Multidisciplinary management of Blepharo-Cheilo-Dontic Syndrome and the role of overdenture in dental management

ABSTRACT

Aim Blepharo-Cheilo-Dontic (BCD) syndrome is a rare condition characterised by abnormalities of the eyelid, lip and teeth. A 12-year-old girl with BCD syndrome presented at the Dental Hospital following referral from the multi-disciplinary cleft lip and palate clinic. She had skeletal Class III relation, with left posterior cross bite, occlusal contacts on the second permanent molars with poor oral hygiene. The permanent units missing were 15, 14, 13, 12, 11, 21, 22, 25, in the upper arch and 35, 34, 32, 44 and 45 in the lower arch. This patient presented a complex aesthetic problem, which through multidisciplinary care resulted in a satisfactory aesthetic outcome. In this case report we present the clinical management and the role of the overdenture in her dental management.

Key words: Multidisciplinary; BCD syndrome; dental management; overdenture.

Introduction

Blepharo-Cheilo-Dontic (BCD) syndrome is a rare autosomal dominant condition of congenital facial clefting, oligodontia, euryblepharon, lagophthalmos, and ectropion [Gorlin et al., 1996]. Although the expressivity is variable, the common manifestations are cleft lip/palate, ectropion and lagophthalmos [Allanson and McGillivray, 1985]. Sporadic cases have also been reported.

BCD syndrome was initially known as Elschnig syndrome, however following the review of cases in the literature, the term Blepharo-Cheilo-Dontic syndrome was considered more descriptive and appropriate [Gorlin et al., 1996].

Less than 25 cases have been reported in the English literature since 1996 [Gorlin et al., 1996; Guion-Almeida et al., 1998; Valdéz-de la Torre et al., 1999; Yen et al., 2001; Martinhago and Ramos, 2004, lida et al, 2006]. The gene responsible for BCD is yet to be identified.

The characteristic abnormalities of the eyelids, lips and teeth include [Gorlin et al., 1996] the following.

• The eyelids; euryblepharon (eyelids with abnormally wide lid opening), ectropion (turning out of the eyelid away from the eyeball) of the lower eyelids, distichiasis (congenitally formed extra row of eyelashes) of the upper eyelids and lagophthalmia (inability to close the eye completely).

• The lips in the form of cleft lip and palate, often bilateral.

• The teeth- oligodontia and microdontia.

BCD syndrome can be described in binary, ternary and quaternary combination. The common features are cleft lip and palate, oligodontia, eyelid ectropion and lagophthalmos. Other features reported are abundant eyelashes, hypertelorism, broad nasal base, hypoplastic malar and maxilla, everted lower lip, and agenesis of thyroid gland, a rare feature [Martinhago and Ramos, 2004]. Oligodontia and microdontia are extremely variable and when present, affect both dentitions [Gorlin et al., 1996].

This case report describes the dental management of a 12 year-old female (GY) with BCD syndrome who was referred to the Newcastle Dental Hospital by the Multidisciplinary Cleft Unit. She presented with concerns about the appearance of her face and the lack of front teeth which resulted in difficulty with her speech and a loose upper partial denture.

Case report

Medical history Initially, the diagnosis of her presenting features at birth was difficult and many genetic tests were carried out before the conclusive diagnosis of BCD syndrome was made. A quaternary combination of features affecting the eyes, lips, teeth and thyroid gland were present, in the form of bilateral cleft lip and palate, hypoplastic maxilla, euryblepharon, ectropion, lagophthalmos, abundant eyelashes, inactive thyroid, oligodontia and microdontia. She had undergone multiple reconstructive surgeries to repair the cleft of the lip and palate, the eyes and nasal base, and at the time of presentation further surgery was planned by the cleft and palate team to improve her facial appearance. In addition to the features of BCD syndrome, her medical history included delayed development with mild cerebral atrophy, petit mal epilepsy, and well-controlled asthma.

Dental history GY was an irregular dental attendee, under the joint care of the community dental service and the cleft team, before her referral to the Dental Hospital. Previous history revealed repair of cleft lip and palate, extraction of hypoplastic upper left central incisor (21) and carious upper left lateral incisor (22) and supernumerary tooth, in the line of the cleft palate. Sulcoplasty had been performed to deepen the upper labial sulcus and aid her prosthetic retention. There was a history of dental caries and restorations under local analgesia. From her clinical records, the upper lateral incisors (52, 62) were missing in the primary dentition. In the permanent dentition, a total of eleven teeth were missing. 11, 12, 13, 14, 15, 25, 35, 34, 32, 44, 45. At the time of her presentation, she was dissatisfied with her upper denture and was not using it because of its poor retention.
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complexity of this case, an interdisciplinary approach to treatment planning and management was adopted. With input from cleft team, restorative, and oral surgery, the following treatment plan was discussed with GY and her mother and consent was obtained.

The aim of the treatment plan was to address the patient's concerns, maintain good oral health and her remaining dentition. The significantly underdeveloped premaxilla and the severity of hypodontia prevented the option of orthodontic expansion anteriorly in the canine region and alveolar bone graft. Maxillary expansion on the right side would be difficult due to absence of teeth anterior to 16. In a conventional cleft lip and palate case with a developed premaxilla, the premaxilla would have been grafted to the lesser segments to achieve stability. Also in view of the recent videofluoroscopy assessment, future maxillary advancement as a part of the treatment option to improve her facial profile was discarded as this would worsen the velopharyngeal dysfunction and further compromise her speech. Provision of an upper overdenture to replace the missing tissue and teeth was then decided upon. Sulcoplasty procedure was rescheduled to deepen the anterior maxillary sulcus and improve retention of a prosthesis.

The dental treatment plan was tailored to the short and long term need of the patient. Introduction to dental treatment was commenced with preventive measures. This along with maintaining the existing occlusion assisted with gaining GY's confidence. Carious teeth were restored using preformed metal (PFM) crowns and upper overdenture fabricated.

Family and social history GY lived with her mother and 2 older siblings. She was of normal intelligence and attended mainstream school. There was no family history of BCD syndrome.

Clinical presentation

At clinical examination, G.Y was very shy and withdrawn and much of the communication was through her mother. The extra-oral examination (Fig. 1, 2) revealed clinical characteristics of BCD syndrome, class III skeletal pattern and severely hypoplastic maxilla.

Intra-oral examination (Fig. 3) showed poor oral hygiene with shallow maxillary labial sulcus. The upper arch was collapsed and v-shaped with deformity and scarring in the premaxilla region. The occlusal molar relationship was Class III, however the incisor relationship was not recorded as the 13, 12, 11, 21, 22 were absent. There were occlusal contacts on second permanent molars with left posterior cross-bite. The erupted teeth were 17, 16, 23, 24, 26, 27, 37, 36, 33, 31, 41, 42, 43, 46, 47 and 36 was heavily restored. Radiographic investigations, dental panoramic tomogram (DPT) and radiograph (Fig. 4) confirmed the missing teeth and the presence of 18, 17, 16, 23, 24, 26, 27, 28, 38, 37, 36, 33, 31, 41, 42, 43, 46, 47, 48 and caries in 46 and 36. The lateral cephalometry view (Fig. 5) revealed severe Class III facial profile. Upper anterior occlusal view was taken to rule out any tooth or root segment in the cleft region. Study models and clinical photographs were taken for clinical records.

Treatment plan Due to the uniqueness and complexity of this case, an interdisciplinary approach to treatment planning and management was adopted. With input from cleft team, restorative, and oral surgery, the following treatment plan was discussed with GY and her mother and consent was obtained.

The aim of the treatment plan was to address the patient's concerns, maintain good oral health and her remaining dentition. The significantly underdeveloped premaxilla and the severity of hypodontia prevented the option of orthodontic expansion anteriorly in the canine region and alveolar bone graft. Maxillary expansion on the right side would be difficult due to absence of teeth anterior to 16. In a conventional cleft lip and palate case with a developed premaxilla, the premaxilla would have been grafted to the lesser segments to achieve stability. Also in view of the recent videofluoroscopy assessment, future maxillary advancement as a part of the treatment option to improve her facial profile was discarded as this would worsen the velopharyngeal dysfunction and further compromise her speech. Provision of an upper overdenture to replace the missing tissue and teeth was then decided upon. Sulcoplasty procedure was rescheduled to deepen the anterior maxillary sulcus and improve retention of a prosthesis.

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Treatment Dental acclimatisation was commenced
with the following preventive measures, oral hygiene and toothbrushing instructions. Dietary assessment and analysis were carried out and dietary advice was given, tailored to GY requirement. Daily fluoride mouthwash was instituted along with the use of fluoride toothpaste (1500 ppm) twice a day.

The carious 46 and 36 were subsequently restored with preformed metal crowns (Ion Ni-Chro Dental Products/3M) under local anaesthetics and inhalation sedation and the premolars and molars underwent fissure sealing (Fig. 6, 7). A maxillary overdenture with flexible soft lining (Eversoft acrylic lining, Eustanel®) overlying all maxillary permanent teeth and engaging the natural teeth undercut to improve retention was fabricated and fitted (Fig. 8). Improvement in the patient's facial appearance was noted after fitting the overdenture, and subsequent improvement in her speech (Fig. 9, 10, 11). Regular recall visits were arranged to review her oral health, the prosthesis and reiterate oral and dietary advice.

At two year recall, the 18 was noted to be erupting into the buccal sulcus and was extracted. A new maxillary overdenture was fitted 28 months after the initial one.

Discussion

This case illustrates the successful interdisciplinary management of a young patient presenting with multiple dental problems. GY presented with a rare syndrome with distinct facial and dental features which resulted in her lack of confidence and her initial withdrawn behaviour. Good oral health was achieved with the introduction of preventive measures and restoration of carious teeth. With the provision of the prosthesis, the improvement in her confidence was phenomenal. GY became a highly motivated young girl, maintaining good oral hygiene and was quite happy to receive dental treatment without sedation towards the end of her treatment. As GY was not keen on a lower denture, it was agreed not to undertake treatment in the mandibular arch.

The provision of a functional and aesthetically pleasing maxillary overdenture was achieved through an interdisciplinary approach. Removal of impacted 18 was required along the line to achieve a satisfactory outcome for this patient. Due to the extent of her dental intervention, GY would continue to be a high priority case for preventive care.

The use of overdentures, a conservative line of approach in cases of severe maxillary hypoplasia, severe hypodontia and cleft lip and palate patients is well documented [Shapiro and Kokich, 1984; Moore and McCord, 2004; Vergo, 2001]. Overdentures can readily restore function, appearance, soft tissue deficit and re-establish positive occlusion as the prostheses replace both the tissue and missing teeth, increase vertical facial height and result in overall improvement in appearance.

The advantages of overdentures [Bassi, 2007] can be classified as follows.

• Psychologic: maintained or even improved self-image.
• Functional: periodontal receptors in the preserved teeth allow occlusal force discrimination to be maintained, and masticatory performance and electromyographic activity remain similar to the dentate population [Rissin et al., 1978]. According to Nagasawa et al. [1979], the retention of only 2 roots is sufficient to obtain good results.
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- Biologic: the resorption of the alveolar ridge is extremely diminished when compared to complete denture wearers [Crum and Rooney, 1978].

The use of overdentures is not without its pitfalls. Possible complications and failures associated with their use include tooth decay, gingivitis, endodontic failure and vertical root fractures. In this case, thorough oral hygiene, fissure sealing, fluoride protection were implemented as control measures against tooth decay. Regular review appointments were also in place to monitor dental health and reiterate oral hygiene instructions. The incidence of gingivitis around retained roots was reported to be between 4 and 13% gingivitis [Budtz-Jorgersen, 1995]. A 12.1% endodontic failure rate was shown in a 23-year longitudinal study [Ettinger and Qian, 2004]. This is commonly in the form of periapical lesions related to both endodontically treated teeth (37%) and vital teeth (19.8%). Vertical root fractures were reported in 30.9% of overdenture abutment failures and occur more frequently in the maxilla when opposed by natural teeth. Most of the failures and complications could have been prevented by excellent oral hygiene home care and professional assistance [Ettinger and Qian, 2004].

Although no endodontic treatment was carried out in our patient, regular monitoring to maintain the remaining natural teeth is crucial.

While undertaking her dental treatment, she underwent further constructive surgeries to her eyes and nose to improve function and aesthetic appearance.

Conclusion

BCD syndrome is a rare condition and the choice of management in this case was the provision of the maxillary overdenture, highlighting the role of overdenture in complex cases. The replacement of missing dental and skeletal structures with a removable prosthesis resulted in remarkable improvement in her aesthetic appearance and function, mastication and speech, and allowed postponement of definitive surgical and restorative treatment until the cessation of her skeletal growth.

Referral to the restorative department for advanced restorative care in the form of assessment for bone graft and provision of implants would be considered in the future, taking into consideration the patient’s request.

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References


