Prevalence of obstructive sleep apnoea in children with syndromic craniosynostosis and outcome of intervention with adenotonsillectomy or nasopharyngeal airway

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Introduction

Obstructive sleep apnoea (OSA) has a reported prevalence of between 40 to 85% in children with syndromic craniosynostosis (SC)[1]. In an otherwise healthy population, adenotonsillectomy (AT) is the primary treatment although information on effectiveness is limited[2] and in SC patients OSA remains in 40-90% of patients[1]. Another option is the use of a nasopharyngeal airway (NPA). This study aims to assess the prevalence of OSA in our population of SC patients and the efficacy of AT and NPA.

Methods

A retrospective review was conducted of patients (0.13-18.25yrs) diagnosed with SC (Apert, Crouzon, Pfeiffer syndromes) and referred for a diagnostic cardiorespiratory sleep study (SS) at Great Ormond Street Hospital from 2012 to 2015. SS were scored for evidence of OSA and categorised as normal, mild, moderate or severe based on their obstructive apnoea/hypopnoea index (OAHI). In the population undergoing AT or NPA following their diagnostic study, OAHI were compared post-intervention.

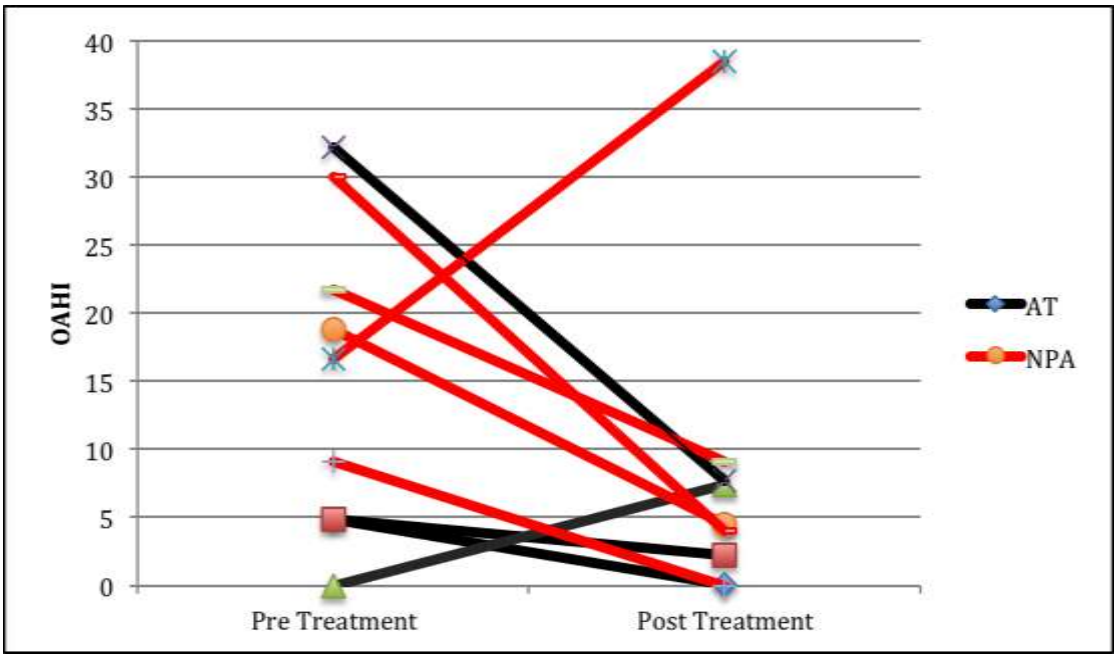
Results

Table 1: Diagnostic cardiorespiratory sleep study results.

	Total	Apert	Crouzon	Pfeiffer
No. of children (%)	30	11 (36.7%)	13 (43.3%)	6 (20%)
Age (median (IQR))	0.92 (0.31-3.78)	0.48 (0.3-6.4)	0.83 (0.44-2.62)	2.49 (0.12-5.12)
Age (range (years))	0.03 – 18.25	0.13 – 8.84	0.17 – 18.25	0.03 – 9.45
OAHI (median (IQR))	0.90 (0-10.2)	5.80 (0.9-15.8)	0.30 (0.0-1.0)	11.15 (0 – 30.6)
OAHI range	0 – 48.4	0 – 18.8	0 – 48.4	0 – 32.2
OSA severity	n (%)	n (%)	n (%)	n (%)
Normal (OAHI<1)	16 (53.3)	3 (27.3)	10 (76.9)	3 (50)
Mild (OAHI 1-5)	4 (13.3)	2 (18.2)	2 (15.4)	0
Moderate (OAHI 5-10)	3 (10)	3 (27.3)	0 (0)	0
Severe (OAHI > 10)	7 (23.3)	3 (27.3)	1 (7.7)	3 (50)

OSA was present in 14(46.6%) patients with higher prevalence in Apert(72.7%) and Pfeiffer(50%) versus Crouzon(23.1%). Following their diagnostic study four underwent AT, resolving obstruction in one on follow-up study. Five had a NPA placed, again resolving obstruction in one (Figure 1).

Figure 1: OAHI Pre and post treatment with either AT or NPA.



Discussion

Consistent with the literature[1], we report a prevalence of OSA in SC greater than the normal paediatric population (OAHI:1.2-5.7)[3]. Despite limited patient numbers undergoing intervention (AT or NPA) in this cohort, results suggest an improvement in OAHI although resolution was rare. Formal statistical analysis was not undertaken due to limited numbers in the intervention groups. This study supports screening for OSA in all SC patients, including multi-disciplinary discussion about the most appropriate treatment modality which is not only limited to AT or NPA but also includes craniofacial surgery and ventilatory support.

References

1. Zandieh. Plast Reconstr Surg.2012Apr;131:847-52.
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