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Letter to Editor

Anaesthetic challenges in a patient with hallermann–Streiff syndrome and cleft palate undergoing cataract surgery

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Dear Editor

The paediatric airway is significantly more versatile and challenging than the adult airway due to distinct anatomical and physiological differences. An experienced anaesthesiologist, equipped with thorough preoperative evaluation and well-planned intraoperative strategies, can achieve better outcomes by anticipating and managing potential complications.[1] Hallermann-Streiff syndrome is a rare congenital disorder featuring craniofacial abnormalities with dental defects and developmental delay. Only 200 cases globally, etiology remains unclear through GJA1 gene mutations which are sporadic involving ectoderm and mesoderm.[2]

A 4 years old female weighting 10kgs with diminision of vision and whitish discoloration of the pupil since 2 months. Birth history revealed preterm vaginal delivery and cleft palate since birth but tolerating oral feeds without regurgitation and mutism and bilateral hearing loss since age of 1 year. Physical examination revealed short stature, frontal bossing, almond shaped eyes, microstomia, retrognathia (**Figure 1**), cleft palate (**Figure 2**) widely spread nipples, loose skin, tapering fingers, syndactyly in left lower limb. Her preoperative blood investigations were within normal limits. 2D echo was suggestive of normal heart study.

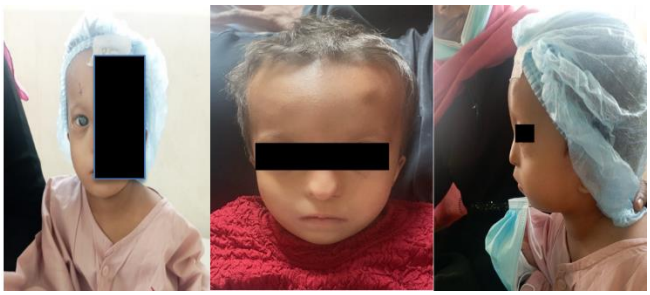

Figure 1: Right eye cataract, frontal bossing, microstomia, retrognathia

Figure 2: Cleft palate

In the operating room, standard ASA monitors (ECG, NIBP and SpO2 probe) were applied. A 24-gauge cannula

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was already secured in the left hand, and ringer lactate was initiated as maintenance fluid. The child was premedicated with 0.04mg of glycopyrrolate, 0.2mg of midazolam. Induction was performed with 20mcg of fentanyl and 20mg of propofol and 20mg of succinylcholine given and ventilated for 1 minute. A senior anaesthetist using the Macintosh size 2 blade with external manipulation the glottic opening was successfully visualized. An endotracheal tube (ETT) of size 4.5 was introduced and fixed at 12cms after confirming bilateral air entry. Muscle relaxant atracurium 5mg given and was ventilated with Oxygen and air 1:1 ratio with 1% sevoflurane. Intraoperatively patient remained hemodynamically stable. The procedure was completed uneventfully within an hour. The neuromuscular blockade was reversed with 0.5mg of neostigmine and 0.08mg of glycopyrrolate. Extubation was smooth and patient transferred to post-operative care unit for monitoring.

1. Discussion

Patient with Hallermann Streiff syndrome due to anatomical structural abnormalities of upper respiratory tract like frequent respiratory infection, feeding difficulties, sleep apnea but as such symptoms are not noted in our patient.[3] Due to upper airway deformities with cleft palate and facial features like microstomia, retrognathia, brittle teeth, anterior larynx contributes for difficulty in laryngoscopy anticipating difficult intubation.[4] Management of difficult airway is challenging more in paediatric age group because of the risk for hypoxia and bradycardia. A video laryngoscope, bougie, and equipment for emergency tracheostomy should be kept ready. Avoiding a reactive airway and ensuring smooth intubation and extubation without eliciting a stress response are crucial.[5] Strict adherence to paediatric difficult airway management guidelines is essential to minimize the risk of complications. Preoperative preparation included a fully equipped difficult airway trolley, immediate availability of emergency resuscitation drugs, and the presence of an experienced senior anaesthesiologist ensured prompt and effective airway management.

2. Informed Consent and Patient Confidentiality

Informed consent was obtained from the parents to use the images for medical publication with measures taken to maintain the anonymity of the patient.

3. Conflict of Interest

None.

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