




**RS Global**  
Journals

**Scholarly Publisher**  
**RS Global Sp. z O.O.**  
ISNI: 0000 0004 8495 2390

Dolna 17, Warsaw, Poland 00-773  
Tel: +48 226 0 227 03  
Email: [editorial\\_office@rsglobal.pl](mailto:editorial_office@rsglobal.pl)

---

<b>JOURNAL</b>	World Science
<b>p-ISSN</b>	2413-1032
<b>e-ISSN</b>	2414-6404
<b>PUBLISHER</b>	RS Global Sp. z O.O., Poland
<b>ARTICLE TITLE</b>	LONG-TERM TEAM DENTAL REHABILITATION OF A YOUNG PATIENT WITH ECTODERMAL DYSPLASIA
<b>AUTHOR(S)</b>	Nataliya Emelyanova, Dmitry Emelyanov
<b>ARTICLE INFO</b>	Nataliya Emelyanova, Dmitry Emelyanov. (2024) Long-Term Team Dental Rehabilitation of a Young Patient with Ectodermal Dysplasia. <i>World Science</i> . 2(84). doi: 10.31435/rsglobal_ws/30062024/8187
<b>DOI</b>	<a href="https://doi.org/10.31435/rsglobal_ws/30062024/8187">https://doi.org/10.31435/rsglobal_ws/30062024/8187</a>
<b>RECEIVED</b>	25 May 2024
<b>ACCEPTED</b>	27 June 2024
<b>PUBLISHED</b>	29 June 2024
<b>LICENSE</b>	 This work is licensed under a <b>Creative Commons Attribution 4.0 International License</b> .

---

© The author(s) 2024. This publication is an open access article.

# LONG-TERM TEAM DENTAL REHABILITATION OF A YOUNG PATIENT WITH ECTODERMAL DYSPLASIA

**Nataliya Emelyanova**

MD, senior research associate of the department of Complex Risk Reduction of Chronic non-Communicable Diseases, Government Institution 'L.T. Malaya Therapy National Institute of the National Academy of Medical Sciences of Ukraine', Kharkov, Ukraine.

ORCID ID: 0000-0001-6089-6206

**Dmitry Emelyanov**

PhD, assistant professor, the Department of Study of Aging Processes and Prevention of Metabolic-Associated Diseases, Government Institution

"L.T. Malaya Therapy National Institute of the National Academy of Medical Sciences of Ukraine"

DOI: [https://doi.org/10.31435/rsglobal\\_ws/30062024/8187](https://doi.org/10.31435/rsglobal_ws/30062024/8187)

---

## ARTICLE INFO

**Received:** 25 May 2024

**Accepted:** 27 June 2024

**Published:** 29 June 2024

## KEYWORDS

Primary Adentiya, Dysplasia, Prosthetics.

## ABSTRACT

Today, one of the important issues in dentistry is the treatment of primary adentia, which has its own characteristics, taking into account the age of the patient, the need for orthodontic preparation for prosthetics, the design of the prosthesis. Primary adentia brings not only quantitative changes in the teeth, but also creates conditions for functional and morphological disorders of speech, breathing, chewing, leads to violations of the development of facial skeleton and causes underdevelopment of the jaws, which reduces the quality of life of the patient. Primary adentia with multiple missing teeth is a genetically determined condition and very often has a hereditary character in the form of hypohidrotic and hydrotic dysplasia. We present to your attention a description of the complex work of dentists on the completed case of a patient with a diagnosis of hypohidrotic ectodermal dysplasia (Christ-Siemens-Touraine syndrome). With multiple adentia, it is necessary to provide denture care as soon as possible, which requires engagement of dentists of various specialties in treatment and rehabilitation: orthodontists, orthopedists, therapists and maxillofacial surgeons.

---

**Citation:** Nataliya Emelyanova, Dmitry Emelyanov. (2024) Long-Term Team Dental Rehabilitation of a Young Patient with Ectodermal Dysplasia. *World Science*. 2(84). doi: 10.31435/rsglobal\_ws/30062024/8187

---

**Copyright:** © 2024 Nataliya Emelyanova, Dmitry Emelyanov. This is an open-access article distributed under the terms of the **Creative Commons Attribution License (CC BY)**. The use, distribution or reproduction in other forums is permitted, provided the original author(s) or licensor are credited and that the original publication in this journal is cited, in accordance with accepted academic practice. No use, distribution or reproduction is permitted which does not comply with these terms.

---

## Background.

Primary adentia, partial or complete, today remains one of the most difficult clinical situations requiring complex dental intervention. According to the literature, the prevalence rate of primary adentia ranges from 1.5 to 8%, with multiple absence of teeth noted in 0.3% of the population (1).

Clinical manifestations of primary adentia correspond to morphological and functional disorders in the dentition, and their severity increases with increase in the number of missing teeth.

In the literature, there are several variants of congenital adentia: hypodontia (the most common pathology, when less than 6 teeth are missing – more often the 3<sup>rd</sup> molars, lateral incisors and lower premolars); anodontia (complete absence of teeth), oligodontia (absence of more than 6 teeth, as a rule, combined with systemic diseases) (2). Primary adentia with multiple missing teeth is a genetically

determined condition and very often has a hereditary character in the form of hypohidrotic (X-linked type of gene expression) and hydrotic (autosomal dominant or autosomal recessive type of inheritance) dysplasia. This can explain the fact that the congenital absence of individual teeth is often a family feature, moreover, it is more often transmitted through the female line (3,4).

The most common hypohidrotic ectodermal dysplasia has a triad of symptoms: hypotrichosis, anhidrosis or hypohidrosis, and primary oligodontia or adentia (5). It should be noted that hypohidrotic ectodermal dysplasia in women often remains undiagnosed due to the partial or complete absence of general symptoms of the disease, in the presence of symptoms in the dentition (6).

To date, there is no single algorithm for treatment of patients with primary oligodontia, and providing care to such patients causes difficulties for dentists. In this regard, each clinical case with coverage of possible approaches to complex rehabilitation of a patient with ectodermal dysplasia is of undoubted interest.

We present to your attention a description of a completed case of a patient with a diagnosis of hypohidrotic ectodermal dysplasia (Christ-Siemens-Touraine syndrome).

## 2. Clinical Case.

*Ethics approval and consent to participate.*

**This study was conducted in accordance with the amended Declaration of Helsinki. The institutional review board approved the study, and participant provided written informed consent.**

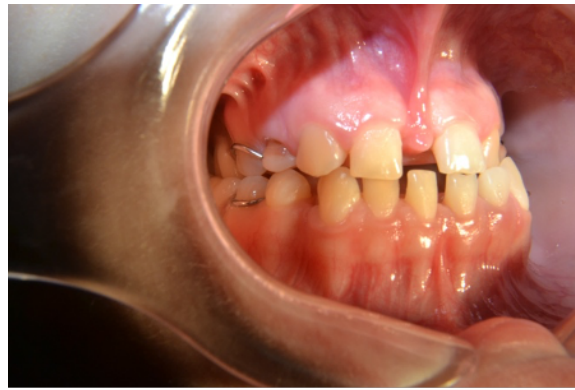
A patient D., born in 2004 (17 years old) complained of multiple absence of teeth in the upper and lower jaws, dysfunction of chewing, speech, aesthetics. The patient noted dryness in the oral cavity.

The patient's parents reported that in childhood there was an absence of certain teeth, but they did not go to the dentist about this. During the mixed dentition, the absence of eruption of the lateral group of teeth caused concern. After referring to a geneticist and a dentist, a diagnosis of hypohidrotic ectodermal dysplasia was made. The patient received partial removable lamellar dentures for the upper and lower jaws, which were replaced as the jaw grew. Despite the long-term wearing of the upper partial removable denture, adaptation to it never occurred, and therefore the patient experiences difficulties in eating and emotional tension. The patient's parents were offered the excision of autologous blocks from the iliac crests, but this proposal was not considered due to financial constraints. It is important to note the hereditary nature of this adentia is along the female line – the mother has no lateral incisors in the upper jaw and first premolars in the lower jaw, while the sibling has all the teeth (including the 3<sup>rd</sup> molars).

**External examination.** The patient is tall (174 cm), normosthenic physique. The hair is long, of normal density. The fingers of the wrist are of the correct form, the nail plates are without any pathological changes.

**Extraoral examination.** There is asymmetry of the face due to the position of the lower jaw in the left lateral occlusion, the bridge of the nose has a saddle shape. Concave facial profile (Ricketts analysis). Deficiency of the middle third of the face, violation of the ratio and closure of the upper and lower lips, the protruding chin are visually determined. Opening of the mouth is free. Movements of the lower jaw are without deviations. The skin of the face is dry, with macerations and hyperemia around the lips, the red border of the lips is covered with scales.

**Intraoral examination.** The depth of the vestibule of the oral cavity is average (7 mm), the mucous membrane of the oral cavity is pale pink, insufficiently moistened. The frenulum of the lower lip has the correct shape and place of attachment. The frenulum of the upper lip is thick, has low attachment and is woven into the alveolar process with a continuous mucous cord (type 3) (Fig. 1). The shape and size of the tongue are normal, the papillary apparatus is moderately pronounced. The frenum of the tongue has the correct shape and size. The gingival papillae are flattened, a thin gingival phenotype is noted (the vestibular relief of the alveolar process resembles a "washboard"). The rate of salivation at rest is reduced – 0.2 ml/min, during stimulation it is normal – 1.7 ml/min.



*Fig. 1. Vestibule of the oral cavity of the patient D. The frenulum of the upper lip is thick, has low attachment and is woven into the alveolar process with a continuous mucous cord, a thin gingival phenotype is noted.*

Cross bite with a tendency to mesial. Dental formula: upper jaw: 15, 53, 13, 11, 21, 23, 63, 25; lower jaw: 35, 33, 32, 31, 41, 42, 43, 44, 45. Temporary teeth are stable, 11, 21, 31, 32, have mobility degree I. Expressed microdentia. The width of the diastema in the upper jaw is 8 mm, between the incisors, canines and second premolars of the upper and lower jaws, there are tremas, as well as a mismatch between the inter-incisor lines and the midline of the face.

The upper jaw has a partial removable lamellar denture with wire retaining clasps at 15, 25. The alveolar process in the area of missing teeth is significantly atrophied and thinned, the tubercles of the upper jaw are made by a movable mucous membrane. A partial removable lamellar denture on the lower jaw is located with support on 35, 45, and the prosthetic bed is represented by a thin drop-shaped alveolar process, sharply tapering to the base and passing into the mobile mucous membrane of the oral cavity bottom. Functions of speech, swallowing, chewing are impaired.

On the orthopantomogram, the absence of impacted teeth or their follicles was noted, in addition to the central incisors, canines and the 2nd premolars, the presence of first temporary molars on the upper jaw with unresorbed roots was noted. An expansion of the periodontal gap in the region of the roots of the teeth was also revealed (Fig. 2). According to computed tomography: the maxillary sinuses are symmetrical, single-chambered, with alveolar bays, the condition of the alveolar bone does not correspond to age, spongy bone tissue has a large-looped pattern, the alveolar process is not expressed in the area of missing chewing teeth (Fig. 3). The spatial position of TMJ heads is symmetrical, intra-articular structures are normal.



*Fig. 2-3. Data of orthopantomogram (fig. 2) and computed tomography of the TMJ (fig.3).*

**Diagnosis:** Based on the results of a clinical examination, a diagnosis was made – primary multiple adentia of the upper (12, 14, 16, 17, 18, 22, 24, 26, 27, 28) and lower (35, 36, 37, 38, 46, 47,

48) jaws. Cross bite with a tendency to grade 3. Violation of the function of chewing, aesthetics against the background of hypohidrotic ectodermal dysplasia.

**Treatment:** after consultation with an orthopedic dentist and discussion with parents, a plan was drawn up for the complex correction of existing disorders. A frenectomy was performed on the upper jaw to create the possibility of subsequent elimination of the diastema.

One of the difficult moments was removal of defects in the lateral incisors. A partial bracket system was installed to level and approximate the central incisors, and distalize the canines with the help of opening springs (creating space for the lateral incisors) (Fig. 4).



Fig. 4. The first stage of treatment, the establishment of a partial bracket system.

Due to impossibility of obtaining sufficient space for installation of intraosseous implants, adhesive structures were made on a metal perforated frame with fixation on the palatal surfaces of the central incisors and canines of the upper jaw, the missing lateral incisors were made of metal-ceramic. Also, at this stage, restoration of the upper central and lower frontal teeth was carried out (elimination of tremas and diastema) (Fig. 5-6).



Fig. 5-6. Next stage of treatment. Adhesive structures were made on a metal perforated frame with fixation on the palatal surfaces (fig.5) and restoration of the upper central and lower frontal teeth was carried out (fig.6).

The choice of the restoration method was due to the pathological mobility degree I of the above-mentioned teeth, the fear of the pathological pockets formation and additional decrease in the interalveolar height. To assess the qualitative and quantitative changes in the alveolar bone, repeated cone-beam computed tomography was performed – no signs of resorption were observed.

The preferred option for replacing defects in chewing teeth was clasp prosthetics instead of existing partial removable lamellar dentures. Based on the obtained impressions, working models were cast and, subsequently, a clasp prosthesis with combined clasps for the upper jaw and a clasp prosthesis with Ney type I clasps for the lower jaw were made, which would improve aesthetics and chewing efficiency (Fig. 7).

However, when fitting the clasp prosthesis to the lower jaw, difficulties arose due to the specific shape of the alveolar process – a drop-shaped form. As a result, after installation of the lower clasp prosthesis on the prosthetic bed, its fixation was weak and only at rest, and there was no stabilization in the process of articulatory movements of the lower jaw (the prosthesis was dropped and fell out). This caused discomfort and the patient could not use such a prosthesis.



Fig. 7. View after replacing missing teeth with a clasp prosthesis.

Advice was given on the use of moisturizers and oral hygiene. The patient was very satisfied with the results in terms of function, aesthetics and phonation level, which allowed her to regain her self-esteem.

### 3. Discussion.

The most common complaint of patients with ED is anxiety about dental anomalies and appearance, which creates discomfort and severely limits communication opportunities (7). With primary adentia, underdevelopment of the alveolar processes occurs, which complicates and sharply limits orthopedic treatment, including the use of intraosseous implants. The use of removable prosthetics allows modification of treatment during periods of jaw growth, which, in fact, was carried out in our patient (8).

Despite the positive result and satisfaction of the patient, certain difficulties and failures in treatment were revealed. When discussing the treatment plan, the patient insisted on aesthetics, because when she smiled, the gingival zone of the abutment teeth was clearly visible. It was decided to make a thermoplastic clasp prosthesis with dentoalveolar clasps for the upper jaw (Fig. 8). Also, certain difficulties were revealed when manufacturing the lower clasp prosthesis due to the peculiarities of the prosthetic bed (drop shape with narrowing towards the base), thin and very mobile mucous membrane (Fig. 9).

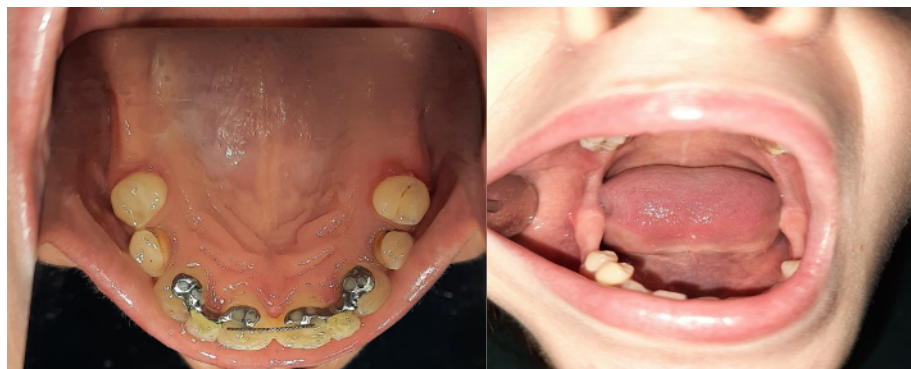


Fig. 8-9. View of the prosthetic bed of the upper and lower jaw.

The lower clasp prosthesis with a standard base plastic had weak fixation and lack of stabilization (the patient threw it off by moving her tongue). A soft relining of the prosthesis was carried out using an elastic material based on acrylic resin, which solved the tasks of treatment.

The bite of a removable clasp prosthesis should be in harmony with the bite of the patient. Since oligodontia or anodontia results in atrophy of the alveolar processes, reduction in vertical size, protruding chin, and class III intermaxillary relations, early prosthetics should be performed as early as possible. This treatment modality improves the patient's quality of life and can be considered an acceptable treatment modality for functional and aesthetic rehabilitation.

The clinical case was investigated for 2 years. This case proves the idea that, using a multidisciplinary approach, severe cases of ED can be successfully repaired and treated in a dental practice.

### Conclusion.

In women who are carriers of the ectodermal dysplasia manifestation gene, the disease can vary from the complete absence of clinical manifestations to partial (adentia) or a complete set of all symptoms.

As a result of the treatment, the aesthetics of the face and smile improved, chewing efficiency was restored, which made it possible to expand the diet, the patient's speech, her mental and emotional state improved.

With multiple adentia, it is necessary to provide denture care as soon as possible, which requires engagement of dentists of various specialties in treatment and rehabilitation: orthodontists, orthopedists, therapists and maxillofacial surgeons.

### DISCLOSURE.

The authors have nothing to disclose.

### CONFLICT OF INTEREST.

No conflict of interest is declared by authors.

### REFERENCES

1. Al-Ani AH, Antoun JS, Thomson WM, Merriman TR, Farella M. Hypodontia: An Update on Its Etiology, Classification, and Clinical Management. *Biomed Res Int.* 2017;2017:9378325. doi:10.1155/2017/9378325.
2. Nunn JH, Carter NE, Gillgrass TJ, et al. The interdisciplinary management of hypodontia: background and role of paediatric dentistry. *Br Dent J.* 2003;194(5):245-251. doi:10.1038/sj.bdj.4809925.
3. Ye X, Attai AB. Genetic Basis of Nonsyndromic and Syndromic Tooth Agenesis. *J Pediatr Genet.* 2016;5(4):198-208. doi:10.1055/s-0036-1592421.
4. Jonsson L, Magnusson TE, Thordarson A, et al. Rare and Common Variants Conferring Risk of Tooth Agenesis. *J Dent Res.* 2018;97(5):515-522. doi:10.1177/0022034517750109.
5. Wohlfart S, Meiller R, Hammersen J. et al. Natural history of X-linked hypohidrotic ectodermal dysplasia: a 5-year follow-up study. *Orphanet J Rare Dis.* 2020;15: 7. <https://doi.org/10.1186/s13023-019-1288-x>.
6. Liu H, Su L, Liu H, Zheng J, Feng H, Liu Y, Yu M, Han D. Rare X-Linked Hypohidrotic Ectodermal Dysplasia in Females Associated with *Ectodysplasin-A* Variants and the X-Chromosome Inactivation Pattern. *Diagnostics.* 2022; 12:2300. <https://doi.org/10.3390/diagnostics12102300>.
7. Deo K, Sharma YK, Shah B, et al. Improvement in the Quality of Life of a Patient of Ectodermal Dysplasia with Reconstructive Surgeries. *J Cutan Aesthet Surg.* 2019;12(4):244-247. doi:10.4103/JCAS.JCAS\_17\_19.
8. Faezeh Atri, Sania Azizi, Sepehr Hatampou. Removable prosthetic treatment of an ectodermal dysplasia patient: A case report. *Journal of Craniomaxillofacial Research.* 2020;7(1):34-39. doi 10.18502/jcr.v7i1.4015.