

**304****HYPERTELORISM AND MIDLINE FACIAL DEFECTS. A CLINICAL STUDY OF 63 PATIENTS.**

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The objective of this study was establish the different conditions in 63 patients with anomalies related to craniofacial midline including the evaluating of the frontonasal process involvement in these conditions and delineating the probable new syndromes. Minimal diagnostic criteria for inclusion in the study were the presence of hypertelorism associated with, at least, one facial midline defects, involving the nose and/or the lips. The results have shown that 3 patients had classical frontonasal dysplasia and 60 had syndromes that overlap frontonasal dysplasia. Primary frontonasal process involvement was present in 20 cases (3 of them with classical frontonasal dysplasia; 4 with acromelic frontonasal dysplasia; 10 with frontonasal dysplasia with corpus callosum agenesis and mental retardation; 2 with frontonasal dysplasia and hemimelia, and 1 case without diagnose). In 43 cases the frontonasal process was secondarily affected probably due to: early fusion of cranial sutures (15 cases); etmoidal or fronto-etmoidal sutures involvement resulting in basal or frontonasal encephaloceles (12 cases); multiple cranial vault bones defects (4 cases); and first branchial arch involvement (8 cases). In 3 cases, the diagnose was syndromic conditions that, usually, have no frontonasal involvement (Robinow syndrome and G/BBB syndrome). Findings from our study included 10 known syndromes with facies similar to the classical frontonasal dysplasia (acrofrontofacionasal disostosis; severe acrofrontofacionasal dysostosis; hypertelorism Teebi syndrome; craniofrontonasal dysplasia; frontofacionasal dysplasia; oculoauriculofrontonasal syndrome; acromelic frontonasal dysplasia; midline facial cleft, ocular anomalies, basal encephalocele, and corpus callosum agenesis syndrome; Robinow syndrome; and G/BBB syndrome) and 4 new recurrent pattern syndromes. Contact: Maria Leine Guion-Almeida E-mail: mlguion@centrinho.usp.br Medical Genetics Department USP / Bauru / SP / BRAZIL

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305**ATYPICAL CRANIOFACIAL CLEFTS: A SYNDROMOLOGICAL APPROACH**

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Cleft lip/palate and cleft palate are the commonest congenital anomalies of the face in human beings. They are usually explained by alterations in the closure of the "natural" facial embryological junctures due to different types of insults. However, in some conditions, the closure of facial prominences does not follow the conventional route, and other more complex and unusual types of facial clefts occur originating the so called atypical craniofacial clefts. Precise figures concerning prevalence are divergent according different authors, and this most likely results from problems involving classification, specially in those cases of very complex types of clefting or different types of clefts occurring in the same patient. Here, for practical purposes, we will follow the Tessier classification that take in to account the description of the facial surface aspect and the involvement of the bones of the face and cranium. These clefts are numbered from 0 to 14 considering two main landmarks: the mouth and the orbit, the later is the main landmark, since it is common to face and cranium. There is an additional cleft, the number 30, that does not fit within this sequence, since it runs through the midline of the mandible, consisting the most extreme "southbound" clefting. The clefts related to the oral region and those that run in direction of the ear, but restricted to the infra orbitary region, are the so called "southbound" clefts, and the clefts that run from the lateral canthi of the orbit in direction to the cranium through the orbital vault are the so called "northbound" clefts. Most of the facial clefts combines these two types, resulting in a large mosaic of craniofacial clefts. Our main purpose is to present the variation existing between and within these types of clefts observed in 70 patients, as well as their relationship with different craniofacial syndromes.

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307**EARLY SPEECH AND LANGUAGE DEVELOPMENT IN CHILDREN WITH CLEFT: EVIDENCE FROM AMERICAN AND SWEDISH CHILDREN.**

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Purpose: The purpose of this study was to compare the early speech and language development of children with unilateral cleft lip and palate (UCLP) and typically developing children from the US and Sweden. In the US children had complete palate repair at a mean age of 12 months of age while the children from Sweden had late hard palate closure. Subjects: 30, 18-month old children with UCLP (10 from Sweden with feeding obturators) and 20 typically developing children were studied from Johnson City, Tennessee and Goteborg, Sweden. Procedures: Language samples were collected on all children as part of comprehensive evaluation of their speech and language performance. Fifty utterances (vocalizations and words) were taken from each sample and analyzed for language and speech performance including: 1) a measure of vocalization complexity: Mean Babbling Level (MBL)(Stoel-Gammon, 1989), 2) phonetic inventory for vocalizations, 3) phonetic inventory for words, and 4) the number of words used in a language sample. Results: Children with UCLP did not differ from typically developing peers on speech measures including MBL and phonetic inventory in vocalizations for children in the US and Sweden. However, the children with UCLP did differ from typically developing children in their phonetic inventory in words and word use. Further, the Swedish children with obturators had fewer consonants than the children without obturators at the prelinguistic level. Conclusions: While the complexity and number of prelinguistic phonetic inventories did not separate the children with clefts from noncleft children in either the US or Swedish children, sound use in words did differentiate the children. Further, use of obturators did not appear to enhance sound development in this group of children. The data point to a complex relationship between early speech and language development.

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308**A PSYCHOSOCIAL STUDY OF BRAZILIAN PATIENTS WITH CLEFT LIP AND PALATE**

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Psychosocial adjustment aspects were evaluated in a sample of 32 patients with cleft lip and palate, 13 of them had bilateral clefting and 19 unilateral. Age range was 15 to 21 years and sex ratio was 1:1. All patients selected had been submitted to primary surgical procedures and some of them were waiting for esthetical surgery. Surgical corrections, orthodontic treatment, speech therapy and others procedures were performed at Hospital de Reabilitação de Anomalias Craniofaciais, USP, Bauru. This study was based on answers of parents and sons to specific questionnaire in elucidative interview. The questions include social characteristic about family, education, employment, relationship; clinical aspects about surgeries and treatments; psychological experience related to cleft, parents satisfaction and self-satisfaction with treatments received, as well as about others interventions they wish. The results of this study showed a relatively good adjustment. All patients were single and had not a well defined profession probably due to age group they belong. In relation to socioeconomic status, the majority belong to low superior class (53,12%) and middle-class (31,25%), conciliating both, study and work. The level education was compatible with chronological ages and nearest-age siblings. Self-satisfaction level in relation to the treatment received ranged from regular to positive, since 56,25% of the sample showed somewhat dissatisfaction with their appearance and 53,12%, with their speech. The results showed the psychosocial situation of the patients and the benefits of the treatment performed.

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