Royal National Orthopaedic Hospital NHS Trust

AIRWAY IMPROVEMENT FOLLOWING HALO-TIBIAL TRACTION IN A CASE OF AXENFELD-REIGER SYNDROME

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We present a case of a 4 year-old child with Axenfeld-Reiger Syndrome referred to our centre for urgent kyphosis correction due to worsening neurological weakness in both legs. She was managed initially with staged halo-tibial traction prior to operative fusion. Progressive halo-tibial traction improved laryngoscopic views from Cormack & Lehane grade III to I in only two weeks between successive proceedures.

Introduction

- Axenfeld-Reiger Syndrome is a rare disorder primarily affecting the eye, particularly the anterior chamber.
- Its prevalence is estimated to be 1:200,000. The condition is inherited as an autosomal dominant condition of which there are 3 genotypes
- Patients may have characteristic facies with a prominent forehead, flattened midfacae and nose, hypertelorism and maxillary hypoplasia. Microdentia and oligodentia may also be a feature.
- ARS can be associated with other defects such as pituitary dysfunction leading to growth retardation.

Case Study

- A 4 year old girl of 20kg presented with bilateral leg weakness. She had severe kyphosis. Previous surgery had been performed at another centre for kyphosis correction one year earlier with the insertion of an extensible magnetic growth rod system from T2-L2. Revision surgery to trim prominent rods was required 8 months later.
- Her condition continued to to rapidly worsen with deteriorating power in both legs leaving her unable to walk. She was then referred to RNOH for ongoing management.
- Her previous anaesthetic history had been uneventful with an intravenous induction of general anaesthesia.
- On general examination she was of small stature and noted to have loose teeth. No other concerns were identified.
- For application of the halo-tibial traction at RNOH anaesthesia was conducted by inhalational induction with nitrous oxide and oxygen with incremental Sevofluarane. On this occasion a size 2.0 LMA was inserted without difficulty.
- She returned for subsequent intervention 2 weeks later. The same technique was used for induction whilst maintained in halo-tibial traction. Direct laryngoscopy was performed for intubation and the best views achieved were noted to be grade III. Intubation was successful with a Bougie on first attempt.
- Two weeks later the patient returned for surgical anterior release of vertebral ligaments. The same operator performed an inhalational induction of anaesthesia in halo-tibial traction. Laryngoscopic views had improved to grade I allowing easy intubation.

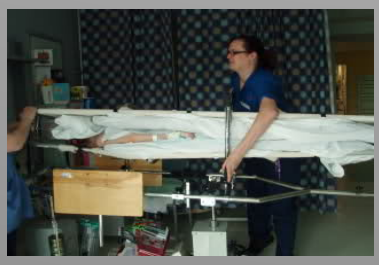

Figure 1. Halo-tibial traction frame

Considerations for anaesthesia

- Preoperative anaesthetic assessment in children with ARS should look for facial features such as maxillary hypoplasia that may impede effective bag-valve-mask ventilation [1].
- Teeth are often of poor quality or easily loosened and patient/parent should be warned of dental damage.
- Cardio-vascular assessment should seek to illicit other congenital defects.
- Extra care should be taken not to exacerbate conditions affecting pressure on the orbit, especially in the prone position as these children may suffer from glaucoma later in childhood.
- Familiarity and training with the apparatus as part of the pre-assessment is helpful. The horizontal bed frame has at one end a halo which is attached by pins to the skull. Traction is supplied in a pulley system distally. The bed frame is turned periodically 180 for comfort.
- Halo-traction therefore often presents the anaesthetist with challenges to intubation as access to the airway is limited surrounding the patient's face and the cervical spine is fully immobilised not permitting neck extension.
- Awake fibre-optic techniques are often not suitable in paediatric patients due to poor co-operation. Asleep fibre-optic techniques and videolaryngoscopy have been described as successful alternatives to managing the difficult paediatric airway in emergency and elective situations

Conclusions

- Axenfeld-Reiger Syndrome may contribute to increased difficulty with airway management due to cranio-facial features.
- The application of halo-tibial traction initially may certainly add difficulty to airway management due to limited access to the patient's airway.
- A difficult airway plan should be prepared and/or rehearsed prior to the start of anaesthesia
- This case report highlights the need for full appreciation of the anticipated difficult paediatric airway caused by a combination of causes. Of particular interest in this patient was the rapid and dramatic improvement in laryngoscopic views from III to I, that may be attributable to halo-tibial traction.

References

1. Baduni N, Pandey M, Sanwal MK, Verma M. Anaesthetic challenges in a patient with Axenfeld-Reiger syndrome. *Anaesth Essays Res* 2012;6:108-9