

## Aesthetic Anxiety in a Child with Cleidocranial Dysplasia

Ayesha Shahid, Arooj Aman, Amna Malik

### ABSTRACT

Cleidocranial dysplasia (CCD) is a rare syndrome that affects the skeleton and teeth. It is characterized by absent or hypoplastic clavicles, mobile shoulder girdles, patent fontanelles, supernumerary teeth, retained deciduous and delayed permanent teeth. A 10 year old boy with CCD is reported with chief complaints of aesthetics and dental pain. The bullying and social agony at a tender age were alarming as the patient was highly distressed regarding his missing teeth. Juvenile aesthetic concerns and the psychosocial impact were emphasized in this case. He was treated with manual scaling, pulp therapy, restoration, and a prosthesis to speedily replace his missing teeth. Long-term orthodontic treatment was suggested. Due to the early diagnosis, a better prognosis exists for multidisciplinary treatment. Counselling was pivotal for dealing with his aesthetic anxiety and oral health. Special attention should be given to the aesthetics and psychosocial state of patients with syndromes in underdeveloped societies.

**Keywords:** Aesthetic anxiety, cleidocranial dysplasia syndrome, dental treatment, psychosocial stigma, young male

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### INTRODUCTION:

Cleidocranial dysplasia (CCD) is a “one in millions” rare congenital disorder commonly caused by RUNX2 gene mutation, in which endochondral and intramembranous bone formation is defective. RUNX2 is involved in the osteoblastic lineage of stem cells.<sup>1</sup> Calvaria and clavicles are primarily affected in this autosomal dominant disorder, with absent clavicles in 10% of the cases.<sup>2</sup> Common findings are aplastic or hypoplastic clavicles, Wormian bones, incompletely closed fontanelles, short stature, retained deciduous dentition, delayed eruption of permanent teeth, supernumerary teeth, brachycephaly, hypertelorism, frontal bossing, etcetera.<sup>3</sup> Other skeletal anomalies may include small and bell-shaped thoracic cage, mobile shoulder girdle, underdeveloped maxilla, malformed paranasal sinuses, and bone defects. CCD patients are easily diagnosed by abnormalities of clavicles, skull, and dentition.<sup>4</sup>

The deranged skeletal features in CCD may cause aesthetic anxiety in patients which may hinder their social integration.

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Missing anterior teeth affects social life and may lead to other clinical issues.<sup>5</sup> The orodontal anomalies are of utmost clinical significance to dentists, who should be able to diagnose the syndrome. Multidisciplinary management is required to restore aesthetics and function. A case of CCD is presented in this report with aesthetic distress at a young age.

### CASE

A 10 year old boy of Pashtun origin presented to the Islamic International Dental Hospital, Islamabad in March 2019, with complaints of both aesthetic distress and dental pain. He complained that children in his madrassa bully him about his lack of anterior teeth, which traumatized him. On examination, he had visibly deficient clavicles and abnormal mobility of the shoulders. He was able to bring the humeral heads close to each other (Figure-1). He had short stature and a very lean body for his age. He had a long and narrow face (dolichofacial) with anterior divergence, mid-face deficiency, frontal bossing, hypertelorism, depressed nasal bridge, and depressed zygomatic bones. He was diagnosed with CCD and had no family history. His father gave consent on behalf of the child to be photographed and published. Chest x-ray posteroanterior view showed the absence of a right clavicle and a hypoplastic left clavicle, bell-shaped thorax, and low-placed scapulas (Figure-2). Upon intraoral examination, a narrow, “V” shaped, and high vault palate was seen. The patient had several impacted permanent teeth, malocclusion class 3, and #46 was in posterior buccal crossbite. Among permanent teeth, the upper arch had 14, 16 and 26 erupted whereas the orthopantomograph showed impacted 11, 12, 13, 15, 21, 22, 23, 24, and 25 (Figure-3). Broken down roots (BDRs) of deciduous 55, 64 and 65 were

present whereas 53, 62 and 63 were retained. The lower arch had entire deciduous teeth retained and only 36 and 46 erupted. There was pain on percussion in #46 and the periapical x-ray showed deep caries. No supernumerary teeth were present. Lateral cephalogram showed a hypoplastic maxilla, long y-axis and large FMA, indicating class 3 mandibular prognathism, and a high vertical growth pattern (Figure-3). An anteroposterior view of the skull showed normal sutures (Figure-3) Dental scaling was manually performed and residual roots were extracted. It was followed by pulpectomy of the carious tooth with a Glass ionomer cement (GIC) restoration. Upper and lower anterior partial dentures were made for aesthetic purposes, with spaces underneath for the permanent teeth to erupt. The patient was called for follow-up visits where the acrylic of the denture was trimmed whenever the teeth erupted further (Figure-4). For the skeletal class 3 mandibular prognathism, occlusal chin cups were presently advised. Orthodontic treatment for the eruption of teeth was also suggested which the patient refused. The patient was counseled for his aesthetic anxieties and oral hygiene maintenance. He was referred to orthopedics for stabilization of his shoulders. The patient was extremely satisfied with the prosthesis.

Figure 1: Frontal profile of patient showing hypermobile shoulders and deficient clavicles



Figure 2: Chest radiograph posteroanterior view showing bell-shaped thorax. The arrow on the right side shows the absence of a clavicle and the arrows on the left side show a hyperplastic clavicle



## DISCUSSION

A child's environment plays a key role in the development of sound social and mental health. McNamara et al. reported that 93.5% of CCD patients present with dental abnormalities.<sup>6</sup> Since the case had visible anomalies, he was subjected to traumatic bullying and psychosocial anxiety even at the tender age of ten. Similarly, Garg and Agrawal reported an adult male CCD patient who was also psychologically traumatized due to missing teeth and societal abandonment. He also reported speech and communication barriers and wanted rapid dental restoration.<sup>4</sup> Alves and Oliveira also reported an adult female CCD patient with aesthetic agony and the need for speedy restoration of teeth.<sup>7</sup> This implies the aesthetic vulnerability of CCD patients, not only in mature adults but juvenile patients as well. Similar to this case, Tristão et al. concluded in their systematic review that dental malocclusion is related to increased bullying in children and teenagers.<sup>8</sup> Aesthetic concerns in children due to syndromes like cleft lip and palate have also

Figure 3: Radiographs of the patient. (a) Anteroposterior view of skull. (b) Lateral cephalogram. (c) Orthopantomograph showing several impacted permanent teeth and few retained deciduous teeth

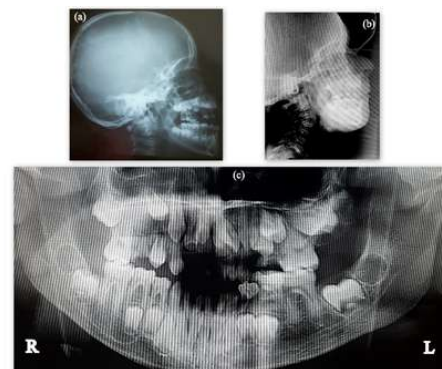
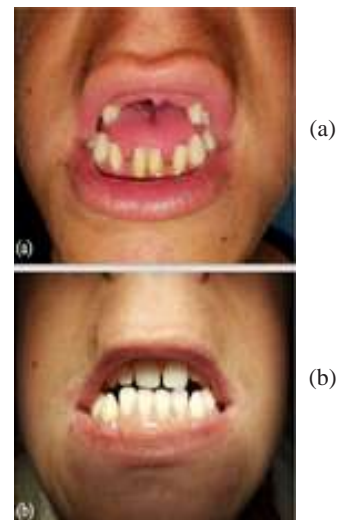


Figure 4: (a) Pre-treatment facial profile. (b) Post-treatment aesthetic facial profile with prostheses



been reported, where the children report psychosocial agony.<sup>9</sup> Surprisingly, some other case reports of pediatric CCD did not evaluate the aesthetics related anxiety, the social or mental health of the children.<sup>10,11</sup> This is alarmingly important in underdeveloped countries where the aesthetic anxieties of children and their mental health and self-esteem are not emphasized. Psychological assessment and counselling of patients with congenital deformities must always be performed.

The treatment planning for CCD should be focused on a functional and aesthetic outcome.<sup>12</sup> Treatment usually consists of extraction of retained deciduous and supernumerary teeth at an accurate time to guide the eruption of permanent teeth with the help of orthodontic traction and elastics. Orthodontics and oral surgery combine the goal of correction of mandibular prognathism.<sup>13</sup> The rapid and non-invasive treatment modality includes tooth-supported prosthesis.<sup>14</sup> This case has been reported due to the rarity of CCD and its associated aesthetic trauma in a child.

### CONCLUSION

A multidisciplinary approach is required for the rehabilitation of function and aesthetics of CCD patients. Even in pediatric patients, aesthetic concerns should be treated earnestly. Counselling the patients about their aesthetic anxieties and healthcare is pivotal for their self-esteem and social life.

#### Authors Contribution:

**Ayesha Shahid:** Research conception, case management, design, writing the final draft, data collection and analysis

**Arooj Aman:** Research conception, design, writing initial draft and data collection and analysis.

**Amna Malik:** Research design, editing the draft data, data collection and analysis.

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