AIRTRAQ FOR DIFFICULT INTUBATION IN MORQUIOS SYNDROME

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ABSTRACT

Introduction: Airway management in pediatric patients with atlanto-axial dislocation in the presence of craniofacial abnormalities is challenging. Moriquo’s syndrome is characterized by high incidence of abnormality at the cranio-vertebral junction with mucopolysaccharide deposit in the airway resulting in difficult airway management. Airtraq as a conduit for fibreoptic intubation in difficult airway is not reported Moriquo’s syndrome. We report successful tracheal intubation in a child with Moriquo’s syndrome with atlantoaxial dislocation using this technique.

Case report: An eleven year old child with Moriquo’s syndrome was scheduled for occipito-cervical fixation for atlantoaxial instability and odontoid hypoplasia. Airway examination revealed macroglossia, prognathism, short neck with restricted movements. General anesthesia with spontaneous ventilation was administered using sevoflurane. Despite a glottic view of grade I we failed to intubate using Airtraq. Using Airtraq as a conduit, intubation was accomplished using fibreoptic endoscope with the endotracheal tube threaded on it. Intubation was accomplished successfully with minimal movement of cervical spine. The postoperative recovery was unremarkable and patient had no fresh neurological deficit postoperatively.

Conclusion: Use of Airtraq as a conduit for fibreoptic intubation is an easy, safe and efficient airway management option in patients with difficult intubation with CV junction anomalies.

Keywords: Moriquo Syndrome, Atlantoaxial Instability, Difficult Intubation, Airtraq, Failed Intubation, Conduit, Fibreoptic Intubation

INTRODUCTION

Craniofacial abnormalities often make airway management difficult. The reported incidence of difficult intubation with mucopolysaccharoidosis (MPS) is high. ¹ There is high incidence of abnormality at the cranio-vertebral junction like odontoid hypoplasia and atlanto-axial dislocation resulting in instability of the cervical spine in these patients.² Cervical instability in the presence of difficult airway management not only poses difficulty to airway management and but also increases the risk of neurological deterioration during airway management. We describe successful management of tracheal intubation in an 11 year old Moriquo’s syndrome with atlantoaxial dislocation using a Airtraq as a conduit for fibreoptic endoscope.

CASE REPORT

Parental consent was obtained for reporting this case. A 11 year old child, born of non-consanginous marriage, diagnosed to have morquio’s syndrome presented to the department of Anesthesia, Nizams institute of Medical Sciences, with complaints of tingling sensation and weakness of both upper and lower limbs since two months. No history of motor weakness, normal bowel and bladder continence. The child was poorly built, underweight and short for his age. He weighed 24kg and his height was 100cm (disproportionate dwarfism). There was no mental retardation. The parents report a history of snoring. Symptoms of obstructive sleep apnoea (OSA) were not forthcoming. His airway examination revealed mild macroglossia, prognathism, short neck with restricted movements. X-ray cervical spine (figure-1) revealed hypoplasia of odontoid and flattening of C3 to C7 vertebral bodies. MRI spine: atlanto-occipital assimilation, basilar invagination, significant compression of cervico-medullary junction. X-ray of forearm showed medial slanting of lower ends of radius and ulna bilateral pointed proximal ends of 2 and 5 metacarpals, with widening of proximal ends of radius and ulna. 2D-Echocardiogram revealed bicuspid aortic valve, myxomatous mitral valve with trivial mitral regurgitation. There was no evidence of pulmonary arterial hypertension (PAH). The biventricular function was normal. Biochemical enzyme assay revealed significant reduction of Galactose -6-sulphate sulfatase2.3nmol/17hr/mg(Normal range: 40-170nmol/17hr/ mg) confirming the diagnosis of Moriquo’s syndrome.

An occipito-cervical fusion was contemplated. In view of the anticipated difficult airway management, intubation using Airtraq endolaryngoscope under general anesthesia and spontaneous ventilation with sevoflurane was planned. The child was co-operative for entering the operating room and allowed cannulation of the vein. Inj. glcopyrrrate 0.05mg, Inj fentanyl 40mcg were administered intravenously. Manual in-line stabilization was used to stabilize the neck. The child was induced with gradual incremental concentrations of sevoflurane while maintaining spontaneous ventilation. Airtraq® video laryngoscope (size 1) was used for intubation. The vocal cords could be visualized but advancing the endotracheal tube was difficult. The endotracheal tube was repeatedly going posteriorly despite manipulation of airtraq and manual posterior displacement of larynx. Patient was ventilated with facemask between attempts to intubate. The haemodyanamics and oxygen saturation were maintained at normal levels. The endotracheal tube was threaded over a nasal endoscope and airtraq was used as a conduit to facilitate intubation. The endoscope was passed through the vocal cords and a cuffed endotracheal tube size 5 was advanced into the trachea. Patient was maintained on volatile induction and maintenance anesthesia (VIMA) with sevoflurane intraoperatively. Surgery lasted for 3 hours. There were no haemodynamic changes during surgery. No neurological monitoring was performed intraoperatively as the institute did not have the capability. After end of surgery patient

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We encountered a similar problem in this case. Difficult intubation despite visualization of glottis is a common problem with videolaryngoscopic devices. Aitraq is a simple, inexpensive equipment suitable for management of difficult intubation in developing countries with paucity of advanced airway management gadgets. Airtraq is known to facilitate difficult intubation in pediatric patients without the need for cervical movement. Though glottis visualization was accomplished easily with airtraq intubation was difficult. The institute did not possess pediatric FOB. Moreover, being a center with low turnover for pediatric patients, the experience with pediatric FOB was also limited. A 30cm length 3.5mm nasal endoscope was available. The flexible tip of the endoscope facilitated easy intubation.

DISCUSSION

Management of difficult paediatric airway is challenging. This child posed an additional problem of moriquo’s syndrome with associated atlantoaxial instability, limiting any neck manipulation during intubation. The mucopolysaccharidoses (MPS) are a group of genetic disorders characterized by deficiencies in enzyme production that lead to accumulation of mucopolysaccharides (Glycosaminoglycans-GAG) throughout the body. The anatomic factors affecting respiratory status and hindering airway management in moriquo’s syndrome include macrocephaly, a prominent forehead, and short neck, upper airway narrowing by hypertrophied tongue, tonsils, adenoids, and mucous membranes, thickening and redundancy of the oropharyngeal mucosa, as well as blockage of nasal passages, lower airway narrowing by deposition within the tracheobronchial mucosa, decreased thoracic dimensions related to scoliosis and thoracic hyperkyphosis; and decreased abdominal dimensions because of lumbar hyperlordosis, gibbus formation, and hepatosplenomegaly. These abnormalities, along with stiff temporomandibular joints and an anteriorly positioned larynx, make mask ventilation and endotracheal intubation extremely difficult. The poor respiratory reserve also reduces the safe apnea period for intubation.

There are several reports of difficult or failed laryngoscopy with MPS. In addition to difficult airway, cervical instability is also a concern during intubation in these patients requiring neck immobilisation during intubation to reduce the possibility of neurological deterioration which also adds to the difficulty in intubation.

Though fibreoptic is the gold standard, however, the deposition of soft tissue in the neck and oropharynx in Morquio syndrome may present difficulties for conventional fibreoptic intubation. Use of videolaryngoscopes is reported in difficult or failed laryngoscopy in MPS. Difficult intubation despite visualization of glottis is a common problem with videolaryngoscopic devices. We encountered a similar problem in this case. Despite adequate glottis visualization tracheal intubation was not possible as the endotracheal tube was repeatedly directed posteriorly. Maneuvers to aid visualization like neck movement were contraindicated due to craniovertebral instability.

Successful use of a supraglottic airway device as a conduit for fibreoptic -guided tracheal intubation has been described in Hunters syndrome. There are no reports of use of airtraq as a conduit for fibreoptic intubation in Moriquos syndrome. Airtraq is a simple, inexpensive equipment suitable for management of difficult intubation in developing countries with paucity of advanced airway management gadgets. Airtraq known to facilitate difficult intubation in pediatric patients without the need for cervical movement. Though glottis visualization was accomplished easily with airtraq intubation was difficult. The institute did not possess pediatric FOB. Moreover, being a center with low turnover for pediatric patients, the experience with pediatric FOB was also limited. A 30cm length 3.5mm nasal endoscope was available. The flexible tip of the endoscope facilitated easy intubation.

CONCLUSION

The combination of airtraq with fibreoptic endoscope were complimentary and enabled easy intubation in moriquo’s syndrome with predicted difficult airway with minimal cervical movement and no deterioration of neurological status.

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