Research Open

Journal of Dental and Maxillofacial Research

Volume 1 Issue 1

Case Study

Rieger Syndrome: A Case with Congenital Absence of Premaxillary Area

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Received: September 03, 2018; **Accepted:** September 10, 2018; **Published:** September 25, 2018;

Case Study

Rieger syndrome is characterized by absent maxillary incisor teeth, malformation of the anterior chamber of the eye, and umbilical anomalies [1]. A case with congenital absent of premaxillary area is presented.

The patient, a 10 7/12 -year-old boy, was born to young (father 25, mother 16), non-consanguineous, apparently normal parents, after an uneventful, full-term pregnancy. Hydramnios and a long-lasting delivery is reported. He weighed 3000 gr. at birth and had choanal atresia, bilateral aniridia, glaucoma and inverted strabismus. His younger 7-year-old brother and 4-year-old sister are reportedly normal. No similar case among relatives is reported.

Physical and radiologic examination showed absence of premaxllary area and incisor teeth hypodontia and delayed eruption of permanent dentition, short facial height (-3.0 SD) highly arched narrow palate, (narrow free border of soft palate, with small uvula, hypertrophic tonsils), severely short palatal plane (–5.4 SD) and concave skeletal profile (–5.6 SD) posterior displacement of maxillary sinuses and projection of the periumbilical skin (dry palmar skin low posterior hairline). Intelligence was normal.

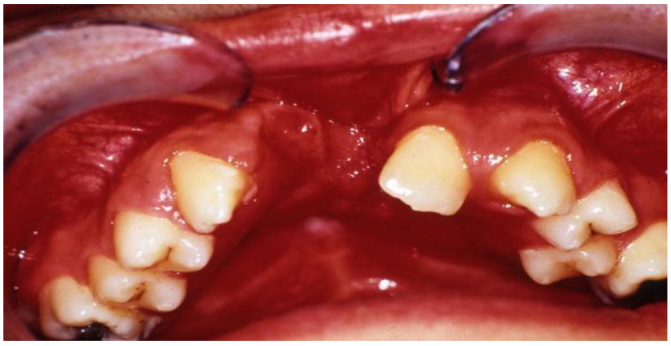

Figure 1. Absence of premaxillary area.

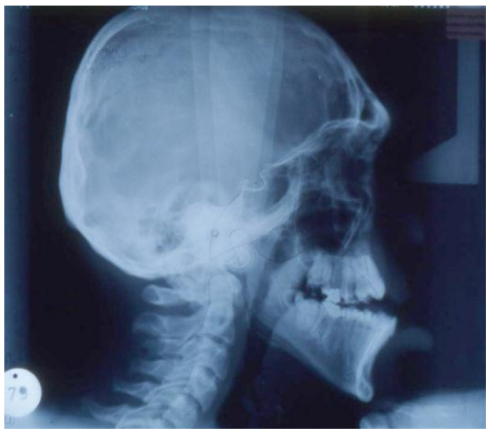

Figure 2. Lateral cephalometric radiography Short Facial Height, concave profile.

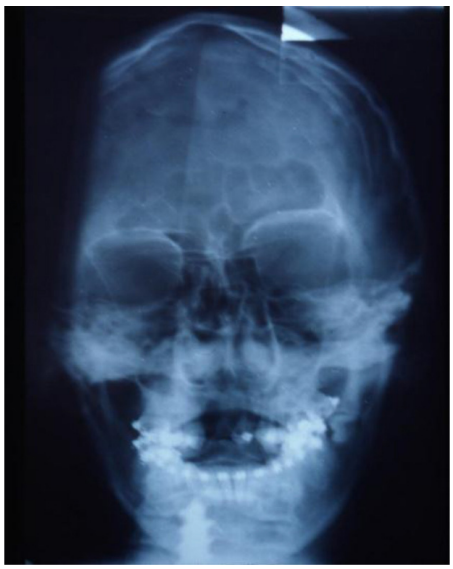

Figure 3. Posterior-front cephalometric radiography. Absence of premaxillary area, infraorbital bony distance.

DOI: 10.31038/JDMR.1000103

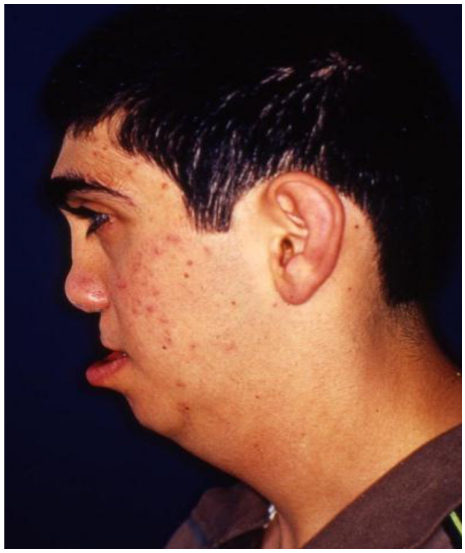

Figure 4. Concave profile.

His karyotype was normal, 46, XY (G-bands).

Panoramic radiograph

Absent teeth

52, 51, 61, 62

18 13, 12, 11, 21, 22, 23 28

48 45, 43, 41 31 33 35 38

Cephalometrics

Patient 10.5 -year-old	Father 35-year-old

Cranial base

S-N 71.8 mm (-2.2 SD)	70 mm -3.5 SD
S-Ba 44 mm (-0.6 SD)	48 mm norm
S-N-Ba 129.6 dg (0.1 SD)	126 dg norm
SN-FH 151 dg (3.1 SD)	
ANS-PNS 43 mm (-5.4 SD)	50 mm -3.0 SD

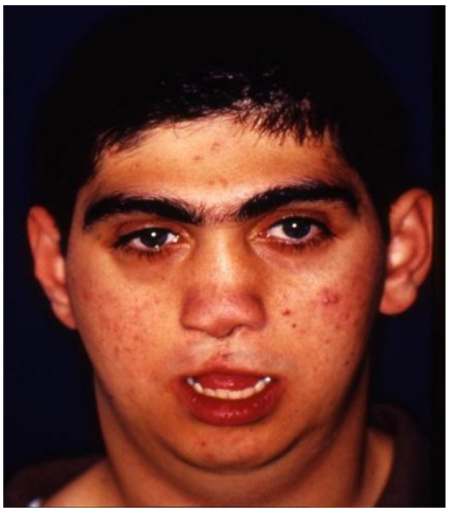

Figure 5. Surgically corrected congenitally absent philtrum.

Skeletal Relations

Facial Angle 90.6 dg (2.3 SD) (PN-FH)	88 dg 3.0 SD
Lande's Angle 81.0 dg (-1.2 SD) (AN-FH)	91 dg 3.0 SD
Convexity -17.8 dg (-5.6 SD) 180-(NAP)	

Vertical Analysis

Mandibular Plane	20.2 dg (-1.9)	23 dg
Y-Axis	51.8 dg (-2.5)	
UFH (N-ANS)	47.1 mm (-2.0)	61 mm norm
TFH (N-Me)	106.7 mm (-3.0)	132 mm norm
UFH/TFH	44.2% 43.93%	46.21% SNA 82 dg norm SNB 80 dg norm ANB 2 dg norm

Anterior Cranial Base: Moderate Short	Severely short
Posterior Cranial Base: Normal	normal
Saddle Angle: Normal	normal
Palatal Plane: Severely short	Severely short

Maxilla: Mildly retruded to forehead severely protruded to forehead well related to anterior cranial base

Mandible: Prognathic to forehead severely protruded to forehead well related to anterior cranial base

Convexity: Severely decreased; concave skeletal profile Overclosure tendency Maxilla and mandible well related to each other Bony interorbital Distance: 18 mm 23 mm

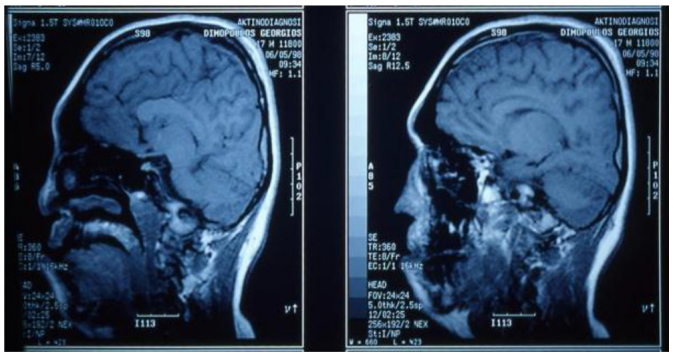

Figure 6. Lateral MRI tomography showing posterior displacement of maxillary sinuses.

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Reference

1. Gorlin RJ, Cohen Jr, MM Hennekam RCM (2001) Syndromes of the Head and Neck, OXFORD Universal Press. Rieger syndrome (hypodontia and primary mesodermal dysgenesis of the iris). Pp: 1181–1183.

Citation:
Euterpe Bazopoulou (2018) Rieger Syndrome: A Case with Congenital Absence of Premaxillary Area. *J Dent Maxillofacial Res* Volume 1(1): 1–3.
DOI: 10.31038/JDMR.1000103