Airway management in children with Pfeiffer syndrome: A retrospective review of 413 anaesthetics at one centre

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Introduction

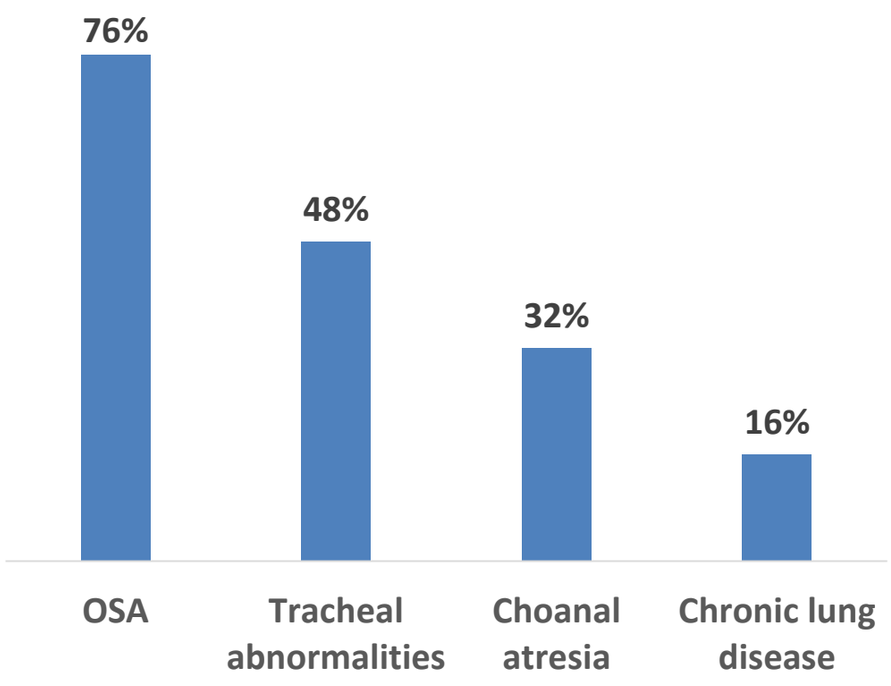
Pfeiffer syndrome is a rare autosomal dominant genetic disorder, characterised by craniosynostosis, maxillary hypoplasia, and upper airway obstruction. Anaesthetic airway management can therefore be challenging. We conducted a retrospective review to determine the nature of preoperative airway pathology and incidence of intraoperative airway difficulty in this group of children.

Methods

Following research governance approval, we used the hospital craniofacial database to identify children with Pfeiffer syndrome. A retrospective review of all their available anaesthetic records was conducted and data on patient demographics, preoperative airway status, and intraoperative airway management was analysed. 25 children and 413 anaesthetic records were evaluated.

Results

Percentage of children with the following airway & respiratory pathologies



Difficulty of BMV	Freq.	%
Easy	126	63%
Moderate (required simple adjuncts)	61	30%
Difficult (2 person technique / LMA)	12	6%
Not Possible	2	1%

C&L Intubation Grade	Freq.	%
1	108	51.7%
2	53	25.4%
3	45	21.5%
4	3	1.4%

68% of children had a tracheostomy:
53% inserted in first year of life
44% were decannulated within 1yr
56% had it for between 5yrs to life

Airway obstruction on induction of anaesthesia occurred 37% of the time. Intubating difficulty varied over time.
48% of patients had a change in C&L grade of 2 or more.
80% were a grade 1 & 56% a grade 3-4 at least once.
20% of difficult intubations were previously easy.

Conclusion

This is the largest review of anaesthetic airway management in children with Pfeiffer syndrome. We found a high incidence of airway and respiratory pathology, including OSA and tracheostomy dependence. Airway obstruction on induction was the most common anaesthetic complication. When compared to Apert syndrome, a similar craniofacial disorder, these children were 15 times more likely to have a difficult (grade 3 or 4) intubation. Importantly, ease of intubation in many children varied considerable over time.