Successful Anaesthetic Management Of Meckel-Gruber Syndrome: A Case Report

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ABSTRACT

Meckel Gruber syndrome (MGS) is one of the rare autosomal recessive inherited disorder affecting several organ systems of the body. It is specifically characterised by occipital encephalocele, bilateral polycystic kidney, post axial polydactyly making the airway management difficult.

We would like to share successful management of a case of Meckel Gruber syndrome with giant occipital-cervical encephalocele posted for excision. Difficult airway cart was kept ready prior to intubation. This includes laryngeal mask airway of appropriate size, high frequency jet ventilation, fibre-optic bronchoscope, a cricothyroid cannula and preparations for tracheostomy.

Anaesthesia to paediatric age group poses many challenges to anaesthesiologist and that for congenital anomalies add to it. Working knowledge of these kind of syndromes and specific considerations for the same should be kept in mind when assessing a case of occipital encephalocele leading to more effective and appropriate management of cases.

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postoperative mechanical ventilation explained to both surgeons and relatives.

**Figure 1- Preop before excision**

![Figure 1- Preop before excision](image1)

Baby posted for excision of encephalocele sac under general anaesthesia. Operation theatre was kept warm and emergency airway cart was kept ready. Baseline parameters of baby recorded in lateral position were within normal limits. Baby received injection glycopyrrolate 0.05 mg and injection fentanyl 15 mcg intra venously and induced with sevoflurane. Only epiglottis was visualised during direct laryngoscopy in lateral position (Cormack Lehane grade 3) [9]. Due to non-availability of paediatric size fiber-optic bronchoscope we passed 1.5 size AMBU LMA™ in lateral position, ventilated the baby and confirmed its correct position by capnography. To secure definitive airway, uncuffed 4.5 mm endotracheal tube was blindly inserted through AMBU LMA™ in trachea and confirmed its correct position by capnography and bilateral auscultation of chest [Figure 3]. Intraoperatively baby placed in prone position and maintained with nitrous oxide, oxygen, sevoflurane and atracurium. Perioperative period was uneventful and post extubation baby shifted to post anaesthesia care unit for further observation. Her body weight was 5 kg in immediate post-op period suggesting the contribution of sac to be 2.5 kg in her presenting weight. Baby got discharged from hospital after 17 days with follow up advised.

**Figure 3- Equipment used for intubation**

![Figure 3- Equipment used for intubation](image3)

**Discussion**

Johann Friedrich Meckel in 1822 first described the syndrome MGS simply termed Meckel syndrome. Due to the involvement of several organ systems Meckel observed death in the pediatric age group [3]. Out of all the organ systems involved the major diagnostic criteria of MGS include at least 2 of these 3 classic manifestations such as cystic renal dysplasia, occipital encephalocele or other anomalies of central nervous system and polydactyly (found in 100%, 90% and 83.3% respectively) [4]. Varying presentations and the multi organ involvement makes this syndrome difficult for diagnosis and management of the case. Many infants die within hours of birth or if survive have poor quality of life. The cause of death could be due to organ dysfunctions like non-functional kidneys, liver and pulmonary hypoplasia. Ultrasound screening perinatally can be used to diagnose these disorders in the early trimisters. MGS can be
diagnosed as early as 14 weeks of gestation by ultrasound by characteristic findings of occipital encephalocele, polycystic kidneys and postaxial polydactyly [5].

Major anaesthetic challenge in management of occipital meningoencephalocele is securing the airway [6]. Because of the mandibular micrognathism and anatomical abnormalities of the larynx, tongue, and cervical vertebrae these patients usually present with difficult airway. Therefore, an alternative airway management plan should be ready prior to commencement of the procedure. Difficult airway cart should be kept ready prior to intubation. This should include laryngeal mask airway of appropriate size, high frequency jet ventilation, fibre-optic bronchoscope, a cricothyroid cannula and preparations for tracheostomy [7-8].

Specific attention has to be given to blood loss, maintenance of body temperature, position of head in prone position and its associated complications and endotracheal tube position [6]. Working knowledge of these kind of disorders should be kept in mind when assessing a case of occipital encephalocele which helps in effective and appropriate management of cases perioperatively.

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