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# ORO-DENTAL MANIFESTATIONS IN A CHILD WITH VISCERAL DWARFISM

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## ABSTRACT

The oral cavity represents an important source of information about the health of a child. Oral lesions may manifest early, sometimes concomitant with a systemic disease, disappearing with general health improvement or later persisting in spite of the disease's remission. Also, these can present developmental abnormalities. Oral mucosal lesions are commonly found in patients with liver cirrhosis. We present the case of a 12 years old boy with liver transplant since he was 9 years old, for the biliary cirrhosis secondary to the biliary hypoplasia. The patient had also very low height and weight prior to liver transplant, advanced osteomalacia and many oro-dental manifestations. Severe growth delay and osteomalacia have multiple causes, due to the primary hepatic pathology and to the chronic malabsorption syndrome, also due to the post-transplant immunosuppressant medication. Oro-dental lesion type and severity depends on the age of the child. It is important to know when the systemic disorders started, their duration and type, as well as their impact. The oral cavity can be significantly affected by immunodeficiency, cholestasis, malnutrition and coagulopathy.

**KEYWORDS:** oro-dental manifestations, visceral dwarfism, child

## INTRODUCTION

The oral cavity represents an important source of information about the health of a patient. Oral lesions may manifest early, sometimes concomitant with a systemic disease, disappearing with general health improvement, or later persisting in spite of disease remission. Also, these can present developmental abnormalities (1).

Patients with liver cirrhosis can develop oral lesions. The occurrence and type of lesion depend on the degree of liver function impairment and its type, also on the severity and duration of systemic diseases. In children, it is important the age at which the early symptoms of liver disease are experienced (2). The prevalence of chronic liver disease in children and adolescents has not been determined. The necessity to perform liver transplants in children and adolescents, was estimated at 2:10 000 births (3).

## CASE REPORT

We present a 12 years old boy who was hospitalized in the pediatric gastroenterology clinic for height and weight hypotrophy, chronic diarrhea, colicky abdominal pain.

The physiological personal history shows that was he was a term baby, eutrophic, weight at birth = 3360g, with Apgar score 9, without neonatal issues. At the age of one month he suffered from severe jaundice and at the age of three months he was diagnosed with biliary hypoplasia (liver

biopsy). The patient developed biliary cirrhosis, portal and parenchymal decompensated secondary to the biliary ways hypoplasia, a condition for which, at the age of 9 years old, he underwent liver transplantation from a living donor (mother).

Prior to transplantation, he suffered from disabling itching, subsequent to the chronic severe cholestasis and he had numerous fractures of the femoral pathological bone with faulty consolidation (at the age of 3, 6 and 9 years old) resulting in the distortion of the legs, especially the left limb, and at the age of 10 years old he developed osteoporosis in the long bones. At the age of 11 years old, he was diagnosed with urinary tract infection with Proteus and Enterobacter and he underwent repeated rounds of antibiotics. Before the liver transplantation the protein-calorie malnutrition emerged.

The clinical examination shows the severely stature-weight drawback H = 105 cm (-7.51 SD), W = 20 kg (-6.8 SD), pale skin, slightly dysmorphic face, scleral icterus, marked dental dystrophy, postoperative front abdominal scar, unequal and deformed legs with the shortening of the left lower limb, globular chest, symmetrical rib movements, normal cardiopulmonary sounds, distended abdomen, meteoroids, sensitive to palpation without movable dullness, accelerated bowel movements with daily watery stools.

The biological tests revealed a microcytic hypochromic anemia, hyposideremia, hypoproteinemia, hepatocytolisis syndrome, but without cholestasis syndrome and with negative immunological tests for celiac disease. Protein electrophoresis, immunogram, urinalysis, stool showed no pathological changes. The digestion sample revealed a stool with acid pH, presence of starch and fats, suggesting the existence of a malabsorption syndrome. The colonoscopy objectified non-specific rectocolitis lesions without abnormalities of caliber, diverticula or polyps.

The endocrine assessment revealed normal thyroid function; normal IGF, PTH and normal Turkish saddle radiography.

Oral clinical examination was performed to assess oral mucosa, hygiene, gingiva and teeth. Thus, the oral clinical examination (difficult due to microstomia) revealed the existence of dental abnormalities regarding the color, position and size (Figure 1). The teeth have a greenish-brown color, drawing the attention to the systemic changes during the creation and mineralization of the enamel. Palpation revealed some tooth areas (porous, with poor mineralization) where enamel was absent, causing the intense brown color of the dentin, a premise of the dental and periodontal lesions, making cleaning difficult. The existence of spaces in frontal jaw drew the attention to the absence of maxillary canines (1.3, 2.3) on the arch and the space necessary for their eruption and alignment. The lack of space was obvious, causing the palatalization of the second right premolar (1.5 is located on the palatine compared to 1.4).

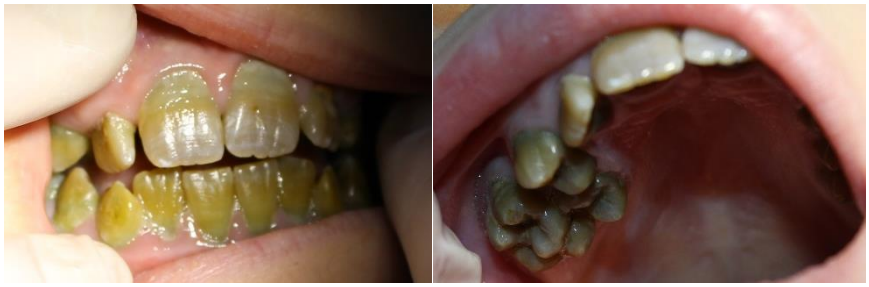

Figure 1. *Dental abnormalities regarding the color, position and size*

The chronic open pulpitis from 4.6, of multifactorial etiology, has been difficult to treat under the systemic treatment conditions. Generalized gingivitis was evidenced by changes in the shape and color of the gums, without the existence of the gum-periodontal sockets, being the outcome of the retaining of the plaque on the rough surface of the affected tooth enamel, given that the self-cleaning was hampered by dental crowding.

The orthopantomography revealed that although there is a slight delay of eruption (at the age of 12 the second molar on the four hemi-arcades should have erupted) there are buds in correct position, potentially eruptive. On the jaw there was evidence of the wisdom teeth buds. The jaw revealed the existence of two fully formed canines that had not enough place on the arch, which leads, once more, to the need of the intervention of an orthodontist (Figure 2).

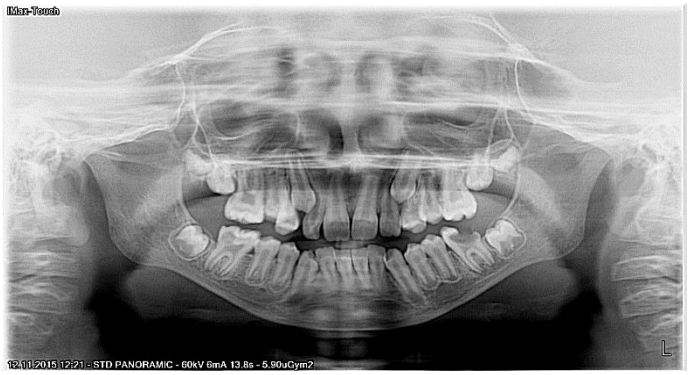


Figure 2. *The orthopantomography*

**DISCUSSION**

Clinical manifestations of liver failure depend on the degree of organ damage. In decompensated cirrhosis, the patient develops jaundice, symptoms of malnutrition, ascites, esophageal varices, edema and coagulation disorders. Underlying diseases in cirrhosis of the liver may manifest, and drug side effects may occur after glucocorticosteroids or other immunosuppressant administration in autoimmune liver disease (4).

In the case of our patient, the severe growth retardation and the osteomalacia have multiple explanations, both through the primary liver pathology and the chronic malabsorption syndrome and the post-transplant immunosuppressive medication.

Lesion type and severity depends on the age of the child, when the systemic disorders started, their type and duration, as well as their impact. The oral cavity can be significantly affected by malnutrition,

cholestasis, immunodeficiency and coagulopathy (5).

In our case, the oro-dental changes are a consequence of the systemic diseases and they have changed the structure of the tooth enamel, along with an underdevelopment of the jaw bones which made impossible the correct eruption of the teed developed correctly from the dimensional point of view. Thus, it was necessary to include an orthodontist in the multidisciplinary team dealing with the child.

The risk of cirrhotic manifestations depends on the severity of liver impairment. Two basic scores, MELD (Model of End-Stage Liver Disease) and PELD (Score of Paediatric End-Stage Liver Disease) and the Child-Pugh score are used to assess liver functions in order to determine organ transplant indications in children under the age of 12 years (6). MELD uses the patient's values for serum bilirubin, serum creatinine, and the INR to predict survival (7). MELD score in our child was 14.

Up-to-date publications suggest that children with liver failure are more prone to oral mucosa lesions than generally healthy children. These might result from malnutrition, hypoproteinemia, coagulopathy, cholestasis or immunodeficiencies (8).

The dental-alveolar incongruence is a consequence of the reduced development of the jaw bone in the context of the dwarfism; the jaw bone supports the teeth which are unaffected dimensionally by the visceral syndrome. In this context, the inter-arch dental relationships are also affected (static and dynamic occlusion) as well as the mandibulo-cranial relationships, the little boy having a posture with a magnified vertical dimension of the lower floor.

In patients with advanced liver disease, coagulopathy associated with thrombocytopenia and plasma deficiency of coagulation factors may cause petechiae and spontaneous gingival bleeding. Bleeding disorders, petechiae, increased vulnerability to bruising, and gingival bleeding were described in children with liver dysfunctions (9).

In our patient, the oral breathing determines the specific facies, elongated, with underdeveloped cheekbones and dry looks of the lips, the upper incisors exposed and unprotected by saliva which also allows the decay of the front teeth.

Malnutrition accompanying liver failure manifests as qualitative deficits and infections. Typical symptoms of malnutrition such as angular fissures, cheilitis and glossitis in children with chronic liver diseases, were reported by researchers (10). The authors of a study confirmed that 58% of adult liver transplant candidates had one or more oral mucosa abnormalities: fissured tongue – 37%, atrophy of the tongue papillae – 18%, angular cheilitis – 4%, candidiasis – 2% (11).

In a study, from 34 children who underwent a liver transplantation under the age of 6 years, 61.3% with congenital biliary atresia presented tooth staining and green gingivae (12). Other authors report that green or greenish brown dental discolouration results from biliverdin accumulation in dental tissues when cholestasis develops during tooth development (13). There were reported frequent cases of green dental discolouration (50–61.3%) in children with hyperbilirubinaemia and biliary atresia (14).

Oral mucosal lesions are frequent found in children with liver cirrhosis. Advanced liver disease promotes oral candidiasis. The severity of gingivitis correlates with the presence of dental plaque (15).

**CONCLUSIONS**

Severe growth delay and osteomalacia have multiple explanations, both through primary hepatic pathology and chronic

malabsorption syndrome and post-transplant immunosuppressive medication.

Lesion type and severity depend on the age of the child when the systemic disorders started, their type and duration, as well as their impact. The oral cavity appears to be significantly affected by immunodeficiency, malnutrition, cholestasis, and coagulopathy.

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