International Congress Series 1254 (2003) 119–121

Multidisciplinary management of children with craniofacial syndromes with particular reference to the airway

H. Nishikawa, K. Pearman*, S. Dover

Children's Hospital Birmingham, Steelhouse Lane, Birmingham B4 6NH, UK

Keywords: Craniofacial syndromes; Airway problems; Tracheostomy

This paper concerns the affect that craniofacial syndromes can have on the upper airways. A brief definition and classification of craniofacial anomalies is presented and the management of airway problems that they can cause is discussed.

Pathology or dysplasia (abnormal development), which affects the brain, cranium and facial skeleton results in craniofacial deformities. Van der Meulen has classified craniofacial anomalies. The most important groups are the craniofacial dysplasias and craniofacial dysplasias of other origin. These dysplasias comprise conditions such as clefts, craniosynostosis (premature fusion of cranial growth plates), dysostoses (abnormal facial bone development) or combinations of these conditions. Craniofacial syndromes such as Apert, Crouzon and Pfeifers come into this category. Vascular anomalies and neurofibromatosis affecting the craniofacial skeleton and soft tissues can also have profound affect upon the airways.

The otolaryngologist's perspective (K. Pearman, FRCS).

Children with craniofacial syndromes present numerous problems requiring input from many disciplines. The principal role of the ENT surgeon is in airway management although otological care must not be overlooked. An analysis was made of the case records of a group of 22 children with Apert's, Crouzon's and Pfeiffer's syndromes treated in our unit.

The structural abnormalities of the upper airway in these children include a hypoplastic maxilla, which is retruded with a short, high arched hard palate leading to a small nasal cavity and nasopharynx often with septal deviation and choanal atresia or stenosis.

* Corresponding author. Tel.: +44-1213-338111.

E-mail addresses: hiroshi.nishikawa@bhamchildrens.wmids.nhs.uk (H. Nishikawa), k@pearman.demon.co.uk (K. Pearman), Stephen@msdover.freeserve.co.uk (S. Dover).

doi:10.1016/S0531-5131(03)01117-8

The soft palate may be abnormal in position hanging almost vertically in the oropharynx and obstructing it. It may also be long and thick or cleft and can be swollen by deposits of mucopolysaccharides.

A hypoplastic mandible further reduces the volume of the oropharynx and mouth in some cases. Adenotonsillar hypertrophy may further obstruct the oropharynx as the child grows.

Various congenital laryngeal abnormalities are described in craniofacial syndromes of which subglottic stenosis is the commonest. The sleeved trachea in which normal tracheal rings are replaced by what is effectively a cartilaginous tube is an interesting finding at tracheostomy but has not caused any functional problems in our group of patients.

The variety of potential causes of airway obstruction in these patients means that the clinical features of upper airway obstruction can be complex and may vary over time. What is primarily nasal in the newborn period may become primarily oropharyngeal as the child gets older, anatomical relationships change, tonsils grow and soft palates thicken. It is therefore important to be vigilant for the late development of obstructive symptoms, particularly sleep-obstructed breathing. The significance of this can easily be overlooked by professionals and carers against the background of the multiplicity of problems which these children face. There is an interaction between obstructive sleep apnoea and the raised intracranial pressure, which these children suffer as a result of craniosynostosis. Both conditions can cause failure to thrive, somnolence, vomiting and behavioural problems; carbon dioxide retention may cause raised intracranial pressure and raised intracranial pressure may affect central control of respiration.

The effects of upper respiratory infections and gastroesophageal reflux may be particularly severe in children with marginal airways.

Other functional problems include swallowing and speech difficulties due to the combined effects of the airway, oral anatomy and, in some cases, pharyngeal dystonia. Hearing loss, due to aural atresia, ossicular abnormalities and effusions may also affect speech development and the tendency to underestimate the potential of these children must be resisted.

Investigations of the airway include imaging and polysomnography. While the latter is essential both diagnostically and in assessing the effects of treatment, it does not necessarily give all the information required about the airway and clinical assessment remains vital.

Supportive management includes oxygen therapy, monitoring and hospitalisation during respiratory infections. Of 22 patients in this analysis, 19 required more than supportive airway management.

Our experience of CPAP is relatively limited with only two patients on long-term therapy. Difficulties with this modality of treatment include problems with use of the equipment at home and during respiratory infections and the potential for the mask to damage prominent eyes.

Nasal stenting has a place in the management of some children. In the early months, in particular, the nasal component of airway obstruction may predominate and its treatment may buy time while other aspects of the care are dealt with. Six of our patients were stented for choanal atresia and stenosis all with initial benefit. Only two required no further intervention. Three requiring tracheostomy at some stage, and one an adenotonsillectomy.

When the obstruction is primarily oropharyngeal, and sleep-obstructed breathing is the primary problem, parents can sometimes manage their children with nightly nasopharyngeal intubation. If a tube is needed during the daytime, however, it can interfere with feeding and is not an effective long-term management. In our four cases, only one used an NPA for a period of months. Long-term management was by CPAP in two cases and by tracheostomy in one; one case required an NPA for a short time for supportive therapy after tonsillectomy.

Adenotonsillectomy was performed for obstructive sleep apnoea in nine cases. Results were good in six; two subsequently required tracheostomy, one temporary, and one long-term. Adenotonsillar surgery should be regarded as helpful rather than curative, reducing symptomatology to a level where the airway is safe and more aggressive intervention can be avoided. There was one postoperative death, probably airway-related, in our series underlining the need for intensive postoperative monitoring in children with complex airway problems.

Our unit has no experience of uvulopalatopharyngoplasty in these cases. While some successful cases have been reported in the literature, the complex nature of the anatomical problems in these cases makes it difficult to guarantee success beforehand and there is a theoretical risk of producing velopharyngeal incompetence when midface advancement is performed later.

Tracheostomy remains the bottom line in airway intervention in these cases. Ten of our patients had a tracheostomy at some stage. Five have been decannulated after interventions of various kinds, three remain tracheostomy-dependent and two died of problems unrelated to the airway. It may be that we should have used tracheostomy earlier in some of our patients. Tracheostomy is not without its problems however and it adds an extra burden to parents who may already be overwhelmed by considerations of their child's deformity, intracranial pressure, feeding, eyes, etc. Our policy has therefore been to manage airways as conservatively as possible consistent with safety.

The airway effects of maxillary and mandibular advancement will be discussed separately.

Many disciplines can be involved in the management of these patients including neurosurgery, plastic surgery, maxillofacial surgery, otolaryngology, hand surgery, ophthalmology, dental surgery, paediatrics, anaesthesia, intensive care, respiratory medicine, speech and language therapy, psychology and audiology. Not every patient will need input from all of these and detailed discussion is beyond the scope of this paper. In airway management, it is essential to have the backup of expert anaesthesia and intensive care if cases are to be managed safely. When multiple systems are affected, it is easy to overlook developing problems such as sleep apnoea and to underestimate a child's potential because remediable problems such as glue ear and speech problems are given inadequate attention. The existence of multidisciplinary teams helps to minimise these difficulties by promoting the assessment of the whole child.