Six patients with midface retrusion secondary to craniofacial dysostoses were evaluated by computer assisted tomography (CT) before and after surgery. Computer reformatted life-size images were generated for linear measurements and for assessing the encroachment upon the orbit of the anterior and middle cranial fossae, the lateral displacement of the ethmoids, and the retrusion of the maxilla. Dynamic ocular CT demonstrated alterations in the angle of divergence of the optic nerve with globe movement which made measurements of that angle an unreliable assessment of hypertelorism. Surgical separation of the midface from the base of the skull was shown to be a fracture within the palatine bones 'between itsh horizontal and pyramidal processes. The integrity of the antral walls was preserved in all patients, in contrast to the comminution seen with traumatic LeFort III fractures. CT evaluation of patients with midface retrusion has further defined the nature of their anomalies, facilitated operative planning, demonstrated the osteotomies in vivo and reduced radiation exposure.

**KEY WORDS:** Craniofacial dysostoses, craniofacial surgery, midface advancement, CT scan

Rene LeFort clarified the osseous anatomy of traumatic craniofacial disjunction in the early 1900's through the study of fresh and prepared guillotined heads (LeFort, 1901). With the popularization of elective craniofacial disjunction in the late 1960's, attention again was focused on the relevant head and neck anatomy by the study of cadaver material (Matras and Perneeczky, 1975). Radiography was employed for in vivo evaluations. Movement of both dentition and standard anthropological landmarks was readily documented with cephalometric radiographs (Firmin, et al. 1974). Details of the cribriform plate and optic foramina were discerned through polytomographic radiography, increasing the safety of midface disjunction and orbital translocation procedures (Tessier, et al., 1967). While these techniques allowed for surgical planning and postoperative evaluation, they failed to completely clarify the in vivo osseous anatomy of elective craniofacial operations, while soft tissues were inadequately assessed. Computerized axial tomography (CT) was added to the preoperative planning armamentarium in the mid 1970's (Becker, et al., 1976). The ability of the CT to display discreet soft tissues as well as bone has significantly increased our understanding of the aberrant anatomy of the craniofacial...
Marsh and Gado: CT Anatomy of the Craniofacial Dysostoses

dysostoses. Furthermore, CT of such patients following craniofacial surgery has improved
definition of the osteotomies and associated
soft tissue changes. Preliminary CT observa-
tions from our first six patients are reported.

Materials and Methods

Six patients with congenital midface retrus-
sions who underwent midface advancement
were studied (Table 1). There were two Crou-
zon Syndromes, two Apert Syndromes, one
Pfeiffer Syndrome and one Saethre-Chotzen
Syndrome. There were three males and three
females. The ages at the time of surgery
ranged from three to fifteen years. Two pa-
tients had synchronous frontal bone advance-
ment and one had hypertelorism correction as
well as midface and frontal bone advance-
ment. The remainder underwent Tessier's
"semi-opened" technique (Tessier, 1976).

All patients received a pre- and postopera-
tive radiologic evaluation. The CTs were ob-
tained initially (Cases 1, 2 and 3) by an EMI
1010 head dedicated unit. High resolution
scans were obtained in the latter three cases.
In two of these (Cases 4 and 5), the high
resolution package of the 5005 EMI scanner
was used. In the most recent case (Case 6),
high resolution scanning was obtained by a
Somotom II Siemens scanner.

Examination of the midface, orbits and
cranium was obtained in cases 1 through 5
using a 4mm thick CT slice. In Case 6, the
facial slice thickness was reduced to 2mm and
cranial slice thickness increased to 8mm.
Scans were routinely obtained in the axial
plane. Coronal scans were obtained in two
patients prior to the acquisition of computer
reformatting of images. Computer reformating
refers to the generation of images in non-axial
projections by the computer from the set of
axial images. All patients were sedated with
parental narcotic and phenothiazine for the
preoperative study. The postoperative study
was performed under barbiturate sedation ad-
ministered for the removal of the intermaxil-
lary and maxillary-cranial fixation 6 to 8
weeks postoperatively. Three patients with
traumatic LeFort III fractures have been evalu-
ated with the same CT protocol for compar-
ison.

Our current technique for facial and orbital
scans consists of 2mm contiguous slices by a
high resolution Somotom II scanner allowing
a picture element with a life size of 0.3mm in
each case. 8mm contiguous slices are obtained
of the cranium on the same scanner. The
scans are obtained in the axial projection.
Computer reformating of the images is done
in each case producing sagittal and oblique
projections. Each CT slice delivers a maxi-
mum radiation dose of 1.5 rad to the skin.
Although polytomography delivers only 1.0
rad per slice, the numbers are misleading since
these radiographic examinations are never
limited to a single slice. The cumulative dos-
age of a multislice CT is less than that of a
complete polytome study. This is because in
the CT only the volume of tissue included in
the slice is subjected to the primary radiation.
The adjacent tissues are exposed only to scatter,
resulting in a much lower additive dose
when multiple slices are exposed. In contrast,
for each polytome slice, the entire volume of
tissues on both sides of the area of interest is
exposed to the same primary radiation. For

<table>
<thead>
<tr>
<th>Case Number</th>
<th>CPCDI #*</th>
<th>Sex</th>
<th>Age**</th>
<th>Syndrome</th>
<th>Operative Procedure</th>
</tr>
</thead>
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<tr>
<td>1</td>
<td>78–1026</td>
<td>N</td>
<td>15-4</td>
<td>Apert</td>
<td>midface and frontal bone advancements</td>
</tr>
<tr>
<td>2</td>
<td>79–1935</td>
<td>F</td>
<td>6-7</td>
<td>Crouzon</td>
<td>midface advancement</td>
</tr>
<tr>
<td>3</td>
<td>78–1013</td>
<td>F</td>
<td>9-4</td>
<td>Saethre-Chotzen</td>
<td>midface and frontal bone advancements; synchronous correction hypertelorism</td>
</tr>
<tr>
<td>4</td>
<td>78–1021</td>
<td>M</td>
<td>10-3</td>
<td>Apert</td>
<td>midface and frontal bone advancements; synchronous independent transposition left orbit</td>
</tr>
<tr>
<td>5</td>
<td>78–1028</td>
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<td>6-6</td>
<td>Crouzon</td>
<td>midface advancement</td>
</tr>
<tr>
<td>6</td>
<td>80–1944</td>
<td>F</td>
<td>3-2</td>
<td>Pfeiffer</td>
<td>midface advancement</td>
</tr>
</tbody>
</table>

* Cleft Palate and Craniofacial Deformities Institute accession number.
** age in years-months at time of operative procedure.
example, a CT of four to ten contiguous slices results in a total dose increase by a factor of 0.5. When the number of slices greatly exceeds ten, the total dose may be doubled. In a polytome examination of four to ten cuts, the total radiation dose increases by a factor from four to ten respectively. The total radiation dose delivered in a complete set of CT of the cranial and facial structures does not significantly exceed the dose delivered with the routine skull series consisting of five conventional radiographs. Furthermore, the capability of the Somotom II for computer reformatting allows generation of additional images with no further radiation exposure of the patient.

Results and Discussion

Cranium: Intracranial Malformations. Identification of intracranial malformations is a major objective of the preoperative scan. The ability of this noninvasive technique to visualize the brain parenchyma and the cerebrospinal fluid space without the need of lumbar or ventricular puncture is an invaluable asset to the modern radiologic evaluation of these patients. In our series of six consecutive patients, hydrocephalus was present in four. The hydrocephalus was severe enough in two patients to have required previously a shunt procedure (Figure 1). In the pre CT era, hydrocephalus was reported to occur in 13 of 25 percent of patients with either one of the craniofacial dysostoses or isolated craniosynostosis who were studied (Bertelsen, 1958; Laitinen, 1956). In these previous reports, hydrocephalus was detected by pneumoencephalography. Due to the invasiveness of the technique, it was natural that only selected cases were subjected to the procedure. Therefore, the previous reports cannot be relied upon to give the true incidence of hydrocephalus with either craniosynostosis or the craniofacial dysostoses. This point was emphasized in a review by Fishman et al., (1971), who reported eight cases of hydrocephalus associated with Apert’s Syndrome. In a non-selected study, such as ours, in which all patients with the diagnoses in question are examined by CT regardless of neurologic status, the true incidence of hydrocephalus associated with or as a complication of the craniofacial dysostoses will be determined. Pneumoencephalography previously diagnosed other intracranial anomalies in patients with the craniofacial dysostoses, including agenesis of the corpus callosum and the septum pellucidum (Warkany, 1971). These structures were present in our six patients. The significance of intracranial malformations found in association with the craniofacial dysostoses is unknown. Longitudinal CT studies of infants with these syndromes may resolve the issue.

Orbits: Congenital Abnormalities. The shallow orbit and its consequent proptosis are hallmarks of the craniofacial dysostoses (Marsh, 1980). Axial CT with multiplanar computer reformatting of images facilitates definition of deformities of all four orbital walls. In all of our cases, the mesial wall of the orbit was displaced laterally by the ethmoid cells. This deformity is presumed to be secondary to pressure upon the sinus by the growing frontal lobes of the brain since bicoronal synostosis forces the anterior growth vector of the frontal lobes into a vertical direction. This displacement is best demonstrated in the axial projection (Figure 2A). In this same projection, the lateral orbital wall is seen to be convex towards the orbital cavity. This abnormal curvature is the result of deformity of the greater wing of the sphenoid caused by pressure from the growing temporal lobe. Synostosis of the sphenofrontal, sphe-
FIGURE 2. 6-7 year old male with Crouzon syndrome (case #2).
A. Preoperative high resolution CT in the axial projection. The medial wall of the orbit is displaced laterally on both sides. The lateral wall of the orbit is also displaced laterally and bowed convexly anterior (large arrow). There is narrowing of the angle between the lateral wall of the orbit and the lateral wall of the middle cranial fossa, causing effacement of the temporal fossa (small arrow).
B. CT six weeks after semi-open LeFort III midface advancement. The medial walls of the orbit are displaced medially. There is residual hematoma in the nasal vestibules and the residual ethmoid cells. The rim of the lateral orbital wall is more anterior on each side and the interposed bone grafts are well seen (arrows).

Deformities of the roof and floor of the orbit are best visualized on the “longitudinal orbital view” (Marsh and Gado, 1982). This view is obtained by computer reformatting of images in an oblique projection, the plane of which corresponds to the longitudinal axis of the orbit. In this view (Figure 3A), the shortness of the floor of the orbit due to the maxillary retraction is well visualized. We have also observed, in this projection, that the concavity of the “S” curvature of the normal orbital floor appears reversed in patients with craniofacial dysostosis resulting in a convex “C” curvature (Figure 3B). In addition to defining the anatomy of the orbital walls, CT can be used to calculate orbital cavity volume. Our method is the subject of a separate communication.

The degree of hypertelorism can be determined by measuring the distance between the mesial walls of the orbits on axial or computer formatted coronal projections. Life-size images are generated, so that direct measurements can be made upon the CT image. Life-size coronal images are also useful in determining the position and configuration of the floor of the anterior cranial fossa. Preoperative identification of nasal prolapse of the cribiform plate is necessary not only for the purpose of planning correction of concomittant hypertelorism, but also to avoid injury to the dura of olfactory and frontal lobes during Tessier’s “semi-opened” technique. While hypertelorism (horizontal orbital displacement) is the most common orbital abnormality associated with the craniofacial dysostoses, dystopia with orbital asymmetry in all three planes may occur. Multiplanar computer reformatting of the CT images adds refinement to the clinical impression of the need for unequal orbital translocation. Life-size computer generated CT images allow direct quantitative measurements for surgical planning.

ORBITS: DIVERSION OF OPTIC AXES. Evaluation of the angle of divergence of the optic axes has been stressed by others as an important aspect of the orbital evaluation (Converse, et al., 1970). Difficulty arises, however, in selecting the points to define the line of the axis to be measured. The two lines most commonly delineated are a tangent to the lateral orbital wall and a superimposition upon the optic nerve. Both of these constructions are
FIGURE 3. Evaluation of the roof and floor of the orbit by reformating of CT images in the longitudinal axis of the orbit. The top image is the actual axial scan with the plane of reformation indicated by the dashed line.

A. Presurgical 21 year old woman with Crouzon syndrome. (case not included in this report). The floor of the orbit is short with a “C” configuration convex toward the orbit. The cornea is anterior to a line drawn between the superior and inferior orbital rims, i.e., the patient has exorbitism. (from Marsh and Gado, Plast. Reconst. Surg., 1982, In press.)

B. Normal patient of same age and sex. Note the greater length of the orbital floor with an “S” configuration having a prominent concavity below the globe. The cornea is behind the superior and inferior orbital rims, i.e. there is no exorbitism. The inferior pole of the globe is supported by bone in contrast to the Crouzon’s patient.

drawn on an axial polytome or CT image at the level of the center of the globe. Neither of these approaches, however, yielded reproducible data in our hands. The lateral orbital wall of patients with the craniofacial dysostoses is usually curved convexly toward the orbit thereby precluding construction of a line parallel to the wall (Figure 2A). The placement of a tangent to the curve, therefore, is at the discretion of the measurer rather than being based upon constant anatomical features. The axis of the optic nerve has proven as unreliable for reproducible measurements as the lateral wall. Our dynamic ocular CT studies of adolescent patients with craniofacial dysostosis have confirmed the impression we gained from patients without skeletal deformities who were evaluated for strabismus: the angle of divergence of the optic nerves is an unsatisfactory measure of globe position. This should not be surprising since the angle of divergence of the optic nerves depends on the degree of abduction of the globe. This can easily be demonstrated in the conscious, cooperative patient by obtaining images with the patient voluntarily looking at an object on the right or left side of the visual field (Figure 4). Our experience with the patients in this study has revealed the presence of variable and unpredictable degrees of lateral deviation of both globes due to the effects of sedation. Therefore, we currently do not measure the angle of deviation of the optic nerves. Other methods for measuring the location of the globes are being investigated. We are also conducting preliminary studies on orbital muscle position.

Orbits: the Middle Cranial Fossa and the Temporal Fossa. As mentioned above, anterior pressure by the growing temporal lobe upon the greater wing of the sphenoid results in bowing of the lateral wall of the orbit. The resultant convexity is directed towards the orbital cavity. This curvature and the lateral displacement of the lateral wall of the orbit, described above, combine with the altered expansion of the middle cranial fossa to result in a narrowing of the angle between the lateral orbital wall and the lateral wall of the middle cranial fossa. The acuteness of this angle reflects the degree of effacement of the temporal fossa (Figure 2A). In extreme cases,
the frontal process of the zygoma and the squamosal temporal bone approximate each other (Figure 4). Preoperative recognition of this gross alteration of periorbital anatomy is essential to avoid inadvertent entry into the middle cranial fossa while performing the lateral orbital wall osteotomy.

**Orbits: Postoperative Evaluation.** Examination of the orbits on the postoperative CT clarifies the nature and amount of bony

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**Figure 4.** Alteration in the angle of diversion of the optic nerve with globe adduction and adduction in a presurgical 16 year old female with Crouzon syndrome (case not included in this report). Both the curvature of the optic nerves with movement and the changing angle between them preclude consistent measurements.

A. Patient is in right lateral gaze.
B. Patient is in left lateral gaze.

**Figure 5.** The effect of midface advancement on the osseous anatomy of the cephalad aspect of the pterygomaxillary fissure (arrow). The patient is a 13-4 year old male with Apert syndrome (case #1).

A. The lateral and posterior walls of the maxillary antrum (M.A.) are closely approximated to the base of the skull (B.S.) preoperatively.
B. Six weeks post LeFort III midface and frontal bone advancements widening of the pterygomaxillary fissure (arrows) is apparent.
movement, orbital volume augmentation and alterations in the position of the orbital contents. Linear measurements are taken directly from lifesized axial CT images or from computer reformatted images obtained in coronal and oblique planes. Volumetric changes are assessed via the computer program for measurement of the orbital volume. The ability of CT to visualize soft tissues allows evaluation of postoperative alterations in the position of the globe, optic nerve and orbital muscles. Alterations in the configuration and position of the orbital walls are noted as well (Figure 2B). Bone grafts are also visualized and their relationship to the orbital contents clarified. Specifically, the grafts placed into the lateral orbital wall are seen not to protrude into the orbital cavity nor do they impinge upon the lateral rectus muscles or alter the course of the muscles. The ability to visualize soft tissues in the postoperative examination excludes other complications such as intraorbital hematoma. Likewise, one is able to assess the degree of correction of exorbitism by evaluating the length and configuration of the floor of the orbit in relation to the globe in the longitudinal orbital projection (Figure 3A).
Midface: In Vivo Anatomy of Midface Advancement. Surgical correction of congenital midface retrusion requires separation of the midface from the base of the skull (Figure 5). Although the anatomy of osteotomies in this area has been studied in cadavers and on prepared skulls (Matras and Perneczky, 1975) definition of the in vivo disjunction remains unclear. This separation is known as “pterygomaxillary disjunction” (Bell et al., 1990). The nomenclature, however, is anatomically inexact since the maxilla does not articulate with the pterygoid plates. The palatine bone is the bridge interposed between...
FIGURE 8. Post-traumatic LeFort III CT at the mid-maxillary antrum level in a 21 year old female. Note the fracture and comminution of the antral walls and left pterygoid-palatine-maxillary articulations (large arrow). The right pterygoid-palatine-maxillary articulations are intact (small arrow) and, in fact, the “pterygomaxillary disjunction” is a transantral fracture.

the midface and the base of the skull (Figure 6). Pre- and postoperative CT’s were obtained in six consecutive patients who underwent midface advancements. Separation of the midface from the base of the skull consistently occurred within the palatine bones as a fracture between the horizontal and pyramidal processes (Figure 7). The palatine crest, which is the anteriormost attachment of the tensor veli palatini, remained in continuity with the midface. Both nondisplaced and comminuted fractures within the pterygoid plates were sometimes observed as well. The posterior wall of the maxillary antrum remained intact in all cases. This is in contrast to the antral comminution seen in traumatic craniofacial disjunction (LeFort III fractures) (Figure 8).

Summary

Computer assisted tomography (CT) was employed pre- and postoperatively in elective craniofacial disjunction to clarify aberrant anatomy and define osteotomies in vivo in six consecutive patients with midface retrusion secondary to several of the craniofacial dysostoses. Four patients had preoperative hydrocephalus. Of these, two had previously required a shunt procedure. No other intracranial anomalies were found except distortion of the cranial fossae. Multiplanar computer reformatted images proved useful to assess encroachment upon the orbit of the anterior and middle cranial fossae as well as the lateral displacement of the ethmoids and retrusion of the superior maxilla. In the longitudinal orbital view, the “S” curve of the floor of the orbit appeared to change into a “C”, convex toward the orbit. Dynamic ocular CT demonstrated alterations in the angle of divergence of the optic nerve with globe movement. This variability made measurements of that angle an unreliable assessment of hypertelor-
ism. CT of the separation of the midface from the base of the skull consistently demonstrated the disjunction to be a fracture within the palatine bones between the horizontal and pyramidal processes. The integrity of the antral walls was preserved in all patients, in contrast to the comminution seen with traumatic LeFort III fractures. Computer reformatted life-size images were generated for direct pre- and postoperative linear measurements for both surgical planning and followup. In addition, the use of reformed images has markedly reduced the total radiation exposure at any single locus from that delivered with biplanar polytomography. CT evaluation of patients with midface retrusion has further defined the nature of their anomalies, facilitated operative planning, demonstrated the osteotomies in vivo and reduced radiation exposure.

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