REVIEW ARTICLE

Prosthodontic Solution for Pediatric Patient with Ectodermal Dysplasia: A Review Article

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Abstract

Background: Ectodermal dysplasia is a category of hereditary disorders characterized by a primary deficiency in the function of the hair, teeth, nails, or sweat glands, as well as another abnormality in any ectodermal tissue. Ears, lips, mouth or nose mucous membranes, and the central nervous system are among them. In ectodermal dysplasia, more than 2 ectodermal structures are deformed and are a related genetic. Past lessons seen from hypohidrotic ectodermal dysplasia act as a model for disclosing the etiology and pathogenesis of other ectodermal dysplasia types by identifying phenotypically similar syndromes can be the result of alteration in various genes that alteration in identical genes could result in different phenotypes and that alteration in the genes downstream the identical signaling pathway which could alter the phenotype significantly. In a letter to Charles Darwin in 1838, Wedderburn described a case of ectodermal dysplasia in 10 Hindu male relatives. Thurnam registered two cases of hypohidrotic type in 1848. In most cases, dentists do not have the thorough knowledge to tackle such cases along with that the inability for the guardians and the child to support the long-term treatment needed for ectodermal dysplasia as the appliances has to keep on changing stands as a blockade in the pathway of a successful treatment. The present analysis focuses on epidemiology, etiology, clinical features, and diagnosis along with different treatment options, on classification, genetic aspects, including clinical manifestation. To restore the esthetic appeal and work of the stomatognathic system, patients also require a multidisciplinary approach for clinical preparation and dental treatment. Follow-ups of the patient with adjustments in their dentures should be made to meet the continuous development occurring in the patient, early dental treatments may enhance the patient’s appearance and psychological problems. Proper communication should be there between the patient and the dentist if the treatment has to be a success. When it comes to children, guardian should actively support the instruction provided by the dentist. Dentist should provide an environment for the child and his/her guardian to convey their problems and an active time allotment should be given to make the treatment successful.

Keywords: Biocon, Fixed prosthodontics, Hypohidrotic ectodermal dysplasia, Over dentures, Removable prosthodontics, Short implant

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Ectodermal Dysplasia

Ectodermal dysplasia is a category of hereditary disorders characterized by a primary deficiency in the function of the hair, teeth, nails, or sweat glands, as well as another abnormality in any ectodermal tissue. Ears, lips, mouth or nose mucous membranes, and the central nervous system are among them. In ectodermal dysplasia, more than 2 ectoderm structures are deformed and are a related genetic. Past lessons seen from hypohidrotic ectodermal dysplasia act as a model for disclosing the etiology and pathogenesis of other ectodermal dysplasia types by identifying phenotypically similar syndromes can be the result of alteration in various genes that alteration in identical genes could result in different phenotypes and that alteration in the genes downstream the identical signaling pathway which could alter the phenotype significantly. In a letter to Charles Darwin in 1838, Wedderburn described a case of ectodermal dysplasia in 10 Hindu male relatives. Thurnam registered two cases of hypohidrotic type in 1848. In most cases, dentists do not have the thorough knowledge to tackle such cases along with that the inability for the guardians and the child to support the long-term treatment needed for ectodermal dysplasia as the appliances has to keep on changing stands as a blockade in the pathway of a successful treatment. The present analysis focuses on epidemiology, etiology, clinical features, and diagnosis along with different treatment options, on classification, genetic aspects, including clinical manifestation. To restore the esthetic appeal and work of the stomatognathic system, patients also require a multidisciplinary approach for clinical preparation and dental treatment. Follow-ups of the patient with adjustments in their dentures should be made to meet the continuous development occurring in the patient, early dental treatments may enhance the patient’s appearance and psychological problems. Proper communication should be there between the patient and the dentist if the treatment has to be a success. When it comes to children, guardian should actively support the instruction provided by the dentist. Dentist should provide an environment for the child and his/her guardian to convey their problems and an active time allotment should be given to make the treatment successful.
Emmanuel, et al. Prosthodontic solution for pediatric patient with ectodermal dysplasia

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Epidemiology and Etiology

Ectodermal dysplasia is transmitted as an X-linked recessive trait where the female acts as a carrier and male is affected. Prevalence in the population has been found to be between 1:10,000 and 1:100,000 male live births. Mutations in EDA, EDAR, and EDARADD genes have found to be the reason behind the occurrence of ectodermal dysplasia.

Classification

It is of two types
- Hidrotic/Clouston syndrome: Hair, skin, and nails are affected, while the teeth as well as sweat glands remain unaffected
- Hypohidrotic/anhidrotic: Rare genetic disorder characterized by decreased sweating capacity, missing tooth, hypodontia, and fine sparse hair (hypotrichosis).

Clinical Manifestation

- Pyrexia of unknown cause
- Patient is not able to sweat
- Heat intolerance
- Normal meal or a normal work could lead to hyperpyrexia
- Seizures
- Other neurological sequelae
- Pharyngitis
- Rhinitis
- Cheilitis
- Dysphagia
- Decreased immunity
- Wheezing and breathlessness
- Skin is dry, pigmented and wrinkles could also be seen
- Frontal bossing
- Prominent supraorbital ridges
- Saddle nose.

Dental Abnormalities

The teeth most commonly present are upper canines, upper central incisors, and upper 1st molar (conical shape, malformed, and widely spaced).

Diagnosis

Diagnosis is mainly based on the dental anomalies and clinical manifestations. Dermatoglyphic analysis, lacrimal secretion type, and hair distribution on the scalp also act as a diagnostic criteria. Genetic testing is available through GeneDx.

Treatment

Removable prosthodontics

The key care options for the management of ectodermal dysplasia in children having missing or lack of tooth are prosthetic rehabilitation using removable dentures and complete dentures. In total anodontia, therapy will involve either traditional or implant-assisted complete denture. Removable partial dentures and overdentures can be considered in patients with partial anodontia. Residual ridges are not developed properly and vertical occlusal height is also lost in this cases. The patient’s full denture prosthesis improves the patients profile by changing the occlusal heights and substantially enhances the role of mastication, esthetics, phonetics, and psychological support. In any case, guardian must be mindful that the child will reject the procedures, some clinicians have recorded one denture at a time. It has been noted that the 2nd denture should be provided at a gap of 2–4 months from the delivery of 1st. It has been noted that 1st denture has good prognosis. Its seen 3 years child who could also have denture, according to Kupietzky and Houpt.

Disadvantages

- Periodic transition due to shifts in development and challenges in having an ideal integrity
- Frequent adaptation is needed, due to development
- Minimum of three replacements is needed in each dentition period.

Advantages

- Without dentures, the mandible anterorelation forces the jaw to shift up and forward and there is a reduction in the lower facial height leading to Class 3 malocclusion
- Dentures cause the mandible to rotate downward with the consequent natural location of the chin in space.

Fixed Prosthodontics

It is not widely used in the treatment of ectodermal dysplasia since, when treated, many patients are often very young. In young, actively growing patients, fixed partial dentures with rigid connectors should be discouraged as it restricts the growth of jaw. Crowns could be beneficial but in primary tooth, large pulp chamber and small crown size are a concern. Suri et al. explains that dentofacial growth pattern can be improved along with aligning of tooth with the use of fixed and functional appliances which significantly enhanced sagittal and vertical dentofacial relationship and facial esthetic.
Implant Prosthodontics

Studies investigated how dental implants affected the development and dentoalveolar growth in pigs shown that dental implant function like ankylosed teeth due to the lack of a periodontal ligament, which could inhibit the dentoalveolar growth if given at the growth period. No standard procedure is recommended for the placement of implants in developing children.

Kearns et al.\cite{Kearns} stated that placing implant in the anterior mandible is not an issue as the symphyseal suture of mandible ceases its growth in the first 6 months of life. The installation of the fixed implant crossing the midpalatal sutures would result in a limitation of the maxilla’s transverse growth in the growing maxilla until early adulthood. Sweeney et al.\cite{Sweeney} suggested that 18 years are the ideal age for the placement of implants.

Biocon

Minidental implants are an alternative to conventional dental implants, but are commonly used in cases where there is less room and it is possible to position a small implant. The head part looks very much like a ball that aids in the retention mechanism. Advantages of Biocon are as follows:

- Biocompatible
- Minimizes the cost for treatment
- Easier cleansing
- No cutting or suture required
- Simple
- Single appointment
- Superior technically.

Conclusion

To restore the esthetic appeal and work of the stomatognathic system, patients also require a multidisciplinary approach for clinical preparation and dental treatment. Follow-ups of the patient with adjustments in their dentures should be made to meet the continuous development occurring in the patient, early dental treatments may enhance the patient’s appearance and psychological problems.

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