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CASE REPORT

Oral and systemic manifestations of congenital hypothyroidism in children. A case report.

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Abstract: Hypothyroidism is the most common thyroid disorder. It may be congenital if the thyroid gland does not develop properly. A female predominance is characteristic. Hypothyroidism is the most common congenital pediatric disease and its first signs and early symptoms can be detected with neonatal screening. Some of the oral manifestations of hypothyroidism are known to be: glossitis, micrognathia, macroglossia, macroquelia, anterior open bite, enamel hypoplasia, delayed tooth eruption, and crowding. This paper briefly describes the systemic and oral characteristics of congenital hypothyroidism in a patient being treated at a dental practice. The patient had early childhood caries and delayed tooth eruption. There are no cases of craniosynostosis related to the primary pathology, which if left untreated, increases the cranial defect. Early diagnosis reduces the clinical manifestations of the disease. Delayed tooth eruption will become a growing problem if the patient does not receive timely treatment and monitoring.

Keywords: *Congenital Hypothyroidism, Oral Manifestations, Neonatal Screening, early childhood caries.*

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INTRODUCTION.

Congenital hypothyroidism (CHT) is the most common pediatric endocrinopathy. It is the result of a congenital tissue decrease in the biological activity of thyroid hormones from the womb, which is essential for many critical processes of fetal brain development, formation of synapses and myelination. Thyroid hormones are important in the development of the nervous system in postnatal life, their absence or deficiency cause alterations in brain development, such as mental retardation and physical development damages, including orofacial complex. CHT is one of the most common preventable causes of learning difficulties[1].

CHT has a worldwide prevalence and its incidence varies according to country, to neonatal screening coverage, coverage methods and ratified examinations. The reported global incidence ranges from 1/3000 to 1/4000 live births. An important aspect is its prevalence in females (2:1). It is more common in Hispanics and Native Americans than in blacks. It is a growing risk in children with Down syndrome and has a fatality rate ranging from 15 to 20% in newborns[2,3].

CHT etiology includes: abnormalities of the thyroid gland in 90 to 95% of cases, hypothalamic-pituitary alterations and thyroid hormone resistance. Primary hypothyroidism is a series of alterations that occur during the embryological development stage of the gland, known as sporadic congenital hypothyroidism, and detected by neonatal screening. Its causes include thyroid dysgenesis and dyshormonogenesis, as is the case with mutations of the thyroid peroxidase gene[4].

Other causes of nenonatal CHT are transplacental passage of antithyroid drugs given to treat maternal hyperthyroidism, maternal exposure to excess iodine and inadvertent administration of radioactive iodine for the treatment of thyrotoxicosis or thyroid cancer during pregnancy. Environmental circumstances, such as iodine deficiency, increa-

se the frequency of primary CHT[5,6].

Clinical diagnosis occurs in less than 5% of newborns with hypothyroidism because the condition shows minimal signs and symptoms. Endocrine disruptions have great medical and dental significance, and it is important to implement the most favorable dental approach to treat these patients[7].

Table 1 shows systemic and oral manifestations of HTC. The objective of this report is to inform and discuss the case of a patient diagnosed with CHT, its systemic and oral manifestations and its dental treatment.

CASE REPORT.

Female patient age 4, middle-class family, preschool student, delivered by cesarean at term and hypoxic during the same period.

Patient was diagnosed with congenital hypothyroidism, detected at birth by neonatal screening. She is under treatment with L-thyroxine, according to her body weight of 1.6 to 2.1µg/kg/day, and in permanent control. The patient goes to a dental practice complaining of pain in one of her tooth. She eats a balanced diet at specific times. The patient maintains a cooperative attitude.

Physical examination showed frontal bone malformation due to anterior plagiocephaly craniosynostosis, tracked in various studies to verify its growth and development (Fig. 1). The patient had a weight of 18kg and a size

Table 1. Clinical and orofacial manifestations of congenital hypothyroidism.

CLINICAL	OROFACIAL
Anemia	Vertical facial growth
Cardiomegaly	Decreased length and angle of the cranial base
Heat sensitivity	Macroglossia
Constipation	Delayed deciduous and permanent tooth eruption
Cretinism	Thick lips
Dry hair	Mouth breathing
High levels of aspartate, transaminase, alanine dehydrogenase, lactate and creatinase	Enamel hypoplasia in deciduous and permanent dentition (less often)
Goiter	Glossitis
Hyperlipidemia and Hypertelorism	Dysgeusia
Lethargy	Micrognathia
Low amplitude of the QRS wave in the electrocardiogram	Compromised periodontal health (bone resorption)
Paresthesia	Anterior open bite
Reduced heart rate and breathing	Enlargement of the salivary glands
Seizures	Short tooth roots
Tachycardia	Large pulp chambers
Weight gain	Permanent open apices

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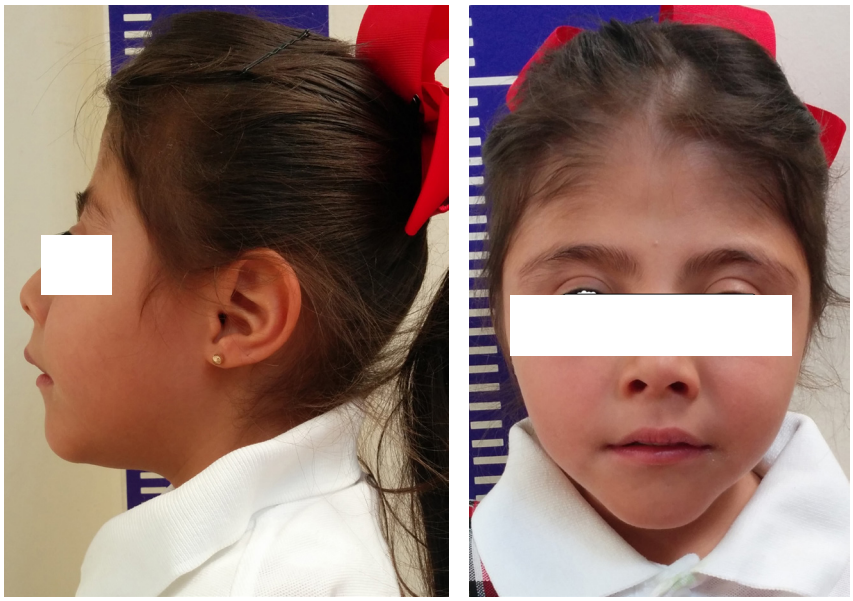
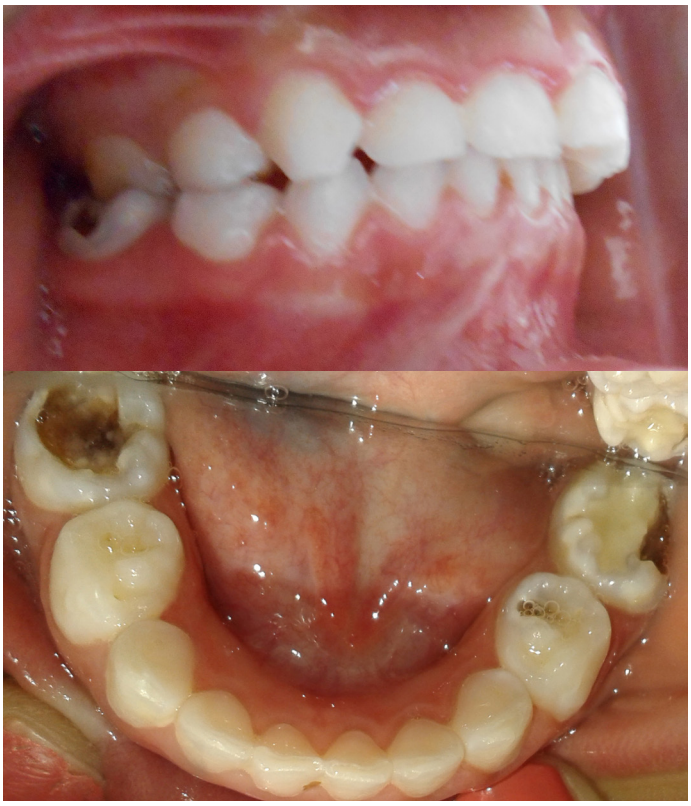
Figure 1. Extraoral view.



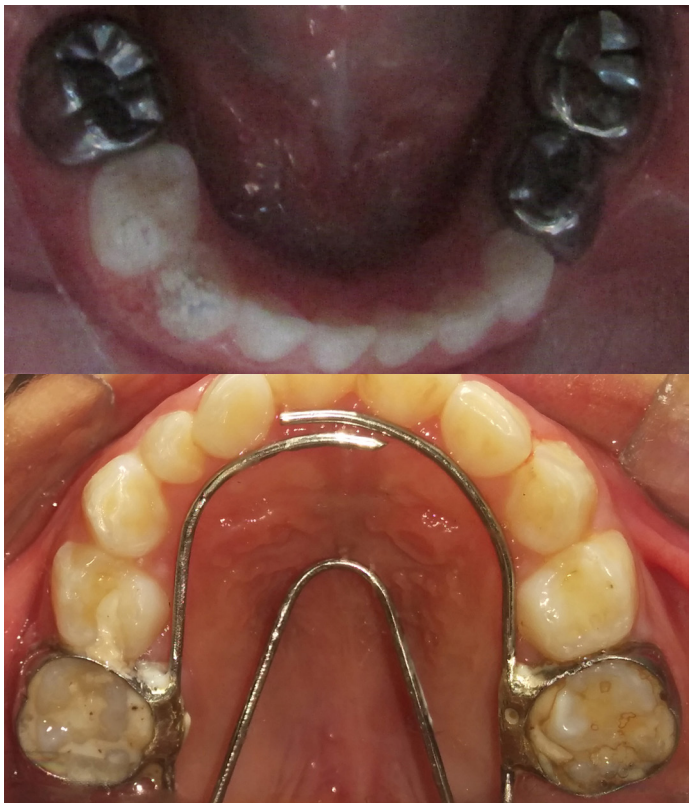
Low hair implantation and bone malformation of the skull, front retracted back by anterior plagiocephaly: craniosynostosis.

Figure 2. Intraoral view



Right unilateral crossbite, caries in teeth 54 and 64, class I relationship on the left canine

Figure 3. Treatment control.



Restoration with ION crowns and "W Porter" appliance to uncross bite

of 105cm, a head circumference of 46cm (decreased), the height/age ratio corresponds to the 50th percentile (normal), the p-15 weight/age ratio is normal, a low body index of 14.5kg/m.

She had a convex profile with the top third increased and the lower third decreased. Her facial type was mesoprosopic and cranial to mesocephalic, her neck allowed sufficient mobility and upper and lower limbs showed proper mobility. Systemic and oral manifestations corresponded to: facial dysmorphology by anterior plagiocephaly craniosynostosis; low hair implantation; hypertelorism; delayed bone age; hypertonic facial asymmetry; delayed tooth eruption; unilateral crossbite; and early childhood caries (ECC).

The patient had a class II relationship on the right canine, class I on the left, straight right terminal plane and left terminal with mesial step. She also had overjet and overbite in tooth 8, an unilateral right posterior crossbite and delayed tooth eruption, ECC in teeth 75, 84, 85, 6/2 according to the International Caries Detection and Assessment System (ICDAS) and irreversible pulpitis in teeth 54 and 64, 2/2 ICDAS, (Fig. 2) with dental pain.

Due to primary diagnosis and extension of treatment stimulating hormone test was requested, showing normal parameters. Preventive treatment performed with the consent of the patient's mother consisted of: sealing of pits and fissures in teeth 65 and 55; brushing technique. Curative: in pulpotomy teeth 74, 84 and 8; placement of resin in teeth 64 and 54. The restoration consisted of: ION crowns in teeth 75, 84 and 85, and finally placement of fixed orthopedics appliance "W Porter" to modify the right unilateral posterior crossbite with subsequent monthly monitoring. Mother and daughter were reminded of the importance of keeping a proper diet and good oral hygiene mainly in the retainers (Fig. 3). Good oral health, free of caries was observed in checkups.

DISCUSSION

By means of diagnostic tools, such as dental X-rays and laboratory analyses it is possible to detect morphological alterations or changes in chemical composition originating genetic and metabolic disorders[8,9]. The mouth shows multiple oral manifestations of various systemic disorders that can be identified by the pediatric dentist[10]. Thyroid disorders are the most common of all endocrine diseases. If the CHT is not treated, it may cause severe mental retardation and stunted growth[11,12].

Patients with untreated CHT with severe symptoms may be at risk if dental treatment is performed without proper medical supervision. Therefore, the main objective of the dentist is to detect CHT in these patients and refer them to medical treatment before performing any dental procedure. In this case the patient was treated with sodium L-thyroxine, since the detection of CHT by neonatal screening, to stimulate growth, proliferation and hormonal maturation[13].

Knowing the patient's medical history in relation to drugs and doses to treat CHT is important because this knowledge contributes to identify aggregate pathologies (congenital heart, kidney or urological abnormalities, mental retardation, stunted growth, deafness or symptoms of hypometabolism) and avoid clinical complications, such as hypothyroid coma, consisting of hypothermia, bradycardia, hypotension and seizures[14]. In the case of CHT, when newborns are diagnosed by neonatal screening and treated in the first 45 days, they generally develop normal intelligence[15]. In this case, treatment with L-thyroxine from birth to date has reduced the main manifestations of CHT and some are not even present, such as cretinism, goitre, glossitis or dysgeusia, enlarged salivary glands, among others.

Once the hypothyroid patient is under good medical treatment, no special problems should arise in the dental treatment, except to treat malocclusion and enlarged tongue, if any. The most reported clinical features described in literature are: delayed physical and mental development, thick lips and macroglossia[16,17]. The lack of growth in terms of length is observed very early. Early diagnosis leads to a good outcome for the child in terms of growth and mental capacity[18].

With respect to clinical characteristics, in this case the patient only presents a decrease in head circumference development caused by mild cranosinostosis and absence of several features that most authors describe, as the condition of this patient was diagnosed and treated at a very early age. The most important oral manifestations related to the primary pathology of the patient are: delayed eruption, crossbite and ECC.

The delay in the ongoing tooth eruption will become a growing problem as the child develops, because the deciduous teeth should remain a longer time before the eruption of the permanent[19]. The treatment plan is to restore the masticatory function in decayed deciduous teeth and restore masticatory function using an orthopedic appliance[20], considering that the defect of thyroid hormone action slows the growth rate. The patient got used to her orthopedic appliance after three appointments.

A patient with CHT and the health risks caused by this condition are a reminder of the importance of having an updated knowledge of pediatric diseases, their dental management regarding etiopathology, drug interactions, medical management, treatment, potential complications and dental approach.

CONCLUSION.

Early detection and treatment of CHT minimizes systemic disorders and the development of complex inherited orofacial manifestations.

Cranosinostosis is a finding unusually reported in this disease, something that increases the craniofacial defect. ECC and delayed eruption should be treated and intercepted waiting for the eruption of the permanent dentition.

Manifestaciones sistémicas y orales del hipotiroidismo congénito en niños. Informe de caso.

Resumen: El hipotiroidismo es el más común de los trastornos de la tiroides, puede ser congénito si la glándula tiroides no se desarrolla correctamente. La predominancia femenina es una característica. Los primeros signos y síntomas prematuros son detectados a la introducción del tamiz neonatal para hipotiroidismo, por ser la enfermedad congénita más frecuente en pediatría. Entre las manifestaciones orales del hipotiroidismo se observan: glositis, micrognatia, macroglosia, macroquelia, mordida abierta anterior, hipoplasia del esmalte, retardo en la erupción dental, y apiñamiento. Mediante revisión sucinta, se describen las características sistémicas y orales del hipotiroidismo congénito en una paciente que acude a la clínica dental, que dio lugar a caries de la infancia temprana y un retraso en la erupción dental. No existen casos de craneosinostosis relacionados con la patología base, la cual si no es tratada, incrementa el defecto craneal. Los pacientes prematuramente diagnosticados ven disminuidos las características clínicas. El retraso en la erupción se volverá un problema creciente si no se da seguimiento puntual.

Palabras clave: *Hipotiroidismo congénito; manifestaciones orales; cribado neonatal; caries temprana de la infancia.*

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