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Developmental facial paralysis: A review

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Asymmetrical crying faces;
CHARGE

Summary The purpose of this study is to clarify the confusing nomenclature and pathogenesis of Developmental Facial Paralysis, and how it can be differentiated from other causes of facial paralysis present at birth.

Differentiating developmental from traumatic facial paralysis noted at birth is important for determining prognosis, but also for medicolegal reasons.

Given the dramatic presentation of this condition, accurate and reliable guidelines are necessary in order to facilitate early diagnosis and initiate appropriate therapy, while providing support and counselling to the family.

The 30 years experience of our center in the management of developmental facial paralysis is dependent upon a thorough understanding of facial nerve embryology, anatomy, nerve physiology, and an appreciation of well-recognized mishaps during fetal development.

It is hoped that a better understanding of this condition will in the future lead to early targeted screening, accurate diagnosis and prompt treatment in this population of facially disfigured patients, which will facilitate their emotional and social rehabilitation, and their reintegration among their peers.

Introduction

Congenital facial paralysis (CFP) refers to conditions that are acquired during or at birth (e.g. from trauma), while developmental facial paralysis (DFP) is the result of developmental mishaps during fetal development.[1] DFP can present in isolation or as part of a recognised syndrome, such as Möbius, Goldenhar, CHARGE, etc. (Figure 1).

Studies on the incidence of facial paralysis in the newborn demonstrate great heterogeneity, (from 1.4 up to 64 per 1000) while the nomenclature has been confusing.[2–5] Given the dramatic presentation of this condition, accurate and reliable guidelines are necessary in order to facilitate early diagnosis and initiate appropriate therapy, while providing support and counselling to the family.

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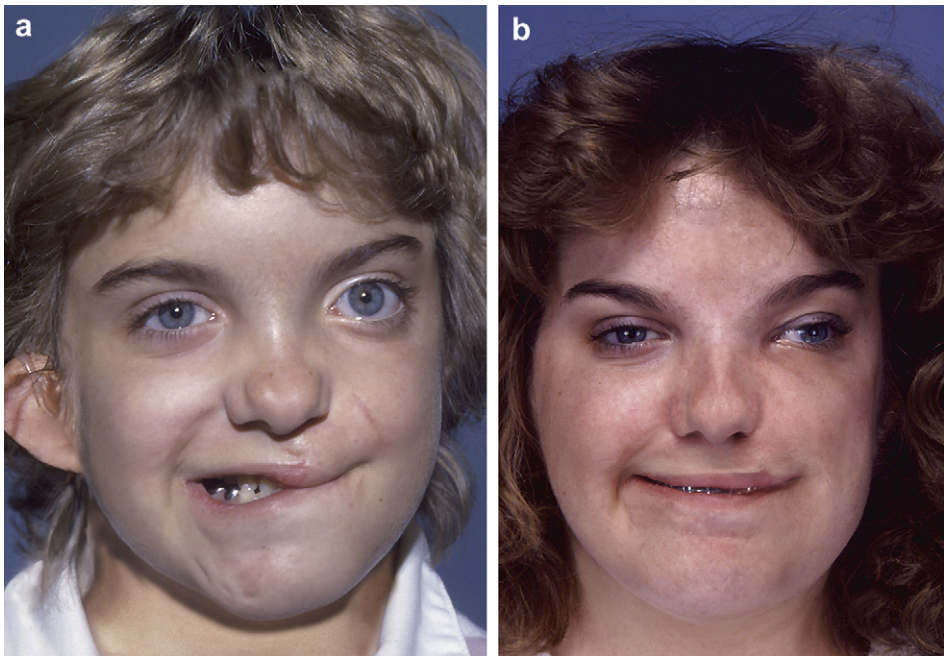

Figure 1 Example of Developmental facial paralysis with multiple anomalies. This 10-year old girl presented with left (L) sided developmental facial paralysis (Figure 1a). She was born at full term gestation with normal delivery and her antenatal and birth history was unremarkable. Left facial paralysis was noted at birth. She also had associated cleft lip and palate anomaly that were repaired at the age of 3 weeks and 11 months respectively, aberrant right subclavian artery, bilateral developmental conductive hearing loss (VIII) and external ear deformities (cupping and constriction) and left spinal accessory nerve (XI) deficit with weak shoulder and left neck webbing. Genetic screening was normal. She had CFNGs X 3 in our Center and fifteen months later she had a right free pectoralis minor transfer to left cheek for smile restoration. Sixteen months later she had a right pedicle frontalis muscle transfer to the left upper and lower eyelid, neurotized by the upper CFNG that carried motor fibers from the upper zygomatic branch of the right facial never, to substitute for the missing orbicularis oculi muscle. Her last surgery involved pedicle transfer of the left digastric muscle to substitute for the left depressor complex, neurotized by the lower CFNG that carried motor fibers from the mandibular branch of the right facial nerve, along with a Rhinoplasty, and scar revision of her previously repaired cleft lip. Patient at her last follow-up visit when she was 16 years old (Figure 1b).

Differentiating developmental from traumatic facial paralysis noted at birth is important for determining prognosis, but also for medicolegal reasons.[6,7] In general, DFP carries a poor functional prognosis and the importance of early recognition and treatment has been stressed by many authors, while most traumatic cases are likely to recover spontaneously.[3—5]

The purpose of this study is a thorough literature review of the pathogenesis and evaluation of Developmental Facial Paralysis, and how it can be differentiated from other causes of facial paralysis present at birth.

Methods

A literature search for the years 1966—2010 using the NLM PubMed using the keyword groups "facial nerve paralysis pediatric, developmental, congenital" was performed. In addition, Embase, CINAHL, Citation Manager and a number of other key biomedical electronic databases were searched. Relevant information was also identified by hand searching all the Plastic Surgery Journals since 2000, by scrutinizing the cited references of all facial paralysis articles and the Conference Proceedings.

Anatomy

The facial nerve arises from the brain stem nuclei.[8,9] The motor fibers of the facial nerve loop dorsally around the abducens nerve nucleus and exit at the cerebellopontine angle. The parasympathetic and sensory fibers form the nervus intermedius which join the motor root of the facial nerve as it exits the brain stem.

The course of the facial nerve is divided into the cisternal segment in the cerebellopontine angle, the intracanalicular segment, the labyrinthine segment and the tympanic segment (separated by the anterior genu), the mastoid segment (separated by the posterior genu), and the extracranial segment.[10,11] Several branches are given off during the intrapetrosal course. The extracranial segment consists mainly of the parotid portion of the facial nerve and its terminal branches that innervate the facial musculature.

Embryology

The facial nerve begins to develop at 3 weeks of life from the facio-acoustic primordium and is intimately related to the structures of the middle ear, external ear, parotid