

EVALUATION OF KNOWLEDGE AND ATTITUDE REGARDING THE PROSTHETIC MANAGEMENT OF ECTODERMAL DYSPLASIA PATIENTS AMONG THE POST GRADUATE STUDENTS IN CHENNAI

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ABSTRACT:

BACKGROUND: Ectodermal dysplasia is a well-recognized syndrome which is characterized by congenital dysplasia of one or more structures derived from embryonic ectoderm. Oral findings are of particular interest since the disease is characterized by hypodontia, oligodontia or anodontia, which can, moreover, affect both the maxilla and mandible and cause delayed eruption, malformed teeth, producing a small, pointed, conical appearance; and resorption or atrophy of the alveolar border, thus complicating the fundamental rehabilitation procedure in these patients. Prosthodontic management of this patient was challenging for all steps right from impression making to use of dentures because of the fragile mucosa, poor alveolar ridges and decreased salivation. Treatment commenced as soon as possible in order to avoid possible resorption and atrophy of the alveolar ridges, and to control the difficulties in eating and speaking. This research articles deals with the recent updates in the prosthetic management of ectodermal dysplasia and review on various related articles.

MATERIALS AND METHOD: A questionnaire based study consisting of 15 questions and was distributed among local population. The sample size was 100. The research was done among the prosthetic postgraduate students in chennai.

RESULT: From the survey we come to know that most of the students were aware of oral and clinical manifestations and the treatment but nearly 70% of the students were not aware of some specific conditions, gene mutations and behaviour management techniques in the treatment of ectodermal dysplasia.

CONCLUSION: It is necessary for a postgraduate students to be aware of prosthetic management of ectodermal dysplasia and some specific conditions so that they will be able to diagnose and treat patients with rare and idiopathic condition better.

KEYWORDS: Hypodontia, anodontia, oligodontia, rehabilitation, salaivation

INTRODUCTION

Hypohidrotic ectodermal dysplasia is a hereditary disorder of two or more ectodermal structures. It may include hypodontia, hypotrichosis and hypohidrosis.[1] Ectodermal dysplasia (ED) is a large group of heterogeneous heritable conditions characterized by congenital defects of two or more ectodermal structures and their appendages: hair (hypotrichosis, partial or total alopecia), nails (dystrophic, hypertrophic, abnormally keratinized), teeth (enamel defect or absent) and sweat glands (hypoplastic or aplastic)[2,3] There is hypoplasia or aplasia of structures such as skin, hair, nails, teeth, nerve cells, sweat glands, parts of the eye, ear and other organs. [1] Ectodermal dysplasia can be inherited in any form of several genetic patterns including autosomal-dominant, autosomal-recessive, and X-linked modes.[4,5] These are a heterogeneous group of disorders characterized by developmental dystrophies of ectodermal structures, which affects the development of keratinocytes and cause aberrations in the hair, sebaceous glands, eccrine and apocrine glands, nails, teeth, lenses and conjunctiva of the eyes, anterior pituitary gland, nipples and the ears.[6]

GENE MUTATION

The EDA, EDAR and EDARADD genes provide instructions for making proteins that work together during embryonic development. These proteins form part of a signaling pathway that is critical for the interaction between two cell layers, the ectoderm and the mesoderm. It is essential for the formation of several structures that arises from ectoderm, including the skin, hair, nails teeth and sweat glands. Mutation in these genes prevents normal interaction between the ectoderm and mesoderm and impairs the normal development of hair, sweat glands and the teeth.

CLASSIFICATION

Clinically there are two major types of ED namely hidrotic and anhidrotic (hypohidrotic form). The hidrotic form, inherited as an autosomal trait, affects teeth, hair and nails but usually spares the sweat glands and was first described by Clouston in 1929.[7] Whereas the hypohidrotic form (Christ-Siemens-Touraine Syndrome) is most common type seems to be an X-linked recessive trait, with an incidence of this syndrome estimated to be 1 to 7 per 10,000 live births.[8,9] Recently, a new classification for ED has been proposed, based on the molecular genetic data by Priolo who divided the ED into two groups. The first group includes disorder in which a defect in the epithelial-mesenchymal interaction and the second group involved defect in cell-cell adhesion and communication.[10]

CLINICAL MANIFESTATIONS

The skin covering majority of the body may be abnormally thin, dry and soft. Fine linear wrinkles and increased pigmentation are often present around the eyes, appearing prematurely aged. Scalp hair, eyebrows, eyelashes, and other body hair may be sparse, poorly developed, or absent. There may be hyperkeratosis of the palms of the hands and soles of the feet. Fingernails and toenails also may show faulty development. Other reported clinical manifestations are frontal bossing; sunken cheeks, prominent supraorbital ridges, a saddle nasal bridge, large low set ears and hypoplastic-appearing alaeque nasi. In addition, the midface is depressed and hypoplastic; the cheek bones are high and broad (malar hypoplasia) and appear flat and depressed, thick everted lips and or a large chin.[11,12,13] Typical general mental development, frontal bossing with characteristics reduction in amount of hair (hypotrichosis), absence of sweat glands (anhidrosis) resulting in temperature elevation, absence of sebaceous glands (asteatosis) resulting in dry skin, depressed nasal bridge, protuberant lips, prominent supra orbital ridges, sunken cheeks, wrinkled hyperpigmented skin around the eyes and large low set ears[14]

ORAL MANIFESTATIONS

Orofacial characteristics of this syndrome include anodontia or hypodontia, hypoplastic conical teeth, underdevelopment of the alveolar ridges, frontal bossing, a depressed nasal bridge, protuberant lips, and hypotrichosis. It can affect both the primary and permanent dentition.[15] There is presence of anodontia, Hypodontia, malformed and widely spaced peg-like teeth, protuberant lips, loss of occlusal vertical dimension, and underdeveloped alveolar ridge. With little or no dental support, a hypoplastic maxilla and mandible result in bite collapse and narrowing of the alveolar ridges.[16,17] In rare instances, one or both jaws may be edentulous and the alveolar processes may not develop due to the absence of teeth.[18] Our case presented with missing teeth more in the mandible than in the maxilla. Most common congenitally missing teeth include third molars and maxillary lateral incisors, which are also seen associated with our case with inclusion of even the second molars. Complete anodontia of both deciduous and permanent dentition is rarely reported.[19] Significant differences in the number of primary missing teeth have been detected between X-linked HED and autosomal-recessive HED in recent investigations.[20]

TREATMENT

Treatment generally includes

- Removable prosthodontics
- Fixed prosthodontics
- Implant prosthodontics

In cases where there is associated cleft lip and palate the treatment may consist of intervention by a plastic surgeon and an oral and maxillofacial surgeon. In such cases a maxillofacial prosthesis may be indicated.

Removable prosthodontics

Removable prosthesis is the most frequently reported treatment modality for the dental management of ED. [21,22,23] In childhood a removable partial denture (RPD) or complete denture, overdenture is often the treatment of choice because of the need to easily modify the intraoral prosthesis during rapid growth periods. These treatment options afford the ED patient and his or her family an easy, affordable, and reversible method of oral rehabilitation. Cooperation of the patient as well as the support of the family is necessary if removable prosthesis is to be successful in young patients. [24] Although complete dentures are an acceptable form of treatment, overdenture or RPD supported by natural teeth are desirable for preservation of alveolar bone. The advantages of preserving the natural teeth are (i) Preservation of alveolar bone, (ii) Preservation of proprioception, (iii) Improved retention (iv) Improved support, and above all (v) Less psychological trauma of losing natural teeth for patients. [25] Problems like loose dentures, loss of proprioception, and bone resorption can be resolved with overdenture. [26] Patients treated with complete maxillary overdenture and mandibular overdenture demonstrates less vertical alveolar bone reduction than patients with conventional complete maxillary and mandibular dentures. [27] Periodic recall of young ED patients is also important because prosthesis modification or replacement will be needed as a result of continuing growth and development. [28] In addition to adjustments related to fit, the occlusion of prosthesis must be monitored for changes because of jaw growth. Other problems related to removable prosthesis are speech difficulties, dietary limitations, and loss of the prosthesis. [29]

Fixed prosthodontics

Fixed Prosthetic Option: It is seldom used because of the presence of only minimal number of teeth. Patients are quite young & FPD's with rigid connectors should be avoided in young, actively growing patients as rigid FPD's interferes with jaw growth, especially if prosthesis crosses the midline. Hence, Individual crown restorations can be done, but larger pulp sizes and shorter crown heights have to be considered for either doing root canal treatment or the reduction of teeth. In early teens, orthodontic treatment is indicated as consolidation of spaces may better prepare mouth for FPD or implants in future, they fall within this parameters[30]

Implant prosthodontics

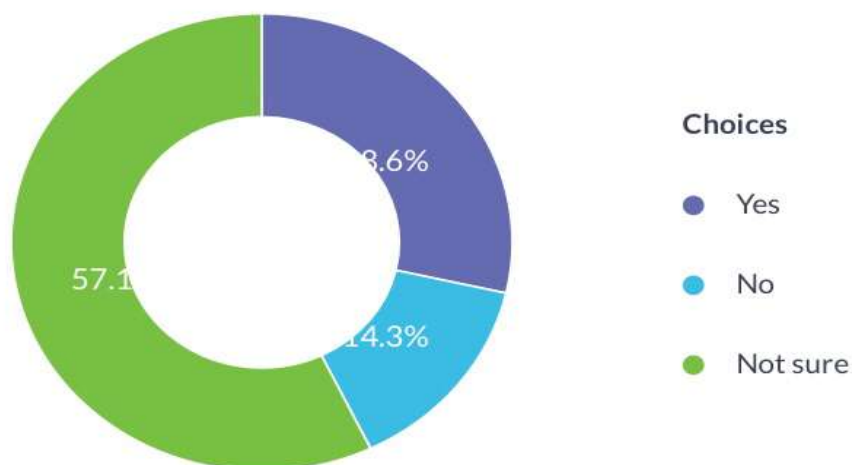
The placement of implants in growing children is not recommended as a routine practice. Experiments designed to study the effect of dental implants on dentoalveolar growth and development in pigs demonstrated that implants, owing to an absence of a periodontal ligament, behave like ankylosed teeth i.e., they remain stationary and do not erupt together with adjacent teeth leading to inhibition of growth and development of the alveolar process. Submergence of an implant is disadvantageous for a number of reasons. First, an infraocclusion occurs, which disrupts carefully constructed occlusal relationships and leads to compensatory eruption of opposing teeth and tipping of adjacent teeth. Second, a vertical discrepancy develops between the mucosal margin of the implant and the gingival margins of adjacent teeth [31].

MATERIALS AND METHODS

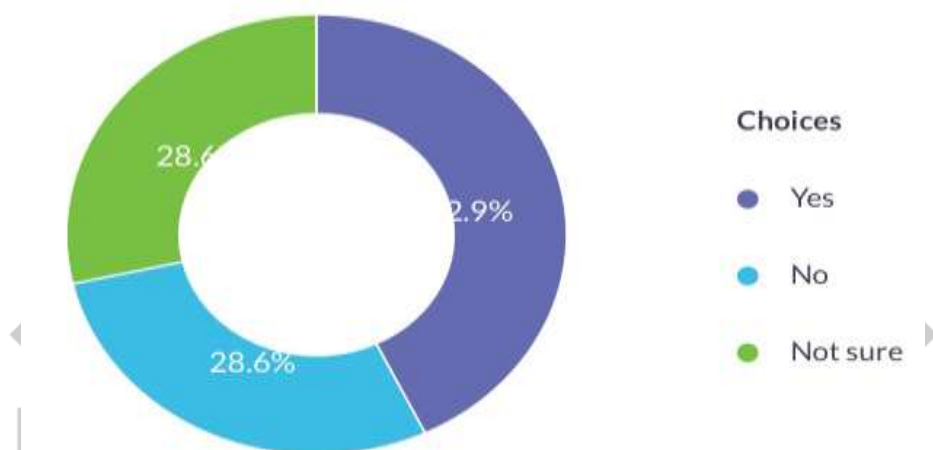
A questionnaire based study consisting of 15 questions and was distributed among local population. The sample size was 100. The research was done among the prosthodontic post graduate students in Chennai. In this survey there were no right or wrong answers and no time limit was given to them. The people were asked to answer only the questions which they know or only the questions which they can understand and the remaining questions were asked to skip. After the completion of questions by the people the responses were interpreted in accordance with the norms.

RESULT

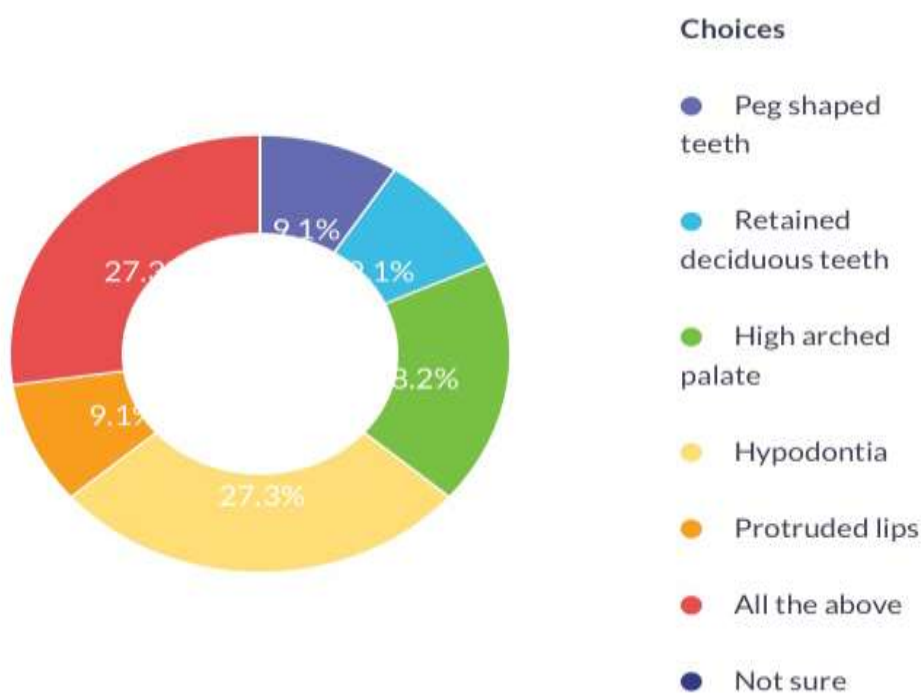
Do you know which gene mutation causes ectodermal dysplasia?



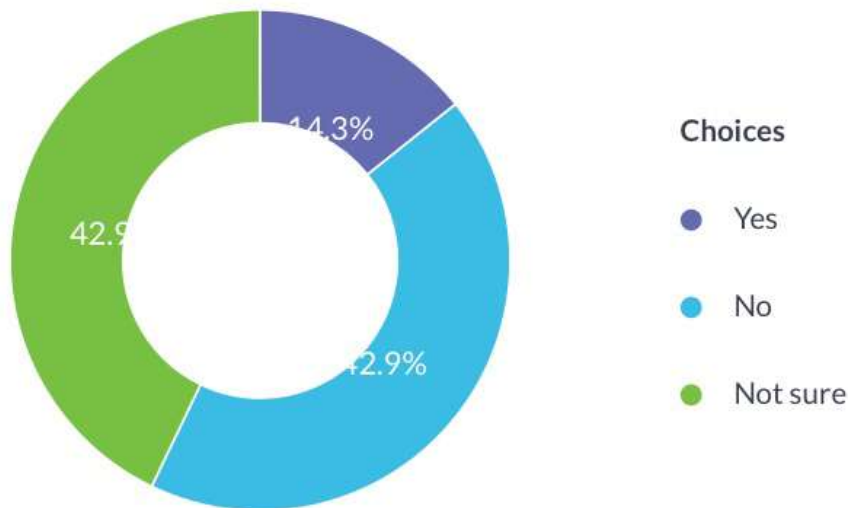
Are you aware of the behavioural management techniques in case of a child with ectodermal dysplasia?



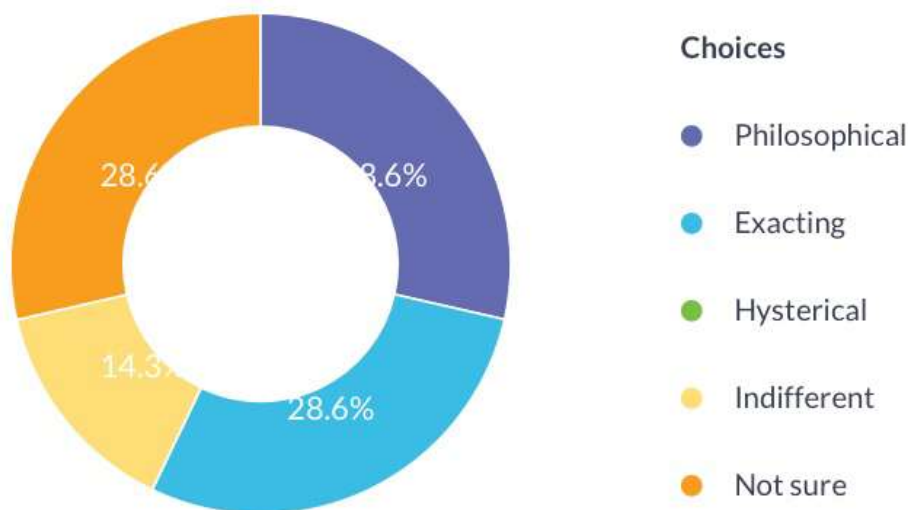
What are the common oral manifestations of ED patients?



Do you know what Freire-maia syndrome is?



If so, what was the mental attitude of the patient towards the treatment?



DISCUSSION

Dental defects associated with ED can cause severe esthetic and functional problems. The treatment objective of this case was to create a more favorable starting point for the prosthodontic phase of rehabilitation by improving the sagittal and vertical skeletal relationships and facial esthetics.[32] Toddlers or young patients afflicted with ED have a marked challenge with the development of speech, poor esthetics, underdeveloped orofacial musculature, poor masticatory function and affected social and psychological development. Prosthetic oral rehabilitation seems highly recommended for these patients as early as 3 years of age to stimulate a normal oral functional development. However, the fit needs to be evaluated frequently and prostheses should be refabricated annually to account for growth. Oral rehabilitation of ED patients with removable prostheses at such a young age can prepare them for more definitive prosthetic treatment once growth is completed. The ability of the young patient to adapt to removable denture prostheses is rapid and the benefits of improved speech, esthetics, self-esteem, psychosocial development and overall function are evident.

Treatment generally includes a removable and /or fixed partial denture, an overdenture, complete denture prosthesis or an implant retained prosthesis. In cases where there is associated cleft lip and palate the treatment may consist of intervention by a plastic surgeon and an oral and maxillofacial surgeon. In such cases a maxillofacial prosthesis may be indicated. In the present case prosthodontic management was done by removable partial denture. The option of complete denture was ruled out to preserve remaining alveolar ridges. Fixed prosthodontic management is seldom used for such type of patients because of minimal number of teeth present and often patient are young in which fixed partial denture with rigid connector will hamper active growth of dental arches. Options which could be considered in our patient were of overdenture after intentional root canal treatment of the existing teeth. But considering the increased number of appointments and disinterest of patient this option was also ruled out. The treatment option preferred was of a removable partial denture considering his present age. Moreover, considering the growth potential of our patient erupting tooth remained as potential overdenture abutment and an option for a fixed partial denture or implants in future.

As the child matures the removable prosthesis needs relining, rebasing or remaking to accommodate growth changes and maintain normal oral functions. When child reaches teenage years, orthodontic treatment may be indicated as better management of spacing may prepare the mouth for a fixed partial denture or implants in future.

In older patients, depending on pattern of missing teeth and the available bone support, osseointegrated implants can be used. If bone support is inadequate then bone grafting may be necessary. Overlay or overdenture may also be indicated as it involves retaining the tooth or tooth root after intentional root canal treatment.

Experiment was conducted to know the percentage of awareness about the prosthetic management of ectodermal dysplasia among the prosthodontic postgraduate students. In this questionnaire questions about gene mutations, classification, behaviour management, oral and clinical manifestations, associated syndromes and treatment plan were also included. From this study we come to a conclusion that more than 90% of dental students were aware about clinical and oral manifestations of the condition and how to treat it and some have treated patients with ectodermal dysplasia during their dental practice. They were also aware of associated syndromes and complications and the behaviour of the children and adult with ED.

CONCLUSION

To summarize, only when the postgraduate students are aware of common features of rage condition and the treatment. But awareness about the specific conditions in ectodermal dysplasia and the knowledge about gene mutations should be created in order to understand the condition better and proceed with better treatment.

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