



Parameters of Oral Health Care for Individuals **Affected by Ectodermal Dysplasias**

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NATIONAL FOUNDATION FOR
ECTODERMAL DYSPLASIAS

DEDICATION

This booklet is dedicated to members of the dental profession who provide beautiful smiles while giving of their time and talents.

OUR MISSION

Our mission is to empower and connect people touched by ectodermal dysplasias through education, support and research. For more information, visit us online at www.nfed.org.

INTRODUCTION

The ectodermal dysplasias are a group of inherited disorders that involve defects of the hair, nails, teeth, and sweat glands. However, the ectodermal dysplasias are a remarkably diverse group of human disorders, so other parts of the body may be affected. The types of ectodermal dysplasias are recognized by the combination of physical features that an affected person has and the way in which they are inherited. More than 100 types of ectodermal dysplasias were first described in a book by Drs. Freire-Maia and Pinheiro from Brazil (*Ectodermal Dysplasias: A Clinical and Genetic Study*. New York, AR Liss, 1984). There are now more than 180 types of ectodermal dysplasias recognized. (Figure 1)



Figure 1: These children are affected by four different ectodermal dysplasias.

SPECIAL NOTE

This document provides guidelines for health care professionals to manage the oral health problems of individuals affected by the ectodermal dysplasias. While these guidelines discuss treatment options for affected individuals, clinical circumstances and individual preferences must be considered to determine an optimal plan for treatment. The guidelines are not intended to establish the standard of care, but to aid dentists and affected individuals in making informed choices about treatment. Other interested parties, including third parties responsible for payment, may find these guidelines useful.

This document discusses, in several sections, the importance of consulting a prosthodontist to oversee comprehensive treatment plans. While this is certainly appropriate, it should be noted that there are a relatively small number of prosthodontists available in many areas. It is recommended that a prosthodontist be consulted, but if there are none available, then it is advisable to see a dentist who has experience in complex and implant prosthodontics.

The mention of any “brand name” drug or product in this booklet is presented as an example only and must not be interpreted as endorsement of a specific product or brand by the National Foundation for Ectodermal Dysplasias (NFED) or its Scientific Advisory Council.

MORE INFORMATION

Medical and dental treatment for people affected by ectodermal dysplasias may be quite complicated. Dental treatment is particularly important since it is critical for a normal diet, facial appearance, speech, and emotional development. This booklet provides helpful information for anyone involved in decisions about dental care for people affected by ectodermal dysplasias.

For more information on dental care associated with the ectodermal dysplasias, contact the NFED (NFED, www.nfed.org, 618-566-2020) to receive an order form for relevant medical articles.

DEFINITION OF ECTODERMAL DYSPLASIA

The ectoderm is one of the three germinal cell layers that form the early embryo. It eventually develops into the epidermis (surface skin), nails, hair, tooth enamel, sweat glands, sebaceous glands, and nerves, among other things. Any tissue, including the derivatives of the ectoderm mentioned above, may form abnormally; that is, any tissue may be *dysplastic*. The phrase *ectodermal dysplasia*, then, simply implies that a derivative of the ectoderm has not properly formed (Toumba & Gutteridge, 1995).

An abnormal derivative of the ectoderm may be the only dysplastic tissue in a given individual, or other parts of the body may function abnormally at the same time. The terminology used when only ectodermal tissue is abnormal, and only one of the many derivatives of ectoderm is involved, is straightforward. The term *trichodysplasia* signifies that the hair is inherently abnormal; the term *onychodysplasia* suggests that the nails are inherently abnormal and the term *enamel dysplasia* is used to signify that tooth enamel is inherently abnormal. Any one of these isolated abnormalities could be called an ectodermal dysplasia; however, this latter term has been used historically for syndromes in which multiple derivatives of the ectoderm, that is, more than one tissue or organ, are affected (Kpietzký & Houpt, 1995).

When more than one derivative of the ectoderm is affected, the best approach is to append the word *syndrome* to *ectodermal dysplasia*; *syndrome* means a pattern of signs and symptoms that occur together as part of an abnormal

developmental process. There are ectodermal dysplasias such as the tooth and nail syndrome, the trichodental syndrome, and the hypohidrotic ectodermal dysplasia (HED). Since only ectodermal derivatives are involved in these syndromes, they may be viewed as pure ectodermal dysplasia syndromes.

The terminology is more complex when derivatives of the ectoderm and derivatives of other tissue are affected at the same time. For instance, a syndrome exists in which the hair, teeth, and sweat glands are abnormal, but the individual also has orofacial clefting (ectrodactyly-ectodermal dysplasia-clefting [EEC] syndrome); and another, in which the hair and teeth are abnormal, but there are also unusual dense areas of bone that appear on an x-ray (tricho-dento-osseous [TDO syndrome]). (Figure 2)



Figure 2: A pre-teen affected by ectrodactyly-ectodermal dysplasia-clefting (EEC) syndrome

These syndromes cannot be called “pure” ectodermal dysplasias, because the bone is derived from the embryonic mesoderm. A better approach might be to call these latter syndromes “complex ectodermal dysplasias”. Finally, in some disorders, the ectodermal derivatives are only occasionally or indirectly affected. These disorders are difficult to classify as ectodermal dysplasias, but they are clearly related and must be considered in differential diagnosis.

Differential diagnosis is a key prerequisite to planning treatment for individuals affected by ectodermal dysplasia and

related disorders. The cause of the observed anomaly and its potential for associated health problems must be addressed before deciding upon a course of action. If an individual has

several missing teeth, for example, one should know whether their absence represents a dysplasia (hypodontia), or is the dental caries (tooth decay) or periodontal (gum) disease a consequence of advanced periodontal (gum) disease. When treatment is planned, the impact of associated health problems must be anticipated. It is also imperative to know if other health issues exist, such as skin erosion or immune deficiencies, which will complicate treatment (Kpietzy & Houpt, 1995). (Figure 3)



Figure 3: A girl affected by ankyloblepharon-ectodermal dysplasia-clefting (AEC) syndrome who has skin erosion.

Although diagnosis is the first step in planning intervention, the broader issues of accurate diagnosis, the possibilities of recurrence, alternatives for intervention or prevention (including genetic counseling and alternative family planning), and possible dental and medical complications should be attended first. Furthermore, the discussions here address only those ectodermal dysplasia syndromes with oral manifestations.

ORAL MANIFESTATIONS OF ECTODERMAL DYSPLASIAS

The many oral manifestations of ectodermal dysplasia may be divided into several categories: numeric, structural, morphological, compositional, and/ or positional abnormalities of the teeth, and other manifestations (Guckes, Roberts, & McCarthy, 1998, Pirinen, 1998).

Abnormalities of number of teeth in ectodermal dysplasias are limited to various degrees of hypodontia (a reduced number of teeth), while hyperdontia (an excessive number of teeth) is not common in the ectodermal dysplasias (Bergendal, Koch, & Kurol, 1998). Hypodontia, best defined as an “inherent absence of one or more teeth”, may be mild, severe, or complete (that is anodontia, the complete absence of teeth). (Figure 4) The pattern of hypodontia is syndrome-specific; that is, within a specific syndrome, the teeth involved are usually the same in one affected individual as in another with the same disorder. There still can be variations within a syndrome.

Since the enamel is the part of the tooth derived from the ectoderm, enamel dysplasias may be expected in the ectodermal

Figure 4: Four pictures demonstrating different degrees of hypodontia in individuals affected by ectodermal dysplasias



dysplasias. The enamel may be hypoplastic (thinner than average or pitted), hypocalcified (poorly mineralized), or hypomature (insufficiently hardened). In some affected individuals, more than one type of dysplastic enamel may coexist in the same person. Dentin dysplasia and pulpal dysplasia are not usually components of ectodermal dysplasias.

Deterioration of the enamel caused by dental caries (tooth decay) and erosion or discoloration of the enamel caused by external agents are not dysplasias and thus not considered integral components of ectodermal dysplasias. Some kinds of enamel dysplasia may, however, predispose to caries or discoloration. In some forms of ectodermal dysplasia, the saliva production may be reduced leading to dry-mouth (xerostomia), which may put the individual at increased risk for tooth decay. It is recommended to have the dentist carefully check the teeth and apply preventive treatments (e.g., fluoride varnish or other caries prevention techniques) as needed.

Morphological abnormalities of teeth in the ectodermal dysplasias may involve the clinical crowns or the roots. The clinical crowns of posterior teeth may be globe-shaped and lack normal occlusal morphology, (otodontal dysplasia) or the clinical crowns may be smaller than average (microdontia). Teeth that are smaller than average often are described as being conical, tapered, or pegged

in shape. There is a positive correlation between hypodontia and microdontia; that is, when some teeth are inherently absent, the teeth present in that individual may be smaller than average.

Teeth affected by enamel hypoplasia are often smaller than average and/or conical because their enamel layer is thin, depriving the teeth of their usual bulky contours. The pulp chambers of teeth in some ectodermal dysplasias are larger than average, giving the roots an abnormal shape (taurodontia). The pulp chambers are affected because Hertwig's epithelial root sheath, an ectodermal derivative, determines the level of the pulpal floor and subsequently the site of root bifurcation.

Delayed eruption of teeth, abnormal patterns of eruption of teeth, abnormal positions of teeth, and malocclusion of the jaws may occur in ectodermal dysplasias. When teeth are absent, the alveolar ridges are often underdeveloped. **Development of the alveolar bone accompanies tooth bud formation, so without tooth bud formation, alveolar bone formation is dysplastic or abnormal.** Other oral manifestations of the ectodermal dysplasias include abnormal speech, difficulty with chewing, and swallowing.

FACTORS INFLUENCING DENTAL CARE

Individuals affected by the ectodermal dysplasias have several difficulties in finding comprehensive treatment for their disorders.

Resources for treatment are limited. Few dentists are familiar with the ectodermal dysplasias and with treatment options for affected individuals. The geographic distribution of dentists familiar with the syndromes is uneven. Furthermore, formal referral networks among the dentists trained to provide the necessary treatment are rare.

Access to comprehensive care is also complicated by socio-economic factors. Affected individuals simply may lack the resources to pay for treatment. The extensive and comprehensive care required is quite expensive, and many families do not have the financial resources or insurance coverage necessary to cover the costs. Some public assistance programs help with payment and the NFED has a Treatment Assistance Program. Some families may not qualify or the assistance provided may not be sufficient for the level of care and restoration needed. For some individuals, travel expenses to centers that provide treatment are prohibitively high or limited access to childcare for other members of the family makes travel impossible.

Cultural factors also influence the access to and availability of care. Families may not be motivated to seek adequate care and/or may have unrealistic outlooks about their obligations for compliance and cost sharing. The success of treatment for some affected individuals may be hampered by their immaturity, behavioral problems, emotions, or physical disabilities.

A risk assessment is necessary for individuals affected by ectodermal dysplasias prior to planning definitive dental treatment. The individual's general health should receive careful consideration, since some ectodermal dysplasias have associated immune deficiencies, recurrent infections, or other conditions that may alter the selection of certain treatment options. Degenerative oral disease, whether coincidental or part of ectodermal dysplasia, must also be considered. If there are no contraindications, a full spectrum of age-appropriate treatment options is available.

TREATMENT GUIDELINES

The goal of dental treatment for individuals affected by the ectodermal dysplasias is to provide an age-appropriate dentition that optimizes chewing function (and thus nutrition), oral/facial development, speech, swallowing, and esthetics. Comprehensive treatment also aims to enhance physical, emotional and psychosocial development for affected individuals. Since the dental manifestations of the ectodermal dysplasias persist throughout life, dentists must anticipate working closely with children, adolescents, and adults. (Nowak, 1988) If treatment for cleft lip and palate is necessary, guidance should be sought from the American Cleft Palate-Craniofacial Association for parameters of care (<http://www.cleftpalate-craniofacial.org/>).

Recommendations about treatment are discussed in this document for specific age categories: preschool (ages 0-6), school age children (ages 7-12), adolescents (13 - skeletal maturity), and adults. In all age groups, all the oral structures should be evaluated for their effects on treatment. Also, risk factors such as oral hygiene and diet that are related to caries (tooth decay) and periodontal disease (gum disease) should be evaluated. After treatment, age appropriate programs for prevention of dental disease should be followed.

In order for the treatment plan to be successful, individuals affected by ectodermal dysplasias should have their dental prosthetic cleaned and maintained as recommended by the provider. In the case, with a fixed implant prosthesis, the cleaning is not “routine” but rather a complex and lengthy process involving provider time and skill. These cleanings are medically necessary to maintain the health and integrity of the prosthesis.

TREATMENT OPTIONS FOR PRESCHOOL CHILDREN (0-6 YEARS)

The American Academy of Pediatric Dentistry (AAPD) recommends that children visit a dentist at age one year or six months after the eruption of the first teeth, whichever comes first, to establish a Dental Home (Nowak & Casamassimo, 2000). **It is recommended that, if a child doesn't have erupted teeth by age one year old, then that child should be evaluated for ectodermal dysplasia.**

This recommendation is also reasonable for children affected by ectodermal dysplasias. If diagnosis is made earlier, the first visit to a dentist should occur at the time of diagnosis. Furthermore, children whose family history places them at risk to be affected should visit a dentist in the first few months of life. The goal of intervention for this age group is to develop a treatment plan and restore as normal a dentition as possible early in life. This is important to facilitate speech, enhance general health through good nutrition, and to improve appearance.

The first visit to a dentist for children affected by or at risk for ectodermal dysplasias should include visual inspection of the oral cavity, digital probing of the dental arches and palate, and appropriate radiographs. Wherever possible, the number of teeth present, erupted or unerupted, should be determined, the morphology of the teeth should be documented in notes or with photographs, and the character of the enamel should be determined. If an adequate number of teeth are erupted, inter-arch occlusion and alignment of the teeth within an arch should be assessed. The integrity of the palate and the amount and consistency of the saliva should be evaluated as well. Appropriate home and professional oral health preventative programs should be initiated and monitored.

In the absence of overt abnormalities of the dentition, age-appropriate dental care and anticipatory guidance following the guidelines of the AAPD, should be provided (www.aapd.org). A program for recall visits should be **instituted** since it is important to retain the dentition for future prosthetic support.

- For children with hypodontia, removable or non-rigid fixed prostheses may be considered, including complete dentures or “over-dentures” (Toumba & Gutteridge, 1995). (Figure 5)



Figure 5: A toddler boy with and without his dentures

- Dentures must be size and age appropriate.
- Provisional non-rigid fixed or removable prostheses may be appropriate.
- Implant prostheses are **not** recommended for children in this age group.
- Any rigid fixed prosthesis is **not** recommended in this age group because it may interfere with craniofacial growth and alignment of the dental arches.

If a removable prosthesis is used, it will be necessary to relines, adapt or remake it periodically to accommodate craniofacial growth and to prevent irritation/inflammation of the underlying soft tissues (Unger, Crabtree, & Meyer, 1990). A survey by the NFED revealed that prostheses, on average, need replacement every 2.5-3.5 years depending on the child's growth patterns. (Figure 6)

- Minor tooth movement, root canal therapy, and selective extraction of teeth may be necessary to allow successful prosthetic treatment.



Figure 6: Dentures made for a child

Remaining teeth used for denture retention are at risk for decalcification leading to caries (decay) and possible loss. To protect these “abutment” teeth, a strict regimen of daily plaque removal, restricted intake of cariogenic dietary substances and optimal topical fluoride exposure (e.g. topical fluorides, such as varnish or gel) must be followed. If decalcification or caries (decay) is noted, full coverage should be considered. With primary teeth a stainless steel crown can be placed. With permanent teeth, a cast full-coverage crown may be appropriate. (Figure 7)

Figure 7: Stainless steel crowns



For children who have enamel dysplasia, esthetic “bonded” restorations or other full crown coverage restorations may be indicated. If removal of enamel is necessary to modify abnormally shaped teeth, bonded veneers or full crown coverage should be used.

Children with HED often have conical shaped incisors that are very pointed and can be extremely sharp. This can cause problems with biting of the lip or lead to other soft tissue trauma. Ideally, the management of this problem involves bonding of the conical shaped teeth to provide a more normal incisor shape. Bonding will make the tooth both less likely to cause soft tissue trauma and be more esthetically normal. When appropriate, bonding is preferred over grinding the pointed teeth.

Reshaping the incisal edge of a pointed tooth is recommended as a last resort only if bonding to reshape the teeth to a morphology is not possible or a reduction in tooth height is indicated for prosthesis fabrication. Grinding off the teeth does not address esthetic concerns to any great extent, it reduces the vertical dimension (which is usually already deficient) and can compromise the ability to bond later. Finally, bonding is typically less traumatic and is reversible.

General anesthesia is not recommended routinely for individuals in this age group. The risk of general anesthesia can outweigh the benefits and treatment usually can be accomplished with local anesthesia. In some cases, for patient comfort, sedation may be beneficial. General anesthesia may be indicated due to the complexity and extent of the oral disease or if the patient can't be managed using non-pharmacological approaches.

TREATMENT OPTIONS FOR SCHOOL AGE CHILDREN (7-12 YEARS)

The goals of dental intervention for affected children of elementary school age, 7-12 years, are to prepare for the transition from primary to secondary dentition, to minimize the influence of existing dental abnormalities on facial growth (thus promoting normal growth and development), and to enhance speech, chewing, swallowing, and esthetics. Prostheses to replace missing teeth are particularly important for this age group. (Figure 8)

In the absence of overt abnormalities of the dentition, age-appropriate dental care and anticipatory guidance, the guidelines of the AAPD should be followed (www.aapd.org). Furthermore, prostheses and restorations provided during earlier treatment should be maintained and enhanced. Children in this age group must be scheduled for periodic assessment of oral health, growth, and development.

Figure 8: A school-age boy without and with his dentures



- Prostheses for replacement of missing teeth are appropriate for this age group. (Figure 9)
- If complete dentures are considered, both maxillary (upper) and mandibular (lower) prostheses are recommended since opposing dentures improve esthetics, function, and retention.
- **Dentures must be age and size appropriate.**
- Fixed prostheses should be used with caution and monitored frequently in this age group because they may interfere with growth of the dental arches.
- Conventional full and partial acrylic dentures may be appropriate in this age group.
- If a prosthesis is used, it must be relined, readapted or remade periodically to accommodate craniofacial growth.
- Minor tooth movement, root canal therapy and selective extraction of teeth should be considered only to allow successful prosthetic treatment.
- Orthodontic evaluation may be needed if it is advisable to move teeth for esthetics and function.



Figure 9: Prosthetic appliance for a preteen and non-rigid prosthetic appliance

Teeth with conically shaped crowns lack suitable undercuts for engaging the clasps of a removable partial denture. Sometimes altering the contours of the teeth by bonding plastic composite to posterior teeth and canines will improve retention sites. Primary teeth should be retained, if possible and only if they are not ankylosed, in order to retain alveolar bone. This bone may be necessary later in life if implants become an option and may allow for less reliance on grafting procedures.

Affected children missing all mandibular teeth, including incisors, canines and premolars, may be candidates for implants. Individuals, whose teeth cannot be used as anchors for prostheses, also may be candidates for dental implants. Implants maintain alveolar bone in the area of the implant, improve function, and are valuable for those who cannot tolerate removable prostheses. Implants should not interfere with growth in the mandibular anterior edentulous region after age seven years.

In this age group, implants are recommended **only** for the anterior portion of the mandibular arch (Cronin & Oseterle, 1998). (Figure 10) Neither they nor “interim implants” are recommended for the posterior portion of the mandibular arch or anywhere in the maxillary arch because substantial growth is still expected.

Grafting of alveolar bone is **not** recommended for affected children in this age group. If grafts are considered necessary for implants in the anterior portion of the mandibular arch, an alternative treatment to implants should be considered until grafts can be accomplished at an older age.



Figure 10: Implant-supported bar in the anterior mandible

For children who have enamel dysplasia, esthetic bonded restorations or other full crown coverage restorations are appropriate. If removal of enamel is necessary to modify abnormally shaped teeth, bonded veneers or full crown coverage should be considered.

General anesthesia is not recommended routinely for individuals in this age group. General anesthesia may be indicated depending on the complexity and severity of the oral health disease or if the patient cannot be managed using non-pharmacological approaches.

TREATMENT OPTIONS FOR ADOLESCENTS (13 YEARS-SKELETAL MATURITY)

Each patient in this age group should be assessed individually because of the wide range of ages when growth is completed.

If craniofacial growth is incomplete, any rigid fixed prosthesis should be used with great caution. Adolescents are concerned with their appearance, and dental treatment should be planned accordingly. There is considerable research illustrating the negative social and educational effects a poor self-image can produce, whether related to dental or other abnormalities. The importance of acceptance is a valid consideration when deciding whether treatment is appropriate.

Without overt abnormalities of the dentition, age-appropriate dental care and anticipatory guidance, following the guidelines of the AAPD, should be provided for adolescents. Furthermore, functional prostheses and restorations incorporated in earlier treatment should be maintained.

- Prosthetic replacement of missing teeth is appropriate for affected adolescents.
- If complete dentures are contemplated, both maxillary and mandibular prostheses are recommended because opposing dentures improve esthetics and function.
- Rigid fixed prostheses are not typically recommended for adolescents because they might interfere with growth of the dental arches.
- Conservative rigid or fixed prosthesis (e.g., adhesive resin restorations or “bonded bridges”) may be used if growth is completed.
- Comprehensive orthodontic tooth movement and/or selective extraction of teeth to allow successful prosthetic treatment may be more extensive for adolescents than for younger children.

(Figure 11)



Figure 11: Comprehensive orthodontics in an adolescent

Because treatment is often complex, a multidisciplinary or team approach is recommended, and decisions should be made jointly by affected individuals and the involved dentists. Because most options for treatment involve prosthetic appliances, prosthodontists should be members of multidisciplinary teams and may assume the role of primary decision maker (Bergendal, 2001).

It is very important to use a multidisciplinary team approach in orthodontic and implant treatment plans. A prosthodontist should oversee tooth movement and implant placement to make sure the teeth and implants are in the most ideal position for the final restorative work to achieve maximum esthetics and function. In many cases, the prosthetic treatment plan cannot be determined at an early age but the involvement of the prosthodontist early in the treatment plan can prevent avoidable, irreversible treatment decisions that might compromise later care.

Affected adolescents who are missing their mandibular (lower) front teeth, or whose mandibular front teeth cannot be used as anchors for prosthetic appliances, may be candidates for implants. Implants maintain alveolar bone and improve function. Implant supported restorations are helpful for individuals who do not tolerate removable prostheses.

Implants are recommended only for the anterior portion of the mandibular arch, unless craniofacial growth is complete. (Figure 12) Neither conventional implants nor “temporary implants” are recommended for the posterior portion of the mandibular arch or anywhere in the maxillary arch, where significant growth is still expected.



Figure 12: Mandibular anterior implants in an adolescent

Grafting of alveolar bone is also not recommended until completion of craniofacial growth. For adolescents who have enamel dysplasia, esthetic bonded restorations or other full crown coverage restorations are appropriate. If removal of enamel is

necessary to modify abnormally shaped teeth, bonded veneers or full crown coverage may be appropriate. Primary teeth should be retained, if possible and only if they are not ankylosed, in order to retain alveolar bone. This bone may be necessary later in life if implants become an option and may allow for less reliance on grafting procedures.

General anesthesia is not recommended routinely for individuals in this age group. The risk of general anesthesia outweighs the benefits in this age group, and compliance with treatment can be accomplished without sedation. Pharmaceuticals should be used only after discussions with the patient and parents about risks and benefits.

TREATMENT OPTIONS FOR ADULTS

The goals of treatment for adults are to restore complete function and to optimize esthetics. Fixed restorations are preferred to provisional or removable alternatives and are the optimal way to restore function. (Guckes, Roberts, & McCarthy, 1998) (Figure 13) Definitive orthodontics, oral surgery to reshape the dental arches, alveolar bone grafting, selective tooth extraction, and periodontal surgery should be considered to meet the needs of the particular patient.

Figure 13: A man without and with his dentures



Because such extensive treatment is complicated, a multidisciplinary team approach is recommended, with decisions being made jointly by the affected individuals and the involved dentists. Because most options for treatment involve prosthetic appliances of one sort or another, prosthodontists, when possible, should be members of multidisciplinary teams and often will assume the role of the primary decision maker. (Bergendal, 2001)

Treatment options depend on the ability of affected individuals to care for and to maintain the permanent restorations. Affected individuals must understand that definitive treatment requires continuing maintenance and probable revisions; they must be prepared for multiple, lengthy visits to complete the initial restoration and then lifelong visits to maintain implants and prostheses.

In the absence of health-related contraindications, age-appropriate dental care is recommended. The prostheses and restorations, which were part of earlier treatment, should be reevaluated. If complete dentures are contemplated, both maxillary and mandibular prostheses are recommended since opposing dentures improve both function and esthetics. Rigid fixed prostheses are appropriate since craniofacial growth is complete. Comprehensive orthodontic tooth movement and selective extraction of teeth will allow optimal prosthetic therapy.

Implants may be used anywhere in the dental arches and, if necessary, bone grafting prior to implants is appropriate. (Figure 14) It is important to talk with the provider and to ask about the type of implant

Figure 14: The patient-dentist relationship is key to achieving treatment success.



system being used and how long they have used the system being recommended. Since implant therapy is intended to last a long time, if not the life of the patient, it is important to select a dental implant system that has good clinical research and clinical documentation along with a reputable company supporting the system. There should be cautious use of new implant designs and new types of clinical procedures in affected individuals. Examples are the use of small diameter or “micro” implants or immediate loading (such as “teeth-in-a-day”).

For adults who have enamel dysplasia, esthetic bonded restorations or other full crown coverage restorations are appropriate. If it is necessary to remove enamel to modify abnormally shaped teeth, bonded veneers or full crown coverage would be appropriate.

General anesthesia is not recommended routinely for individuals in this group. The risk of general anesthesia may outweigh the benefits, and treatment can be accomplished without sedation. Pharmaceuticals should be used only after discussion about benefits and risks.

OTHER CONSIDERATIONS: INSURANCE

All affected individuals, their families, dentists and other health care providers, insurance companies, and governmental agencies must recognize that the oral manifestations of the Ectodermal dysplasias are intrinsic components of genetic and congenital medical disorders. Thus, the costs for evaluation and treatment of the oral manifestations of these disorders should be covered by generic medical insurance policies. This is not to say that the cost for treating degenerative oral diseases, such as dental caries and periodontal disease, should be borne by medical insurance.

If the oral diseases result from congenital disorders, medical insurance should cover the associated costs. Examples of oral diseases that are a consequence of congenital disorders are caries caused by enamel dysplasia, periodontal disease caused by a deficient immune system, and malocclusion caused by hypodontia. Medical insurance companies are unlikely to cover expenses for such dental diseases, unless they are forced to do so through vigorous campaigns from interested parties. In some states, these activities resulted in specific changes in state medical insurance regulations to mandate additional coverage for the management of ectodermal dysplasias.

OTHER CONSIDERATIONS: PROVIDERS

The oral manifestations of the ectodermal dysplasias are often complex, and affected individuals might benefit from a team approach to evaluation and planning for treatment. (Figure 15)

Figure 15: Implants in mandibular arch in an adult



Affected individuals or their guardians, however, should not accept all team members as being equally trained and experienced.

Questions should be asked about

- The training of key individuals on the team
- The format for team meetings
- Intra-office communication among team members
- The pattern of referrals to specialists not on the core team
- The policies for payment and submission of insurance claims

Individual providers should be asked about their training and experience in treating patients affected by ectodermal dysplasias or related ectodermal dysplasias. It would be appropriate to seek input from other people who have been treated by this team or individual provider.

The issue of provider competency is especially critical for affected individuals whose treatment plan includes dental implants. Dentists who are placing implants or doing the subsequent prosthetic treatment should have been trained at programs accredited by a nationally recognized body or academic institution. The dentist should have, whenever possible, experience managing patients affected by ectodermal dysplasia.

The American Dental Association (ADA) Commission on Dental Accreditation reviews and accredits educational programs in the dental specialties. Programs in oral and maxillofacial surgery, periodontology and prosthodontics have specific educational requirements for the placement of implants. Programs in prosthodontics have specific guidelines in the prosthetic treatment of implant cases.

Oral health providers should give individuals a detailed treatment plan, with specific cost estimates, and alternatives or options for care. Oral health care providers should encourage individuals to seek independent opinions and should be receptive to alternative opinions from other providers. Records, especially radiographs, should be shared among providers. The written estimates of cost should stipulate clearly which aspects of treatment are directed toward or necessitated by the underlying disorder and therefore should be covered by medical insurance.

Providers should provide multiple treatment plan options that include the advantages and disadvantages of each plan. All plans may be acceptable but alternative treatment plans allow the patient and or parents to make an informed consent considering the cost and personal commitment.

OTHER CONSIDERATIONS: TIMING AND DURATION OF DIAGNOSIS AND TREATMENT

Although precise diagnosis of a specific ectodermal dysplasia is not always possible at birth, anyone at risk should be evaluated as early in life as possible. At-risk individuals are those with a family history of an ectodermal dysplasia or those who had suspicious findings on sonograms before birth. Evaluations at birth should include assessments of hair density and distribution, skin color and moisture, skin pigmentation, surface characteristics of toenails and fingernails, alveolar arch configuration, and secretions (tears, sweat, saliva, and nasal discharge).

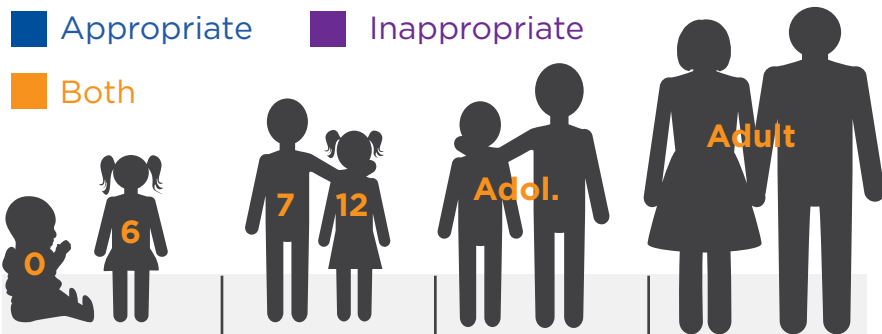
Frequently, however, the diagnosis of an ectodermal dysplasia is not suspected until heat intolerance appears, or until tooth eruption or tooth shape is abnormal. When one of these

problems is noted, assessment of the various ectodermal derivatives is appropriate, and diligent radiographic (panoramic radiography of the jaws, for example) or physiological assessment (scalp biopsy, for example) should be done.

Early treatment of the dental components of ectodermal dysplasia is possible. Many parts of the treatment program, however, may be delayed: teeth may not erupt sufficiently for restorative treatment; early treatment might compromise craniofacial growth; occlusion is not established; the alveolar ridges may be insufficiently developed to retain dentures; or the affected individual may not be able to cooperate with treatment. Treatment, then, must be done in stages.

Affected individuals and their families must understand that some treatment is temporary and that definitive resolution may be delayed until later in life, as long as one does not neglect periodic dental care. Even then, treatment may be a prolonged process and diligent compliance and many follow-up visits may be necessary to attain optimal results (Toumba & Gutteridge, 1995). Treatment for affected individuals must always be tailored to the age and to the individual, with appropriate acknowledgement to the costs and to the patient's physical, emotional, and behavioral development.

SUMMARY OF AGE SPECIFIC TREATMENT GUIDELINES

■ Appropriate ■ Inappropriate ■ Both			
			
Restorations	Restorations	Restorations	Restorations
Ortho Limited	Ortho Limited	Ortho Limited	Ortho Limited
Ortho	Ortho	Ortho	Ortho
Dentures	Dentures	Dentures	Dentures
Prosthesis Fixed	Prosthesis Fixed	Prosthesis Fixed	Prosthesis Fixed
Prosthesis Non Fixed	Prosthesis Non Fixed	Prosthesis Non Fixed	Prosthesis Non Fixed
Implants Mandible	Implants Mandible	Implants Mandible	Implants Mandible
Implant Other	Implant Other	Implant Other	Implant Other
Bone Graft	Bone Graft	Bone Graft	Bone Graft
General Anesthesia	General Anesthesia	General Anesthesia	General Anesthesia

OTHER MEDICAL CONSIDERATIONS:

This document has referred primarily to dental aspects of care provided for individuals affected by the ectodermal dysplasias. Dental providers are often the first professionals to entertain the diagnosis of one of the ectodermal dysplasias, and are often the professionals that interact most consistently with the affected individuals.

It is important, therefore, that dental professionals be aware of the medical problems that might be involved in ectodermal dysplasia patients, and the potential of referral to medical professionals that might collaborate in the care of these individuals. Regular communication with the patient's primary care provider--family physician, pediatrician, and internist--is crucial, so that potential medical complications of dental interventions might be discussed.

If the dental professional is the first to make the diagnosis, discussion with the primary care physician regarding referral to a medical geneticist is warranted. Dr. Kathleen Motil and collaborators have published on the nutritional concerns of children affected by ectodermal dysplasias. It may be important for the dental professional to speak with the child's pediatrician to suggest the benefit of involvement of pediatric nutritionists or feeding teams in the early care of children with ectodermal dysplasia.

Similarly, children affected by ectodermal dysplasia have been shown to have issues related to speech, hearing and language (hypodontia, cleft palate, abnormal saliva/mucous, recurrent ear infections, abnormally dry cerumen in external ear canal, etc.) Therefore, consideration of collaboration with or

referral to audiologists, speech and language pathologists and otolaryngologists should occur early in the course of dental and medical management.

Other health professionals that might participate in the treatment team could include dermatologists (abnormalities of skin, hair, nails); ophthalmologists (dry eyes, ankyloblepharon); allergy/immunology specialists (possible increase in prevalence of atopic disorders, known immune deficiency risk in some syndromes); and/or psychologists (self-esteem, quality of life). As one of the key professionals in the life of an individual with ectodermal dysplasia, the dental specialist could play a key role in education and referral of the patient for other medical evaluations and management.



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GLOSSARY OF TERMS

Alveolar: referring to the bony prominences in which teeth are anchored

Ankylosed: a tooth which has become fused to the jawbone

Anodontia: intrinsic absence of all the teeth

Bifurcation (of tooth roots): splitting of roots into two or three parts

Caries (dental): tooth decay

Cleft Lip: defect in the fusion of the upper lip during prenatal development

Cleft Palate: defect in the fusion of the roof of the mouth during prenatal development

Craniofacial: referring to the head and face

Crowns (of teeth): the part of teeth that is visible in oral cavity

Dental arches: the ridge of alveolar bone in which teeth are anchored

Dentin: intermediate cell layer of teeth

Dentures: artificial replacements for teeth

Dysplasia: abnormal organization of cells into tissues

Dysplastic: referring to dysplasia

Ectoderm: embryonic cells that form the outer covering of the body

Ectodermal: referring to the ectoderm

Embryogenesis: prenatal development between 4-7 weeks gestation

Enamel: the hard outer covering of the crowns of teeth

Eruption: the emergence of teeth through bone and gums

Genetic: pertaining to inheritance

Hyperdontia: intrinsic excess in the number of teeth

Hypodontia: intrinsic reduction in the number of teeth

Malocclusion: poor occlusion of teeth in opposing arches

Mandible: lower jaw

Mandibular: referring to lower jaw

Maxilla: upper jaw

Maxillary: referring to upper jaw

Mesoderm: the embryonic cells that form muscle and bone

Morphological: pertaining to morphology

Morphology: shape

Occlusal: referring to occlusion

Occlusion (dental): the meeting of teeth in opposing arches

Orofacial clefting: see cleft lip and cleft palate

Orthodontist: a dentist who specializes in the irregularities of the tooth alignment or occlusion

Periodontal disease: disease of gums and structures that support teeth

Prosthesis: dentures and other artificial appliances to replace teeth

Prostheses: plural of prosthesis

Prosthodontist: dentist who specializes in dentures and other prostheses

Pulp: innermost cell layer of teeth

Radiographs: x-rays

Radiodensity: opacity of bones of x-ray pictures

Roots (of teeth): the parts of teeth that anchor them in bone

Syndrome: coexistence of two or more physical features

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