fashion across the posterior aspect of the skull. Premature fusion or synostosis of this suture is rare but may be unilateral or bilateral.

Characteristically in unilateral synostosis the posterior skull is flattened on the involved side. Displacement of the ear with compensatory skull changes may be present in severe cases. Bilateral synostosis is characterized by extensive flattening across the posterior skull and increased vertical growth to accommodate brain enlargement. In mild cases the deformity can be covered by hair and surgical repair may not be indicated. When there is significant flattening or asymmetry, we recommend release of the fused sutures and posterior skull remodeling. Surgical treatment consists of extensive posterior skull craniectomies with remodeling of both sides of the occiput and posterior advancement of the affected side. Rigid fixation with microplates is used to hold the reconstructed bones in position and maintain the contour until healing takes place.

Occipital plagiocephaly without synostosis must be distinguished from lambdoid synostosis. Most patients referred to our center for posterior skull deformities are deformational or positional plagiocephaly. In these infants, the skull becomes misshaped from repeated pressure on the same position, without the premature fusion of the lambdoid suture. Positional plagiocephaly usually improves without surgery. Once the child’s sleep patterns change and he spends more time awake, the brain’s normal growth forces help to reshape the posterior skull. Some centers recommend a custom fitted helmet to mold the head back into position over a period of several months. However, in cases where the posterior skull deformity progresses or is severe, we recommend surgical remodeling as described, even though a true synostosis may not be present.

CRANIOFACIAL DYSOTOSIS

Craniofacial dysotosis (Crouzon’s and Apert’s diseases) are characterized by craniosynostosis with cranial dysmorphia and facial deformities (hence, the term craniofacial dysotosis).

APERT SYNDROME

Apert syndrome or acrocephalosyndactyly syndromes are rare conditions. In 1906, Apert described the skull, facial, and hand deformities of several patients characteristic of this syndrome that now bears his name. The incidence of infants born with Apert syndrome is one for every 100,000 to 160,000 live births. Many of the infants born with this syndrome show a sporadic transmission, which means that a family may have a child with Apert’s when no other members of the family are affected. The recurrent risk of having another child with Apert’s for two unaffected parents is negligible. However, if the parent is affected there is a 50% chance of each offspring having Apert syndrome with both males and females affected equally.

Clinical Features: Patients with Apert syndrome have very distinct facial and extremity features. Abnormal skull shape is due to craniosynostosis or premature fusion of the sutures or soft spots. The skull usually demonstrates a short anteroposterior diameter (brachycephaly) and may be excessively tall (turricephalic) or abnormally wide (euryprosopia). The forehead is generally always retruded, but this may not be obvious due to the hypoplasia of the midface. The orbits or bony sockets which contain the eyes are very shallow causing a bulging or proptosis. The orbits are usually rotated downward and lateral causing a downward shape to the lateral corners of the eyes. There may be a moderate increased distance between the eyes (hypertelorism) with muscle imbalance.

The middle of the face in Apert’s is both retruded and very hypoplastic. This causes the central midface to have a characteristic sunken-
in appearance with the nose being thick and beaked. The upper jaw or maxilla characteristically shows a narrow arch with an open bite and dental crowding. The maxilla is significantly retruded compared to the mandible with the teeth of the lower jaw projecting in front of the upper teeth. Other possible clinical features include moderate hearing loss, speech impairment, acne, and decreased mental capability in some individuals. Intellectual potential may be difficult to evaluate due to communication problems. Some patients with Apert syndrome may have normal intelligence. All patients with Apert syndrome demonstrate a unique hand malformation. This is characterized by a complex syndactyly or fusion of the skin, soft tissue, and bones of the fingers. Both hands are affected equally, as are the feet. This unusual variation of syndactyly can be used to identify Apert’s from other similar syndromes.

**Treatment:** The treatment of patients with Apert syndrome is not uniform due to significant variations in the facial anomalies, age of patients when first seen, and previous operations. Our primary concern of the infant born with this syndrome is: compression of the brain, breathing problems, protruding eyes with corneal exposure, and lack of facial growth. The surgical plan must be flexible and individualized to the patient. Multiple stages or operations at different ages are usually necessary.

When we see these patients as infants, the first stage is treatment of the craniosynostosis with total
calvarial reshaping. We prefer to do this procedure at four to six months of age. A frontal-orbital advancement is performed which increases the intracranial space and size of both orbits. Total skull reshaping helps correct the tower skull problem which is not addressed by frontal-orbital advancement alone. A frontal-orbital advancement is performed which increases the intracranial space and size of both orbits. Total skull reshaping helps correct the tower skull problem which is not addressed by frontal-orbital advancement alone. A ventriculo-peritoneal shunt may be needed for treatment of a hydrocephalus. This is performed prior to skull remodeling. Occasionally a repeat craniotomy is needed to further reshape the calvarial vault and advance the orbits. The next stage of the reconstruction is midfacial advancement. We usually perform this procedure between the ages of four to six years old prior to starting school. If necessary, an intra/extracranial advancement of the entire face and forehead (monobloc) can be performed with correction of mild hypertelorism or lateral rotation of the orbits. This procedure, as described by Tessier, is called a facial bipartition and corrects several deformities at once. It effectively widens the maxilla and derotates the orbits, and narrows the upper face. In milder cases an extracranial LeFort III advancement may be used.

The final steps in the reconstruction are maxillary/mandibular osteotomies to correct the maxillary/mandibular osteotomies to complete the correction of any further dental discrepancies. These procedures are usually performed after eruption of permanent dentition and completion of growth (teen years). Additional procedures such as rhinoplasty, genioplasty and eyelid surgery may be beneficial. Surgical separation of the fingers is usually started in the first year of life and completed by three to four years of age.

The patient with Apert syndrome represents a complex combination of multiple deformities. Our goal is to treat not only the function and physical problems but the psychosocial issues as well.
CROUZON SYNDROME

This syndrome was originally described in 1912 by a French neurosurgeon. He described four essential characteristics: exorbitism, retromaxillism, inframaxillism and paradoxical 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-

well. We feel these individuals are best evaluated and treated by a craniofacial center that utilizes the multi-disciplinary team approach providing a coordinated, comprehensive, long-term treatment plan with careful monitoring of growth and development.

Apert Syndrome

Four year old girl with Apert syndrome.

Postoperative result after monobloc facial advancement with facial bipartition (medial rotation of orbits).

Preoperative

Postoperative

Apert Syndrome

Four year old girl with Apert syndrome.