediatrics can be handled by adequate preparation and detailed planning and discussion of the course of action with each team member. Unfamiliar or rarely seen cases are easier to handle when protocols are followed and senior clinicians are actively involved in the case. Hydrocephalus with cleft lip although may have increased the probability of difficult or failed intubation, proper counter measures to the anticipated difficulties ahead of time mitigated any possible complications.

References


Fig 1

Fig 2

Background

Paediatric neuro-anaesthesia practice has seen many cases of hydrocephalus. The dilatation of the ventricular system within the brain causes compressive effects on the parenchyma. Due to the larger diameter of the cranium in these children, visualization of the glottis and often ventilation is relatively more difficult than in otherwise normal children. A cleft lip is defined as a congenital deformity of the primary palate located anterior to the incisive foramen. Whereas cleft palate is defined as a congenital abnormality of the secondary palate i.e. soft and hard palate. Due to the anatomical abnormality, laryngoscopy and ventilation are a challenge in this group of patients. Individually these conditions pose difficulty in airway management in patients. Together, the risk of difficulty in ventilation and securing an airway increases and adequate precautions need to be taken. A combination of hydrocephalus with midline structural anomalies is uncommon. We report a case of a 2-years-old child who presented with hydrocephalus, cleft lip and cleft palate. The clinical condition of the child posed a difficult airway management challenge.