

CASE REPORT

Dental manifestation and management in a patient with combined pituitary hormone deficiency: a two-year case report

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Abstract

Combined pituitary hormone deficiency (CPHD) is a rare disorder caused by a complete absence of the anterior pituitary gland hormones. The Dental manifestation, management and two-year-follow-up of a case of a 12-year-old patient with CPHD is reported in this paper. A 12-year-old male patient with medical history of congenital CPHD and vitamin D deficiency presented for dental treatment. The patient looked tired and younger than his chronological age. Intraoral examination revealed good oral hygiene, generalized edematous gingiva, physiological pigmentation and high maxillary labial frenum attachment. The occlusion examination, showed class I molars, 2 mm overjet and 90% overbite. Single anterior tooth #21 crossbite with 1.5 mm diastema. Also, Miller's class I recession in tooth #31 with a probing depth of 1 mm, and grade I mobility. The treatment plan was formulated in phases, and the primary physician consultation was obtained and a clearance for dental treatment with a recommendation to double the hydrocortisone dose before a stressful dental procedure was recommended. Preventive and restorative programs were planned and provided. Then, the lower lingual holding arch was provided after the extraction of mandibular primary canines with composite turbo, and tooth #21 crossbite was corrected using a nance appliance with Z spring. Finally, the patient was recommended to attend follow-up visits every three months. During which the preventive program was reinforced and the restorative treatments were reevaluated. At the 2-year-follow-up satisfactory and stable aesthetic and functional results were obtained. This report highlights the oral manifestations, the value of establishing a preventive program, and restorative, and orthodontic interventions among patients with CPHD.

Keywords

Adrenal insufficiency; Growth hormone; Hypothyroidism; Oral manifestation; Panhypopituitarism

1. Introduction

Panhypopituitarism or combined pituitary hormone deficiency (CPHD) is a rare disorder caused by a complete absence of the anterior pituitary gland hormones, including growth hormone (GH), thyroid hormone (TH), and Adrenocorticotropic hormone (ACTH) [1, 2]. The incidence of CPHC is estimated to be 4.2 cases per 100,000 per year worldwide [3]. According to the National Institute of Health, there are less than 200,000 diagnosed patients with CPHD in the United States [3].

Many etiologies were reported to lead to CPHC, including congenital and environmental factors. Genetic mutations in the (*PROPI*) gene were found to contribute to approximately 50% of the familial cases of CPHD [4–6], while environmental factors include any diseases of the hypothalamus or in the pituitary gland itself [7]. The symptoms of CPHD can range from mild to severe, depending on which hormones are affected, and

patients usually need lifelong hormonal replacement therapy [8–10].

The clinical presentation of CPHD is strongly related to which hormone is deficient [8, 9]. But they usually present with small and younger facial appearances for their age due to the delayed growth of the skull and facial skeleton due to GH deficiency [11]. In comparison, individuals with TH deficiency are usually overweight, lack muscle tone [12]. Finally, individuals with ACTH deficiency present with anorexia, muscle weakness, and infection resistance, and stress [10].

Recently, Nota *et al.* [13] 2021 reported on the oral manifestations of an 11-year-old female patient diagnosed with CPHC. That patient was diagnosed with severe skeletal class III due to maxillary hypoplasia, mandibular protrusion, and as well as Solitary Median Maxillary Central Incisor syndrome.

However, based on our knowledge, no case report in the lit-

erature explains the oral manifestation and dental management with long-term follow-up of pediatric patients diagnosed with CPHC. Therefore, the dental manifestation, management, and two-year follow-up of the case of a 12-year-old male patient diagnosed with combined pituitary hormone deficiencies are described in this paper.

2. Case description

A 12-year-old male patient presented with his mother at the Pediatric dentistry clinics at King Abdulaziz University dental hospital in November 2020 with a chief complaint of “I don’t like the appearance of my teeth”.

Medical history revealed that the patient was diagnosed with congenital CPHC and vitamin D deficiency since birth, and he is taking GH injections, levothyroxine, hydrocortisone and vitamin D supplements daily. No known drug or food allergies were reported, and his vaccinations are up to date. The mother reported that her son was delivered through Cesarean section, with no complications during pregnancy and delivery. No family history of CPHC was also reported.

Upon general physical examination, the patient looked tired and younger than his chronological age, and his movements were slow. Based on his physician, the patient’s height reached to below the 5% percentile of his age group. At the same time, his weight and intelligence quotient (IQ) is conceded with in the normal range.

Upon extraoral clinical examination, his skin, hair and temporomandibular joint were within normal limits, with non-palpable lymph nodes. The patient had a symmetrical face with average mandibular plane angle and competent lips. Intraoral examination revealed good oral hygiene, generalized edematous gingiva, and physiological pigmentation. No pathology or abnormalities were detected in the labial and buccal mucosa, soft and hard palates, tongue and floor of the mouth. High maxillary labial frenum attachment was detected according to Placek Mirko [14]. The patient was in the early mixed dentition stage with multiple occlusal and proximal carious teeth. Occlusion examination showed class I molars classification, class III primary canine classification on the right side, class I primary canine classification on the left side, 2 mm overjet and 90% overbite. Single anterior tooth #21 crossbite, 1.5 mm diastema, rotated permanent maxillary central incisors, and retained tooth #72 were also detected. Based on Tanaka and Johnson’s mixed dentition space analysis, the patient had maxillary and mandibular mild crowding of 4 and 5 mm respectively. Due to the traumatic occlusion caused by the anterior tooth #21 crossbite, Miller’s class I recession in tooth #31 [15], was detected with a probing depth of 1 mm and grade I mobility. Fig. 1 demonstrates the intraoral clinical pictures of the patient at the initial visit.

Radiographically, the dental age of the patient was delayed and did not coincide with his chronological age, and the distal roots of the mandibular first permanent molars were dilacerated. Fig. 2A–D represents the patients’ radiographs at the initial visit. Also, the cephalometric analysis revealed a class I skeletal relationship.

A treatment plan was formulated in phases and started by consulting the patient’s primary physician to validate the safety



FIGURE 1. Intraoral clinical pictures of the patient at the initial visit.

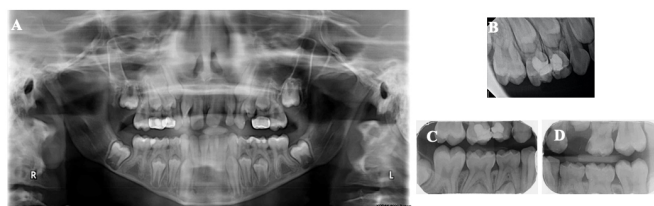


FIGURE 2. Patient’s radiographs at the initial visit. (A) Panoramic radiograph, (B) Periapical radiograph of the maxillary right posterior teeth, (C) Right bitewings, and (D) Left bitewings.

of the planned dental treatment and to review the recommended precautions needed before providing any of the required dental treatment. The dental treatment was cleared by the physician with a recommendation to double the hydrocortisone dose before a stressful dental procedure, including extraction. Before initiation, the mother signed a treatment informed consent form after discussing the treatment plan. The risks, advantages and alternative treatment options were also introduced to the mother.

In the preventive phase, a program was planned, including oral hygiene instructions for brushing using fluoridated toothpaste twice daily, and flossing daily, especially in tight interproximal areas. All of the instructions were given to the patient and his mother using a tooth model. Diet counseling and prophylaxis with 5% sodium fluoride (NaF) topical fluoride were also performed in the preventive phase of the treatment.

For the restorative phase, multiple restorations, stainless steel crowns, and extractions were performed. Post-restorative phase intraoral clinical pictures are presented in Fig. 3. During the treatment, the patient was potentially cooperative (tense-cooperative) according to the Wright classification [16] and Positive according to the Frankl Classification [17].

Then, the single anterior tooth #21 crossbite was corrected using a nance appliance with Z spring and a lower lingual holding arch after the extraction of mandibular primary canines to resolve lower anterior crowding and maintain the leeway space. A composite turbo was placed on the mandibular first molars to open the bite. At the one-month follow-up visit, correction of anterior cross bite was achieved with satisfactory



FIGURE 3. Post-restorative phase intraoral clinical pictures.

overjet and overbite. The occlusal composite turbo and the Z spring in the nance appliance was removed, and the appliance was re-cemented as a space maintainer. The lower lingual holding arch and the re-insertion of the nance appliance at the one-month follow-up visit are presented in Fig. 4A–C.

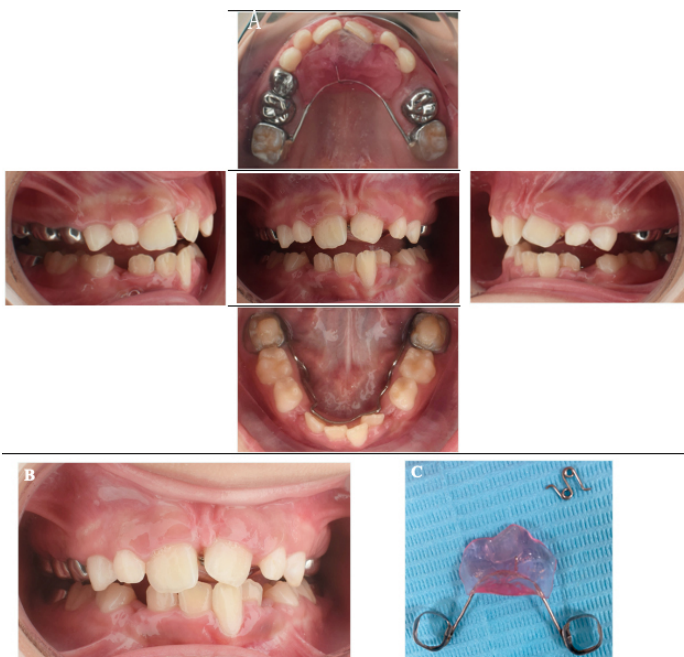


FIGURE 4. Intraoral clinical pictures at the one-month follow-up visit. (A) Insertion of the maxillary nance appliance with Z spring and lower lingual holding arch after the extraction of mandibular primary canines, (B) One-month follow-up visit, in which the occlusal composite turbo and the Z spring in the nance appliance were removed, and the nance appliance was re-cemented as a space maintainer. (C) The Z spring in the nance appliance was removed.

Finally, the patient was recommended to attend follow-up visits every three months, and the patient attended the 3, 6, 9, 12-, 15- and 24-month follow-up visits. During the follow-up visits, oral hygiene instruction, diet counseling, and prophylaxis with 5% NaF topical fluoride were performed.

Also, all the restorations, stainless steel crowns, and appliances were examined and were found to be intact. Intraoral clinical pictures at the 6- and 12-month follow-up visits are presented in Fig. 5A, B. At the 15-month follow-up visit, the primary maxillary canines were extracted, and a labial bow was attached to the nance appliance to retain the maxillary anterior teeth and to maintain the space. The Intraoral clinical pictures at the 15- and 24-month follow-up visits are presented in Fig. 6A, B. Also, the patient's radiographs at the 24-month follow-up visit are shown in Fig. 7A–C.



FIGURE 5. Intraoral clinical pictures at the 6- and 12-month follow-up visits. (A) at the 6-month follow-up visit, (B) at the 12-month follow-up visit.

Both the nance appliance and the lower lingual holding arch will be removed after the complete eruption of the second premolars to ensure minimal loss of leeway space. At the permanent dentition stage, the patient will undergo phase II orthodontic treatment to close the diastema and level and align the teeth. The diastema closure will take place after the eruption of the maxillary canine followed by frenectomy of the upper high labial frenum attachment based on current guidelines [18].



FIGURE 6. Intraoral clinical pictures at the 15- and 24-month follow-up visits. (A) At the 15-month follow-up visit, the primary maxillary canines were extracted; labial bows were attached to the nance appliance, (B) At the 24-month follow-up visit.

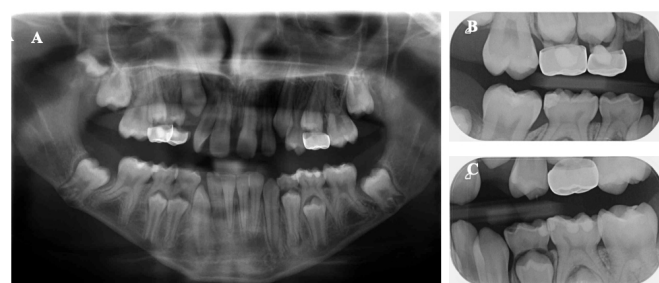


FIGURE 7. Patient's radiographs at the 24-month follow-up. (A) Panoramic radiograph, (B) Right bitewings, and (C) Left bitewings.

3. Discussion

Combined pituitary hormone deficiency is a rare disease that affects the anterior pituitary causing a lack of all hormones and affecting the patient's physical and dental health. The literature is limited on oral cavity effects and safe dental management among patients with CPHD. Therefore, oral manifestations and dental management with a two-year follow-up period are reported in the current case.

According to the literature, the clinical presentation of CPHD is strongly related to which hormone is deficient [8, 9]. Growth hormone is essential for average growth and

development. Individuals with GH deficiency usually suffer from delayed skull growth and facial skeleton growth, giving them a small facial appearance for their age [11]. Orally, these individuals typically present with delayed eruption and shedding of deciduous teeth. Crowding, malocclusion, and retarded growth of the mandible are also common among individuals with GH deficiency [11]. The patient presented in this report was diagnosed with GH deficiency since birth and took 1.2 mg of GH hormonal replacement daily. Clinically he presented with short stature and looked younger than children who are the same age. Also, he presented with malocclusion and crowding.

Individuals with TH deficiency are usually overweight, lack muscle tone, and have an expressionless face. Broad and flat noses and dry, pale, wrinkled skin are also common characteristics among individuals with TH deficiency [12]. The delayed eruption, enamel hypoplasia in both dentitions, macroglossia, micrognathia, gingival edema, and mouth breathing are the most common dental features among individuals with TH deficiency [19]. In this report, the patient looked tired, and his movements were slow.

Finally, ACTH deficiency manifests as adrenal insufficiency; clinically, patients can present with anorexia, nausea, weight loss, muscle weakness, diminished resistance to infections, and stress [10]. The melanic pigmentation of the skin and oral mucosa is the most specific sign of primary adrenal insufficiency. This patient presented clinically with muscle weakness and with pigmentation of the oral mucosa [20, 21].

In the 11-year-old female patient diagnosed with CPHC reported by Nota *et al.* [13] in 2021, severe skeletal class III due to maxillary hypoplasia and mandibular protrusion with anterior and posterior crossbite were among the oral manifestations. But in our case, the cephalometric analysis revealed a class I skeletal relationship. Also, the patient had an anterior single-tooth crossbite causing traumatic occlusion and gingival recession in one of the mandibular incisors.

The correction of anterior crossbite should be as early as possible because, if left untreated, it could cause recession and mobility to the lower anterior teeth [22, 23]. There are several acceptable treatment options to resolve anterior single-tooth crossbite including, nance appliance with Z spring, composite as an inclined plane [24], removable appliances [25], and fixed devices [26]. In the current case, the nance appliance with Z spring was preferred to correct the anterior single-tooth crossbite, and satisfactory results were obtained.

The keys to management begin by taking a proper medical history, obtaining medical consultation from the patient's physician, and applying stress reduction protocol in the clinic, like early short appointments. Finally, encourage the child and reinforce oral hygiene to achieve optimal treatment outcomes.

4. Conclusions

This report highlights the oral manifestations and the value of establishing a preventive program, early restorative, and orthodontic interventions among patients with CPHD.

ABBREVIATIONS

CPHD, Panhypopituitarism or combined pituitary hormone deficiency; GH, Growth hormone; TH, Thyroid hormone; ACTH, Adrenocorticotropic hormone.

AVAILABILITY OF DATA AND MATERIALS

The data presented in this study are available on reasonable request from the corresponding author.

AUTHOR CONTRIBUTIONS

SDA—Providing the treatment and follow-up to the patient; SDA and SMB—Initial drafting of the manuscript; SMB—Critical reviewing and editing the manuscript.

ETHICS APPROVAL AND CONSENT TO PARTICIPATE

Before treatment initiation, the mother signed a treatment informed consent form after discussing the treatment plan. The risks, advantages, and alternative treatment options were also introduced to the mother.

ACKNOWLEDGMENT

Not applicable.

FUNDING

This research received no external funding.

CONFLICT OF INTEREST

The authors declare no conflict of interest.

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How to cite this article: Sara D. Alzanbaqi, Sara M. Bagher. Dental manifestation and management in a patient with combined pituitary hormone deficiency: a two-year case report. *Journal of Clinical Pediatric Dentistry*. 2023; 47(5): 181-185. doi: 10.22514/jocpd.2023.069.