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SPECIAL REVIEW

Marathon of eponyms: 3 Crouzon syndrome

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The use of eponyms has long been contentious, but many remain in common use, as discussed elsewhere (Editorial: Oral Diseases. 2009: 15; 185). The use of eponyms in diseases of the head and neck is found mainly in specialties dealing with medically compromised individuals (paediatric dentistry, special care dentistry, oral and maxillofacial medicine, oral and maxillofacial pathology, oral and maxillofacial radiology and oral and maxillofacial surgery) and particularly by hospital-centred practitioners. This series has selected some of the more recognised relevant eponymous conditions and presents them alphabetically. The information is based largely on data available from MEDLINE and a number of internet websites as noted below: the authors would welcome any corrections. This document summarises data about Crouzon syndrome.

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Also known as

Craniofacial dysostosis

The condition

Crouzon syndrome is a rare first branchial arch syndrome, whose inheritance is autosomal dominant, affecting about one in 60 000 individuals. Mutations on chromosome 10q, in fibroblast growth factor receptor (FGFR) genes, may be responsible (often *FGFR2*) for the onset of the syndrome.

In Crouzon syndrome, the fibrous sutures in the skull and face are abnormal, fuse early and distorted bone growth ensues, resulting in craniofacial anomalies – termed craniostenosis by Virchow. During infancy, the coronal, the sagittal and, occasionally, the lambdoidal sutures begin to fuse early and this is complete by the second or third year. The order and rate of suture fusion determine the degree of deformity and disability. Premature fusion of the coronal and lamboidal sutures causes oxycephaly (turricephaly); premature fusion of coronal and lamboidal sutures unilaterally causes plagiocephaly; sagittal suture fusion causes dolicocephaly; fusion of the coronal suture causes brachcephaly; fusion of the metopic suture results in trigonocephaly and premature fusion of all skull sutures causes Kleeblattschaedel (clover leaf skull).

In Crouzon syndrome, not only do the calvarial and cranial base sutures fuse prematurely, but so also do those of the orbit and maxillary complex, leading also to high and wide forehead, ocular hypertelorism, exophthalmos, nasal septum deviation, narrowed or obliterated anterior nares, and wide beaked nose, low-set ears and severe maxillary hypoplasia. The upper lip is shortened and sometimes cleaved.

Without treatment, blindness, deafness and learning disability are potential outcomes. Nasal septal deviation, mid nasal abnormalities, choanal abnormalities and nasopharyngeal narrowing can obstruct the airway.

Craniofacial surgery can prevent the suture closure from damaging the developing brain; orthognathic surgery can correct the midface deficiency. Multiple staged surgery is normally needed. In addition, many patients are given a custom-fitted helmet (or cranial band) to be worn for several months after surgery.

Acrocephalosyndactyly (Apert syndrome) is comparable, with a similar inheritance and involvement of FGFR, similar craniofacial features but additionally with syndactyly of the hands and feet. Other syndromes that may need to be differentiated include Muenke craniosynostosis (*FGFR3*-associated coronal synostosis), Pfeiffer syndrome, Saethre-Chotzen syndrome, Jackson–Weiss syndrome and non-syndromic craniosynostosis.

Crouzon syndrome is occasionally associated with acanthosis nigricans appearing in childhood in the axillary fossa, angles of the mouth and on the lips. *FGFR3* mutations appear responsible for the onset of this syndrome.

The main person

Louis Edouard Octave Crouzon was born in 1874 in Paris, France, and studied Medicine there, graduating in

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1900 and moving on to postgraduate training with Philippe Charles Ernest Gaucher (1854–1918) and Marie, amongst others. He became chef de clinique et de laboratoire at the Hôtel Dieu in 1906 and was promoted to Médecin des Hôpitaux in 1912, and was on the staff of the Salpêtrière. During the Great War, he was responsible for a military neurological service and subsequently continued investigations in that field, especially in hereditary dystrophies, in particular into the hereditary cerebellar ataxias, which led him to consider cervical and lumbar spine deformities. Crouzon became president of the Paris Neurological Society and secretary of Revue Neurologique. He based several articles upon his observations of long-stay patients in the Salpêtrière. Crouzon first described his eponymous syndrome in a mother and a son with abnormal facial features presented to the Medical Society of Paris in 1912, and later reported a second kindred, involving seven persons. He died in Paris, in 1938.

Source internet sites (accessed 21 February 2009) and further reading

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