

## Complex oral rehabilitation of a patient with Witkop's syndrome: an interdisciplinary approach

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### Abstract

**Witkop's syndrome** - a gentle form of ectodermal dysplasia - is a rare autosomal dominant disease manifested by defects of the nail plates of the fingers and toes, hypodontia or anodontia with normal hair and sweat gland function. Other ectodermal tissues, organs do not show any alteration.

Oral manifestations may include incomplete development of both primary and permanent tooth germs. The location and form of erupted teeth could be irregular. This case report of a 23-year-old woman reveals the importance of a complex interdisciplinary dental treatment.

Clinical examination ascertained lack of numerous permanent teeth: besides the numerous deciduous teeth, only maxillary first incisors, first molars and mandibular first molars were present in the mouth.

Authors, an interdisciplinary team of surgeons, orthodontists, and prosthodontists proposed the complex rehabilitation of the patient with fixed prosthesis, and emphasize the importance of the long term follow up of the patient.

**Key words:** Witkop's syndrome, interdisciplinary dental treatment, oligodontia, genetic disorders, complex rehabilitation, fixed prosthesis

### Introduction

Ectodermal dysplasia (ED) is a hereditary condition characterized by abnormal development of the skin, hair, nails, sweat glands, and the stomatognathic system. A genetic disorder with congenital abnormalities of two or more ectodermal structures, commonly affecting the development of teeth, accompanied underdevelopment of the alveolar ridges [1,2]. Dental abnormalities and abnormal facial appearance are of major concern in childhood and adolescence, since they can restrict the individual socially and affect his or her self-confidence.

Witkop's syndrome (a gentle form of ectodermal dysplasia), a rare autosomal dominantly inherited developmental ectodermal tissue anomaly, manifesting in defects of the nail plates of the fingers and toes, and in hypodontia or anodontia with normal hair and sweat gland function [1, 2].

Oral manifestations can be incomplete development of both primary and permanent tooth germs, malalignment and anatomical irregularities of the erupted teeth. A number of factors can lead to congenital oligodontia: intrauterin problems, space limitation, physical obstruction or disruption of the dental lamina, failure of initiation of the underlying mesenchyme [3, 4, 5].

Oral rehabilitation in the early stages of the patient's life may provide functional and esthetic restoration as well as psychologic health [6]. Partial or total anodontia may result in functional defects, such as decreased chewing capacity, speech disorder and aesthetic problem. With any form of ED, young patients may display a range of symptoms and challenging rehabilitation. The quality of life of the affected individuals can be improved by an interdisciplinary approach of the treatment plan [7]. Prosthetic treatment may include removable, fixed, implant-supported prostheses, or a combination of

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these options [8, 9]. For the rehabilitation, it is crucial to know the age, number and condition of the present teeth, alveolar ridges and the growth status of the patient. The type of dental treatment depends on the severity of the disease (tooth size, tooth stability, and amount of available alveolar bone). Removable partial dentures can be used to replace congenitally missing teeth, and composite resin materials can be applied to restore conical-shaped maxillary teeth to achieve a favorable esthetic result [6]. These options minimize the sacrifice of healthy dental tissues [6].

### Case report

A 23-year-old female patient with partial anodontia was referred to the Department of Prosthodontics, Dental Faculty, Semmelweis University. Her chief complaints were difficulty with mastication and problems with her facial appearance. General history revealed that she had not any other disease. Signs of koilonychia and onychorrhexis and moderate disturbances within the plates could be detected on her nails. Other ectodermal tissues or organs did not show any lesions.

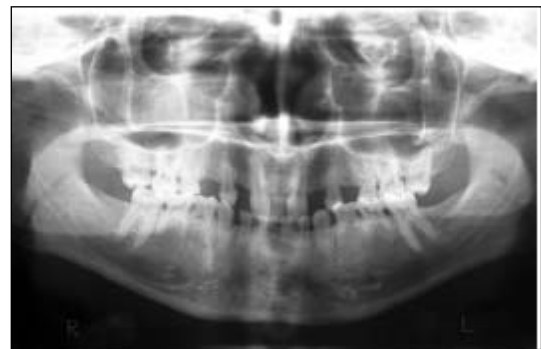
The craniofacial status of the patient was analyzed. The face showed to be a dolyccephal type. Undersized conical teeth, retained deciduous teeth, an enlarged maxillary buccal frenum with a midline diastema, a decreased occlusal vertical dimension (OVD) and underdeveloped alveolar ridges could be detected during the oral examination. A buccal crossbite on the left side and an increased (grade 3) attrition of the primary teeth (*Fig 1*) could be evidenced. Radiographic examination confirmed an oligodontia within the permanent teeth (*Fig 2*). As permanent dentition there were only the maxillary first incisors, maxillary first molars and mandibular first molars, the following remaining primary teeth were present too: maxillary canines and molars, mandibular incisors, mandibular canines and molars. All the teeth showed a positive vitality test. According to the clinical examination the occlusal relationship belonged to Class I. according to the Angle Classification. With the help of a cephalometric radiograph a deep bite with a skeletal origin could be recognized.

The final diagnosis was skeletal deep overbite (increased by the attrition of primary teeth) in Angle-Class I relationship of the jaws, resulting in a deep mentolabial sulcus, a decreased facial height, supraocclusion of the front teeth and

infraocclusion of the permanent first molars (*Fig 3*). It must be noted that the patient's brother had a similar dental status, thus the genetical examination suggested Witkop's syndrome.



*Figure 1. Diagnostic casts*



*Figure 2. Orthopantomographic image before the treatment*



*Figure 3. Deep overbite, supraocclusion of frontal teeth*

Periodontal and conservative treatment included professional dental hygienic treatment, (scaling and polishing), occlusal composite filling of the

bilateral first molars of the lower jaw and fissure sealants of the bilateral first molars in the upper jaw. The patient was then referred to oral surgery to carry out the frenulectomy of the enlarged maxillary buccal frenum. The orthodontic treatment was performed with a fixed appliance system for 6 months because of her labial median diastema (*Fig 4*). Patient came for a monthly regular control during the orthodontic treatment.



*Figure 4. Localised space closure using a sectional upper fixed appliance.*

Prosthetic treatment was started with an occlusal analysis, including the registration of intercuspal position, anterior and peosterior determinants of occlusion, and a possible reorganized occlusal scheme was recorded. Diagnostic casts were then made, duplicated, and mounted in a semiadjustable articulator (Dentatus, Svedia, Sweden) at the proposed OVD. Occlusal rim was used as a guide for determining an accetable plane of occlusion. All the teeth were prepared with paragingival shoulders and fitted with immediately fabricated crowns to protect the exposed teeth, and to restore the reorganized OVD. The temporary restoration served as a splint too, to protect the retention after the local orthodontic tretment in the maxillary incisor region. After the new jaw registration with wax rims, the temporary bridges were set up with wax in the new OVD (*Fig 5*). After try in, the full-arch fixed partial dentures were fabricated from a light cured acrylic resin (Gradia, GC Europe, Leuven, Belgium), and fitted temporarily in the mouth of the patient. OVD was raised with 2 mms. The provisional restorations were worn by the patient for two months, before final impressions were taken, allowing sufficient time for evaluation of the OVD. The patient was really satisfied with the provisional prosthesies, although additional vis-

its were needed to refine the occlusion.



*Figure 5. Temporary bridges wax-up model in semiadjustable articulator set on average values in the rearranged occlusal vertical dimension*

The definitive treatment contained fixed partial denture included a fixed porcelain fused to metal fixed partial denture both in the maxillary and on the mandibular arches to establish acceptable occlusal vertical dimension, esthetics, oral function and a therapeutic function of splint. In the maxillary jaw a 14-unit, while on the mandible a 12-unit porcelain fused to metal full house bridge was designed (Wirobond C, Bego, Germany; and Noritake, EX-3, Japan). After two months waiting period the permanent appliances were finished with the use of a semiadjustable articulator setted on individual values (Dentatus, Svedia, Sweden) and with face-bow transfer. Impression were taken with special trays using an A silicone impression material (Elit H-D+, Zhermack, Elite Mono Maxi, Italy). The maxillary and mandibular completed restorations were evaluated in bisquit stage, and a clinical remount was performed. Following the bisquit probe, a group function occlusal scheme was created (*Fig 6*). All restorations were charazterized, glazed and polished, and then luted with resin modified glass ionomer cement (Ketac TM Cem radiopaque, 3M Espe, USA). The patient has been wearing the fix prostheses without any complaints as well with acceptable occlusal vertical dimension, esthetics and oral function for two years (*Fig 8*). Home care instructions were given, and the patient was scheduled for a periodic maintenance care.



*Figure 6. The ready maxillary fixed porcelain fused to metal bridges mounted in the Dentatus articulator*



*Figure 7. Final radiographic image*



*Figure 8. Final satisfied picture of the patient with the fixed denture*

## Discussion

Our case-report evaluated a young female patient with severe oligodontia with Witkop's syn-

drome in the background. In case of oligodontia the advantages of existing teeth regard with retention, stability, function and the phonetics should be considered. In this case, the abutments of the fixed bridges were both deciduous and permanent teeth, prepared to a paragingival bevel form, which provided both esthetically and functionally a satisfactory result. Authors believe, that this kind of prosthetic appliance can help to protect the proprioceptive mechanism, and to prevent the resorption of the residual alveolar ridges.

Endosseous implants could also be considered as an alternative treatment [9, 10, 11], but in this case tooth supported prosthetic appliances were preferred because of the retention and stability of the remaining deciduous teeth and in order to prevent the alveolar bone resorption. Authors also considered the young age of the patient in the selection of the treatment modalities, and believe that good cooperation, proper oral hygiene and maintenance care (with what the patient was familiar) the patient will be able to retain her remaining teeth for a long period of time. Implants, placed into extraction sites, proximity to the buccal alveolar crest, appear as a major consideration too. The observations suggest that the width of the buccal alveolar ridge should be at least 2 mm to maintain the alveolar bone level especially for higher values of buccal resorption [13]. Osseointegrated implants might be a solution for the future, with bone augmentation, when primary teeth lose their stability.

According to the results, firm deciduous teeth with a relatively normal anatomy and complete roots can serve as excellent abutments for fixed a prosthetic appliance. The patient was also very satisfied by the excellent esthetic results.

## Conclusion

This case revealed that extensive prosthodontic treatment in young adults should preferably be performed with an interdisciplinary team approach. This interdisciplinary approach had the advantages of continuity and shared responsibility for therapy decisions

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