Implants in children with ectodermal dysplasia: a case report and literature review

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Abstract: The replacement of teeth by implants is usually restricted to patients with completed craniofacial growth. Implant insertions in children or adolescents are circumvented due to several unfavorable potential effects including trauma to tooth germs, tooth eruption disorders and multidimensional restrictions of skeletal craniofacial growth. Moreover, the functional and esthetic results of the oral rehabilitation are only temporary acceptable. However, to a small number of pediatric patients suffering congenitally from severe hypodontia caused by syndromes such as ectodermal dysplasia, conventional prosthodontic rehabilitations are insufficient. We report the case of a boy with ectodermal dysplasia who exhibited a severe hypodontia and who was treated with implants inserted into the anterior mandible at the age of 8 years. The implants were functionally loaded and resulted in a high patient satisfaction. We recommend the early insertion of dental implants in children with severe hypodontia. Reviewing the current literature, several aspects of syndromic hypodontia, patient selection and implant planning are discussed.

Implants inserted into pediatric patients do not follow the regular growth process of the craniofacial skeleton and are known to behave similar to ankylosed teeth, resulting in both functional and esthetic disadvantages [OpHeji et al. 2003]. Additionally, they can interfere with the position and the eruption of adjacent tooth germs, thus resulting in potential severe trauma of the patient [Rossi & Andreasen 2003]. These and other adverse effects have resulted in a very restrictive indication for dental implants in those individuals who have not completed craniofacial growth yet [OpHeji et al. 2003].

However, in the last few years, an exception of this restriction was reported in those children who suffer from extended hypodontia or even adontia. While oligodontia with deficiency of only a few teeth is a relative common finding some populations, the absence of numerous teeth occurs only rarely [Silverman & Ackerman 1979; Matteeiuws et al. 2004]. Most often, these children suffer from congenital syndromes such as ectodermal dysplasia [ED; MIM #305100] [Percinoto et al. 2001], a rare congenital disease characterized by an aplasia or dysplasia of tissues of ectodermal origin such as hair, nails, skin and teeth [Kere et al. 1996]. In affected patients, the extensive lack of both deciduous and permanent teeth results in an atrophy and a reduced growth rate of the affected alveolar processes [Johnson et al. 2002]. Recent reports suggest that these pediatric patients can benefit remarkably from an implant-supported oral rehabilitation already during childhood [Guckes et al. 2002]. The aim of the present study is to present a report on a boy with hypodontia caused by hypohydrotic ED, who received dental implants at the...
age of 8 years. Special considerations of implant insertions in growing individuals are discussed and the current literature is reviewed.

Case report

The proband (male, white, Caucasian) exhibited the full-blown condition of hypohidrotic ED. At the age of 3 years, clinical evaluation revealed a hyperactive child with a dry hypopigmented scaly skin. The hair was thin, sparse and blond with sparse eyelashes and eyebrows [Fig. 1]. Intraorally, a severe hypodontia in both the deciduous and permanent dentition was found; in the deciduous dentition, only the central maxillary incisors were present and in the permanent dentition only the central incisors and the germ of the first right mandibular molar was found [Fig. 2]. Following a skin biopsy, hypohidrotic ED was diagnosed. At age five, dentures made of heat-cured acrylic resin were integrated in both the upper and lower jaw. During the following years, the mandibular rehabilitation became increasingly difficult due to growth of the mandible and deficient height of the alveolar processes. This resulted in recurrent functional insufficiencies of the lower denture.

At the age of 8 years it was decided to insert two implants into the anterior mandible to improve oral functions and to reduce potential psycho-social handicaps of the patient. The implant positions were planned at articulated cast models and transferred into surgery by an individual waiver that was fixed at the maxillary denture [Fig. 3a–c]. Following regional anesthesia, two implants were placed ad modum Branemark in the canine region of the anterior mandible [Fig. 3d–g]. Despite a remarkable multi-dimensional atrophy of the mandibular alveolar process, the insertion of cylindric screw implants [Nobel Biocare MK III; diameter 3.75 mm, Nobel Biocare, Gothenburg, Sweden] with a length of 13 mm was easily possible and resulted in safe primary stability [Fig. 4]. After a submerged healing period of 3 months, the implants were exposed and abutment connection was performed. Prosthodontic procedures were started 2 weeks later as soon as the soft tissues around the abutment cylinder had healed. An impregum (3M Espe AG, Seefeld, Germany) impression was taken of the lower jaw with abutment copings and an alginate impression of the maxillary denture. Models were made and mounted in an articulator after bite registration.

In the mandible, a bar-based denture was integrated [Fig. 5a–d]. In the edentulous maxilla, a new conventional denture was integrated, allowing satisfactory function [Fig. 5e, f]. The boy got easily accustomed to the new rehabilitation [Fig. 6a–c] and performed an excellent oral hygiene. The parents reported that after the insertion of the restoration, the dietary intake of the patient had changed significantly. The boy began to eat meat and apples, food he avoided before. The denture has now been in situ for 2 years and has functioned well.

Discussion

Ectodermal Dysplasia

Severe hypodontia or even adontia in children are very rare conditions, most often associated with congenital syndromes such as Down syndrome (trisomy 21) [Mestrovic et al. 1998] or ectodermal dysplasia [Silverman & Ackerman 1979]. There exist more than 170 clinically distinct hereditary syndromes in which ED is present [Kere et al. 1996; Lamartine 2003]. They are caused by an impaired development of epidermal appendages and are characterized by a primary defect at least in one of the following tissues: nails, hair, teeth or sweat glands [Priolo et al. 2000]. EDs are rare diseases with an estimated incidence of seven in 100,000 births for all EDs [Itin & Fistarol, 2004]. The pattern of inheritance is different, including Mendelian modes and sporadic cases. Several classifications of EDs have been proposed from a clinical point of view [Pinheiro & Freire-Maia 1994], with molecular genetic attributes [Priolo et al. 2000] and based on identified causative genes that most often are involved in processes of intercellular communication and signalling [Lamartine 2003].

The clinical diagnosis of hypohidrotic EDs in the neonatal period and in early infancy is sometimes difficult because sparse hair and absent teeth are often
normal findings at this stage. Dysmorphic facial features such as prominent supra-orbital ridges, frontal bossing and a depressed nasal bridge might be interpreted as normal variants. However, as seen in the presented case, the diagnosis becomes more easy during childhood based on the medical history and clinical examination. Sparse hair growth and deficient teeth become increasingly recognized, probably associated with hypoplastic mucous membranes and a decreased mucous production in the aerodigestive tract due to absent mucous glands. This can lead to chronic upper respiratory tract infections, otitis, dysphagia, hoarseness, bronchitis and sometimes hemoptysis [Siegel & Potsic 1990]. Facial symptoms become increasingly manifest and include maxillary hypoplasia, ‘saddle’ nose formation, prominent lips and linear wrinkles around the eyes. The hair is fine, dry, brittle and sparse, and the skin is thin and dry with hypohidrosis. Absent or decreased sweating in patients with anhidrotic, respectively, hypohydrotic ED is caused by absence of sweat glands. Affected children have difficulty controlling fevers and already mild illness may produce extremely high fevers because of absent temperature regulation by sweating. Affected adults are unable to tolerate a warm environment and require special measures to maintain a normal body temperature [Pinheiro & Freire-Maia 1994]. The diagnosis of hypohydrotic ED is usually performed by a biopsy of the skin that shows absent or hypoplastic sweat glands. Additionally, for some subtypes of EDs genetic testing is available [Vincent et al. 2001]. Currently, there is no causative treatment available.

Intraorally, besides mucous symptoms, patients with hypohydrotic ED suffer from both an altered shape and a reduced number of teeth. In clinical examinations [Glavina et al. 2001] and experimentally in knock-out organisms representing the phenotype of ED [Kristenova et al. 2002], the teeth were often found to be smaller in size and exhibited a conical peg-like shape. Hypo-respectively adontia related to ED can affect both the deciduous and permanent dentition [Nunn et al. 2003]. The most conserved teeth in hypohydrotic ED include in the permanent dentition the maxillary central incisors, maxillary first molars, mandibular first molars and maxillary canines [Guckes et al. 1998].

It was reported that especially in mild forms of ED, the most common complaint of childhood and adolescence is concern about the dental anomalies and facial appearance [Siegel & Potsic 1990]. Consequently, the dentist, orthodontist or the maxillofacial surgeon are probably the first medical professionals to be confronted with complaints of EDs.

**Implant positioning**

The conventional prosthodontic treatment of patients with severe hypodontia presents considerable problems [Ekstrand & Thomsson 1988; Boj et al. 1993]. The irregular distribution and abnormal shape of teeth often limit the integration of bridges or crowns in patients with ED [Nunn et al. 2003]. Especially, the hypodontic mandible of children with ED exhibits underdeveloped alveolar ridges [Rashedi 2003] and is therefore an area in which it is very difficult to gain adequate retention and support for conventional prostheses [Oesterle 2000]. Concerning Oesterle, a prudent clinician should always attempt to use a conventional prosthesis to gather functional and esthetic information to aid in the design of the final prosthesis and to allow as much growth as possible before initiating the implant-assisted phase of treatment [Oesterle 2000].
The insertion of dental implants in children or adolescents before completion of craniofacial growth is related to several problems. Experimentally, it was shown that endosseous implants placed in young pigs have imitated the effects of ankylosed teeth (Sennerby et al. 1993). Placed in alignment with adjacent teeth, the implants did not participate in growth processes, resulting in an infraocclusion and multidimensional dislocation when compared with the developing teeth (Sennerby et al. 1993). Additionally, adjacent tooth germs exhibited morphologic changes and disorders of eruption (Thilander et al. 1992). In the nearly anodontic child, however, these problems can be neglected.

Several aspects of craniofacial skeletal growth seem relevant for implant insertions in hypodontic children. Both the maxilla and the mandible are dynamically changing during childhood (Björk 1963, 1977; Skieller et al. 1984; Enlow 1990). Behaving similar to ankylosed teeth implants cannot participate with the maxillary growth processes of drift and displacement (Enlow 1990), resulting in unpredictable implant dislocations during growth or, if implants are fixed together, maxillary growth disturbances. Because of the resorptive aspects of maxillary growth at the nasal floor and the anterior surface of the maxilla, unpredictable...
able implant dislocations in vertical and anteroposterior direction can occur and even implant losses have to be expected. Transversal growth of the maxilla occurs mostly at the midpalatal suture. Consequently, fixed implant constructions crossing the midpalatal suture will result in a transversal growth restriction of the maxilla. All in all, the insertion of implants in the growing maxilla should be avoided until early adulthood [Cronin & Oesterle 1998].

In the mandible, however, the transversal skeletal or alveolo-dental changes are less dramatic as in the maxilla. In the posterior mandible, growth changes occur predominantly in late childhood with large amounts of anteroposterior, transverse and vertical growth [Enlow 1990]. Additionally, the mandible undergoes rotational growth, resulting particularly in vertical alterations [Skinner et al. 1984; Enlow 1990]. When several teeth are present, vertical growth is a major aspect of dental height increase and results in anteroposterior compensatory changes in the dentition. Consequently, implants would remain in an infraocclusal position and would probably be displaced in the anteroposterior direction [Oesterle 2000]. To our knowledge, there exist in the literature no reports on implant insertions in the posterior mandible in pediatric patients.

In the anterior mandible, however, alveolar growth seems relatively small when teeth are missing [Oesterle 2000]. The majority of transversal growth of the mandible occurs quite early in childhood; the anteroposterior growth occurs mainly at the posterior mandible [Skinner et al. 1984]. The use of implants to replace single teeth in the anterior mandible is not advisable due to the compensatory anteroposterior and the vertical growth in this area, however, in children with severe hypodontia, the anterior mandible might represent probably the most suitable site of implant placement [Oesterle 2000].

In the last few years, several case reports of implant insertions in the anterior mandible of children have been published [Bergendal et al. 1991; Bergendal 2001; Kargul et al. 2001; Giray et al. 2003]; most of the authors agree that the mandibular anterior area seems to hold the greatest potential for very early use of an implant-supported prosthesis. In a monocentric prospective study, the survival rate of implants placed in the anterior mandible of pediatric patients with ED was reported with 91% [Guckes et al. 2002]. Interestingly, some reports have demonstrated that craniofacial morphology did not differ significantly between implant-treated and non-treated children with ED, suggesting that treatment with intraosseous dental implants did not necessarily interrupt normal craniofacial growth as assumed before [Johnson et al. 2002]. In the long run, implants located at the anterior mandible probably seem affected by the mandibular growth rotation, which can result in a change in implant angulation [Becktor et al. 2001].

Implant timing
The finding of the ideal time of implant treatment in children seems quite difficult because many different aspects have to be considered while finding the best individual treatment strategy. It seems clear that the placement of implants in children with ED must be a team effort consisting of a surgeon, prosthodontist, orthodontist and periodontist. In younger ages, additionally, a pediatric dentist might also be required. First, the medical diagnosis of ED must be confirmed to avoid mistreatment. Profound dental clinical and radiographic examination to determine the extent of hypodontia is fundamental for further planning. The role of the dental team includes the preservation of the existing dentition, especially in patients with xerostomia, and maintenance of an adequate oral hygiene. If some teeth are present, their prognosis might be estimated. The orthodontist might use the intended implants as an orthodontic anchor to obtain the best position for the existing dentition. Particularly important is an adequate prosthodontic rehabilitation in order to ensure functional, esthetic and psychological well-being of the pediatric patient. Interim restorations may be indicated before definitive care and orthodontic treatment may be necessary to optimize dental position and axis. To date, there is only little research of the impact of extensive hypodontia on a young person [Hummel & Guddack 1997]. It was reported that children with disabilities realize at the age of 9 years their specific conditions when they compare themselves with other children [Hogberg et al. 1986], probably resulting in a state of depression [Nussbaum & Carrel 1976]. Therefore, the dental team also has to support the child in coping with issues of attractiveness during the formative years of childhood [Hogberg et al. 1986].

There are reports that implant treatment should be ended before puberty for optimum functional and psychosocial development [Giray et al. 2003]. Nevertheless, reports in the literature describe placement of implants as early as 3 years [Guckes et al. 1997] or 5 years of age [Smith et al. 1993]. From the orthodontic view the safest time to place implants seems to be during the lower portion of the declining adolescent growth curve at or near adulthood that can be determined by cephalographic radiographs, serial measure of stature or hand-wrist radiographs [Oesterle 2000]. Other relevant aspects to consider include the individual status of the existing dentition, the functional status of mastication and phonetics, esthetic aspects and emotional/psychological well-being [Nunn et al. 2003]. Finally, both the parents and the child have to be compliant to implant treatment and implant hygiene [Kearns et al. 1999].

Conclusion
Following the presented case and the reviewed literature, we recommend the insertion of implants in those pediatric patients who suffer from extended syndromal hypodontia such as seen in ectodermal dysplasia. The most suitable site of insertion seems to be the anterior mandible; insertions in the maxilla should be avoided or at least should not cross the midline. In order to determine the optimal individual time point of implant insertion, the status of skeletal growth, the degree of hypodontia and extension of related psychosocial stress should be taken into account in addition the status of the existing dentition and dental compliance of the pediatric patient.
References


