CAPDEPON SYNDROME

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ABSTRACT

Capdepon syndrome, also known as Stenton–Capdepon syndrome, is a hereditary disease that affects the normal development of teeth, particularly disrupting the formation of enamel and dentin. In this syndrome, changes in tooth color, thinning and rapid wear of enamel, as well as the appearance of short teeth may be observed. It is also referred to as "decorative root teeth" or "mesoectodermal odontopathy." This article provides a scientific analysis of the pathogenetic mechanisms, clinical manifestations, diagnostic criteria, and preventive measures of Capdepon syndrome. The results of the study highlight the significant scientific and practical importance of early detection and effective control of Capdepon syndrome in promoting a healthy lifestyle among the population..

Introduction

Capdepon syndrome (Stenton–Capdepon syndrome) is a hereditary, congenital dental disorder observed during tooth development, primarily characterized by defects in dentin and enamel structure. The disease is inherited in an autosomal dominant manner and is most often associated with mutations in the DSPP gene. In this syndrome, the external shape of the teeth is usually normal; however, the enamel color changes and may appear gray, brown, or opalescent. The enamel becomes fragile, thin, and prone to rapid cracking. Due to insufficient formation of the dentin layer, teeth are susceptible to early fracture, reduction in occlusal height, aesthetic defects, and functional impairments. Radiographic examinations clearly reveal obliteration (narrowing or disappearance) of the pulp chamber and root canals.This article scientifically analyzes the main clinical features, developmental mechanisms, diagnostic criteria, and modern treatment approaches of Capdepon syndrome. In addition, it highlights the importance of a healthy lifestyle in the prevention of this syndrome and presents scientific conclusions aimed at improving public medical awareness.

Main Part

Stainton–Capdepon dysplasia is a genetic disorder that manifests with significant deviations from normal dental development. These conditions are characterized by changes in tooth color, alterations in tooth shape, and rapid degradation of hard dental tissues; as a result, masticatory efficiency decreases and aesthetic problems arise. Deficiency of dental

tissues leads to early degeneration of the pulp and, against this background, to the development of periapical inflammation. All of these factors negatively affect general health and quality of life.

The treatment of this condition is mainly symptomatic, very prolonged, and complex. Etiological treatment of the above-mentioned disorder is almost impossible, as it would require modification of the patient's genome. A thorough study of treatment methods serves to further improve the quality of care provided to children and young people and to enhance their quality of life. [1]

Pathogenesis. Dentinogenesis imperfecta type II (Capdepon syndrome) develops as a result of mutations in genes encoding matrix proteins of dentin, namely phosphoproteins and collagens. When the condition is isolated (i.e., affects only the teeth), it is caused by mutations in the DSPP gene (4q21.3). This gene belongs to the SIBLINGs family and encodes three major non-collagenous matrix proteins of dentin:

- Dentin sialoprotein (DSP)
- Dentin glycoprotein (DGP)
- Dentin phosphoprotein (DPP) [2–8]

When dentinogenesis imperfecta occurs in association with osteogenesis imperfecta, the disease develops due to mutations in type I collagen genes or other closely related genes. Intraoral clinical signs are highly characteristic and facilitate diagnosis. Both primary (deciduous) and permanent teeth are affected, with primary teeth usually showing more severe involvement. The color of dentin is altered, appearing dull and bluish-brown, amber, or opalescent. Due to defects at the enamel–dentin junction, enamel rapidly chips away, and the exposed discolored dentin becomes susceptible to bacterial contamination in the oral cavity. The resistance of dentin is reduced; therefore, teeth wear down very quickly as a result of chewing and biting forces.

Radiographic examination shows bulbous (enlarged) tooth crowns with a marked cervical constriction. The pulp chamber and root canals are markedly narrowed or completely obliterated, and the roots are shortened. Repeated pulp exposure and multiple abscess formations may occur. Osteogenesis imperfecta is not characteristic of this type. In addition to dentinogenesis imperfecta and DSPP gene mutations, enamel defects have also been reported in some families [9, 10]. At the ultrastructural level, surface irregularities and cracks in enamel have been observed [11].

In one family with a nonsense mutation in the DSPP gene, the following features were described:

- irregular arrangement of dentinal tubules,
- an abnormal (smooth) dentino-enamel junction,
- altered enamel structure,
- increased amounts of fibrillar bundles around the dentinal tubules [11].

Case Report

A 7-year-old child visited the Department of Pediatric and Preventive Dentistry. The chief complaint was a brown discoloration of all teeth present since their eruption, along with the absence of several teeth over the past two years.

The patient's history revealed:

- delayed eruption of primary (deciduous) teeth,
- difficulty in chewing due to sensitivity to hot and cold foods.

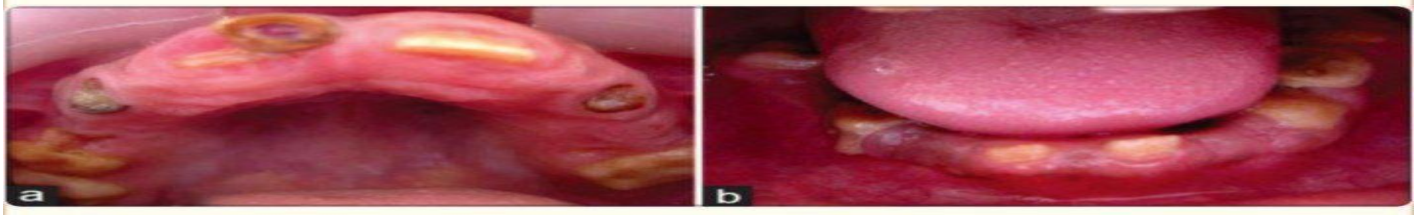
The medical history also indicated that:

- five years earlier, the child had undergone cardiac surgery, and a pacemaker had been implanted to correct an atrial septal defect,
- no other systemic skeletal disorders were reported.

Clinical Examination Findings. Mixed dentition was observed, with the presence of both primary and permanent teeth. The teeth were “tulip-shaped,” exhibited an opalescent brown discoloration, and showed a marked constriction at the cemento-enamel junction (CEJ). These features were present in both primary and permanent teeth. In addition, several missing teeth, deep carious lesions, and pulp-involved defects were noted in all primary teeth.

Detailed Clinical Findings. The primary maxillary right central incisor was severely worn and over-retained, while the permanent maxillary right central incisor was erupting in a palatal position [Figure 1a]. The primary maxillary left and right canines were severely worn, with only the root portions remaining [Figure 1a]. The primary mandibular left second molar and right first molar were affected by deep caries and exhibited Grade III mobility [Figure 1b]. The permanent mandibular right first molar had been restored six months earlier at a private dental clinic.

Figure 1.



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(a) Preoperative view of the maxillary arch.
(b) Preoperative view of the mandibular arch

Panoramic radiographic examination revealed involvement of both dentitions. The crowns appeared enlarged (bulbous) and the roots were shortened. All teeth showed enlarged pulp chambers with very thin dentin, giving the characteristic “shell tooth” appearance [Figure 2]. Based on the clinical and radiographic findings, a provisional diagnosis of Dentinogenesis Imperfecta Type II (DGI-II) was established.

Treatment plan and performed procedures. The treatment plan was developed in accordance with the clinical guidelines of the American Academy of Pediatric Dentistry (AAPD) [12].

The plan included the following:

- Extraction of non-restorable teeth,
- Preventive measures,
- Functional and esthetic rehabilitation.

Dietary counseling was provided because the child was on a highly cariogenic diet. In addition, oral hygiene instructions were given as preventive measures to be carried out at home.

Figure 2.

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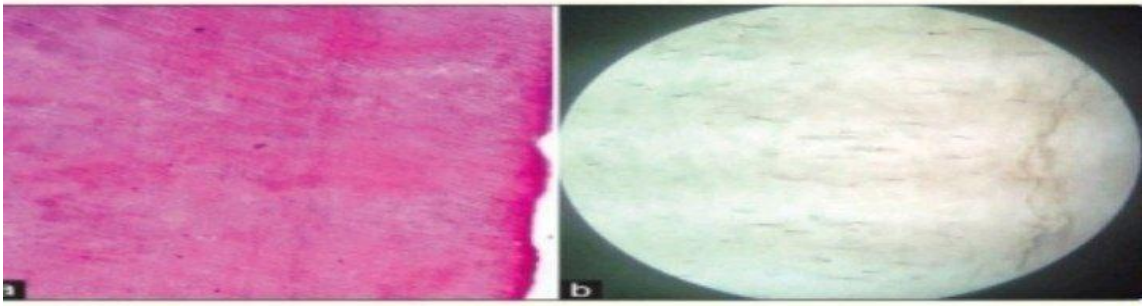
Orthopantomogram revealing enlarged pulp chambers giving characteristic Shell tooth appearance

Behavior management during treatment was carried out using the following methods: the tell–show–do technique, distraction by showing cartoons, positive reinforcement, and reassurance. The patient was treated on the dental chair under local anesthesia. Prior to extraction, antibiotic prophylaxis was administered for the following teeth: primary maxillary right central incisor; primary maxillary left and right canines; primary mandibular left second molar; and primary mandibular right first molar. The primary mandibular first molar was sectioned and subjected to histopathological examination to establish the final diagnosis. A decalcified dentin specimen stained with H&E revealed irregular and sparse dentinal tubules, as well as large areas of hypocalcified matrix. Empty spaces within the dentinal tubules were also observed [Figure 3a]. Ground section analysis of the lesion demonstrated irregular and sparse dentinal tubules throughout the specimen [Figure 3b]. The final diagnosis was Dentinogenesis Imperfecta Type II (DGI-II).

Figure 3.



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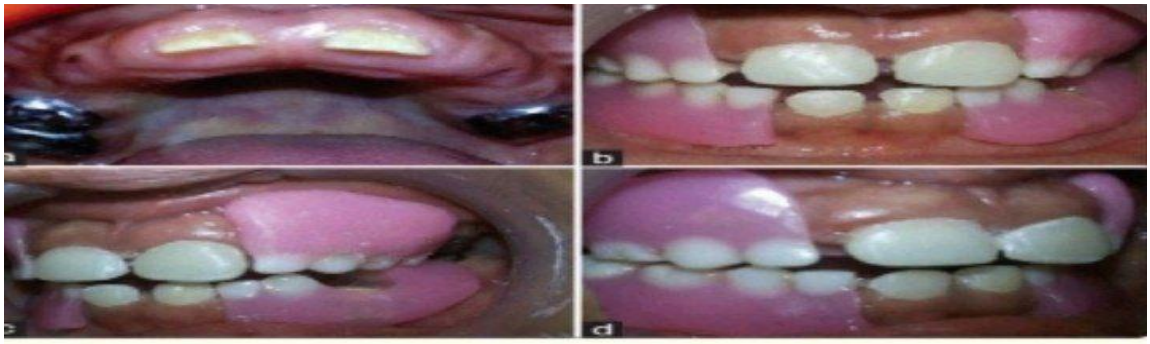
(a) Decalcified H&E-stained section of dentin. (b) Ground section showing irregular and fewer dentinal tubules

The next phase of treatment and rehabilitation involved the placement of stainless-steel crowns on the primary maxillary right second molar and the permanent maxillary left and right first molars. This measure was undertaken to prevent their future breakdown [Figure 4a]. Placement of stainless-steel crowns on the permanent mandibular first molars was also part of the treatment plan; however, the parents requested that esthetic corrections be performed first, including management of the spaces created by several missing or extracted teeth. Maxillary and mandibular impressions were taken using alginate, and the resulting dental stone casts were mounted on a semi-adjustable articulator using a centric record. Removable functional space maintainers for the maxilla and mandible were fabricated and delivered to the patient to improve masticatory function and restore the clinical vertical dimension [Figure 4b–d]. Esthetic rehabilitation was carried out as follows: Pedoform strip crowns were placed on all four permanent maxillary and mandibular central incisors. Minimal tooth preparation was performed, and care was taken to prevent pulpal exposure [Figure 4b–d]. After completion of the esthetic corrections and restoration of the child's masticatory function, the patient did not return for the planned prophylactic measures for the permanent mandibular first molars.

One-year follow-up and ongoing treatment. The patient returned after one year. The chief complaint was recurrent pain in the lower left and right posterior teeth for the past two months. Intraoral examination revealed deep carious lesions in the permanent mandibular first molars, with positive tenderness to percussion.



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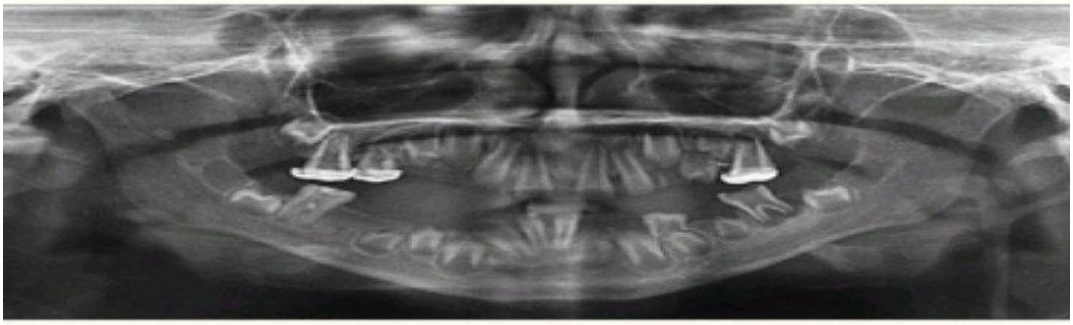
(a) Postoperative view showing stainless steel crown placed. (b) Pedoform strip crowns placed on permanent maxillary and mandibular central incisors. (c and d) Restored vertical heights by placing maxillary and mandibular removable functional space maintainers

Feedback

The permanent maxillary left lateral incisor and the maxillary left first premolar were erupting into the oral cavity. Panoramic radiographic examination revealed the following findings: deep carious lesions involving the pulp in the permanent mandibular first molars, and the presence of a dental abscess (suppurative inflammation) associated with the permanent mandibular left first molar [Figure 5]. The child continued to wear the functional space maintainers; however, the Pedoform strip crowns on the permanent maxillary and mandibular central incisors were no longer present. The functional space maintainer was modified for the following purposes:

Figure 5.



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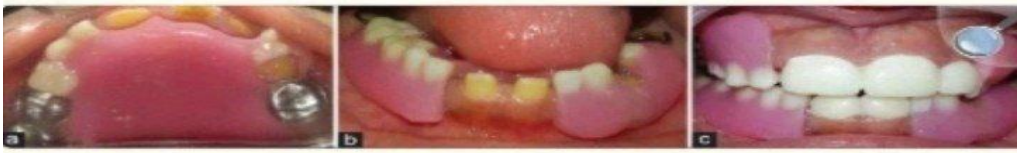
OPG revealing deep caries involving pulp in permanent mandibular first molars

Space was provided to allow the eruption of the permanent maxillary left lateral incisor and the maxillary left first premolar [Figures 6a and 6b]. Apexification using calcium hydroxide was performed on the permanent mandibular first molars. Laboratory-fabricated, heat-cured acrylic crowns were placed using an indirect technique on the following teeth: the permanent maxillary central incisors, the permanent maxillary left lateral incisor, and the permanent mandibular central incisors. This approach was undertaken to ensure better retention and stability [Figure 6c].

Figure 6.



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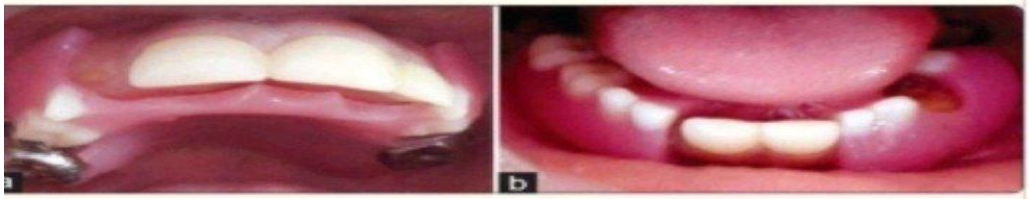
(a and b) Functional space maintainer modification done. (c) Laboratory fabricated heat-cured acrylic crowns placed

Three-month follow-up

The patient was recalled after three months to monitor the eruption of the permanent teeth. Intraoral examination revealed that the permanent maxillary right lateral incisor was erupting. The design of the functional space maintainer was modified to accommodate the eruption of the permanent maxillary right lateral incisor [Figures 7a and 7b]. The heat-cured acrylic crowns and stainless-steel crowns were completely intact. Follow-up visits were scheduled for the patient every three months.

Figure 7.



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(a and b) Functional space maintainer modification done

Conclusion

Early management of teeth affected by Dentinogenesis Imperfecta (DGI) is crucial and includes the following:

Behavior management

Comprehensive, functional, and esthetic rehabilitation

Pedoform strip crowns and other composite restorations may not provide long-term results in teeth affected by severe DGI due to biochemical alterations in the tooth structure. Frequent and long-term follow-ups are required to prevent and monitor future dental anomalies..

References:

1.ВА Еловиков, АБ Виноградов, НХ Горст - МНО Inter-Medical, 2014 - inter-medical.ru

2.Kim JW, Hu JC, Lee JI, Moon SK, Kim YJ, Jang KT, et al. Mutational hot spot in the DSPP gene causing dentinogenesis imperfecta type II. Hum Genet. 2005;116(3):186–91.

3.Lee KE, Kang HY, Lee SK, Yoo SH, Lee JC, Hwang YH, et al. Novel dentin phosphoprotein frameshift mutations in dentinogenesis imperfecta type II. Clin Genet. 2011;79(4):378–84.

4.Lee SK, Hu JC, Lee KE, Simmer JP, Kim JW. A den- tin sialophosphoprotein mutation that partially dis-rupts a splice acceptor site causes type II dentin dysplasia. J Endod. 2008;34(12):1470–3.

5.Ахроров, А. А., & Пулатова, Б., Назарова, Ш. (2021). Усовершенствование тактики хирургического лечения больных с травмой средней зоны лица. *Медицина и инновации*, 1(4), 199–204. [5]

6.Ахроров, А.Ш. (2024). Одонтогенные инфекции головы и шеи. *World Scientific Research Journal*, 1 (Volume-25), 60–70. [5]

7.Shavkatovich, A. A. (2023). Modern Aspects of Treatment of Odontogenic Sinusitis. *Genius Repository*, 25, 23–26. [5]

8.Shavkatovich, A. A., & Davronovich, M. D. (2023). Peculiarities of the treatment algorithm for the congenital defect that crosses the palate. *Intent Research Scientific Journal*, 2, 101–107. [5]

9.Ахроров, А.Ш., & Усмонов, Р.Ф. (2023). Лечения больных с травмой средней зоны лица путем виртуального моделирования. *Образование, наука и инновационные идеи в мире*, 13(6), 16–23. [6]

10. АШ Ахроров (2024). FEATURES OF THE USE OF A BONE BLOCK TAKEN FROM THE ZONE OF THE ZYGOMATICAL-ALVEOLAR BUTTRESS. *Web of Medicine: Journal of Medicine, Practice and Nursing*, 2 (ISSN (E): 2938 …), (страницы указаны в исходных данных). [7]

11.БАМ Ахроров Алишер Шавкатович, Усмонов Рахматилло Ф. (2024). Особенности применения костного блока, взятого из зоны скуло-альвеолярного контрфорса. *Journal of New Century Innovations*, 47, 149–157. [7]

12.AA Shavkatovich (2024). New Technique for Gum Plastic Sugery with Single-Stage Implantation. *Eurasian Scientific Herald*, 1 (2795-7365), 8–12. [8]

13.AA Shavkatovich, PB Juraxanovna (2021). Optimization Of Surgical Tactics For Treating Patients With Midrace Trauma. *The American Journal of Medical Sciences and Pharmaceutical Research*, 3(02), (страницы указаны в исходных данных). [9]

14.Lee SK, Lee KE, Jeon D, Lee G, Lee H, Shin CU, et al. A novel mutation in the DSPP gene associated with dentinogenesis imperfecta type II. J Dent Res. 2009;88(1):51–5.
15.Lee SK, Lee KE, Song SJ, Hyun HK, Lee SH, Kim JW. A DSPP mutation causing dentinogenesis imper-fecta and characterization of the mutational effect. Bio Med Res Int. 2013; 2013:948181. [Research Support, Non-U.S. Gov't].
16.MacDougall M. Dental structural diseases mapping to human chromosome 4q21. Connect Tissue Res. 2003;44 Suppl 1:285–91.
17.Zhang X, Zhao J, Li C, Gao S, Qiu C, Liu P, et al.DSPP mutation in dentinogenesis imperfecta shields type II. Nat Genet. 2001;27(2):151–2.
18.Lee SK, Lee KE, Hwang YH, Kida M, Tsutsumi T, Ariga T, et al. Identifi cation of the DSPP mutation in a new kindred and phenotype-genotype correlation. Oral Dis. 2011;17(3):314–9. [Research Support,Non-U.S. Gov't].
19.Wang SK, Chan HC, Rajderkar S, Milkovich RN, Uston KA, Kim JW, et al. Enamel malformations associated with a defi ned dentin sialophosphoprotein mutation in two families. Eur J Oral Sci. 2011;119 Suppl 1:158–67. [Research Support, N.I.H., Extramural Research Support, Non-U.S. Gov't].
20.Wieczorek A, Loster J. Dentinogenesis imperfecta type II: ultrastructure of teeth in sagittal sections. Folia Histochem Cytobiol. 2013;51(3):244–7.
21.Sodiqov, I. (2025). The social and political role and educational significance of public holidays in Uzbekistan. Western European Journal of Linguistics and Education, 3(03), 99-102.
22.Xusanov, H. (2025). Yangi O 'zbekistonda milliy musiqa madaniyatining ahamiyati. Inter education & global study, (1), 503-510.
23.Mo'minova, L. R. (2023). Results of focusing on art and culture in the new Uzbekistan. Экономика и социум, (10 (113)-2), 244-247.
24.Sodiqov, I. (2025). San'at oliygohlarida yosh rejissyorlarni kasbiy mahoratini va innovatsion kompetentligini rivojlantirish. Mahalliy va xalqaro konferensiyalar platformasi, (2), 336-341.
25.American Academy of Pediatric Dentistry. Guideline on Dental Management of Heritable Dental Developmental Anomalies. Pediatr Dent. 2016;38:302–7. [PubMed] [Google Scholar]