Orthodontic management of achondroplasia in South Africa

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To the Editor: Achondroplasia is a relatively common genetic skeletal dysplasia that manifests with stunted stature and disproportionate limb shortening. Characteristic craniofacial features include a prominent forehead, depressed nasal bridge and maxillary hypoplasia.

It is probable that there are between 500 and 1 000 persons with achondroplasia in South Africa, and it is inevitable that they will seek consultation and care in general and specialised dental practices. In this context, it is relevant that dental and orthodontic management is constrained by practical problems associated with upper airway obstruction and other primary and secondary syndromic components.

In order to provide a perspective on the situation in South Africa, we assessed the special oro-dental needs of 10 affected children. Our findings are presented here.

Patients and methods

Ten children with a firm diagnosis of achondroplasia were documented at the Genetic Clinic of Red Cross Children's Hospital, Cape Town, in 2003. After preliminary appraisal, they were seen in the Faculty of Dentistry, University of the Western Cape, for detailed oro-dental assessment and the implementation of ongoing specialised dental management. Investigations included Panorex views of the jaws and lateral skull radiographs for anthropometric analysis.

General observations

Apart from small stature and short limbs, the affected children had good general health, normal intelligence and buoyant personalities. There were no internal ramifications, no bleeding tendency and no morbid obesity. Several had leg malalignment but retained their agility. The majority had large heads, with possible arrested hydrocephalus, and in 2, shunts had been implanted.

The ambulant children had a mild lumbar lordosis but none had any significant spinal malalignment other than a thoraco-lumbar gibbus in one child. None had any evidence of spinal cord compression, although several had chronic or intermittent backache.

Oro-facial observations

The affected children all breathed by mouth because of upper-airway obstruction. The teeth were normal in shape, texture and mechanical properties, and there was no impaction or malalignment. The mouth was wide, and the upper dentition prominent. Wide spacing of the teeth, which is often prominent in adulthood, was not seen in the children.

Problems in oro-facial management

Short limbs, stunted stature and occasional spinal malalignment made it difficult for the children to sit comfortably in a conventional dental chair. Chronic backache may compound this problem.

Upper airway obstruction due to depression of the nasal bridge, variable choanal atresia and enlarged adenoids resulted in mouth breathing. A small nasal pharynx and larynx posed additional problems for anaesthesia.

The possible presence of cranio-cervical instability necessitated special precautions in head control during dental intervention.

A large heavy head, sometimes with arrested hydrocephalus and an implanted shunt, made head control difficult.

Discussion

According to Dunbar *et al*.[1] the main orthodontic problem in achondroplasia is class III malocclusion consequent upon inherent shortening of the base of the skull; this abnormal configuration leads to retraction and decrease in vertical height of the maxilla.

Although the clinical features and natural history of achondroplasia have been documented extensively, there is a paucity of information on the dental manifestations. For instance, in a multicentre review of 193 affected persons, only brief mention was made of potential dental problems.[2] However, it was recorded that only 3% of the children in this series were regarded as having significant orthodontic needs.

In the light of our experience in the orthodontic management of 10 affected children, the main problems were related to the practical aspects of dental care, rather than any specific abnormalities of the teeth and jaws. These difficulties concerned mouth breathing because of the reduced calibre of the upper airways. In this context, 8 of the 10 children had

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experienced repeated upper respiratory tract infections, with recurrent otitis media and enlargement of the adenoids.

The large heavy head, the small foramen magnum and the possible abnormality of the cranio-cervical junction pose a potential threat of spinal cord compression.[3,4] It is likely that this special risk applies to dental procedures, with or without general anaesthesia, and careful control of the patient's head is warranted. In South Africa, the presence of a thoraco-lumbar gibbus in black children represents an additional potential threat to the spinal cord.[5]

Cephalometric analysis of the craniofacial skeleton was undertaken in 5 of the affected children in order to identify any specific features that might reflect ethnic diversity and impact on dental management in the South African situation. Our findings were essentially in accordance with those previously documented in a thesis by Pedersen,[6] and by Cohen *et al*.[7] in an extensive series of affected persons. In particular, maxillary hypoplasia and relative mandibular prognathism were consistent features.

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