



## DIFFERENT ORTHODONTIC TREATMENT APPROACHES FOR CLEIDOCRANIAL DYSOSTOSIS – A REVIEW

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### ABSTRACT

Cleidocranial dysostosis is a rare congenital skeletal disorder, associated with clavicular hypoplasia or aplasia, delayed closure of cranial fontanelles, brachycephalic skull, delayed exfoliation of primary dentition, eruption of permanent teeth, and multiple supernumerary and morphologic abnormalities of the maxilla and mandible. The disorder is caused by mutation in the CBF1 gene, on the short arm of chromosome 6p21. The prevalence of cleidocranial dysostosis is estimated one per million, without sex or ethnic group predilection. The purpose of this paper is to describe the orthodontic treatment in a patient with cleidocranial dysostosis. Therapy may include removal of supernumerary teeth, surgical exposure of impacted teeth, and orthodontic treatment.

**KEYWORDS :** Cleidocranial dysostosis, Clavicular hypoplasia, Impacted teeth, Supernumerary teeth, Brachycephalic skull.

### INTRODUCTION

Cleidocranial dysplasia (CCD), also known as Marie and Sainton disease<sup>1</sup> or cleidocranial dysostosis, could be a rare congenital abnormality of autosomal dominance inheritance that's characterized by persistently open sutures or delayed closure of sutures, hypoplasia or aplasia of the clavicles, conical thorax, short stature, supernumerary teeth, delayed eruption of permanent dentition and alternative skeletal anomalies.<sup>2</sup>

Dental anomalies and a few degrees of clavicular hypoplasia appear to be consistent characteristics of the disorder.<sup>3</sup> A variety of dental issues might occur in CCD. This condition is of clinical significance to dentistry due to the involvement of the facial bones, retained deciduous teeth and many impacted permanent successors and supernumerary teeth, sometimes presence of follicular cysts and eruptive pseudocysts.<sup>4</sup>

CCD manifests itself as a condition within which teeth fail to erupt, that is believed to ensue to the absence of cellular cementum and a rise within the quantity of a cellular cementum of the roots of the affected teeth.<sup>5</sup> For these reasons, dental management could be an important facet of the health care of affected persons.<sup>6</sup> This disorder is transmitted as an autosomal dominant trait or it's caused by a spontaneous genetic mutation and is present at a frequency of one in one million individuals.<sup>7</sup> It affects both males and females equally.<sup>8</sup> It has been incontestable that mutation of the runt-related transcription factor 2 gene (RUNX2), situated at chromosome 6p21, is related to CCD.<sup>9</sup>

In CCD dental disability begins in late youth, with the

progressive morbidity of the deciduous dentition. Deterioration becomes apace progressive over the amount of a some years, giving the patient an edentulous and aged facial look. Accordingly, the aim of any dental treatment has to include elimination of those disturbances, provision of a functioning masticatory mechanism, and improvement of the patient's appearance. To attain these goals, (a) prosthetic replacement by means that of dentures, with or without previous extraction of the impacted teeth. In some cases, the impacted teeth are exposed and brought in the arch and used to support overdentures.<sup>10</sup> (b) A surgical approach consisting of the extraction of supernumerary teeth, which is followed by surgical repositioning or transplantation of the permanent teeth.<sup>11</sup> (c) Combined approaches of surgical and orthodontic treatment with the aim of actively erupting and aligning the impacted permanent teeth.

### DENTOALVEOLAR CHARACTERISTICS<sup>12</sup>

The following dento-alveolar characteristics of cleidocranial dysplasia is probably present in all cases, to a greater or lesser extent : (1) Over-retained deciduous teeth without resorbed roots. (2) Supernumerary teeth that displace the developing permanent teeth and obstruct their pathway to erupt. (3) Retarded eruption because of a lessened eruptive potential, although it is not entirely missing. (4) Reduced lower third of the facial height and a skeletal Class III tendency due to underdevelopment of the maxilla and forward mandibular rotation. The vertical development of alveolar bone is markedly reduced, with a shallow buccal and lingual sulcus. (5) A late, however spontaneous eruption of first and, usually, second permanent molars in both upper and lower jaws,

(6) Delay in the root development of the permanent teeth which is serious in nature. (approximately 3 years)

### ORTHODONTIC TREATMENT STRATEGY

Space for the unerupted teeth is provided by anteroposterior expansion of the dental arches and, in the vertical plane, by the extraction of deciduous and supernumerary teeth. For efficient orthodontic force application : (a) More number of erupted anchor teeth. Typically, the permanent molars erupt normally and, in many cases, one or two incisors may present in both jaw. (b) A rigid appliance frame. This is often required to withstand the distortion from oral function, considering the long spans of free, unattached arch wire, mesial to the few erupted anchor molar teeth. (c) The application of light forces with a vary of action to individual and groups of unerupted teeth. (d) The planning of the appliance should take under consideration the requirement for it to be sufficiently versatile to hold out its 3 distinct task efficiently, namely, to use forces in the vertical plane to correct the multiple impactions, Help to adapt to the changing environment from a partial to a full permanent dentition, including reapportioning space for further unerupted teeth and establishing interproximal contacts and arch form, and to bring about an occlusion of these teeth, along side with a corrective realignment of their long axes.

### SURGICAL MEASURES<sup>12</sup>

Two surgical interventions are generally planned and their timing is governed by the acceptable root development.

Intervention 1: Dental age should be 7-8 years

- a. Extraction of the anterior deciduous teeth,
- b. Removal of all supernumerary teeth
- c. Surgically exposure of permanent incisor teeth (root developments should be two-thirds their expected length)
- d. Bonding of the attachments immediately
- e. Closure of the surgical flaps fully.

The canines and premolars will be at an early stage, with the roots between a third to a half their expected final length. Surgical intervention in this region is therefore restricted to the removal of any existing supernumerary teeth with their associated deciduous teeth. The immature posterior permanent teeth are not exposed otherwise their dental follicles disturbed at this time.

Intervention 2: Dental age 10 to 11 years (chronological age 13+ years)

- a. Extract remaining deciduous teeth,
- b. Expose unerupted premolars and canines,
- c. Bond attachments immediately,
- d. Close the surgical flaps fully.

### TREATMENT STRATEGY TODAY

Distinct and contrasting approaches have been suggested over the past few years and each of these has its relative merits:

The Toronto-Melbourne approach.<sup>13</sup> This technique advocates a series of surgical procedures, at first involving the removal of the deciduous teeth under endotracheal general anesthesia, with its timing dependent on appropriate root development of the permanent teeth. In the first stage at age 5 to 6 years, deciduous incisors are extracted, whereas the deciduous posterior teeth are removed in the second stage, at age 9 to 10 years. At each intervention, supernumerary teeth and bone overlying the crypts of the corresponding, unerupted, permanent teeth are removed. Only much later, after the spontaneously erupted first molars have been banded, a more localized surgical re-exposure of the permanent incisors is performed and the exposed area packed. Attachments are placed on the permanent incisors

after healing has occurred. Later still, between 9 to 12 years, the premolars are surgically exposed, the supernumerary teeth removed, and the exposed area packed. After complete healing and removal of the packs, brackets are placed on the premolars and canines.

The Toronto-Melbourne authors present no details of the mechanics of the orthodontic treatment that they use in the encouraged eruption of the teeth and their eventual alignment, although the reader is left to assume that a full multibanded edgewise appliance is used in the latter stages of alignment, judging from the illustrations in the latter publication.

The Belfast-Hamburg approach.<sup>14</sup> One of the principal aims of this approach is to limit the inescapable need for extensive surgery to a single episode, designed to remove all deciduous and supernumerary teeth and to expose all unerupted teeth simultaneously. Under general anesthesia and in operating theater conditions, all the deciduous and unerupted supernumerary teeth are removed, the permanent teeth of the normal series are widely exposed, and a surgical pack is placed over them to prevent bony healing and soft-tissue closure over the teeth. Healing is by secondary intention. Over an extended period, these surgical packs are changed frequently, until such time as orthodontic brackets may be bonded to each of the unerupted teeth, under conditions that are less likely to lead to bond contamination than those present during the actual surgical exposure procedure. Often, spontaneous eruptive movements occur with some of the teeth, although it is never sufficient or reliable enough to eliminate the need for extrusive mechanics. Orthodontic appliances must be placed on the few teeth that are fully erupted and elastic thread is tied between the bonded brackets on the unerupted teeth and the arch wires to encourage eruption.

The Jerusalem approach.<sup>15</sup> This method offers a different modus operandi, based on a rationale that is directly related to dentoalveolar development and the factors causing its aberration, which lead to the creation of the clinical picture seen in this syndrome. The method takes into account four principal aspects that should be considered in its comprehensive approach to treatment. These aspects may be listed as follows: (1) the clinical features of the dentoalveolar structures in the disease, (2) the surgical measures needed to overcome them, (3) the planning of appropriate orthodontic treatment states to meet the demands of the unusual circumstances, and (4) concentrating initial efforts towards bringing anterior teeth into the mouth early, for the patient's self-image.

The Bronx approach.<sup>16</sup> As in the Toronto-Melbourne and Jerusalem techniques, deciduous teeth and underlying supernumerary teeth are removed under general anesthesia and surgical flaps are closed. Unlike the previous techniques, this approach uses the placement of a removable partial overdenture for esthetic and functional purposes. As with the Toronto-Melbourne and Jerusalem techniques, the age at which the management commences depends on the stage of root development of the underlying permanent teeth. If necessary, an intermediate operation is undertaken so as to expose unerupted teeth and place orthodontic brackets over fully erupted molars. A transpalatal arch appliance is welded to the brackets and these are used in conjunction as a base for an artificial dentition. After the natural eruption of the permanent teeth with sufficient posterior support, orthodontic appliances are used to bring the teeth into occlusion. Finally as per case requirement, a Leforte-I osteotomy-orthognathic procedure is performed and dental implants are placed.

### DISCUSSION

Surgical and orthodontic difficulties and complications

abound throughout the treatment of cleidocranial dysplasia and there is a risk for the failure of one or other of the different aspects of the treatment or of the prognosis of the result. Tooth buds may be damaged by the trauma of their exposure while they are in an insufficiently developed state. The Toronto-Melbourne approach suggest a series of several extensive and minor surgical procedures, over a long period. By contrast, the Belfast-Hamburg approach offers a one, all-inclusive surgical procedure to eliminate the superfluous (deciduous and supernumerary) teeth and to expose the remainder.<sup>12</sup>

The bonding of a little metal attachment to an impacted tooth immediately after the surgeon has exposed the tooth and prior to flap closure, Ligation is made with a fine wire that extends from the attachment in the direction of the dental arch or vertically through the fully replaced surgical flap. The utilization of surgical packs permits the unkeep of patency of the exposure sites. Their presence and the necessity for their frequent changing causes pain and nuisance and it compromises oral hygiene and normal function.<sup>12</sup> Sound appliance fabrication is needed to beat the problems with the use of fine gauge arch wires to effect wide-ranging extrusive, rotating, and aligning movements in the presence of long edentulous spans. Where more teeth are erupted, the mechanics that may be used is more efficient; but in the absence of erupted incisors, the means to apply augmented eruption forces (anchored on the molar only) becomes very problematic. In general, neither has it been described in the step-by-step or in the single-surgical-episode approaches.<sup>12</sup> Hence a conservative surgical policy is usually recommended in which only enough bone is removed to permit access for the placement of a small eyelet attachment on the minimally exposed tooth surface. The surgery is aimed at preserving rather than removing bone, because the presence of bone does not seem to hinder the orthodontically encouraged eruption of the teeth in these cases.<sup>12</sup>

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