CASE REPORT

Dental prosthetic rehabilitation of a child with ectodermal dysplasia: A case report

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Abstract

Ectodermal dysplasias (EDs) form a part of the complex group of rare congenital diseases, primarily observed as a result of the abnormal development of embryonic ectodermal derivatives. A 5-year-old male child reported to the Department of Pedodontics and Preventive Dentistry, with the typical features of hypohidrotic ectodermal dysplasia (HED), i.e., diffusely sparse hair, eyelashes and eyebrows, subsequent problems with thermoregulation and dry skin. The facial profile showed a prominent forehead, sunken nasal bridge (“saddle nose”), and everted lips. Oral prosthetic rehabilitation was planned for the child to improve both the vertical and sagittal skeletal relationship, additionally providing better esthetics, improved speech, and masticatory function. The case report thus presents the early prosthetic oral rehabilitation of a young boy with HED associated with severe anodontia in the primary dentition.

Keywords: Anodontia, ectodermal dysplasia, male child, primary dentition, prosthetic rehabilitation

Introduction

Ectodermal dysplasia (ED) is one among those large group of conditions resulting from the failure of development of two or more ectodermally-derived anatomical structures.[1] ED may be inherited in any of the genetic patterns, i.e., autosomal-dominant (AD), autosomal-recessive (AR), or even X-linked variant.[2] Although numerous subtypes of ED exists, a low estimated incidence of 1 in 1 lakh population has been reported.[3,4] Two major groups of ED have been distinguished based on the involvement of sweat glands. One among them is the hypohidrotic or anhidrotic variant (also termed as Christ-Siemens-Touraine syndrome), where the number of sweat glands is either reduced significantly or absent. The other variant is hidrotic form of ED (also called as Clouston syndrome), where the sweat glands are relatively normal. Hypohidrotic ED (HED) is the most common type, and it seems to be exhibiting the X-linked inheritance pattern; thus, the males showing more susceptibility compared to females. Hidrotic type of ED is inherited as an AD pattern.[5]

Those affected with ED typically show hypoplasia or aplasia of skin, hair, nails, nerve cells, sweat glands, parts of the eye, ear and most relevant to dentists, i.e., the teeth.[1]

Oligodontia or hypodontia is a common feature, and the teeth that are present often manifest abnormal morphology. Incisors often appear tapered and conical while the molars might be dimensionally smaller.[6] Lack of tooth bud formation leads to the hypoplastic alveolar bone, which in turn causes diminished vertical dimension of occlusion giving them a senile or aged appearance.[5]

Hence, an early dental prosthetic rehabilitation (generally recommended from the age of 5 years) in affected children would avert the psychosocial stigma and give them more self-confidence, also helping in facial esthetics, improving their masticatory function and speech process. If child co-operation is not a hurdle, dentures are even recommended as early as 3-4 years of age.[7] A few clinicians may opt for implant therapy, but it is not feasible for young growing children. Non-compliant bone, the implant itself behaving like an ankylosed tooth, problem with occlusion, tilting or drifting of teeth and implants, and cosmetic reasons are some of the unfavorable factors attributed for it.[8,9]

The present study deals with the early dental prosthetic rehabilitation in a male child aged 5 years affected with HED.

Case Report

A boy aged 5 years was referred to the Department of Pedodontics and Preventive Dentistry, KLE Society’s Institute...
of Dental Sciences, Bengaluru, Karnataka, India, presenting
with the absence of teeth as the chief complaint. Parents came
to know about the medical problem of the child when he was
2 weeks old. Diagnosis of ED was confirmed by a pediatrician.
Family history revealed that none in the family were afflicted
with the same condition. Parents had consulted the dentist when
the child was 2 years old, but no treatment could be done as he
was too young to co-operate.

The child suffered from social stigma from his peers at school.
Frequent episodes of fever which would regress only with cold
sponge bath were reported. Less sweating was noted as the cause
for this by the physician as a part of symptoms of ED. The child
also had a difficulty in chewing food, probably because of dry
mouth associated with the low salivary flow.

On extraoral examination, typical features of ED such as dry
and rough skin, sparse fine hair, lack of eyelashes and eyebrows,
hyponyehrosis, hypoplastic mid-face, prominent lips, visible loss
of facial vertical dimension giving the child a senile appearance
were observed [Figure 1].

Intraoral examination showed completely edentulous
mandibular arch, oligodontia in the maxillary arch with only
two deciduous maxillary second molars (55 and 65) clinically
erupted. Decreased vertical bone height, loss of vestibular depth,
and thin alveolar ridge, especially in the mandibular arch, were
noticed [Figure 2].

Panoramic radiograph taken when the child was only 2 years
old [Figure 3] revealed four tooth germs in maxillary arch,
resembling deciduous maxillary molars and deciduous maxillary
central incisors/canines.

Panoramic radiograph taken when the child was 4½ years
[Figure 4] revealed two deciduous maxillary second molars
(55 and 65) at Nolla stage 9 (root completed with open apex).
Two tooth germs resembling deciduous maxillary central
incisors/canines were also observed. No other tooth germs
were noted. No other pathological changes associated with
the alveolar bone/arches were noticed, except that the vertical
height of lower alveolar bone was decreased.

To fabricate removable partial dentures, two impressions
were made, one with hydrocolloid material (alginate) with stock
tray and later using an elastomeric material with custom tray
[Figure 5]. As two maxillary molars were clinically present, it was
decided to fabricate over-dentures in relation to maxillary arch,
and a complete denture was fabricated for the mandibular arch
[Figure 5]. Removable partial dentures were thus delivered to
the child [Figure 6], which significantly improved esthetics and
helped in mastication. Recall visits were scheduled to evaluate
the compliance of the dentures for relining if required and to
assess the growth changes in the developing jaws.

Figure 1: Extraoral view – front profile (before treatment)

Figure 3: Panoramic radiographic image – when child was
2 years old

Figure 2: Intra-oral view – maxillary and mandibular arch

Figure 4: Panoramic radiographic image – when child was
4½ years old
In the particular case, implant therapy was excluded as a treatment option citing the above-mentioned reasons and also because of the insufficient alveolar bone available.

Although removable dentures are commonly advised, growth changes in children may underline the need for frequent repairs/adjustments such as relining of removable partial or complete dentures. Replacement by a new denture is considered when there is a gross discrepancy in patients with reduction in vertical dimension of occlusion and associated abnormal mandibular posture.[13]

Retention and stability of these dentures are another concern, as it is common to witness dry mucosa and under-developed alveolar ridges.[14] Methods to prevent mucosal dryness and wider coverage of the denture base would help to overcome the problems associated with denture retention and stability.

In children suffering from ED, lack or absence of teeth with abnormal craniofacial vertical and sagittal skeletal relationships hampers the esthetics, masticatory ability, speech and temporomandibular joint function.[15] Thus, it is valid to tell that the most widely followed treatment plan of fabricating removable prosthesis can also be regarded as the best available option for our young growing patients, with the added advantage of early age intervention providing long-term psychosocial benefits.

Conclusion

Children affected with ED often present with a compromised dental status. The overall management includes restorative and prosthetic therapy to implant placements, frequently needing multidisciplinary assistance. Pediatric dentists are imposed with a challenge to treat them with a prime concern to delicately handle the psychosocial implications of the child patient as well as to treat them at the earliest so as to minimize further complications.

References

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