Facial deformity in Crouzon syndrome is characterized by exophthalmos, exorbitism, mild hypertelorism, and maxillary hypoplasia with a Class III occlusion. The Göteborg craniofacial unit corrects this deformity in selected patients by the 2-stage procedure of “spectaclesplasty” followed by a Le Fort I maxillary osteotomy at skeletal maturity. “Spectacles” is a reference to the bilateral circumferential periorbital bony skeleton, and a spectaclesplasty is a differential rotation and advancement of this complex. Spectaclesplasty has been integrated as part of our protocol in managing Crouzon syndrome from birth to maturity.

All patients who have undergone spectaclesplasty were retrospectively reviewed. A composite scoring system analyzing periorbital anatomy and aesthetics was used to compare the preoperative and most recent postoperative photographs. Complications and the need for ancillary procedures were recorded.

Twenty-one patients have undergone spectaclesplasty since this technique was introduced in 1984. Mean follow-up time is 5 years 10 months. Compared with their preoperative aesthetics, the improvement was rated as excellent for 8 patients (38%), very good for 7 patients (33%), good for 5 patients (24%), and minimal for 1 patient (5%). No patient was rated as having no improvement. Mean perioperative blood loss was 111% of estimated red cell mass. Mean operative time was 6.4 hours. The mean duration of stay in the intensive care unit was 28 hours, and the mean hospital stay was 11 days.

There were few complications and no incidences of mortality.

Spectaclesplasty yields high-quality aesthetic results in most cases. It is our impression that spectaclesplasty en bloc rotation advancement of the periorbital bony skeleton can be safely performed before skeletal maturity of the lower face. Correction of the bony periorbital anatomy in early adolescence is important in alleviating psychosocial distress in this age group. In our hands, spectaclesplasty produces a more normal anatomic position of the periorbital soft tissues facilitating both function and aesthetics.

Key Words: Spectaclesplasty, Crouzon syndrome, orbital osteotomies, Le Fort I

In 1912, Crouzon described a familial syndrome characterized by exorbitism, retromaxillism, inframaxillism, and craniosynostosis. The prevalence of Crouzon syndrome at birth is 15 to 16 per million and accounts for 4.5% of all cases of craniosynostosis. Crouzon syndrome is inherited as a highly variable autosomal dominant condition, and approximately two thirds of cases are familial. Approximately 30 different mutations of fibroblast growth factor receptor II have been identified in Crouzon syndrome to date. The Crouzon phenotype is highly variable and ranges from ocular proptosis and midface hypoplasia with no craniosynostosis to a cloverleaf skull deformity. Unlike most other craniofacial syndromes caused by fibroblast growth factor receptor mutations, the limbs are typically unaffected. It remains unclear whether various phenotypic subgroups are caused by specific mutations.

The calvarial deformity is typically brachycephaly secondary to bicornal synostosis; however, mesocephalic and scaphocephalic forms are also seen. The cranial base and midfacial sutures are usually involved, giving rise to a short anterior cranial fossa, shallow orbits, and maxillary hypoplasia. The degree of exorbitism seen is variable and may occasionally endanger vision. Exposure conjunctivitis

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is common. Palpebral over retraction giving the appearance of globe dislocation may occur in extreme cases. When present, excyclorotation of the extraocular muscles causes a V pattern of divergent strabismus. The maxillary hypoplasia is associated with an anterior open bite, crowded maxillary dentition and a class III molar relationship with relative mandibular prognathism. The short anterior fossa creates and obtuse cranial base and in severe cases, the posterior nasal airway may be markedly restricted. Crouzon patients may have obstructive sleep apnea that goes largely unrecognized.

The Göteborg craniofacial unit has developed a protocol for the surgical management of Crouzon syndrome from birth through to maturity. Crouzon syndrome is treated in a staged fashion based on the differential growth of the calvarial vault and brain, the midface, and the mandible. Because of the phenotypic variability of Crouzon syndrome, surgical management is tailored according to individual requirements, and throughout growth, different priorities are present. Cranial decompression is performed early in all but the mildest cases. In bicoronal synostosis, the dynamic cranioplasty for brachycephaly is performed. Details of the operative technique and long-term follow-up have been reported previously. The dynamic cranioplasty for brachycephaly procedure does not involve osteotomies inside the orbit, and the lower margin of the frontal bone flap is approximately 1 cm above the supraorbital rim thereby preserving both the intracranial and the extracranial bony anatomy in virgin condition for the later planned “spectaclesplasty.” Throughout early childhood, patients are monitored regularly, in particular, for signs of raised intracranial pressure and obstructive sleep apnea. At times, a repeat cranial decompression is required. Midfacial advancement is occasionally required for airway maintenance; however, it is usually our preference to use continuous positive airway pressure to maintain the virgin anatomy of the midface until definitive correction. When midfacial advancement is imperative, we have performed a monobloc procedure with spring-assisted expansion as this preserves the periorbital bony anatomy.

The periorbital, midfacial, and dental occlusal deformities are managed as a 2-stage procedure at the commencement of adolescence and at the completion of skeletal growth. The midface deformity in Crouzon syndrome has been traditionally managed with a Le Fort III osteotomy, monobloc, or variations of these procedures. Because of the dissatisfaction with the results of these procedures in our unit including the periorbital aesthetics and the tendency to relapse, the technique of spectaclesplasty was developed in 1985 by the senior author (C.L.). The term “spectacles” denotes the bony periorbital rims, which determine the position, relationship, and function of the canthi, eyelids, and globes. The spectacles are differentially advanced and rotated upward en bloc to a more normal anatomic location with respect to the globes. Mild hypertelorism when present is corrected with a transnasal medial canthopexy in the bone gap created by the advancement. Once skeletal maturity is attained, a Le Fort I maxillary osteotomy and repositioning is performed. At times, a bimaxillary osteotomy is required when significant mandibular disproportion exists. Secondary procedures may include rhinoplasty, genioplasty, and fat grafting of local areas as deemed necessary.

**Patients and Methods**

All patients who have undergone a spectaclesplasty procedure since the technique was developed in this unit were retrospectively reviewed. Technical details, outcomes, and complications were recorded.

A composite scoring system of patient photographs was devised to assess the aesthetic improvement rather than the final aesthetic outcome. This is because some patients achieve an excellent result but preoperatively had a relatively mild deformity. Other patients start with marked disfigurement and multiple previous surgeries and are unable to achieve the same final result. However, they may still have a very marked and worthwhile improvement. There is no completely objective scientific method for analyzing the functional and aesthetic outcome achieved by this procedure, and there is no similarly published procedure with which our data can be compared.

Immediate preoperative photographs were compared with the photographs taken at the last follow-up. Frontal, three quarters, and profile views were assessed and compared. The most recent photograph was used because we believe that any deficiencies in the procedure such as thinning of the subcutaneous tissue and bony relapse become more marked with time.

A score from 1 (poor) to 5 (excellent) was assigned to the following 10 categories to rate their preoperative presence and degree of improvement attained: proptosis, lateral canthal dystopia, hypertelorism, eyelid ptosis, forehead shape, periorbital soft-tissue atrophy, temporal muscle wasting, brow position, malar position, and nasal and overall aesthetics. These scores were added and then averaged to give an overall average composite score for each
patient. Because several patients are still awaiting their orthognathic correction, only the periorbital region was analyzed. Because of the technical nature of the variables being scored, the assessment was performed independently by 2 consultant craniofacial surgeons including the senior author (C.L.). In order that readers may appreciate the composite scores assigned, examples are presented (Figs 1–6).

Operative Technique

Surgical approach is through a coronal scalp and an upper buccal sulcus incision. Subperiosteal dissection of the entire orbital skeleton is performed taking care not to disrupt the medial canthal tendons unless correction of hypertelorism is planned. The anterior temporalis muscle is elevated sufficient to visualize the lateral wall osteotomy. A large frontal bone flap is raised, and the osteotomies are placed bearing in mind that this bone flap will be replaced in an advanced and slightly retrorotated position. The lower osteotomy is at least 1 cm above the supraorbital rim to preserve the strength of the spectacles unit (Fig 7). Extradural dissection of the orbital roofs and foramen cecum then allows safe osteotomy across the roof just behind the bandeau. This osteotomy is continued circumferentially within the bony orbit approximately 1 cm behind the rims and posterior to the lacrimal sac. A vertical osteotomy at the lateral orbital passes through the frontozygomatic suture and stops midway into the zygomatic body. From here, the lower osteotomy across the maxilla is performed from the upper buccal sulcus incision where the infraorbital nerve and roots of the canine may be adequately visualized. Lateral nasal side wall

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Fig 1  A and B, Three years after spectaclesplasty and Le Fort I advancement. Composite score improvement was rated as “excellent.”

Fig 2  A and B, Three years after spectaclesplasty and later Le Fort I advancement. Secondary lateral canthopexy and rhinoplasty were performed. Composite score improvement was rated as “excellent.”

Fig 3  A and B, Six years after spectaclesplasty and Le Fort I advancement. Le Fort III advancement had been performed before spectaclesplasty. Secondary rhinoplasty has been performed. Composite score improvement was rated as “very good.”

Fig 4  A and B, Six years after spectaclesplasty and later Le Fort I advancement. Secondary rhinoplasty was performed, and fat grafting to the cheeks has been subsequently performed. Composite score improvement was rated as “very good.”
Osteotomies are performed with a small osteotome after nasal mucosal elevation. Finally, an osteotome is used from above to separate the nasal septum and the nasal side walls, allowing the nose to come forward with the periorbital bone. A small inspection window is usually removed from the central bandeau to allow the osteotome to be placed as low as possible and at the appropriate angle. Some fragments of the bone within the ethmoid sinuses are the only remaining bony connection. The bony periorbital complex is then mobilized and rotated forward assessing the relationship of the globe to the position of the eyelids and orbital rims. The supraorbital rim typically does not require as much advancement as the infraorbital rim, and so the whole complex is differentially advanced and rotated. A typical movement is a 6-mm advancement superiorly and a 12-mm advancement inferiorly. Cranial bone grafts behind the lateral orbital wall and the anterior maxillary wall hold the advancement and are secured with miniplates. The bone gaps in the socket are not grafted to allow extraorbital volume and thereby assist in exophthalmos correction. The periorbital is released to allow the fat to bulge into the enlarged orbital cavity.

A degree of hypertelorism is common in Crouzon syndrome. In mild-to-moderate cases, the medial canthal tendons are detached, and the lacrimal glands are dissected subperiosteally from its fossa, preserving the nasolacrimal duct. The medial bony orbital margins are then reduced with a burr. The medial canthal tendons are wired together in the gap created by the advancement of the bony spectacles complex. In 1 case where the hypertelorism was particularly marked, the bone was resected either side of the nasal midline and the orbits were formally medialized. In general, however, we avoid this to avoid weakening the periorbital framework. If there is any suspicion of a cerebrospinal fluid leak, galeal frontalis flaps are raised and secured with fibrin glue to the anterior cranial base. Lateral suspension canthopexies are performed before closing the incisions.

**RESULTS**

During the 10-year period from 1985 to 2004, 21 patients underwent a spectaclesplasty at our unit. There were 12 females and 9 males. The mean...
age was 18 years (range, 12–44 years). Four patients were first presented to our unit as adults, and if they were excluded, the mean age was 14 years, which is about the time we would normally prefer to do this procedure.

The averaged composite score system rated the degree of improvement of their periorbital anatomy and aesthetics. The improvement was rated as excellent for 8 patients (38%), very good for 7 patients (33%), good for 5 patients (24%), and minimal for 1 patient (5%). No patient was rated as having no improvement. The results of the composite scoring of periorbital aesthetics are shown in Table 1 and are illustrated with examples in the figures.

The mean operative time was 6.4 hours (range, 3.1–8.1 hours). The mean forward movement of the superior aspect of the spectacle complex (measured at the superior orbital rim) was 9 mm (range, 5–15 mm), and the mean forward movement of the inferior aspect (measured at the inferior orbital rim) was 13 mm (range, 10–21 mm). Mean blood loss was 2070 mL (range, 500–4000 mL). Expressed as a percentage of estimated red cell mass, the mean blood loss was 111% (range, 21%–247%).

There were no intraoperative complications. Two patients had evidence of a cerebrospinal fluid leak postoperatively, and they were kept in the intensive care unit for a mean period of 28 hours (range, 22–44 hours). The mean hospital stay was 11 days (range, 9–34 days). Mean follow-up time was 5 years 10 months (range, 6 months to 15 years).

Sixteen patients had subsequently undergone second-stage Le Fort I maxillary advancement and midfacial vertical lengthening. In 1 male, a bimaxillary osteotomy was performed with mandibular setback. Two patients had normal preoperative occlusion, and a further 3 patients are awaiting skeletal maturity. The mean maxillary movements were 10.3 mm advancement (range, 9–17 mm) and 3.5 mm vertically down at the level of the anterior nasal spine (range, 2–7 mm).

### Table 1. Results of Composite Analysis of Improvement in Periorbital Aesthetics

<table>
<thead>
<tr>
<th>Score</th>
<th>Patients, n (%)</th>
</tr>
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<tbody>
<tr>
<td>5 – Excellent improvement</td>
<td>8 (38)</td>
</tr>
<tr>
<td>4 – Very good improvement</td>
<td>7 (33)</td>
</tr>
<tr>
<td>3 – Good improvement</td>
<td>5 (24)</td>
</tr>
<tr>
<td>2 – Minor improvement</td>
<td>1 (5)</td>
</tr>
<tr>
<td>1 – No improvement</td>
<td>0</td>
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</tbody>
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See text for definition of graded variables in composite analysis.

Sixteen patients had undergone cranial decompression procedures in infancy. Additionally, 3 patients had undergone midfacial advancement in the first 5 years of life (2 monoblocs and 1 Le Fort III). Intraoperative blood loss for these 3 patients was higher than the remainder of the group, although there is insufficient power to determine statistical significance. Twelve patients have had additional procedures to refine their facial aesthetics after spectaclesplasty, and these are listed in Table 2.

Long-term complications have been a palpable plate in the periorbital region in 2 patients. Two patients in this series are no longer alive. One male patient committed suicide 15 months postoperatively. He had a longstanding history of depression and previous suicide attempts. A female patient died in a motor vehicle accident several years after her procedure.

### DISCUSSION

Midfacial advancement to improve the facial aesthetics in Crouzon syndrome has its roots in the first Le Fort III advancement performed by Gillies and Harrison in the early 1940s. Osteotomy was performed anterior to the orbital rims, lacrimal sac, and the medial canthal ligament. In 1958, Tessier performed his first Le Fort III advancement for a Crouzon syndrome patient, in which the osteotomy was performed posterior to the lacrimal crest allowing the en bloc advancement of the lacrimal apparatus and eyelids with the midfacial skeleton. Modifications of the lateral orbital wall osteotomy, sequentially called the Tessier I, II, and III osteotomies, were made improving the lateral rim contour and allowing self-retention. Converse and Wood-Smith advanced the complete orbits and face in 1 piece to obtain better correction of the exorbitism. In 1978, Ortiz-Monasterio reported the monobloc procedure for the treatment of Crouzon syndrome, an advancement of the midface and orbits in 1 piece combined with frontal repositioning. Numerous modifications have been proposed, which advance a large proportion of the periorbital rims but avoid such a nasocranial communication.

### Table 2. Adjunctive Facial Procedures Following “Spectaclesplasty”

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fat grafting malar</td>
<td>5</td>
</tr>
<tr>
<td>Rhinoplasty</td>
<td>4</td>
</tr>
<tr>
<td>Bone graft malar</td>
<td>2</td>
</tr>
<tr>
<td>Removal of plate</td>
<td>1</td>
</tr>
</tbody>
</table>

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Tessier\textsuperscript{13,14} described a subcranial osteotomy advancing three quarters of the orbital rims and bone grafting the remaining supraorbital rim. He further describes advancing the periorbital skeleton only in those patients with Crouzon syndrome who have normal occlusion. Posnick et al\textsuperscript{15} describes a transcranial osteotomy that advances the upper three quarters of the orbital rims and bone grafts the infraorbital rim. With the advent of distraction osteogenesis, distraction of the monobloc segment has been described in an attempt to limit the creation of extradural dead space adjacent to the nasal cavities, and this technique may limit the frequency of severe complications.\textsuperscript{18}

Spectaclesplasty is an attempt to overcome the drawbacks associated with previously described techniques. Intraoperatively, the exact movement is a matter of clinical judgment and is not preplanned by cephalometrics of any other method. Of the deformities seen in Crouzon syndrome, spectaclesplasty specifically addresses (1) the forehead shape and its retropositioning, (2) mild hypertelorism if deemed necessary, (3) exophthalmos, and (4) the "dish face" deformity with malar advancement. Vertical midface lengthening and dental occlusion are addressed at the second-stage procedure.

The maxilla is hypoplastic in all dimensions, and increasing the vertical height of the middle third is usually required in Crouzon syndrome. This is unable to be achieved with the monobloc procedure. Both the monobloc and the various modifications of the Le Fort III fail to correct the anterior open bite deformity when the midface is advanced to a position that optimizes periorbital aesthetics. The forehead and periorbital bony rims reach nearly adult proportions well before the cessation of maxillary and mandibular growth. For this reason, surgery of this region may be contemplated in early adolescence at a time when psychosocial issues are becoming more pronounced. Skeletal correction of the jaws is performed once skeletal maturity has been demonstrated as is the general principle in orthognathic surgery. The cranial cavity is close to adult proportions at age 6 years, and further brow advancement relative to the globe is limited mostly to that achieved with pneumatization of the frontal sinuses. Consequently, the forehead position is relatively stable at a time when significant vertical and midfacial growth has yet to occur.

In the Crouzon syndrome, the nasofrontal angle is usually relatively normal, although the nasal base is retruded as a consequence of maxillary hypoplasia. Optimal correction of the typical dish face deformity involves advancing the infraorbital rim forward significantly more than the supraorbital rim. If this advancement rotation movement is performed with the Le Fort III segment, the nasal aesthetics are usually improved; however, the anterior open bite deformity is exaggerated, and there is no ability to correct the forehead retrusion. Advancement of a brow that is retruded with respect to the globe assists exophthalmos correction and improves periorbital aesthetics. Although onlay bone grafting to the forehead has been practiced, a degree of graft resorption is inevitable. If the Le Fort III segment was advanced along the vector required to correct the occlusal discrepancy, the orbits become vertically elongated, the nose excessively long, and the infraorbital rim is usually often still relatively retruded.

Both the Crouzon phenotype and its associated treatment may give rise to nasal deformities ideally treated with a rhinoplasty. Because spectaclesplasty is usually performed well before the cessation of nasal growth, final correction is not performed until this time. Although 9 of our patients had a rhinoplasty, it is also our impression that the nasal deformity is not as marked as we have seen after our Le Fort III advancements.

There are 2 main concerns with the spectaclesplasty technique, namely, the creation of retrofrontal dead space and the creation of a communication between the nasal and cranial cavities. Nasocranial communication is also created by the monobloc procedure. Galeal frontalis flaps were used to minimize complications when this was felt necessary. Retrofrontal dead space has been cited as the cause of frontal bone resorption in the absence of vascularized dura against the posterior surface. Extradural dead space is an added concern when it is adjacent to a communication with the nasal cavities. Because the mean advancement of the supraorbital bar in our series was 8 mm, the actual retrofrontal dead space is minimal. This consideration is a further advantage in performing spectaclesplasty in early adolescence rather than waiting till adulthood when brain expansion to fill the dead space is more prolonged.

It is our impression that spectaclesplasty en boc rotation advancement of the periorbital bony skeleton can be safely performed before skeletal maturity, and compared with other techniques in our hands, this produces a more normal anatomic position of the periorbital soft tissues facilitating both function and aesthetics.

**References**