Introduction

INTRODUCTION

Congenital abnormalities of the head and neck are complex and often confusing. For these reasons it is helpful to have a classification system that helps to understand the variety of conditions. For any classification system to be useful it should ideally help to explain the aetiology and pathogenesis of the abnormality and to determine treatment. For these multi faceted and multifactorial conditions an ideal classification system is not available. Consequently , there are a number of di ff erent systems available: some are purely descriptive (e.g. Tessier' s classification of clefts), while others apply only to single conditions, such as the OMENS (O, orbital abnormal ities; M, mandibular deformity; E, ear deformity; N, nerve /uni25CF /uni25CF /uni25CF Paul Tessier , 1917–2008, French maxillofacial surgeon, considered the 'father of modern craniofacial surgery'. Jacques C H van der Meulen , 1929–2017, Professor in Plastic and Reconstructive Surgery , Erasmus University , Rotterdam, The Netherlands. Kar l Heinz Binder , 1923–2016, German dentist, documented the facial features of three children with the condition that now bears his name. Louis Edouard Octave Crouzon , 1874–1938, neurologist, Paris, France, described this syndrome in 1912. Eugene Apert , 1868–1940, physician, L'Hôpital des Infants Malades, Paris, France, described this syndrome in 1906. Rudolf Arthur Pfei ff er , 1931–2012, geneticist, Münster, Germany , described this syndrome in 1964. involvement; and S, soft-tissue abnormalities) classification of hemifacial (craniofacial) microsomia, which has utility in instituting treatment protocols.

TABLE 50.1 Types of developmental abnormalities of the face, mouth and jaws. Type Examples Cerebrocranial dysplasias Anencephaly, microcephaly Cerebrofacial dysplasias Rhinencephalic and oculo-orbital dysplasias Craniofacial dysplasias with clefting Lateronasomaxillary, medionasomaxillary, intermaxillary, maxillomandibular clefting Craniofacial dysplasias with dysostosis Sphenoidal, sphenoidal frontal, frontal, frontofrontal, frontonasoethmoidal, internasal, nasal, premaxillomaxillary, nasomaxillary, maxillozygomatic, zygomatic, zygoauromandibular, temporoauromandibular, mandibular, intermandibular Craniofacial dysplasias with synostosis Craniosynostosis: lambdoid and sagittal Craniofaciosynostosis: metopic, coronal, bicoronal Faciosynostosis: vomeropremaxillary (Binder syndrome) Craniofacial dysplasias with dysostosis and Crouzon, Apert and Pfeiffer syndromes synostosis Craniofacial dysplasias with dyschondrosis Achondroplasia After van der Meulen JC, Mazzola R, Vermey-Keers C et al. A morphogenetic classi /f_i cation of craniofacial malformations. 1983; 71 (4): 560–72. In more depth the epidemiology, pathogenesis and • management of cleft lip and palate

Introduction

Cleft lip and/or palate is the most common congenital abnormality a ff ecting the orofacial region. These conditions most commonly occur as isolated deformities but can also be associated with other medical conditions, e.g. congenital heart disease. They are also an associated feature in over 300 recognised syndromes. All children born with a cleft are screened for other congenital abnormalities. Where the cleft is thought to be associated with a syndrome any appropriate further

investigations, including genetic counselling, will be organised.

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