

CROUZON SYNDROME

Description

Crouzon syndrome involves premature fusion (joining) of skull bones (craniosynostosis). Distinguishing characteristics include bulging eyes as a result of abnormal mid-face growth. A receding upper and protruding lower jaw are also found. Crouzon's is distinguishable from other craniosynostosis syndromes by the *lack* of hand and/or foot abnormalities. There is wide variability in the physical appearance of Crouzon patients with the shortened distance between the front and back of head (brachycephaly) being most prevalent.

The most distinguishing facial feature of Crouzon syndrome is bulging eyes that appear to protrude outward due to eye sockets being particularly shallow. Eyes pointing slightly outward account for the visual problems often reported in Crouzon syndrome.

Abnormalities of the cervical spine are prevalent, with some Crouzon's patients also exhibiting subtle hand, elbow and musculoskeletal variations.

Crouzon children may also have one or more of these abnormalities:

- Optic nerve deterioration
- Hearing loss
- Prominent, beaked nose
- High, narrow palate
- Teeth crowding

Prevalence/Causes

Crouzon syndrome occurs in approximately 1:25,000 live births. It is caused by a genetic mutation with evidence that the mutation occurs more frequently with the father's advancing age. Crouzon may be inherited from a parent as well, with a 50% chance existing within each pregnancy that the mutation will be handed down to the offspring.

Treatment

The treatment of Crouzon syndrome involves multiple surgeries:

- Frontal-orbital procedure to enable the skull to grow properly and increase eye socket size
- Surgery on the jaw
- Orthodontics
- Mid-face surgical advancement

A craniofacial team approach is recommended so that various problems can be comprehensively addressed. The team includes: plastic surgeon, neurosurgeon, orthodontist, geneticist; pediatric ophthalmologist, dentist and otolaryngologist; psychologist and speech therapist.

Associated Conditions

Eyes: Prominent eyes and eyelids that do not completely cover the eyes during sleep result in dryness and eye irritation.

Cleft lip/palate: This condition occurs in a small percentage of Crouzon patients.

Skin Coloring: Acanthosis Nigricans describes darkened skin coloration and soft, velvet-like patches of skin on neck, eyelids and surrounding the mouth.

