This syndrome was originally described in 1912 by a French neurosurgeon. He described four essential characteristics: exorbitism, retromaxillism, inframaxillism and paradoxic retrogenia. The incidence of this syndrome appears to be approximately one in 25,000 in the general population. It is inherited as an autosomal dominant pattern with variable expression. However, approximately 25% of reported cases have no family history and represent a new mutation.

Clinical Features: Patients with Crouzon syndrome have very distinct facial features with craniofacial bone morphology similar to Apert syndrome. Premature fusion of the bicoronal suture is the most common type of craniosynostosis with a brachycephalic or oxycephalic appearance to the skull. The extent of the craniosynostosis may be variable as well as the age at onset. There is retrusion of both the forehead and brow, with midface hypoplasia and shallow orbits with bulging eyes (proptosis). Orbital hypertelorism is less commonly seen in Crouzon’s than Apert’s syndrome. The lateral canthi may be slanted downward with some degree of upper eyelid ptosis. The proptosis is usually more prominent in Crouzon’s than Apert’s; however, in Apert syndrome, the other craniofacial deformities are more complex and severe. In addition, hydrocephalus and developmental delay are less frequently seen in Crouzon’s than Apert’s. The nasomaxillary retrusion may cause some degree of nasal airway obstruction with mouth breath-
ing. The chin is usually large with increased vertical height but often retracted resulting in a paradoxical retrogenia. The maxillary teeth are retracted compared to the mandible with a class III malocclusion. Other possible abnormalities include conductive hearing loss, inverted V-shape to palate and exposure keratitis. Variations of this type of craniofacial dysostosis are seen in the syndromes of Carpenter, Pfeiffer and Saethre-Chotzens.

**Treatment:** Our approach to the treatment of Crouzon syndrome is similar to that described for Apert syndrome. The first stage involves treatment of the craniosynostosis with total calvarial reshaping as needed and frontal-orbital advancement. This procedure is usually performed at four to six months of age and is effective in increasing the intracranial space and enlarging the orbits. It is not unusual for a ventriculo-peritoneal shunt to be needed for treatment of a hydrocephalus. Occasionally, a repeat craniotomy will be needed in infancy to further reshape the calvarial vault and advance the orbits. The next stage of reconstruction is midface advancement performed at four to six years of age. This type of advancement can be accomplished with either a LeFort III or a monobloc (entire face and forehead) osteotomy. The position of the forehead and brow determines which procedure would be the best alternative. If necessary, the monobloc osteotomy can be further split in the midline to correct orbital hypertelorism (facial bipartition).

The final stage of the reconstruction is management of the dental class III malocclusion. A LeFort I osteotomy is used to correct the dental discrepancies in combination with orthodontic intervention. This is usually performed after facial growth is completed and may be combined with a genioplasty (chin reduction/advance-ment). Additional procedures such as rhinoplasty may be needed.