Oral rehabilitation of a patient with Kenny–Caffey syndrome using telescopic overdenture

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Abstract
Kenny–Caffey syndrome (KCS) is a rarely reported autosomal disorder characterized by skeletal, ocular, and oral manifestations. Oral features such as microdontia, hypodontia, malalignment of teeth, bone loss, and difficulty in mastication results in serious esthetic and functional handicap. The prosthetic rehabilitation of such patients is challenging, especially when implant placement is not a good choice due to poor Vitamin D levels. The existing literature is scarce in describing the treatment options. This case report describes the oral rehabilitation of a patient affected with KCS using telescopic overdenture.

Keywords: Hypodontia, microdontia, Kenny–Caffey syndrome, telescopic overdenture

INTRODUCTION
Kenny–Caffey syndrome (KCS) is a rare autosomal disease characterized by short stature, cortical thickening and medullary stenosis of tubular bones, delayed closure of anterior fontanelle, ocular abnormalities such as microphthalmia and papilledema, hypoparathyroidism, and associated hypocalcemia.[1‑4] It can be classified into KCS type 1 and KCS type 2. KCS type 1 is an autosomal recessive form caused by mutations of the tubulin-specific chaperone E gene, and KCS type 2 is an autosomal dominant form caused by mutations of the FAM111A (family with sequence similarity 111 member A) gene.[5] The patient discussed in this case report is already diagnosed with KCS type 2.

Patients show normal intelligence in KCS type 2. The dental abnormalities associated with the disease are hypodontia, microdontia, abnormally shaped and positioned teeth, and associated difficulty in mastication.[6]

The prosthodontic rehabilitation of KCS affected patients is not adequately reported in the literature. This article aims in reporting a case of successfully rehabilitated prosthodontic intervention for a KCS type 2 affected young patient.

CASE REPORT
A 26-year-old male patient diagnosed with KCS type 2 reported to the department of prosthodontics with the...
chief complaint of difficulty in chewing due to missing teeth [Figure 1].

The medical history revealed low serum calcium, phosphorous, Vitamin D, parathormone, and growth hormone levels and raised alkaline phosphatase levels. The patient underwent growth hormone therapy but found futile and discontinued. The patient was supplemented with calcium and Vitamin D tablets. The bones of the extremities appeared normal except for minimal increase in bone density. Chromosomal analysis was done (Giemsa banding) indicating a male karyotype (46, XY) with an interstitial deletion in 22q.

The patient underwent root canal treatment (RCT) for 46 and extraction of 43, 4 months before reporting to the department. Intraoral examination revealed the presence of following permanent teeth – 11, 13, 16, 17, 21, 22, 23, 26, 27, 36, 37, 41, 46, and 47 and following retained deciduous teeth were also present – root stump of 52, 64, 65, 73, and 74. Grade 3 mobility, deep periodontal pockets, and poor bone support were noted for 11, 21, 22, 23, 41, and 46. All the retained deciduous teeth were found short. Orthopantomogram confirmed the above findings [Figure 2].

Both the function and esthetics were matter of concern for the patient. Since serum Vitamin D levels were very low, implant supported prosthetic rehabilitations were excluded from the treatment plan.[6] After discussing advantages and the disadvantages of various treatment options with the patient, following final treatment plan was made.

Since the deciduous teeth and periodontally week teeth had poor prognosis and could interfere with the final outcome, extraction of all retained deciduous teeth and following permanent teeth – 11, 21, 22, 23, 41, and 46 were planned. RCT of 13 was proposed. It was decided to prosthetically rehabilitate the patient with a tooth supported telescopic overdenture. Primary copings were planned on all remaining teeth – 13, 16, 17, 26, 27, 36, 37, and 47. Secondary coping was planned on all except 27 as this could interfere with path of placement. The secondary copings on maxillary and mandibular right quadrants were planned to give composite facing. Since the interarch distance between the second and third quadrant was more, acrylic teeth were the better option than composite facings on the secondary copings.

Once extraction and RCT were completed and healing was satisfactory, the patient reported to the department of prosthodontics [Figures 3 and 4]. Diagnostic impression was made using irreversible hydrocolloid (alginate) impression material. Diagnostic mounting was done on a nonadjustable articulator after recording the jaw relation. Loss of vertical dimension of 7 mm was identified.

Tooth preparation of the entire remaining teeth was done. Final impressions of maxillary and mandibular arch were made using addition silicone putty-light body dual impression technique. After fabrication of full metal primary coping, try-in was done in patient’s mouth and found to have perfect fit. Pickup impression for fabrication of secondary coping was made using addition silicone putty-light body dual impression technique. Fit of primary and secondary coping was checked; face bow transfer and the final jaw relation were recorded with corrected vertical dimension.

After mounting in a semi-adjustable articulator, maxillary and mandibular teeth setting was done. Try-in was
performed. Occlusal and esthetic evaluations were found satisfactory both for the patient and the operator. On the day of try-in, 1.23% acidulated phosphate fluoride (APF) gel was applied. The acrylization was done using heat-cured acrylic resin. The final acrylic denture with embedded secondary copings was finished and polished [Figures 5 and 6].

After cementing the primary copings, the final denture was inserted and evaluated for any occlusal errors [Figures 7 and 8]. Once corrections were complete, postinsertion instructions were given. The patient was reviewed after 1 week, 1 month, 6 months, and 1-year intervals. Fluoride application was repeated every 6 months. The esthetics and masticatory efficiency were

**Figure 3:** Maxillary arch – after extraction

**Figure 4:** Mandibular arch – after extraction

**Figure 5:** Telescopic overdentures – intaglio surface

**Figure 6:** Telescopic overdentures – cameo surface

**Figure 7:** Intraoral view of both arches after cementation of primary coping

**Figure 8:** Postinsertion view – with telescopic overdentures
satisfactory and it drastically improved the quality of life of the patient.

DISCUSSION

All cases of KCS are manifested with oral manifestations such as short roots, hypo/oligodontia, microodontia, dental caries, enamel defects, delayed eruption, tooth agenesis, and micrognathia. However, there is a lack of literature regarding the oral rehabilitation procedures for patients with KCS. Only one case has been reported in literature that mentions prosthetic rehabilitation in KCS patient which was treated with simple tooth retained overdenture.

Telescopc overdentures are well known for its advantages such as retention, stability, esthetics, and patient satisfaction. Although disadvantages such as cervical caries, fracture of denture, loss of composite facing, and decementation of copings were reported associated with telescopic overdentures in literatures, no such complications were noted in the presented case until completion of 1-year follow-up due to meticulous care of the denture and oral hygiene measures practiced by the patient. The use of 1.23% APF gel also contributed to the prevention of caries.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the legal guardian has given his consent for images and other clinical information to be reported in the journal. The guardian understands that names and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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