Cleidocranial Dysostosis: A Case Report

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Introduction

Though a definition of cleidocranial dysostosis can be found in orthodontic literature, to the best of the author's knowledge a report of an orthodontically treated case is not available.

In the treatment of this case, the accomplishment of the attained result is so dramatic and the improvement so marked and noticeable that here is another opportunity for testing the validity of the implications and inferences in connection with the relationship of the development and growth of the jaws to/and the development and growth of the teeth and the influence of orthodontic therapy on this relationship individually and collectively.

Cleidocranial dysostosis,[1] in addition to the anatomic anomalies of the clavicle and possible delayed closure of the cranial sutures, presents for orthodontic consideration the distinctive occlusion associated with this condition, particularly manifested in the morphology and growth of the jaws as well as the development of the dentition.

Salzmann[2] describes cleidocranial dysostosis as a "disease of bone growth which occurs with different degrees of intensity. The most commonly encountered defects have been defective or missing clavicles, delayed eruption of the teeth, retarded development of the bones of the jaws and face, and extreme enlargement of the lateral dimensions of the skull. The deciduous teeth erupt late and may be retained in the jaws throughout the entire childhood, the adolescent period, and sometimes even into the third or fourth decades of life. The permanent teeth develop within the jaws, but are impeded in their eruptive progress. There may be many supernumerary tooth follicles formed, with the resultant development of many supernumerary teeth and tooth-like bodies, especially in the region of the maxillary central incisors or the mandibular premolar segments."

History

Inspection of the photograph in Figure 1 reveals a white male, age 15 years, with a dentofacial deformity resembling the skeletal pattern sometimes seen in an edentulous individual at rest. There are prominent frontal bosses and slight supraorbital prominences. The premaxillary area is reduced in size, the zygomas flattened, and the upper lip in a retruded position. According to the parents the patient, at age twelve, had some supernumerary teeth lingual to the upper incisors removed, as well as all remaining deciduous teeth.

In the intraoral photograph of May 1962 (Fig. 2), the erupted teeth can be seen. These teeth are two maxillary central incisors, upper left first molar and upper right first molar. In the mandibular denture, the teeth present

Fig. 1

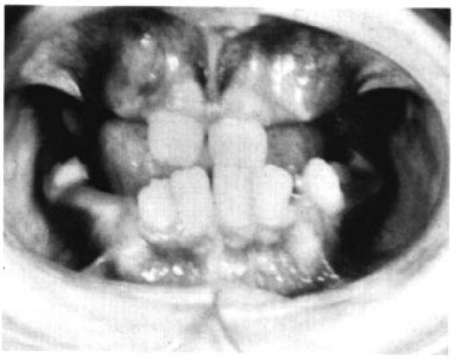

Fig. 2

are the four lower anterior teeth, lower right first molar, lower left first and second premolars and first molar. At this time the remaining teeth were surgically exposed.

The models of the case (Fig. 3) four months after exposure of the teeth illustrate: 1) Class III skeletal and dental relationship, 2) lack of growth of maxilla, 3) lack of dental development in both jaws, particularly in the maxilla, 4) crowded and blocked-out teeth throughout both arches as well as transposition of upper right lateral incisor and canine, 5) the lack of alveolar bone deposition at the alveolar crests, and 6) the lack of occlusal contact of opposing teeth.

Figure 4 is a tracing for use of the Riedel analysis. Attention is directed

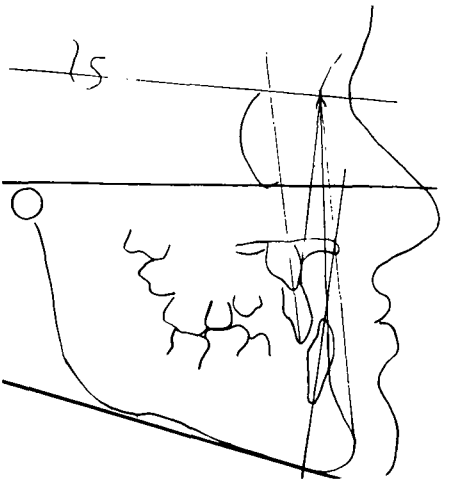

Fig. 4

to the unusual position of sella relative to the Frankfort plane and nasion. This unusual relationship will affect the metric values compared to Riedel's means and standard deviations, but will not affect the comparative biostatistical evidence revealed by records taken on the patient before and after treatment in assessing both the skeletal and dental status. The cephalometric appraisal before and following treatment provides a graphic description of the skeletal and dental deformity, their mutual relationships and the changes resulting from treatment.

Figures 5, 6 and 7 are of records

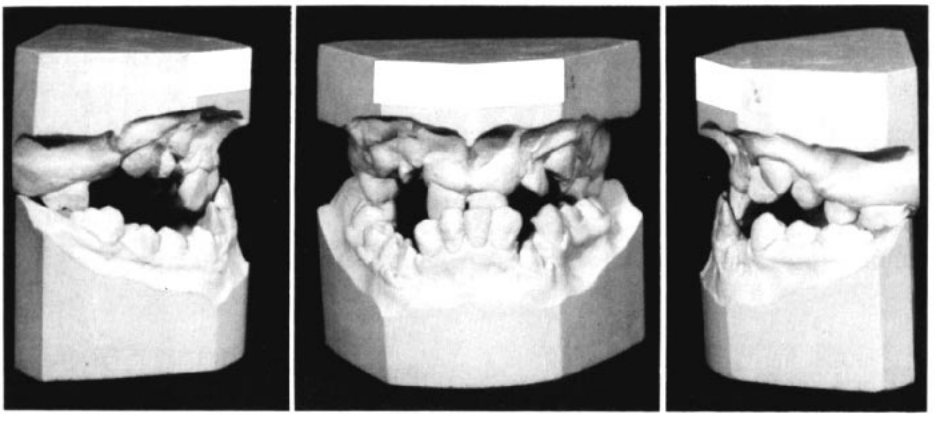

Fig. 3

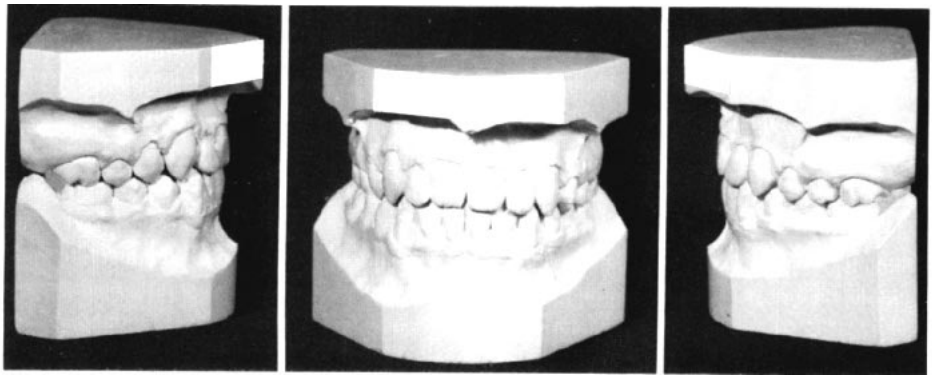

Fig. 5

Fig. 6

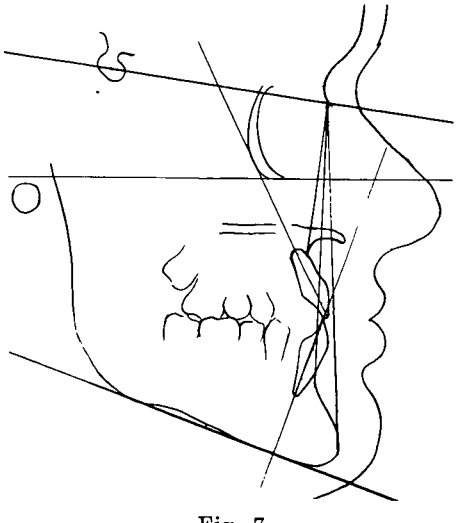

Fig. 7

made March, 1965 upon completion of treatment. Observe the correction of the maxillary right lateral and canine transposition.

Skeletal Changes

S-N-A, the angular measurement indicating the relative anteroposterior position of the maxilla at its apical base to the cranial base before treatment is 89 degrees, a difference of 7 greater than the mean. The after treatment value is 91. In view of the radical dental changes the difference of 2 degrees is insignificant.

S-N-B, the angular measurement, before treatment, of the mandibular base in its anteroposterior cranial base relationship is 97, 17 degrees greater than the mean. The after treatment measurement is 96, no change.

The angular difference between S-N-A and S-N-B, which is normally about —2, shows a significant difference between the maxillary and mandibular apical bases of —8 degrees before treatment as compared to 5 after.

S-N-P, the relative position of pogonion to the cranial base is 101 degrees and is not altered.

S-N-GoGn, the relationship of the mandibular plane to the cranial base is 12 degrees, a figure indicative of good ramal height and condylar growth. The

relationship was the same following treatment; it is this skeletal growth pattern that made possible the attainable optimum in treatment.

Denture Analysis

S-N$\underline{1}$, axial inclination of upper central incisor relative to sella-nasion is 102 degrees, well within the mean. Following treatment this inclination was increased to 124, or 22 beyond the mean.

GoGn$\bar{1}$, lower incisor to mandibular plane reading 82 degrees, well below the mean of 92 before treatment, was increased to 90 after treatment.

N-P$\underline{1}$, the horizontal linear measurement from incisal edge of the maxillary incisor to the facial plane, is —13 mm, compared with a mean of +5.51 mm for the standard, a tremendous difference of 18 mm. Following treatment the linear measurement was —2 mm.

N-P-$\bar{1}$, the anteroposterior relation of the lower incisor to the facial plane, is —5 mm; its usual position is a few mm anterior to the facial plane. The linear measurement following treatment was —3 mm.

The entire face is posterior to the normal limits, the upper face and its dentition to an extreme degree, and the lower face and dentition slightly so.

Following treatment, in spite of the tremendous amount of orthodontic therapy and tooth movement in both dental arches, no change, relatively speaking, other than that attributable to experimental error was found in the skeletal measurements.

The significant changes which took place were purely dental. The inclination of the upper central incisor actually increased 22 degrees, the inclination of the lower incisor increased 8, the linear relationship of the incisal edge of the upper central incisor to the facial plane increased 11 mm, and the relationship of the incisal edge of the lower incisor to the facial plane increased by 2 mm.

Treatment objectives as related to the dentition have been attained, but apparently little has been changed in the skeletal pattern.

Discussion

A discussion and evaluation of the cephalometric analysis obviously should include attention to the unique case history.

Particular interest focuses attention on the genetic and morphologic characteristics of this malocclusion. Genetically it appears unique insofar as the family history indicates. Morphologically, as mentioned above, the unusual anatomic position of sella makes the comparison of angular measurements before treatment with the sample mean of little value. Much is to be gained from both a gross and statistical comparative evaluation of all the records taken before and after treatment.

Conclusion

The attainable optimum was achieved as evidenced by the articulated dentition and functional result. Gross examination of the records prior to treatment readily reveals that the process of eruption of the teeth and associated bone formation at the alveolar crests had not occurred concomitantly; occlusal contact of normally erupting antagonizing teeth was lacking. Nevertheless and fortunately for this young man, the mandible had grown in all directions with commensurate increase in height of the ascending ramus, thus space had developed to accommodate the growth of the maxilla as well as the growth and eruptive development of the teeth and permitted the development of the facial pattern and the attainable optimum in treatment. This is additional evidence that the height of total face is to a large extent determined

by the developmental growth of the mandible and that no absolutely fixed relationship exists between the growth of the jaws and the growth and eruption of the teeth, and that eruption of the teeth and growth of the jaws are largely independent.[3]

This separation of the jaw into two morphologic components is, as Allen[4] emphasized, an important one to remember for the following reasons:

(1) The alveolar process is developed pari passu with the teeth.

(2) The process is an outgrowth from the jaws for a specific purpose. John Hunter (1728-1793) declared that the "Alveolar process of both jaws should rather be considered as belonging to the teeth than as part of the jaws."[5]

The significance attached to this case is of the utmost importance because it vividly illustrates what happened to the dentofacial complex of this patient as a result of the noneruptive pattern of the teeth in the jaws and what followed when orthodontic therapy supplied the stimulus which unlocked and permitted the genetic potentialities to approach their optimum in jaw growth and dental development. What is important is that orthodontic treatment did nothing for jaw growth in spite of the tremendous amount of tooth movement. Had growth of the jaws reached maturity at 15 years? This case had a Class III tendency due to lack of maxillary or premaxillary growth rather than aberrant mandibular growth.

This report would not be complete unless attention is directed to the benefit accomplished for the patient in relieving him of the neurosis caused by his dentofacial deformity. Treatment resulted in a personality change from withdrawal to desire to actively participate as a member of the community.

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