Conclusions: This study suggests that nasopharyngeal airway insertion (compared to tracheostomy) may be associated with improved outcomes regarding feeding in children with Pfeiffer syndrome.

OBJECTIVES

Pfeiffer syndrome is characterised by craniosynostosis and midface hypoplasia. Airway management includes the use of a nasopharyngeal airway (NPA) and tracheostomy. These patients can also have nutrition difficulties and dysphagia due to their abnormal anatomy requiring percutaneous gastrostomy (PEG) in severe cases. The aim of this study was to describe the nature of feeding difficulties in children with Pfeiffer syndrome, and to assess if there is any relation with management of their airway symptoms.

METHODS

Retrospective review of patient records diagnosed with Pfeiffer syndrome over a 20-year period (January 1998 - January 2018) at Alder Hey Children’s Hospital. The method of airway management and feeding support was recorded.

RESULTS

13 patients were included. There were 6 males and 7 females. The median age was 6 years (range 2-18 years). Five patients required tracheostomies (38%) and three patients required NPA (23%). Eight patients required PEG feeding (62%). Six patients (46%) had videofluoroscopy (VFSS) performed, of which 2 demonstrated aspiration. Five patients (38%) were orally fed, of which one (8%) had no problems with feeding whatsoever. One patient had feeding difficulties due to the nasopharyngeal airway with feed being drawn up the NPA, necessitating PEG feeding eventually. One patient had food suctioned from the tracheostomy, although VFSS did not demonstrate aspiration. Tracheostomy for airway management was more likely to be associated with PEG feeding than NPA (p=0.02).

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