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Case Report

Oral Manifestation in a Patient with Bilateral Duane Syndrome: A Case Report

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ABSTRACT

Duane syndrome is a rare retraction anomaly characterized by congenital non-progressive horizontal ophthalmoplegia and other systemic signs. No data has been yet registered about oral manifestation of Duane syndrome. In this article we present a six years old male patient was diagnosed as having Duane retraction syndrome. He presented skeletal Class III with both maxillary and mandibular protrusion, counter clock-wise mandibular rotation, lingualized maxillary and mandibular incisors, first mandibular molar agenesis, asymmetric morphology of the mandibular condyles and multiple decay lesions. Postero-anterior x-ray showed an asymmetrical craniofacial structure. The aim of this case report was to describe the oral signs of a patient affected by DRS.

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Introduction

The Duane Retraction Syndrome (DRS) is a congenital disorder typically characterized by difficulty abducting and adducting one or both eyes. DRS is due to miswiring of the eye muscles and in patients affected by this syndrome, the sixth cranial nerve (that controls the lateral rectus muscle) does not develop properly. This condition was first described by Sinclair, Turk and Stilling in 1895 and in literature it is reported a frequency of 1-5% in general population with strabismus [1]. Previous study associated DRS with multiples anomalies such as crocodile tears, mental retardation and spina bifida occulta [2-4]. No data has been yet registered about oral manifestation of Duane syndrome. The aim of this case report was to describe the oral signs of a patient affected by DRS.

Case Report

Our patient was a little male child. He was 6 years old, born out of a non-consanguineous marriage with an euthotic birth after a pregnancy affected by oligoamnion and old placenta. During the perinatal period the patients reported cardio-respiratory depression, cyanosis, transitory itterus. At the age of 6 years old he underwent the genetic evaluation that putted in light auxological parameters 10°pc, facial dimorphism, ipoacusia and no cardiovascular affections. SNP-Array underlined two microdeletions: one located in the long arm of chromosome 2 of 2q22.3

region and one located in the long arm of chromosome 17 of 17q23.1q23.2 region. These two chromosomal defects confirmed the genetic diagnosis of Duan Syndrome [5].

On examination, his height was found to be 110 cm, with a weight of 18 kg. Extra-oral examination revealed facial asymmetry with reduced intercanthal distance and facial dimorphism (Figure 1 & Figure 2). Intraoral examination revealed bald and fissured tongue with short lingual frenulum and hypotrophic gingiva. The patient presented microdontia with oligodontia; teeth presented severe enamel defects as *amelogenesis imperfecta* that conducted to destructive decay lesions of deciduous teeth. In order to evaluate dental arch dimension, the Korkhaus' index was performed on patient's dental casts showing a considerable reduction of pre-maxilla diameter [6-8]. The patient underwent to X-rays ortopantomography and teleradiography exams in order to assess cephalometric evaluation.

Cephalometric analysis was conducted according the Ricketts and Tweed norms (Table 1 & Figure 3) [9, 10]. His skeletal pattern was skeletal Class III with both maxillary and mandibular protrusion (probably due to the point Nasion retrusion), counterclockwise mandibular rotation, lingualized maxillary and mandibular incisors. Dental x-ray showed first mandibular molar agenesis, asymmetric

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morphology of the mandibular condyles and multiple decay lesions. Postero-anterior x-ray showed an asymmetrical craniofacial structure.



Figure 1: Frontal extra-oral view.



Figure 2: Lateral extra-oral view.



Figure 3: Cephalometric analysis.

Table 1: SNA, SNA angle; SNB, SNB angle; ANB, ANB angle; FMIA, Frankfort – Mandibular Incisor Angle; FA, Facial Axis angle; LFH, Lower Facial Height; FD, Facial Depth angle; II, Interincisive angle.

VARIABLE	NORMAL	REPORTED	DIFFERENCE
SNA	82±2	86.1	4.1
SNB	80±2	87.1	7.1
ANB	2±2	-1	-3
FMIA	67±3	91	24
FA	90±3	96.6	6.6
LFH	47±4	39.8	-7.2
FD	89±3	102.7	13.7
II	130±8	158.3	28.3

Discussion

Duane’s Retraction Syndrome (DRS), also named Stilling-Turk-Duane syndrome, is a congenital oculomotor disease caused by the hypoplasia or the absence of the abducens nucleus. DRS is a rare syndrome (represent approximately 1% of total cases of strabismus). Previous studies associated this syndrome with ocular (nystagmus, anisocoria, ptosis, optic nerve colobomas, etc.), cardiac (auricular septal defect), skeletal (scoliosis, spina bifida, limb hypoplasia, etc.), genitourinary (renal agenesis and vesicoureteral reflux) anomalies [3, 4]. DRS has also been associated with a number of specific congenital syndromes including Klippel-Feil syndrome, Wildervanck syndrome (Duane syndrome, Klippel-Feil anomaly, and deafness), Holt-Oram, morning-glory syndrome, Goldenhar syndrome, Okihiro syndrome (Duane syndrome and radial ray defects), and Usher Syndrome Type 2. No oral manifestation related to DRS have been described earlier in literature.

Our patient presented stomatognathic defects as micrognathia, teeth agenesis, maxillary and mandibular prognathism (probably due to the hypoplasia of frontal sinus that retruded the nasion and microcephaly) and short tongue frenulum. The Korkaus’ Index was performed to verify micrognathia and, despite the teeth defects, reported a great reduction of premaxilla diameter. Dental panoramic x-ray showed an abnormal growth of mandible and particularly of mandibular branch with also an alteration of TMJ heads (Figure 4 & Figure 5).

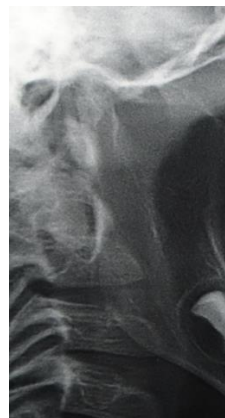


Figure 4: Right mandibular branch.



Figure 5: Left mandibular branch.

Conclusion

DRS is a genetic condition present from birth, and usually recognized in early childhood. It shows usually audiologic, otologic and ocular signs and symptoms. Our observation of oral manifestations in a boy with DRS may prompt a prospective and retrospective review of other patients affected by this syndrome in order to establish if these anomalies are part of it.

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