International Journal of Dentistry and Oral Health

Volume 7 Issue 7,
August 2021

Citation
Ohoud Tawfig Zain Elabdeen et al. (2021), Oro Facial Digital Syndrome (MOHR SYNDROME) Int J Dent & Ora Hea. 7:7,

ISSN 2471-657X

Published by
Biocore Group |
https://www.biocoreopen.org/ijdoh/archive.php

Case Report

Oro Facial Digital Syndrome (MOHR SYNDROME)

Ohoud Tawfig Zain Elabdeen*[1], Nuha Fathelrhman Yahya[2]

[1]Pediatric dentist, BDS: University of Medical Science and Technology, Sudan, Msc of pediatric dentistry: Cairo University
[2] BDS: National Ribat University, Sudan, Resident in pediatric dentistry : Sudan medical specialization board.

Corresponding author: Ohoud Tawfig Zain Elabdeen
Pediatric dentist, BDS: University of Medical Science and Technology, Sudan, Msc of pediatric dentistry: Cairo University. Tel: +249915164019 E-mail: Oya_sharfy@hotmail.com

Article History: **Received:** July 27, 2021;
Accepted: August 05, 2021;
Published: August 16 2021.

Abstract

Oro-facial digital syndrome is a group of related conditions that affect the development of oral cavity, facial structures and digits; along with different degrees of mental retardation and Additional signs involving (CNS) and visceral organs; there are at least 13 clinical types of OFDS. This is the case report of afive year old boy with OFDS type II (Mohr syndrome) presented with oral, facial and limbs deformities. Medical history, physical examination and genetic analysis play a major role in recognizing which type of OFDS. Management of patients with OFDS type II need multidisciplinary team approach (plastic and orthopedic surgeon, pediatric dentist and speech therapist).

Keywords

Oro-facial Digital Syndrome, Mohr Syndrome, Polydactyly, Sunductyly.

Declaration of Conflicting Interest

The authors declare that they have no conflict of interest.

Introduction:

Oro-facial digital syndrome is group of related conditions that affect the development of oral cavity including the mouth, tongue, teeth, and jaw and the development of the facial structures including the head, eyes, nose; fingers and toes (digits); along with different degrees of mental retardation and Additional signs involving the central nervous system (CNS) and visceral organs(1,5). The oral signs and symptoms include unusual lobed tongue, teeth anomalies, hyperplastic frenula, Cleft lip &palate, wide nose, flat nasal bridge, hypertelorism ,syndactyly, brachydactyly ,clinodactyly and polydactyly (1).

Currently, at least 13 clinical type of Oral-facial-digital syndrome has been identified which can be classified according to the existing anomalies into: -
OFDS type I; Papillon-Leage/Psaume syndrome.
OFDS type II; Mohr syndrome .
OFDS type III Sugarman syndrome.
OFDS type IV; Mohr-Majewski Baraitser syndrome.
OFDS type V Thurston syndrome.
OFDS type VI Varadi syndrome.
OFDS type VII.
OFDS type VIII; Edwards syndrome.
OFDS type IX.
OFDS type X; Figuera syndrome.
OFDS type XI; Gabriellisyndrome.
OFDS type XII; Moran Barroso Syndrome.
OFDS type XIII; Degner syndrome.

Oro facial digital syndrome has incidence of 1 in 50,000 to 250,000 newborns (2). Type I accounts for the majority of cases of this disorder and can be easily recognized by its typical X. Linked dominant pattern of inheritance in familial cases (3). Most of other types are transmitted as autosomal recessive syndrome .recently , 11 genes have been identified known to be responsible for OFDS which allow better clinical and genetic definition for this heterogeneous condition(3)(4).

Case presentation:

A five years old Sudanese boy, who happened to be a twin for a girl and was born of consanguineous parents, came to our dental center referred from a university clinic, with his father complaining of multiple carious teeth. History was taken and parents were healthyas well as his twin sister, they had no history of medical problems or history of congenital anomalies.This child was the last of his siblings. Upon examination the patient presented with typical features of OFDS typeII (mohr syndrome). Which include some oral, facial and limbs deformities.

Oral and facial deformities found were: high arched palate, premature eruption of the lower central incisors,missing upper central incisors, midline cleft lip, lobulated tongue or so called tongue nodules, broad nose, in addition to low set ears.

Limbs deformities found were: bilateral polydactyly of hands and feet as well as brachydactyle of the fingers, sunductyly of the big toes in both feet. He was able to move all the limbs symmetrically and no muscular hyotonia was recorded.

Patient was un-able to communicate with us, his father said he did not speak yet, and he had mild degree of mental retardation.

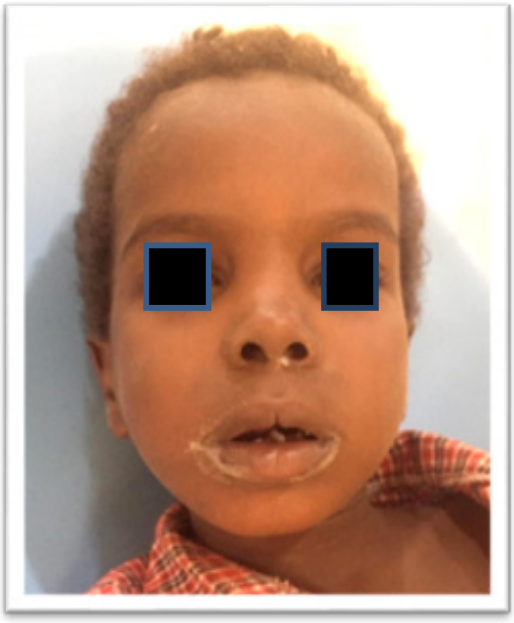

Figure (1): picture shows midline cleft lip, broad nose, low set ears and hypertelorisim.

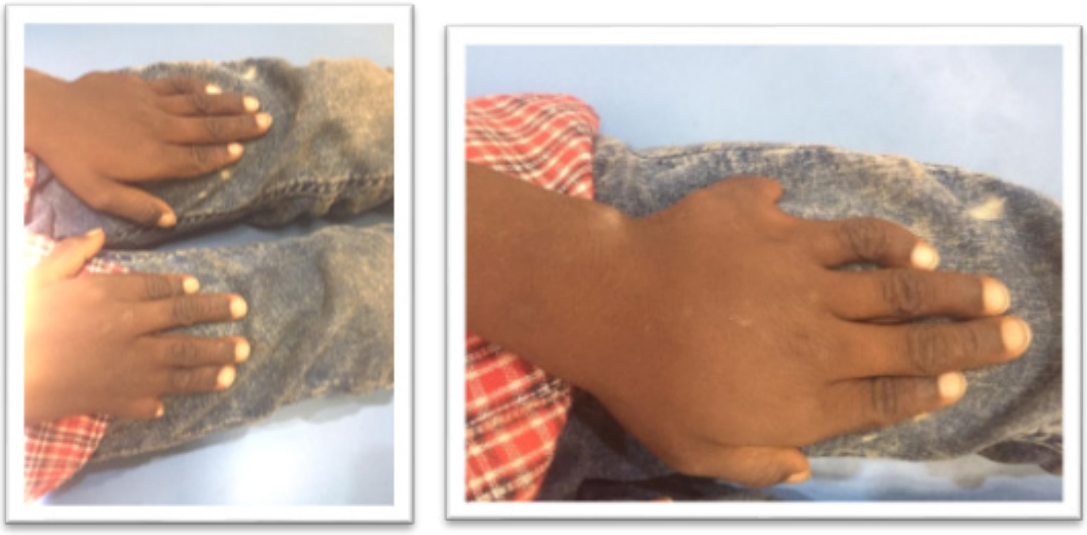

Figure (2): shows polydactyly of the fingers plus brachydactyly (fingers appears short).

Figure (3): shows polydactyly and sunductyly of the big toe in both feet.

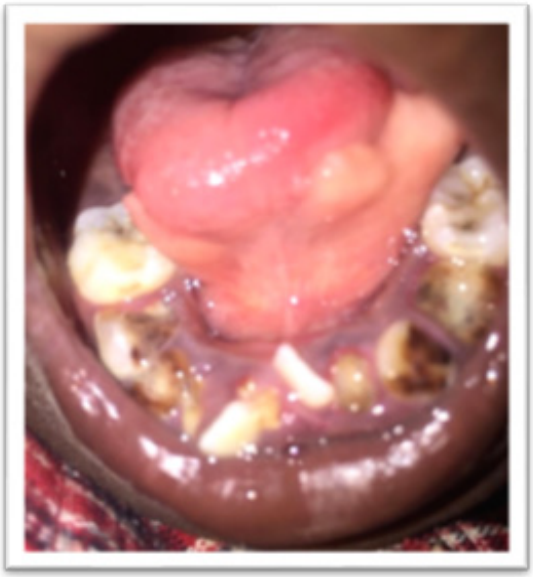

Figure (4): shows tongue nodules, premature eruption of lower central incisors and severely carious primary teeth.

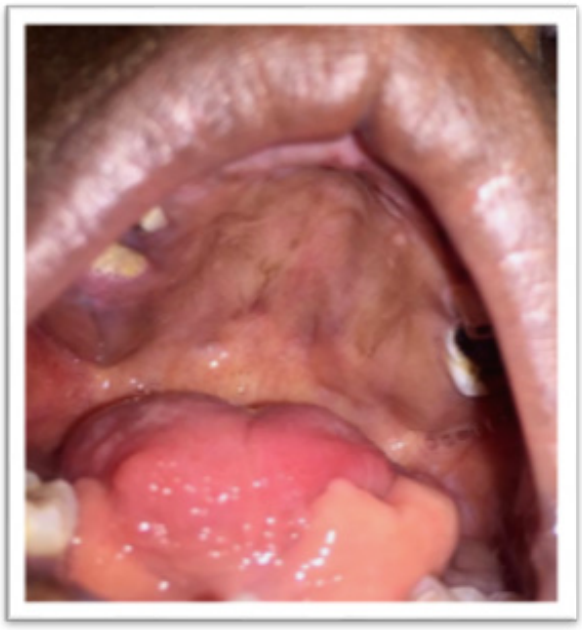

Figure (5): shows high arched palate, tongue nodules, and missing upper central incisors

Discussion:

Oro-facial digital syndrome includes many subtypes with wide variety of signs and symptoms that makes it hard to diagnose. so good medical history and physical examination of patients as well as genetic analysis may play a major role in recognizing which type of OFDS the patient suffers from(5).

In this case diagnosis of OFDS type II (mohr syndrome) was mainly clinically and by taking the medical and family history of the patient plus thorough physical examination that showed typical symptoms of oral, facial and limbs deformities.

Since Mohr syndrome is a rare disease that may require several surgical intervention and reconstruction of the limbs, facial and oral deformities, we referred this patient to a plastic and orthopedic surgeonsfor further evaluation. Also close follow up with a pediatric dentist was done for full mouth rehabilitation.

Regarding the patient un-ability to speak which is alarming around this age he was referred to a speech therapist and behavioral analyst to evaluate his condition.

Patients with OFDS type II need multidisciplinary team approach to follow up their condition and development throughout their life(6).

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