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# Multidisciplinary team approach in the oral rehabilitation of patients with cleidocranial dysplasia to achieve a functional aesthetic outcome

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## Abstract

Cleidocranial dysplasia is a hereditary congenital disorder that results in delayed ossification of midline structures, and is caused by mutations in the RunX2 (runt-related transcription factor 2) gene located on the short arm of chromosome 6. Successful treatment depends on multidisciplinary assessment and a comprehensive staged treatment plan. We present a case series of 12 patients who were managed with a specifically tailored combination of surgery, orthodontics, and prosthodontics to provide a functional dentition and restore their smile and facial contour. Successful dental rehabilitation can be challenging in this group because patients often have multiple dental anomalies and a reduced quantity and density of alveolar bone. Rehabilitation with early intervention and a carefully planned multidisciplinary approach has been successful in the long term.

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**Keywords:** cleidocranial dysplasia; MDT; oral surgery; orthodontics; oral rehabilitation

## Introduction

Cleidocranial dysplasia is a hereditary congenital disorder that results in delayed ossification of the midline structures. It is transmitted as an autosomal dominant trait and is caused by a mutation in the runt-related transcription factor (RunX2) gene on chromosome 6p21, which is essential for the terminal differentiation of osteoblasts.<sup>1</sup> The condition is characterised by defective development of the cranial bones and by complete or partial absence of the clavicles.<sup>2</sup> Patients often develop a short stature with hypoplastic facial bones and a resultant prognathic mandible, and dental anomalies include prolonged retention of the primary dentition, failure of the secondary dentition to erupt, and multiple supernu-

merary teeth.<sup>3</sup> As a result patients present with masticatory hypofunction and poor aesthetics.<sup>4</sup>

A report in 1992 suggested that the optimal age for treatment is between five and seven years, as supernumerary incisor teeth can be diagnosed at this stage with supernumerary canines and premolars diagnosed later.<sup>5</sup> Several treatments that were subsequently proposed (including those from Toronto-Melbourne,<sup>6,7</sup> Belfast-Hamburg,<sup>8</sup> Jerusalem,<sup>9,10</sup> and the Bronx<sup>11</sup>) all adopted timed operations, but we know of no protocol for patients who seek treatment at different ages, many of whom present in their early teens.

We have therefore adopted a holistic multidisciplinary approach that focuses on operation, orthodontic alignment, and prosthodontic rehabilitation. Treatment is governed by the severity of the condition and the needs of the patient, and must accommodate alveolar growth and the development and eruption of the permanent dentition. Successful management

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will provide patients with an optimal functional dentition and where possible, restore dental and facial aesthetics.

## Material and methods

### Assessment and diagnosis

We present a series of 12 patients who were successfully treated with a staged multidisciplinary approach. Initially, they were all assessed by a team of specialists from oral surgery, orthodontics, and restorative dentistry. Dental anomalies varied in severity, and the patients presented at different stages of development. All patients had an orthopantomogram (OPG), and those with severe dental anomalies also had cone-beam computed tomography (CT). After consultation they all had genetic tests to identify the RunX2 gene.

## Results

Our aim was to treat each patient according to their needs and to focus on their dental development, age, and alveolar growth. Table 1 shows the different phases of the treatment regimens. Eight patients were successfully managed with orthodontic and surgical treatment, but as orthodontic treatment was not possible in those with severe dental anomalies, prosthodontic rehabilitation was required to achieve a functional dentition. The type of restoration depended on individual needs. Two cases show the combined approach to treatment.

Case 1 presented with moderate dental anomalies. The primary issue was retention of the deciduous dentition and failure of the permanent dentition to erupt. After extraction



Fig. 1. Case 1. Intraoperative treatment with orthodontic fixed appliance and orthodontic traction with gold chains.

of all retained deciduous teeth, orthodontic treatment enabled the eruption of the permanent dentition through orthodontic traction. The UL3 was transplanted into the arch as it failed to erupt (Fig. 1). Subsequent orthodontic alignment achieved a functional and aesthetic outcome, and a good facial profile (Fig. 2).

Case 2 presented with severe dental anomalies. OPG and cone-beam CT showed 20 unerupted maxillary teeth and 24 unerupted mandibular teeth (Fig. 3), and further disease included cystic lesions associated with the unerupted lower right and left third molars, consistent with dentigerous cysts. The patient was managed solely with surgical and prosthodontic rehabilitation.

The mobile deciduous teeth and all unerupted supernumerary teeth were removed in stages under general anaesthesia. Coronectomy of both lower right and left third molars was done because of their position and relation to the inferior dental nerve, together with enucleation of associated cystic lesions. Bovine bone grafts were used to assist regeneration and to preserve the alveolar ridge during healing, and the

Table 1

Multidisciplinary treatment regimens for rehabilitation of function and aesthetics in the 12 patients. Phase one was early operation in all cases.

Case No.	Phase 2	Phase 3	Phase 4	Phase 5
1	Orthodontic alignment	Exposure and bonding of unerupted dentition	Autotransplantation of UL3	Orthodontic alignment
2	Orthodontic alignment	Exposure and bonding of unerupted dentition	Orthodontic alignment	–
3	Orthodontic alignment	Exposure and bonding of unerupted dentition	Orthodontic alignment	–
4	Interim prosthesis	Implant placed	Final prosthesis	–
5	Interim prosthesis	Implant placed	Final prosthesis	–
6	Implant placed	Final prosthesis	–	–
7	Orthodontic alignment	Exposure and bonding of unerupted dentition	Orthodontic alignment	–
8	Orthodontic alignment	Exposure and bonding of unerupted dentition	Orthodontic alignment	–
9	Orthodontic alignment	Implant placed	Final prosthesis	–
10	Orthodontic alignment	Exposure and bonding of unerupted dentition	Autotransplantation of UR3	Orthodontic alignment
11	Orthodontic alignment	Exposure and bonding of unerupted dentition	Orthodontic alignment	–
12	Orthodontic alignment	Exposure and bonding of unerupted dentition	Orthodontic alignment	–

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