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Letter to Editor

Anaesthetic considerations in a rare case of Bardet Biedl syndrome patient scheduled for adenotonsillectomy

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Dear Editor,

We report a case of 17-year-old female patient who presented with dysmorphism since birth and was diagnosed as Bardet Biedl syndrome (BBS) by genetic testing, with a positive family history. Abnormalities included retinitis pigmentosa leading to reduced vision in the night, obesity (Body mass index - 32kg/m^2), developmental delays, hypogonadism (secondary amenorrhea), spasticity of lower limbs, dental crowding, and high arched palate. The patient presented to the hospital with complaints of recurrent throat pain, associated with fever on and off for the past 2 years, and was diagnosed with adenotonsillitis, scheduled for adenotonsillectomy.

BBS is a rare autosomal recessive ciliary disorder with a prevalence of 1 in 140,000. The disease involves multiple systems,[1] although renal and cardiovascular manifestation account for the greatest morbidity and mortality.[2] Craniofacial and airway abnormalities are not diagnostic but are highly likely to be present and are frequently associated anesthetic morbidity and challenge.[3–5]

In our preoperative assessment a multidisciplinary approach was employed. Due to patient's low cognitive function, her mother provided the medical history. On examination of the patient, we found the patient to have difficult airway, reduced mouth opening (2.5cm), short neck, high arched palate, Mallampati class 3 and difficult venous access due to obesity. Cross reference was obtained from the cardiologist to rule out structural heart diseases, ophthalmologist regarding retinitis pigmentosa of both eye and gynaecologist for secondary amenorrhea due to hypogonadotropic hypogonadism. Investigations ruled out renal dysfunction and diabetes. (Figure 1)

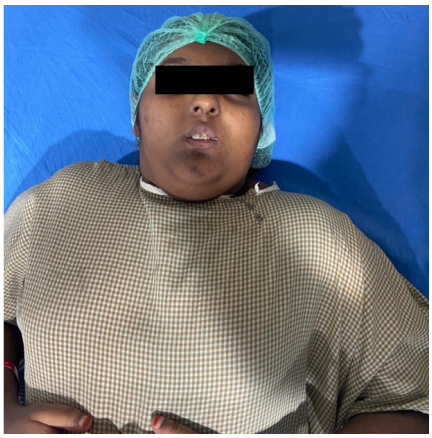

Figure 1: Patient of Bardet Biedl syndrome with craniofacial abnormality and obesity

IV access was secured in the pre operative holding area with USG guidance. In the OT difficult airway cart was kept ready. In order to optimise the first attempt of

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laryngoscopy we planned to position the patient in Head End Elevated Laryngoscopy Position (HELLP), to align the external auditory meatus to the suprasternal notch in an imaginary straight line and to use C-MAC video laryngoscope. (Figure 2)

Figure 2: Positioning the patient for intubation in head elevated laryngoscopy position (HELP)

Patient was preoxygenated for 5 minutes and then induced with Fentanyl and Propofol. Upper airway obstruction was present as anticipated. Use of oropharyngeal airway was preferred over nasopharyngeal airway due to the presence of hypertrophied adenoid. After confirming the ability to ventilate (using two hand mask ventilation), muscle relaxant Vecuronium was administered. Use of succinylcholine was avoided due to the presence of spasticity in the lower limb in preoperative assessment. Sugammadex was kept ready to reverse the neuromuscular blockade of Vecuronium in case of a Can't Intubate and Can't oxygenate (CICO) situation.

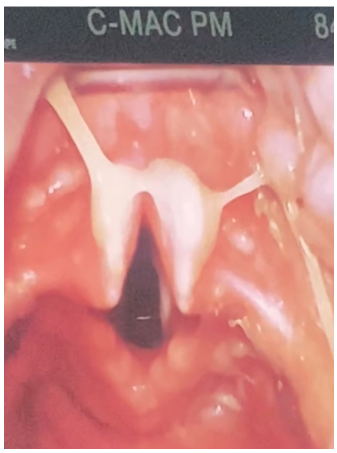

Figure 3: CL- 2B view of glottis C-MAC videolaryngoscope

Patient was intubated with 7 size RAE tube, with C-MAC video laryngoscope. Intraoperatively neuromuscular monitor was used to administer muscle relaxant when required. Intraoperative period was uneventful. Patient was extubated, after adequate reversal with Neostigmine (with glycopyrrolate) and fully awake. Postoperative analgesia was managed with Paracetamol. (Figure 3).

To conclude, patients with BBS may present for various procedures and is important to recognise these patients. A multispeciality approach is needed to assess and optimise these patients. Careful consideration is essential in the intraoperative and postoperative period for managing difficult airway.

1. Conflict of Interest

None.

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