A retrospective review of perioperative complications in children with Pfeiffer syndrome at one centre

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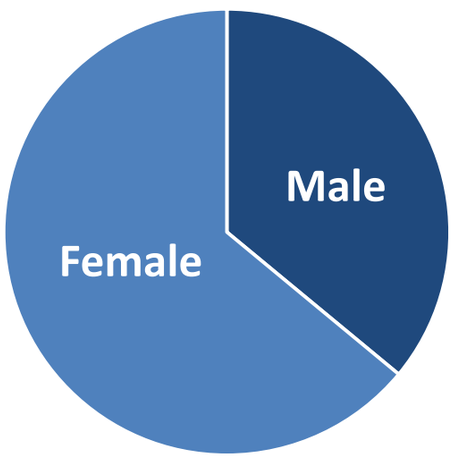
Introduction

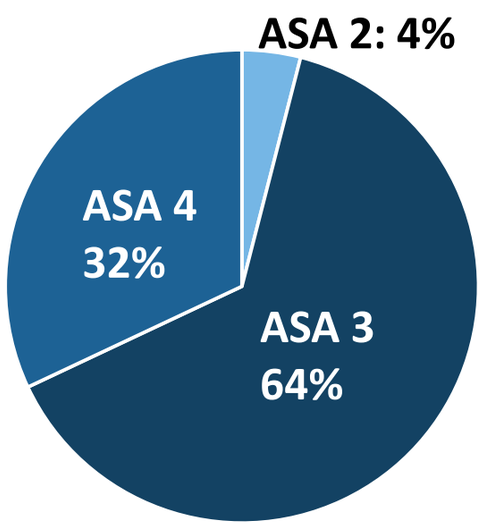
Pfeiffer syndrome is a rare autosomal dominant genetic disorder, characterised by craniosynostosis, maxillary hypoplasia, and upper airway obstruction. Given the limited published data in this cohort, we conducted a retrospective review to determine the incidence of perioperative complications in these children.

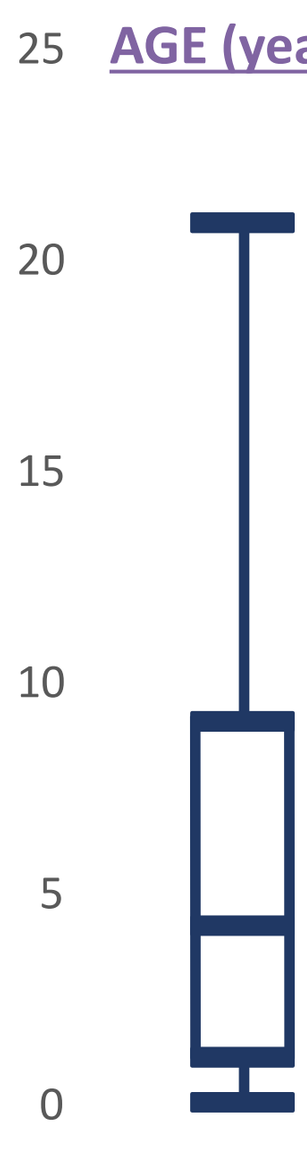
Methods

Following research governance approval, we used the hospital craniofacial database to identify patients with Pfeiffer syndrome. A case-note review was then conducted to collect data on patient demographics, surgical procedures, incidence of perioperative cardiovascular and respiratory complications and ease of obtaining peripheral intravenous access. A total of 25 children and 413 anaesthetic records were analysed.

Results







18 cardiac & respiratory complications (4.4%):

- 1 cardiac arrest – 15wk infant, posterior vault expansion surgery, secondary to haemorrhagic shock (successfully resuscitated).
- 5 episodes of desaturation due to mucous plugging (managed with chest physiotherapy & suctioning)
- 4 instances of dental damage on intubation
- 1 severe bronchospasm (responded to salbutamol)
- 1 laryngospasm leading to hypoxia and bradycardia (resolved with propofol, atropine & PEEP)
- 1 allergic reaction on induction of anaesthesia
- 2 tracheostomy's dislodged in recovery (reinserted)
- 3 re-intubations in recovery:
  - Twice following prolonged apnoea
  - Once due to airway obstruction from facial swelling post fronto-facial advancement.

Difficult IV access in 5% of cases. 9 times peripheral access was not possible (6 had central lines, 3 proceeded without).

Conclusion

From 413 anaesthetics, we recorded 18 cardiovascular and respiratory complications, giving an incidence of 4.4%. This was not higher than the rate previously reported in the overall paediatric population (1), and also comparable to that published in Apert syndrome, a similar craniofacial disorder (2). This demonstrates that in a specialist institution, anaesthesia for children with Pfeiffer syndrome can be conducted safely with a low complication rate.

1) Von Ungern-Stenberg BS, Boda K, Chambers NA et al. Risk assessment for respiratory complications in paediatric anesthesia: a prospective cohort study. Lancet 2010; 378: 773-783
2) Barnett S, Moloney C, Bingham R. Perioperative complications in children with Apert syndrome: a review of 509 anesthetics. Pediatric Anesthesia 2011; 21:72-77.