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Upper airway obstruction in the syndromal craniosynostoses

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SUMMARY. **This series of consecutive cases details the prevalence and management of upper airway obstruction in the syndromal craniosynostoses (Crouzon, Apert and Pfeiffer syndromes). Upper airway obstruction presents more frequently in Crouzon and Pfeiffer syndrome when presenting early and during the intermediate years. Those patients with Apert syndrome appear relatively free of this problem.**

Management has been directed toward increasing the size of the nasopharyngeal space by soft tissue alterations (uvulopalatopharyngoplasty, soft palatal split and adenotonsillectomy) with success. Le Fort III advancement osteotomy has been reserved for those more extreme cases, again with objective airway improvement. These techniques have removed the necessity for the progression to tracheostomy in these cases.

Airway obstruction in patients with the syndromal craniosynostoses has only received limited documentation to date,[1,2] whereas the assessment and early intervention for craniosynostosis, hydrocephalus and exorbitism is detailed.

The advent of improved functional airway assessment by oximetry and formal sleep studies, in concert with 3-D imaging of the skeleton and soft tissues of the head and neck, permits objective recording of the potential multi-level airway obstruction in syndromal craniosynostosis. Just as the expression of the craniosynostosis is not static, so airway compromise in these cases is dynamic, increasing or decreasing in severity with the growth changes in both craniofacial skeleton and soft tissues.

Significant shortening of the anterior cranial base, a less obtuse cranial base angle and retruded midface and mandible set a skeletal scene for early (less than 1 year old) upper airway obstruction.[3] Add to this a long, floppy soft palate, normal sequential changes in the adenotonsillar lymphoid ring and widespread cartilaginous abnormalities, and the problem of airway obstruction presents early and magnifies through the intermediate years.

Where management to date has centred on non-operative support (continuous positive airway pressure) or bypassing the presumed level of obstruction with tracheostomy, neither will consistently and safely maintain a secure airway except in the presence of sophisticated medical support. The isolated reports of early Le Fort III osteotomy and midfacial advancement in syndromal craniosynostosis have been similarly inconsistent in terms of the effect on the airway, with risk of not inconsiderable morbidity.[2]

The evolution of an alternate approach to management of the airway in syndromal craniosynostosis is presented based on the experience of 41 cases treated in the last 3.5 years.

Materials and methods

Included are all paediatric new referrals with syndromal craniosynostoses over the last 3.5 years (n = 38), either unoperated (n = 33) or having been treated elsewhere previously (n = 5) and those old unit patients presenting again during their intermediate years for further operative intervention (n = 4). The latter were among those seen in the unit prior to 1989 for their primary cranial surgery.

All patients underwent a detailed multidisciplinary assessment by members of the craniofacial unit.

Table 1 Airway management in syndromal craniosynostosis—less than 1 year of age 1989–1992

	Palatal Surgery	*Adeno-tonsillectomy*	*Le Fort III*	*Tracheostomy*	*Number of patients requiring surgery*
Crouzon (n = 4)	3	2	—		3
Apert (n = 14)	2	—	—	—	2
Pfeiffer (n = 7)	3	1	1	2	5

Special attention was accorded to a history or signs of airway obstruction, apnoeic episodes, daytime fatigue, failure to thrive, persistent or recurrent upper or lower airway infection, impaired mental capacity and permanent brain damage.

Preoperative investigation by overnight monitored pulse oximetry, or formal overnight sleep studies was performed where the history and clinical findings suggested airway obstruction. In a number of cases presenting in extremis, surgical treatment of the upper airway was undertaken as an emergency before formal quantitative airway studies could be preformed. Post operatively, assessment of the airway by respiratory physicians, together with sleep oximetry was performed. The interval between surgery and these sleep studies varied, with several patients having their follow-up quantitative assessments at their referring institution. The reporting and presentation of the airway studies varied with the institution and the individual respiratory physicians. All cases deemed to have an abnormal preoperative study were so judged by the respiratory physician.

The approach to management of documented airway obstruction evolved during the study period. Stimulated by the death of a child with Pfeiffer syndrome who is included in this series as a consequence of tracheostomy-related complications, a variety of surgical options, both soft tissue and skeletal, have been employed either alone or in combination.

Uvulopalatopharyngoplasty (UPPP)

As previously reported for obstructive sleep apnoea and snoring, this technique involves resection of the soft palate behind the levator palati and a portion of the lateral pharyngeal wall.[4]

Soft palatal split

In those cases where the palate is intact a soft palate cleft has been created by a midline split. This has been performed in isolation and in concert with resection of the posterior soft palate.

Adenotonsillectomy

Where there is a significant contribution to upper airway obstruction from these structures either early or in the intermediate years, adenoidectomy and/or tonsillectomy has been performed.

Le Fort III advancement osteotomy

Le Fort III osteotomy and midface advancement has been routine during the intermediate years in the syndromal craniosynostoses for correction of midfacial deformity, inferior support of the globes, occlusal improvement and social or psychological reasons. The effect of such surgery on upper airway obstruction has been controversial.

In this series Le Fort III advancement osteotomy was performed both during the intermediate period, and early in conjunction with fronto-orbital advancement. The latter fronto-facial advancement was reserved for those cases with extreme life-threatening upper airway obstruction. Soft palatal surgery and adenoidectomy was also added in these cases where airway compromise was critical.

Tracheostomy

Tracheostomy in an early case was followed by infective complications and death on return to his home in the country. Subsequently this procedure has been avoided in all cases referred to this unit.

Continuous positive airway pressure (CPAP) has not been available to this young population and has accordingly never been used.

In a number of cases soft tissue manipulations of the airways (UPPP, soft palatal split etc) were used initially in an attempt to defer or delay midfacial skeletal surgery.

Results

Of the 38 new cases seen in this unit 1989–92 with craniosynostosis syndromes, the numbers by diagnosis are as follows: Crouzon's (n = 11), Apert (n = 19) and Pfeiffer (n = 8). Most presented early (less than 1 year) having had no previous operative intervention of any type (neurosurgical, cranio-orbital or airway) elsewhere. A smaller group presented after 1 year of age, due to delayed, or failure of, diagnosis. Some of these had undergone prior neurosurgical or cranio-orbital surgery by other surgeons. Another four older patients, who had had prior cranio-orbital surgery in this unit during the early period, presented again during their intermediate years and were candidates for midfacial advancement on the basis of the reasons listed previously.

All patients presenting early underwent a conventional fronto-orbital advancement and where lambdoid synostosis was evident, a lambdoid craniectomy.

Early (less than 1 year)

Only four new early cases of Crouzon syndrome were seen. Three patients manifested quantifiable airway obstructive symptoms, with one having a tracheostomy placed prior to coming under our management. Both this case and another had soft palatal splitting with resolution of airway symptoms (Tables 1 and 2).

Of the 14 new cases of Apert syndrome only two presented with objective evidence of airway obstruction and in both, this was corrected by splitting the soft palate in concert with resection of the posterior soft palate margin.

Airway obstructive symptoms were seen in five of

Table 2 Airway assessment and response after surgery—less than 1 year of age 1989–1992

	Initial presentation	*Surgery*	*Follow-up*
Crouzon			
S.H.	Frequent desaturation to less than 40%	Palatal surgery and adenoidectomy	No significant desaturation to less than 90%
A.T.	In extremis	Tracheostomy Palatal surgery later	No desaturations to less than 90%
E.K.	In extremis	Palatal surgery and adenotonsillectomy	No desaturations to less than 90%
Apert			
C.B.	Frequent desaturations to less than 90%	Palatal surgery	No desaturations to less than 90%
A.H.	Frequent desaturations to less than 90%	Palatal surgery	Desaturations to less than 90% for 1% of the time (OA/OH index 0/hour)
Pfeiffer			
G.B.	In extremis	Tracheostomy	No desaturations to less than 90%
A.F.	Frequent desaturation to less than 90%	Tracheostomy	No desaturations to less than 90%
R.A.	Frequent desaturation to less than 50%	Le Fort III Palatal surgery	No desaturations to less than 90%
J.V.	Frequent desaturation to less than 50%	Palatal surgery and adenoidectomy	No desaturations to less than 90%

(OA/OH index = apnoea/hypopnoea index, records the number of apnoeas or hypopnoeas of greater than 10 seconds duration in each hour of observed sleep)

the seven new cases of Pfeiffer syndrome (Fig. 1). Two of these cases, presenting early in the evolution of the treatment approach, underwent tracheostomy with one of them subsequently dying of tracheostomy-related complications. The other underwent Le Fort III advancement later at age 3.5 years, permitting removal of the tracheostomy (Fig. 2). An early primary frontofacial advancement, UPPP and soft palate split was required in one case, producing resolution of extreme upper airway obstruction. A further patient responded to UPPP, soft palate split and adenoidectomy whilst one patient with mild symptoms resolved with growth.

Intermediate (1–10 years)

Airway obstruction was seen in only three of the nine Crouzon syndrome patients treated during this period. Seven of these cases were new referrals, the remaining two having undergone prior cranio-orbital surgery in this unit (Table 2). One new referral, without any previous treatment, underwent adenoidectomy and fronto-facial advancement at age 5 years with resolution of airway symptoms. The other case was of a patient with cloverleaf skull anomaly who presented with sleep apnoea at age 6 years. A UPPP relieved these symptoms and permitted delay of his inevitable Le Fort III midfacial advancement.

In the Apert syndrome group, eight patients were seen in this age range, with five of these being new referrals. Three patients previously managed in this unit demonstrated airway obstructive symptoms. Tonsillectomy at age 2 years followed by fronto-facial advancement at 4.5 years corrected the symptoms in one, whilst soft tissue surgery sufficed in the others.

Only one new case of Pfeiffer syndrome presented during the study period. This patient with cloverleaf skull anomaly had had previous cranial vault surgery and an adenotonsillectomy elsewhere. Severe symptoms of airway obstruction were treated initially by UPPP without improvement. Subsequent Le Fort III advancement osteotomy produced demonstrated correction of the airway obstruction.

The Pfeiffer patient has been detailed above—with decannulation of his tracheostomy after Le Fort III midface advancement.

Discussion

The syndromal craniosynostoses manifest temporally-dependent abnormalities of the cranium, orbit and face. Where the cranio-orbital anomalies are immediately visually striking, the clinical expression of faciostenosis is generally perceived as the later presenting midfacial retrusion. This series suggests otherwise. The three-dimensional midfacial hypoplasia of faciostenosis presents early as upper airway obstruction, the management of which requires early recognition and treatment to avert the attendant cardio-respiratory and neurological sequelae.

In Crouzon, Apert and Pfeiffer syndrome the skeletal basis exists for the development of early and persistent upper airway obstruction.

Peterson-Falzone *et al.*[5] employed cephalometric radiography to demonstrate the abnormal naso-

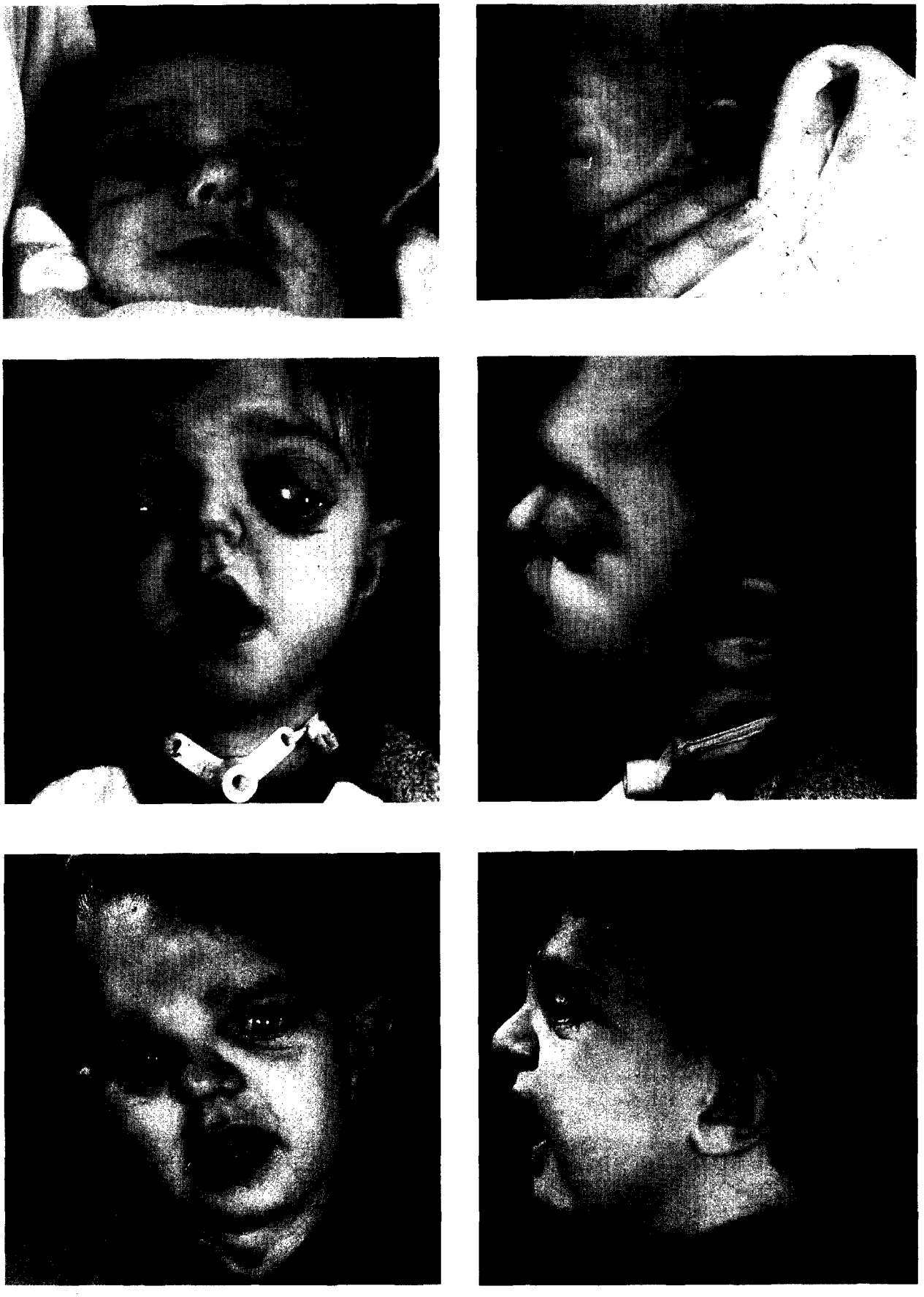

Fig. 1

Figure 1—(A) Male Pfeiffer syndrome infant presenting early prior to cranio-orbital surgery. (B) Presentation at $3\frac{1}{2}$ years prior to midfacial surgery, with tracheostomy in place. (C) Appearance following fronto-facial advancement with resolution of airway obstruction and removal of tracheostomy.

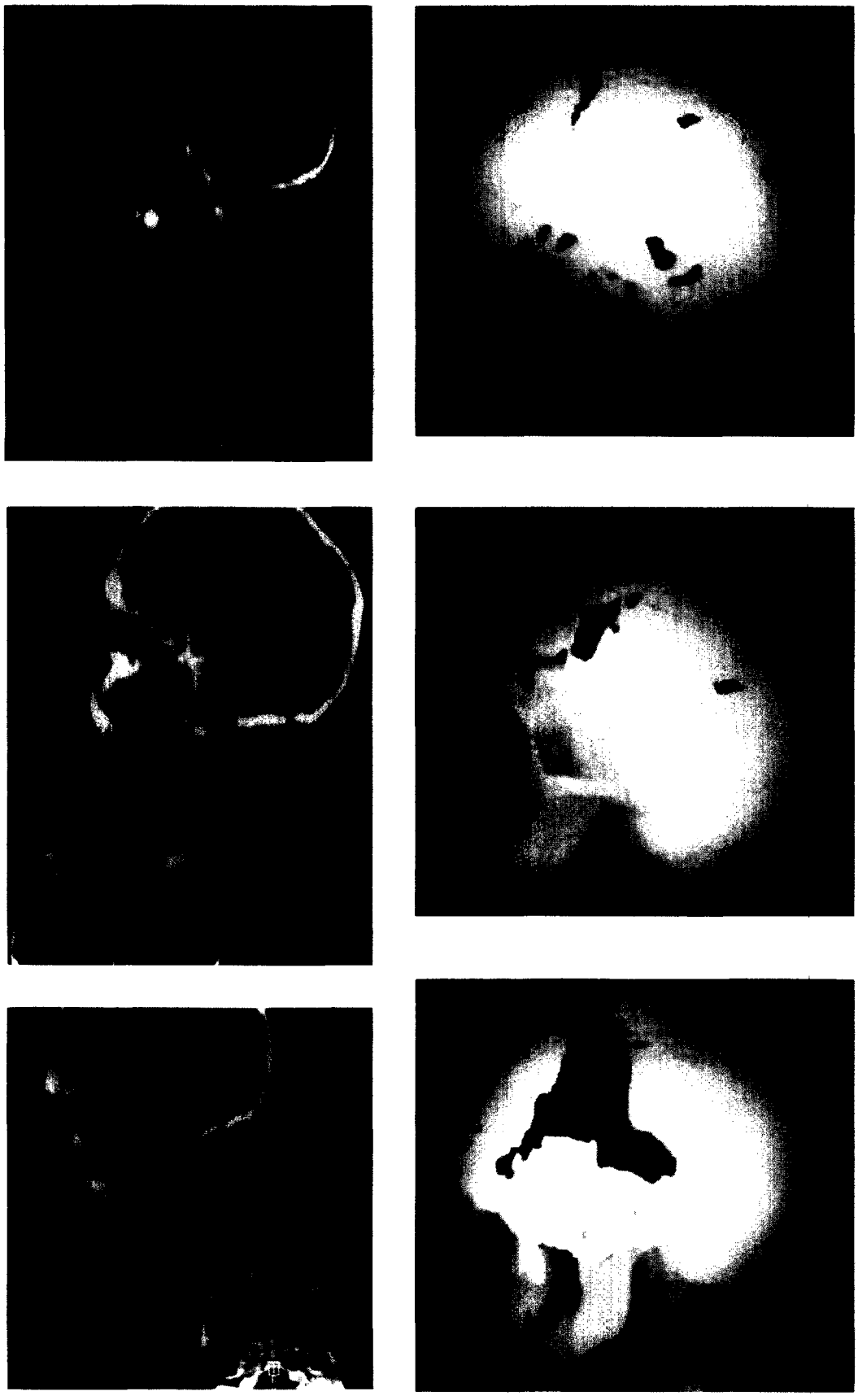

Fig. 2

Figure 2 Sagittal 2-D CT reformats and lateral 3-D CT reconstructions of the above patient show: (A) At early presentation, extreme obstruction of nasopharyngeal airway. (B) At $3\frac{1}{2}$ years persistent airway obstruction and midfacial retrusion. (C) Following fronto-facial advancement, dramatic increase in nasopharyngeal free airway.

Table 3 Airway management in syndromal craniosynostosis—1–10 years of age 1989–1992

	Palatal Surgery	*Adeno-tonsillectomy*	*Le Fort III*	*Tracheostomy*	*Number of patients requiring surgery*
Crouzon (n = 9)	1	1	2	—	3
Apert (n = 8)	2	2	1	—	3
Pfeiffer (n = 2)	1	1	2	—	2

Table 4 Airway assessment and response after surgery—1–10 years of age 1989–1992

	Initial presentation	*Surgery*	*Follow-up*
Crouzon			
B.P.	Desaturation to less than 90% for 78% of the time	Palatal surgery	Desaturations to less than 90% for 0.3% of the time (OA/OH index 2.4/hour)
A.K.	Desaturations to less than 90%	Le Fort III Tonsillectomy	No desaturations to less than 90%
A.A.	Frequent desaturations to less than 90%	Le Fort III	No desaturations to less than 90%
Apert			
S.D.	Frequent short apnoeas and hypopnoeas	Palatal surgery and adenotonsillectomy	No apnoeas or hypopnoeas
D.W.	Desaturations to less than 94% (OA/OH index 6.2/hour)	Le Fort III	No desaturations to less than 94% (OA/OH index 0/hour)
E.T.	Frequent desaturations to less than 90%	Palatal surgery and adenotonsillectomy	No desaturations to less than 90%
Pfeiffer			
J.B.	Desaturations to less than 90% for 17% of the time	Palatal surgery Le Fort III	No desaturations to less than 90%
A.F.	Desaturation to less than 90% for 40% of the time with tracheostomy excluded (OA/OH index 26.5/hour)	Le Fort III Removal tracheostomy	No desaturations to less than 90% (OA/OH index 0/hour)

(OA/OH index = apnoea/hypopnoea index records the number of apnoeas or hypopnoeas of greater than 10 seconds duration in each hour of observed sleep)

pharyngeal morphology in both Apert and Crouzon syndrome. A foreshortening of both the anterior and posterior cranial base, reduction in pharyngeal depth, height and width and reduction in height and width of the posterior choanae is present early and progresses with time. A long flap-like velum, and mandibular body which is short in length add to the pharyngeal crowding in three dimensions and set the scene for early and clinically significant physiological disturbance manifesting as upper airway obstruction. The limited reports on Pfeiffer syndrome document a similar trend to nasopharyngeal morphologic disturbance.

A single-minded focus on this sub-region as the cause of airway obstruction in these complex craniosynostosis syndromes is too simplistic, as there is input to airway maintenance, both structural and functional, from the cerebral cortex above to the terminal alveoli below. The naso- and oro-pharynx is, however, one area where morphological alterations can be safely made in an attempt to rectify airway obstruction.

Clinically significant airway obstruction, on history and formal, quantitative airway assessment is not uniformly prevalent across all the major craniosynostosis syndromes, but rather more frequently seen in Crouzon and Pfeiffer syndromes, with relative sparing of Apert syndrome. This is evident both in those who present early (less than 1 year) and also where diagnosis and referral is delayed.

The basic morphological data for Pfeiffer syndrome

are sparse, but in most craniofacial features these patients mimic the Crouzon syndrome individuals. In Crouzon syndrome the cranial base angle is decreased (basilar kyphosis) in most cases, with an associated, significantly smaller mandibular body length. These features, in combination with the three dimensional constriction of the nasopharynx, constitute an ideal situation for the production of upper airway obstruction. The apparently increased incidence in Pfeiffer syndrome in this series may be accounted for by the fact that four of the seven cases in this series had features of the cloverleaf skull anomaly and hence were at the more severe end of the spectrum.[6]

Apert syndrome has significantly fewer upper airway problems in this series. Morphologically, most Apert syndrome cases show cranial base angles which are close to normal, and may even be increased (platybasia), and mandibular body lengths which are more normal than those seen in Crouzon syndrome. The nasopharyngeal airway in these cases is therefore less constricted and more likely to accommodate normal upper airway function.

The approach to management of the upper airway in the syndromal craniosynostoses has to date been ad hoc and its reporting anecdotal. Lauritzen *et al.*[1] in a small series of patients of mixed ages and diagnoses, suggested that tracheostomy was the only consistent means by which to achieve airway control, and that tonsillectomy or Le Fort III midfacial advancement were ineffective.

More recently Mixter *et al.*,[2] reviewing a series of Apert and Pfeiffer syndrome patients of all ages, concluded that surgical alteration of upper airway anatomy by dilatation of choanal stenosis and Le Fort III midface advancement was ineffective. Conservative management by postural positioning, with procession to tracheostomy where necessary in children, and continuous positive airway pressure (CPAP) or tracheostomy in adults was advocated. In our series, by closely documenting and recording the airway status at all ages, it has been possible successfully to alter the nasopharyngeal space by surgical means to produce clinically significant improvement in airway function, without resorting to tracheostomy. The ideal must be to tailor the variety of the surgical approaches to the temporally changing morphology and function of the nasopharyngeal airway in these syndromal craniosynostosis cases.

Early presentation with airway obstructive symptoms within the first 6–12 months may be managed best by UPPP with or without adenoidectomy, as this has few potential adverse effects. Where this is inadequate, progression to soft palatal splitting is logical, albeit with some potential for increased middle ear effusions and later speech disorder and velopharyngeal incompetence. This latter potential complication may be more theoretical than real in light of the usual problem of the obstructed nasal airway and hyponasal speech in these patients. Later repair of these soft palate "pseudoclefts" is envisaged once the airway is safely maintained, even if this is at the completion of all other facial skeletal surgery. Such a palate repair, in association with pharyngoplasty as necessary, would provide few difficulties as there is no true cleft hypoplasia. No increased problem with middle ear disease has been noted to date, but this warrants close follow-up and investigation.

The volume of adenotonsillar tissue during this period is frequently small and unlikely to contribute significantly to airway obstruction. The technical difficulty of performing an adenotonsillectomy in these young children, where the mandible is small and retropositioned, is quite considerable.

Where primary presentation is delayed beyond one year a similar approach is logical. In the presence of severe airway obstruction resistant to these soft tissue techniques, progression to early Le Fort III midfacial advancement is possible with a degree of safety and stability, producing a further increment in nasopharyngeal airway.

Later, between three and five years of age, adenoid and tonsillar growth may further compromise the nasopharyngeal airway. Symptomatic presentation during this time requires initial consideration for adenotonsillectomy. This may be performed alone, in combination with, or followed by surgery to the soft palate. Both these manoeuvres may help to delay the requirement for midfacial advancement, as in one of our Crouzon patients. Failure to improve then demands progression to early Le Fort III advancement osteotomy, with the understanding that this may need to be repeated later for facial form and occlusion.

Late presentation with upper airway obstruction and sleep apnoea in the teenage and adult patient demands a similar approach, keeping in mind their potential requirement for combinations of Le Fort I and Le Fort III midfacial osteotomy for correction of occlusal and facial aesthetic disturbances.

Employing these approaches it has been possible to overcome the faciostenosis, and to increase the calibre of the upper airway as documented symptomatically and on quantitative testing, without the need for significantly altering the airway at another level by tracheostomy.

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