Hypodontia and Microdontia Associated with Hereditary Ectodermal Dysplasia
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ABSTRACT
Hypodontia and associated conditions like Hereditary Ectodermal Dysplasia (HED) and microdontia markedly influence on physical, functional and psychosocial maturation of the affected individuals. Thorough evaluation, proper counseling and careful treatment planning employing a multidisciplinary approach are keys to a successful, long-term management. This case report describes the prosthodontic management of a young man with hypodontia and microdontia.

Key words: Hypodontia. Hereditary Ectodermal Dysplasia (HED). Microdontia. Odontogenesis. Prosthodontic management.

CASE REPORT
A 22-year-old man referred to the Department of Prosthodontics, de'Montmorency College of Dentistry/ Punjab Dental Hospital, Lahore, from his general dental practitioner with chief complaints of missing teeth and small sized existing teeth.

The patient was well-educated and aware of his problem for which he had consulted many general dental practitioners. He appeared to be psychologically depressed, reluctant and lacked confidence in moving around in his society. His mother and one brother were affected but with lesser severity and no extraoral symptoms. One sister was normal. Similar history was positive in his maternal relatives with cousin marriages for the last four generations.
On general examination, he had thin, sparse hair on extremities. The palms and soles showed marked hyperkeratosis. There was lack of sweating and complaints of hyperthermia in warm conditions.

On extraoral examination, lips appeared protuberant and lower facial height was reduced, giving the patient a concave facial form and an overaged look. On intraoral examination, severe hypodontia with marked microdontia was present. Teeth were small, spaced and conical shaped with deep overbite. Salivary secretions were also reduced and the patient had never experienced the normal salivary flow. Oral hygiene was well-maintained but alveolar processes showed marked undercuts, prominent tuberosities and enlarged tongue (Figure 1).

Radiological examination revealed retained deciduous teeth. Roots were conical shaped with compromised periodontal support. Lower left premolar was endodontically treated previously and showed a periapical radiolucent area (Figure 2). The routine laboratory examinations were normal except that serum alkaline phosphatase level was raised.

After history and examination, patient was given a detailed overview of the condition and all treatment options were discussed. Finally, a removable conventional complete overdenture for the upper arch and individual separate crowns for lower standing teeth were finalized after discussion with his parents.

Study cast was obtained, surveyed and articulated to make-up the final plan. Upper standing teeth were modified and prepared for metal copings (Figure 3).

Lower teeth were prepared for individual crowns. Final impressions were recorded and lower porcelain fused to metal crowns were fabricated in a conventional way.

The upper and lower prosthesis were tried in and finally made available for insertion. The prostheses were inserted and the patient was instructed about their use and maintenance (Figure 4).

Regular follow-ups were planned every week and minor adjustments were done whenever needed.

The patient is now satisfied and comfortable with his aesthetics. He has adult looking appearance and can chew properly with his prostheses. He showed reservations concerning his upper dentures earlier but now has coped with it.

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**DISCUSSION**

HED and hypodontia, though not a life-threatening condition, can have great impact on the physical, intellectual and psychological maturation of the patient. Good communication between health professionals’ interdisciplinary team (Paediatric Dentists, Orthodontists and Prosthodontists) and also with patient/parents/care-takers is crucial for successful outcome for these young patients.

Quality of life in the present context depends both on the inherent severity of a genetic disorder and also social resources available for a person to cope with that abnormality.

Aims and objective of the treatment modalities are improvement of aesthetics, phonetics, function and mastication, which improve the tone of facial and masticatory muscles to compensate for the reduced vertical dimension.

Due to absence of a number of teeth, there was lack of sufficient bone for reliable implant placement. This might be due to localized or generalized decrease of growth stimuli of the jaw bone. Such bone defects can be rectified by augmentation procedures.

Extensive prosthodontic treatment in growing individuals should preferably be performed with a multidisciplinary team approach. In treatments that are planned and performed over long periods, this approach has the advantages of continuity and shared responsibilities for therapeutic decisions.

Growing patients with congenitally missing teeth often need dental implants, even before puberty, for optimum functional and psychosocial development. From a developmental perspective, dental implants can not accompany the physiologic differentiation of the alveolar bone because of difference in anchorage between osseo integrated implant and a tooth in bone.

Dental implants can be used successfully in partially and completely edentulous arches affected by congenital disorders. Joint orthodontic/restorative diagnostic clinics provide the ideal basis for successful treatment and should be considered the most appropriate mechanism for providing patients with high quality care.7
Individuals with congenital craniofacial anomalies report greater dissatisfaction with their facial appearance, lower self-esteem and quality of life. These dental genetic disorders are not associated with significant mortality; however, there is significant morbidity. The effect on individuals and families should not be underestimated.

Because early intervention and sustained treatment are required if a patient with ED is to receive optimal dental treatment, the costs incurred by these families are a considerable financial burden and play a vital role in planning the treatment design.8

Early consideration of the likely final restoration and the maintenance of the appearance and function are areas of primary concern in the management of hypodontia. Necessary restorative treatment will reflect the decision to accept, close or redistribute spacing resulting from the absence of the teeth.

It is important to realize that the treatment aims should be realistic without overburdening the patient, with excessively protracted treatment time fraught with iatrogenic complications.

Young people with hypodontia need early referral to a hypodontia team for optimal management. Specialist paediatric dental care is essential to ensure retention of the reduced number of teeth. Optimization of the spaces orthodontically combined with composite additions, resin retained bridges, veneers, onlays and tooth transplants contribute to an improvement in aesthetics and functions.

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REFERENCES