Unanticipated difficult intubation in a child with Beals-Hecht syndrome presenting for emergency surgery

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Beals–Hecht syndrome is characterized by multiple joint contractures, arachnodactyly and kyphoscoliosis. We report the successful management of a patient diagnosed with Beals-Hecht syndrome who had micrognathia, a high arched palate and restricted mouth opening, who presented to us in the emergency for exploratory laparotomy. Oral intubation was successfully performed using aI-gel supraglottic airway as a conduit for intubation.

Keywords: Beals-Hecht syndrome; general anaesthesia; endotracheal intubation; i-gel; conduit

Introduction

Beals-Hechtsyndrome or Congenital Contractural Arachnodactyly (CCA) is classified as distal arthrogryposis Type 9.[1] Beals-Hecht syndrome is associated with difficult intravenous access, difficult airway, difficult positioning due to multiple contractures. We successfully managed a five year old who presented for exploratory laparotomy with a clinical diagnosis of peritonitis.

Case report

A 5 year old, 14kg male child diagnosed with Beals-Hecht syndrome (BHS) was posted for exploratory laparotomy with a diagnosis of peritonitis. On general physical examination the child had a mouth opening of 2.5cm, a high arched palate, contractures of elbow and hip joint, flexion deformity at elbows, wrists and ankle. The airway was Mallampati class 2.Cardiovascular system of the patient was normal.

His preoperative investigations were within normal limits. X-ray chest revealed thoracolumbar scoliosis.

After premedication with atropine 0.01mg/kg, anaesthesia was induced with propofol 2mg/kg and fentanyl 2µg/kg i.v. Cricoid pressure was applied as it was an emergency laparotomy. Following muscle relaxation with 1.2 mg/kg of rocuronium,larynx was visualized and classified as Grade III Cormack and Lehane. Intubation could not be accomplished even after releasing cricoid pressure. Since Intubating LMA or Proseal of appropriate size was not available, i-gel airway size 2 was inserted. Ventilation was satisfactory as seen by adequate chest rise and a normal capnography waveform.A cuffed tracheal tube (5.5mm ID) was successfully inserted through the i-gel in the first attempt. During removal of the i-gel, the ETT was stabilized with another smaller sized uncuffed red rubber endotracheal tube, by inserting its distal end into the proximal end of the cuffed ETT having removed both connectors. The i-gel was pulled in a cephalic direction and ETT was pushed caudally along with the uncuffed red rubber tube, and once the proximal end of i-gel neared the edge of the mouth the ETT was grasped with fingers inside the mouth. Now the red rubber tube was removed and subsequently the i-gel was also removed. The cuff of the ETT was inflated and its position confirmed. Anaesthesia was maintained with oxygen-nitrous oxide (33%:66%), rocuronium, and sevoflurane. End tidal capnography,arterial oxygen saturation, urine output and non-invasive blood pressure were monitored. Appendicectomy was done in one hour. Rectal diclofenac (25 mg) was inserted for post-operative analgesia. Neuromuscular blockade was reversed and extubation was done when the child was fully awake.

The child was shifted to postoperative recovery ward for observation.

Discussion

There have been few case reports of anaesthetic management of children with Beals-Hecht syndrome.[2,3] It is a rare autosomal dominant

disorder. Usually the patients have a marfanoid appearance, are tall and slender,their arm span exceeding their height measurement by 5cm.[4] Craniofacial features include abnormally shaped ears, mild micrognathia and a high arched palate.[5] Our patient had scoliosis, mild micrognathia, restricted mouth opening and a high arched palate.

A relatively short neck and micrognathia are associated with difficult laryngoscopy or intubation. Usually the line of vision of the larynx is not affected by a restricted mouth opening.[6] The restricted mouth opening in Beals syndrome appears to be due to an abnormal ligament which stretches from maxilla to the mandible anterior to masseter muscles.[7] Our patient was a case of unanticipated difficult intubation. The ASA difficult airway algorithm[8] recommends use of a laryngeal mask airway for adequate ventilation and oxygenation and also as a conduit for intubation. Since we were more familiar with the use of i-gel as a conduit for intubation, we used the i-gel with ease.[9] Literature search revealed the use of i-gel as a conduit for intubation in unanticipated difficult airway in few patients. Michalek et al described successful fibreoptic-guided tracheal intubation through the i-gel in two adults with genetic syndromes and intellectual impairment who had anticipated difficult airways.[10]

We were hence successful in using the i-gel as a conduit for intubation in a child with an unanticipated difficult airway, undergoing emergency surgery. Equipment for dealing with difficult airway should be available when dealing with children with various congenital anomalies.

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