Results of the Tessier Integral Procedure for Correction of Treacher Collins Syndrome

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A new procedure is proposed for correction of the malformations of severe cases of Treacher Collins syndrome in the areas of the maxilla, mandible, and orbit. The operation essentially consists of a rotation of the midfacial segment around a transverse axis at the frontonasal angle. The midface is rotated forward anteriorly and downward posteriorly. It is stabilized by cranial grafts impinged into the temporal bone and the maxilla. The mandible is lengthened either by V shape osteotomy or bone grafting of the ramus. The mandible is fixed in an overjet class III dental occlusion. A posterior bite wafer is maintained for 2 months. The construction of the orbital cavity is completed with cranial bone grafts.

The Tessier integral procedure for Treacher Collins syndrome has been done in one stage in six patients and in two stages in five patients. Sufficient data were available for analysis of only four patients.

Treacher Collins syndrome is characterized by lack of development of the zygomas, maxilla, and mandible, with associated soft tissue anomalies. The zygomatic bones and arches may be totally absent. The mandible is underdeveloped. The maxilla also is retracted, even though it may look protrusive because of the small mandible. Lack of bone in the orbital region is specific to Treacher Collins syndrome.

The orbital osseous and soft tissue defects have attracted the primary attention of craniofacial surgeons (Tessier, 1976). After two or three operative stages, including bone and sometimes cartilage grafts, it is possible to achieve a proper position of the eyelids and lateral canthus. Until now, the retrusion of the lower face has always been corrected by an advancement genioplasty in our experience. This may be sufficient when the mandible is not too hypoplastic. In more severe cases, however, the hypoplasia and backward rotation of the midface and mandible results in choanal atresia and pharyngeal obstruction producing breathing difficulty, which is the main functional anomaly in these patients. As demonstrated on a lateral radiograph, there is no space between the sphenoid and the palate; there is a complete choanal atresia. The method and results of a new surgical procedure to manage the maxillary and mandibular deformities and compromised nasopharyngeal airway of patients with Treacher Collins syndrome is reported.

SURGICAL PROCEDURE

In 1982, a radical procedure was developed by Paul Tessier to correct at the same time the choanal atresia, mandibular hypoplasia, and pharyngeal obstruction and to rebuild the zygomatic bones (Tulasne and Tessier, 1985). It consists of a Le Fort II type osteotomy and total mandibular osteotomy and is performed under tracheostomy. Cranial bone grafts are used to build the orbital regions and to stabilize the midfacial and mandibular segments (Fig. 1).

The Le Fort II type osteotomy permits caudal displacement of the maxillary tuberosities and the posterior part of the palate by a midface rotation around the frontonasal angle. This rotation also enlarges the choanae. The anterior midfacial segment is impacted into the frontal sinus. It is stabilized by strong bony cranial grafts between the maxilla and temporal bone, reconstructing the absent zygomatic arches. The lengthening of the ra-
FIGURE 1 Tessier's integral procedure for correction of Treacher Collins syndrome: A, tracings of the midfacial and mandibular osteotomies. B, Bone grafts are taken in the parietal area and split into halves. C, Following the Le Fort II osteotomy, the midfacial segment is rotated anteriorly and impacted into the frontal sinus. D, It is stabilized by strong bony cranial grafts between the maxilla and the temporal bone. E, The lengthening of the ramus is made by an inverted L-shaped osteotomy and interposition of bone graft. F, The construction of the zygoma, orbital floor, and lateral orbital wall is also made by cranial grafts and additional iliac or tibial grafts.
mus is made by an inverted V-shaped osteotomy and interposition of bone graft. The construction of the zygoma, orbital floor, and lateral orbital wall is also made by cranial grafts and additional iliac or tibial grafts. Intermaxillary fixation is applied on a splint, which maintains an anterior crossbite and posterior openbite. The fixation is discontinued after 2 months.

Because of the backward stress of the mandible, the stability of the midfacial segment is of paramount importance. Therefore, it may be prudent to perform the radical procedure in two stages, starting with the rotation of either the midface or the mandible. The second stage is performed at least 6 months after the first.

CLINICAL RESULTS

The Tessier integral procedure for Treacher Collins syndrome has been done in one stage in six patients and in two stages in five patients. Sufficient data were available for analysis of only four patients. Serial cephalometric tracings were superimposed according to the technique of Baumrind (1976). A fixed triangle defined by the points Basion (Ba), Sella (S), and Sphenoethmoid intersect (Se) is used for superimposition. This area is not affected by the surgery. Furthermore, after six years of age, little if any incremental increase is noted in S-Se distance, and the angle Ba-S-Se shows no change. Tracings of the palate and mandibular contour allow for study of changes following surgery (Fig. 2).

Case 1

The first patient to undergo the integral procedure was a 12-year-old girl. Photographs, radiographs, and cephalometric tracings of this case are shown in Figures 3 to 5. She was operated upon in one stage by Drs. Tessier and Tulasne in April, 1982. A midface anterior rotation was achieved through a Le Fort II osteotomy. The mandible was lengthened through an inverted L-shaped osteotomy. Fixation of the fragments and orbital construction was done with cranial and tibial bone grafts. The supraboral region was released with a large Z-plasty. The palatal plane was rotated 15 degrees. The chin was advanced 34 mm. Radiographic control after releasing the intermaxillary fixation showed a slight posterior rotation of the midface and a 6 mm backward displacement of the mandible. One year later, the position of the fragments was stable. Fourteen months after the radical procedure, additional bone grafts (cranial and tibial) were placed in the orbital region. Genioplasty and extraction of four bicuspsids were performed at that time. Radiographic evaluation 6 months later demonstrated extensive resorption of the chin fragment, as is usually observed in children. At age 14½ years, 31 months after the radical procedure, a third set of bone grafts (cranial and iliac) were placed in the orbital region and a second genioplasty performed. Comparison of serial cephalometric tracings from the first postoperative radiograph (May, 1982) to the radiograph control, 44 months later, showed vertical growth and progressive horizontalization of the palatal plane.

Case 2

A 7-year-old boy underwent the integral procedure in two stages by Dr. Tessier. In October 1979, midface rotation and orbital construction with cranial grafts was performed. One year later, at age 8 years, mandibular lengthening was performed. During the subsequent 4 years from 1981 to 1985, the patient had four operations including bone grafts of the orbits, full-thickness cutaneous grafts of lower eyelids, and three other mandibular lengthenings. Postoperative radiographs were available for study from April, 1980, November, 1982, and June, 1983. From their evaluation, we can conclude only that the position of the midfacial segment was stable 3½ years following operation.

Case 3

A 7½-year-old boy underwent the integral procedure in two stages by Drs. Tulasne and Tessier. In July, 1984, midface rotation and orbital construction with cranial grafts was performed. Five months later, the mandible was lengthened, and additional cranial and tibial grafts were added to the zygomas. The previous cranial grafts had resorbed almost totally. Postoperative radiographic tracings from September, 1984 to January, 1986 showed a stable midfacial segment, but there was a complete relapse of the mandibular advancement.
FIGURE 3 Case 1: A, Patient at 11 years of age before treatment. B, Patient at 16 years of age following a one-stage integral procedure and two additional operations on the orbits and chin.
FIGURE 4  Case 1:  A, Cephalometric radiograph before surgery.  B, Cephalometric radiograph 12 months after the one-stage integral procedure (age 12 years).  C, Cephalometric radiograph following the second operation (age 13 years).  D, Cephalometric radiograph 14 months after the third operation (age 16 years).
FIGURE 5 Case 1: A, Superimposition of tracings before and after the one-stage integral procedure. B, Superimposition of tracings immediately and 44 months after the one-stage integral procedure. Two genioplasties were performed in 1983 and 1984. C, Superimposition of tracings before and 44 months after the one-stage integral procedure.
Case 4

A 16-year-old girl underwent the integral procedure in two stages by Dr. Tulasne. Photographs, radiographs, and cephalometric tracings are shown in Figures 6 to 8. In July, 1982, the mandible was lengthened with tibial grafts and a genioplasty performed. The chin was advanced 30 mm. One year later, she underwent Le Fort II midfacial rotation. The midface was stabilized and orbits constructed with cranial and iliac bone grafts. In 1985 and 1986, additional iliac bone grafts were added to the zygomas and upper blepharoplasties, and lateral canthopexies were performed. Serial cephalometric tracings from July, 1982 to March, 1986 showed excellent stability of both the midfacial and mandibular segments. The 25 percent resorption of the chin fragment is consistent with that usually observed after genioplasty in adults.

FIGURE 6 Case 4: A, Patient at 16 years of age before operation. B, Patient at 20 years of age following a two-stage integral procedure with genioplasty and two additional operations on the orbits and eyelids.
FIGURE 7  Case 4:  
A. Cephalometric radiograph before surgery.  
B. Cephalometric radiograph after the first operation which consisted of lengthening of the mandibular rami and genioplasty.  
C. Cephalometric radiograph in March, 1986 after midface rotation and orbital construction times two.
FIGURE 8  Case 4: A, Superimposition of tracings before and after the first operation. B, Superimposition of tracings 2 months and one year after the first operation, showing a good stability of the mandible and a partial resorption of the chin fragment. C, Superimposition of tracings before and 6 months after the second operation (midfacial osteotomy) showing the lowering of the palate and additional resorption of the chin area.
FIGURE 8 (Continued)  

D, Superimposition of tracings 6 months (January, 1984) and 20 months (March, 1986) after second stage showing good maxillary stability. E, Superimposition of tracings before and 44 months after the first operation.

**DISCUSSION**

This radical procedure can be performed either in one stage with tracheotomy or in two stages separated by an interval of 6 months or more. In principle, this one- or two-stage procedure is reserved for the most severe cases of the syndrome with respiratory compromise due to choanal atresia and micrognathia. However, the operation can be extended to less severe cases who require lowering of the maxillary tuberosities to increase the posterior maxillary height to stabilize lengthening of the mandibular rim.

Eleven patients have been operated in one or two stages according to this new pattern. The breathing problems have been solved totally in all of them. In four patients, serial cephalometric studies have shown good stability of the mid-facial advancement. There is a strong tendency for relapse of the mandibular advancement, however, in young patients.

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**REFERENCES**

