Crouzon Syndrome

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Introduction

Crouzon’s syndrome was first described by a French neurologist, Octave Crouzon (1874-1938) in the year 1912 as one of the varieties of craniosynostosis. It is also known as craniofacial dysostosis. It has prevalence of 15-16% in one million new born and 4.5% of all craniosynostosis [1]. The known race or sex predilection exists [2]. It may be transmitted as an autosomal dominant inheritance but 25% of cases represent fresh mutations [3]. The vast majority of cases of Crouzon syndrome are caused by a genetic mutation, specifically in a gene called FGRF2 (fibroblast growth factor receptor 2) on chromosome 10 and a mutation in FGRF3 on chromosome 4. Cranial malformation in Crouzon syndrome depends on the order and rate or progression of sutural synostosis [4].

Crouzon’s syndrome presents similar craniosynostosis as in the Apert, Pfeiffer and Saethre-Chotzen syndromes except with no digital abnormalities [5]. The appearance of CS can vary in severity from a mild to severe forms with multiple fused cranial sutures and marked midface and ocular defects. Mental retardation is not a hallmark feature unless premature closure of the cranial suture lines impairs brain development [6].

Surgical Treatment

Treatment of children with Crouzon syndrome is complex and is aimed at correcting the skull and midface abnormalities and treating obstructive sleep apnea.

Most children with Crouzon syndrome will need 2–4 skull operations over a lifetime. The earliest skull surgery is frequently done in the first 18 months of life.

The most common surgery for moving the bones of the midface forward in Crouzon syndrome is called a LeFort III operation. This surgery is typically not done before your child is 6–8 years of age. The primary indications for performing a LeFort III operation include severe obstructive sleep apnea which cannot be improved without surgery or significant patient concerns about appearance. During this operation, the bones of the midface are cut across the top of the nose, along the floor of the orbits and across the cheekbones. Most of these cuts may be made through the same incision used for your child’s other skull surgeries. An additional incision may be placed on the inside of the mouth. No scars are placed on your child’s face. After making these cuts, the bones are either moved immediately into their final position and secured in place with plates and screws or moved gradually by a process called rigid external distraction. Which of these techniques is used depends on the age of your child at the time of surgery and the distance that the bones must be moved.

Children with Crouzon syndrome frequently have obstructive sleep apnea due to the underdevelopment of the midface. As a result, your child should be monitored for sleep apnea by your craniofacial team. In mild cases of sleep apnea, medications may be sufficient to improve breathing. If more significant obstruction occurs, tonsillectomy or continuous positive airway pressure (CPAP) masks may help alleviate symptoms. Rarely, a tracheostomy may be required in infants with Crouzon syndrome in order to ensure adequate breathing. Midface surgery, such as a LeFort III operation, may be needed as your child gets older in order to fully treat obstructive sleep apnea or allow for eventual removal of a tracheostomy place in infancy. Ultimately, the goal in all of these interventions is to ensure a good airway so that your child can get enough oxygen to help her develop to her fullest potential.

In the treatment of children with Crouzon syndrome, the following health disciplines are included: craniofacial surgery, neurosurgery, oral surgery, orthodontia and ophthalmology. Upper jaw surgery performed by a plastic surgeon, and for correction of other deformities within the oral cavity, such as high arched palate, bilateral cross bite, hypodontia or crowding of teeth by orthodontist.

Since the condition tends to affect more than one suture, an open surgery is preferred. Subsequent re-correction surgery is also a part of the Crouzon Syndrome treatment regimen. Surgery takes place in multiple stages and may begin by addressing the craniosynostosis to prevent the closure of gaps between the skull bones. It is important to keep these gaps from fusing together, as premature fusion can cause brain damage or result in underdevelopment. In addition, extensive facial and orthodontic surgeries typically are required.

The treatment should be aimed at stimulating the development and expansion of the upper jaw in the sagittal and transverse direction. The orthodontic mobile appliance type “Y” and Delaire-mask should be used for ante-position of the upper jaw. The leveling of dental chains, as well as wearing fixed orthodontic appliances in the upper and lower jaw are indicated, because of the impacted upper canine teeth that occur as a result of underdevelopment of the upper jaw. When orthodontist creates a space for impacted teeth, oral surgeon frees the impacted canine teeth, and they are placed in dental chain. After complete leveling of dental chains and preparation of the patient, maxillofacial surgeon access to the surgical therapy. It is necessary to make ante-position of the upper jaw, with or without using distractors [7–9]. High LeFort I osteotomy is effective for midfacial deformity correction in patients with Crouzon syndrome [10]. After this stage, orthodontist definitely regulate vertical dimension of the bite, because after surgical treatment to a soft opening of the bite may occur. At this stage, orthodontist use inter-maxillary rubbers of varying diameters. Differential diagnosis has to be made from Apert syndrome, Carpenter syndrome and Pfeiffer syndrome.

References


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