Joint Event

38th International Conference on

Advanced Pediatrics and Neonatology

29th Global Dentists and Pediatric Dentistry Meeting

8th International Conference & Expo on

Euro Optometry and Vision Science

March 20-21, 2023

Rome, Italy

Andres Salazar et al., Gen Med 2023, Volume 11

Pfeiffer syndrome: A middle face distraction treatment by a modified Lefort III technique

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Pfeiffer Syndrome: Discovered by Arthur Pfeiffer in 1964, with a prevalence of 1:100,000. It is characterized by the clinical presentation of craniosynostosis, hypoplasia of the middle third of the face, syndactyly of the hands and feet. It usually presents with a bilateral coronal synostosis associated with one or more sutures. The dysmorphic skull presents with a transverse magnification greater than anteroposterior, reflecting an enlarged frontal region, depressed midface with a small nose and low nasal bridge with moderate to severe ocular propotosis with the presence of ocular hypertelorism. Cohen classified it in 1993 into 3 types: Type I is a mild skeletal <u>craniofacial</u> disorder with normal neurological development; Type II is a pansynostosis-type craniofacial disorder, with severe proptosis, skeletal abnormalities in stiff fingers and elbows and limited delayed neurological development; Type III is the same as Type II without pansynostosis. Genetics is associated with the FGFR2 gene mutation in 95% of cases and is represented by types II and III, while the FGFR1 gene mutation is found in 5% represented by type I.

The treatment of these patients is the correction of the present signs and symptoms, for which initially the cranial correction of the synostoses is performed by means of cranioplasty at one year of life, followed by distraction or craniofacial advances at 6 to 8 years of life, later management of sequelae in facial projection, especially in nasal management. The emphasis on ocular proptosis is important since surgical treatment is focused on reconstructing the orbital continent.

Treatment is always multidisciplinary, involving <u>neurosurgery</u>, craniomaxillofacial surgery, otorhinolaryngology, oculoplasty, internal medicine.

Biography

Andres Salazar is a new maxillofacial surgeon recently graduated from Pontificial Javierian University located in Bogota, Colombia. His principles based on his career are to prevent, diagnose and treat all disease of the <u>maxillofacial complex esqueletal</u> and soft tissues with the unique goal that is to give back quality of life to his patients. This workshop about the Middle Face Distraction has been done when he was a resident of four year guided by their professors Dr. Andres Duque and Dr. Jaime Jimenez who authorized to him to be the speaker of their clinical case done together.

Received: February 27, 2023; Accepted: March 01, 2023; Published: March 20, 2023

General Medicine: Open Access Volume 11

ISSN: 2327-5146