The Use of a Temporal Osteoperiosteal Flap for the Reconstruction of Malar Hypoplasia in Treacher Collins Syndrome

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The Treacher Collins syndrome is an uncommon hereditary syndrome seen in many degrees of severity ranging from the complete form to the incomplete form, including the abortive form. Malar hypoplasia and an eyelid coloboma are the "hallmark" of this malformation, the incomplete form of which was first described by Berry.2

The complete form of the syndrome, also known as mandibulofacial dysostosis, was first reported by Franceschetti and Klein3 and involves malformations of the temporoaural complex and the mandible.

In the past, correction of the malar bone has been obtained by using different materials such as silicone,4 dermis fat,5 cartilage6 (modeled or diced, as autograft, homograft, or xenograft), and split rib.7 Currently, layered split-rib and ilium grafts are used by many surgeons.

The disadvantages of inorganic implants such as silicone are well known: dislocation and extrusion may occur due to capsule contraction. Autologous costal cartilage seems to retain some growth potential,8 but unfortunately, it is not available in sufficient quantities. Free split-rib and ilium grafts finally may atrophy, necessitating a series of repeated corrections. No technique is therefore totally satisfactory.

These considerations and the fact that, particularly in children, there is only a limited amount of bone available for reconstruction purposes have stimulated us to look for other solutions. Osteocutaneous flaps incorporating a distal phalanx,9 the ulna,10 frontal bone,11,12 clavicle,13-17 and rib,15-17 and osteomyocutaneous flaps incorporating clavicle17 and rib18 have been used successfully for the reconstruction of skeletal facial defects.

Medgyesi19 compared osteocutaneous and osteomuscular flaps in goats by ink perfusion of the vascular connection between soft tissues and bone and observed poor filling in the osteocutaneous flaps. Despite this, viability of these flaps could be documented by sequential tetracycline labeling. An osteomuscular flap, pedicled on the pericranium and the temporal muscle, was used by Watson-Jones20 for closure of a cranial defect and by Conley17 for reconstruction of an orbit following severe trauma. Siemssen et al.21 recently recorded their excellent results in a large series of mandibular reconstructions with sternomastoid flaps, including the clavicle. Encouraged by these reports and by the result of a recent experimental study22 in which we could demonstrate the osteogenic capacity of temporal periosteal flaps in young pigs transferred from the cranium to the facial skeleton, we have used a temporal osteoperiosteal flap for the reconstruc-
tion of the malar bone in a child with Treacher Collins' syndrome. Our experience with this technique is presented here.

**Case Report**

A 2½-year-old child with Treacher Collins syndrome was admitted for bilateral malar reconstruction. The skeletal defect was associated with an antimongoloid slant of the palpebral fissures and notching of the lower lateral eyelids. With the exception of an external and middle ear underdevelopment, there were no other abnormalities (Fig. 1).

On September 22, 1982, the frontalperiosteum, the temporal muscle, and the skeletal defect in the malar area were exposed by means of a coronal incision and the orbital contents were dissected free. On each side of the skull, a periosteal flap based on the temporal muscle was outlined and raised from its bed in continuity with a tripod-shaped bone graft, split thickness on the left side full thickness on the right side.

This composite flap (Figs. 2 and 3) was transposed into the infraorbital region and fixed to the maxillary periosteum through a subciliary incision. The bone and periosteum were used for reconstruction of the infraorbital rim and orbital floor and gave the malar the required prominence.

The defects in the skull were left to heal spontaneously. The postoperative period was uneventful.

**Fig. 1.** Preoperative view of the patient with Treacher Collins syndrome. Note malar hypoplasia and coloboma of the lower eyelid.

**Fig. 2.** Schematic illustration of the transposition of a temporal osteoperiosteal bone flap to the malar region (modified after Tessier).

On February 28, 1983, the remaining malformations of lateral canthus and lower eyelid were corrected in the following manner. A palpebrotemporal flap was raised, and the lateral canthus and lower eyelid were released from their tethering structures by means of a subciliar incision. An intact orbital rim was thus exposed, and its viability was demonstrated by bleeding from a burr hole. The lateral canthus was fixed in the planned position by a canthopexy, and the palpebrotemporal flap was transposed into the lower eyelid by rotation and advancement (Fig. 4). A surplus of skin was thus produced in the area over the malar bone, where it was most atrophic and deficient. This surplus was trimmed and the wound was closed.

Tangential x-rays made more than 1 year after the first operation produced an image similar to that seen in the pictures that were taken after transposition of the compound flap. There was no sign of resorption and perhaps even an indication of growth (Fig. 5). The appearance of the patient remains satisfactory (Fig. 6).

**Anatomy**

Directly between the skin and firmly united by fibrofatty tissue one finds the temporoparietal fascia and the galea aponeurotica, which together form the subcutaneous fascia. The temporoparietal fascia has its origin in the fascia of the temporal muscle beneath the temporozygomatic arch. Distinct from and superficial to the fascia of the temporal muscle, it becomes contiguous with the galea aponeurotica in the vicinity of the temporoparietal suture. The fascia of the temporal muscle is contiguous with the pericranium, covering the frontal and parietal bone.

Pericranium and galea are separated by a layer of loose areolar tissue. The blood supply in the temporoparietal area is provided by the two deep temporal vessels on the inner deep surface of the temporal muscle and the superficial temporal arteries. The latter vessels run in the subcutaneous fascia and can be traced from the level of the root of the ear to a point approximately 10 cm above the crus helicis, where they become invisible. An anterior branch of the superficial...
FIG. 3. (Left) Design of the compound flap. (Right) Mobilization of the compound flap.

FIG. 4. Schematic illustration of advancement rotation flap for the correction of the eyelid coloboma.

temporal artery splits off at the level of the tragus.

Examination of the pericranium reveals that the outer layer consists of loose connective tissue with fibroblasts and the inner layer of osteoblasts. Vascularity is more extensive in the inner than in the outer layer, and communication between periosteal and endosteal vessels has been demonstrated by angiography and by ink perfusion techniques.

The role of the periosteal vessels in the nutrition of the cortex is not an important one, and their nourishing function is restricted to the outer third of the cortex. The blood for the inner two-thirds of the cortex and the medulla is supplied by the nutrient arteries.

DISCUSSION

The Osteomuscular Flap

The construction of a malar bone with an osteomuscular flap is a major procedure involving an extra operative risk. The question whether the proposed technique is justified will therefore have to be answered first. In our opinion, this risk is acceptable when it is balanced by obvious advantages.

The first advantage concerns the survival of bone when it is transferred as part of an osteomuscular flap. Comparing the fate of pedicled and free bone grafts by histologic sections, autoradiography, fluorescence microscopy, and injection of vessels, Baadsgaard and Medgyesi were able to show that pedicled bone grafts survive while free bone grafts resorb. Brookes and Rhinelander et al. explained the survival of
these flaps by a reversal of the normally centrifugal blood supply into a centripetal one that will maintain the viability of the bone. Brookes added the provision that revascularization of the periosteum from capillaries at the receptor site occurs. His view is consistent with the observation that grafts with periosteum undergo less resorption than grafts stripped of periosteum. Conley et al. studied the biological activity of bone by the uptake of fluorine 18 after transfer of osteomuscular flaps and concluded that the balance and proportion of the changes that occur are dependent on the volume and distribution of direct nourishment after transfer. Conley et al. based their temporal bone flap on the superficial and deep temporal arteries and stated: "The temporal bone flap, based as it was on the superficial and deep temporal arteries, was the composite flap with the more generous blood supply and one in which the nutrient artery was preserved."

Tessier believes that osteomuscular flaps are viable on the basis of his experience in a large number of cases. However, he refrained from using this technique in patients with the Treacher Collins syndrome in order not to weaken the already hypoplastic temporal muscle. Curioni et al. have recently described a technique for reconstruction of the orbital floor with the temporal muscle including the coronoid process. They feel that the vitality of the bone may thus be ensured.

In our patient, the bone flap was designed on both sides of the temporal crest; one half covered

FIG. 5. (a and b) Tangential projections of the malar bone made shortly after its reconstruction showing the full-thickness bone segment on the right side. (c and d) Tangential projections made more than 1 year later showing split-thickness fragment on the left side.

Fig. 6. The appearance of the patient more than 1 year postoperatively.
Fig. 7. Fluorescence microscopy shows osteoid seams covered by layers of very active cylindrical osteoblasts near large round lacunae.

by the aponeurosis of the temporal muscle and the other half by the pericranium. By choosing this location we felt that maximal benefit could be drawn from the circulation provided by the deep temporal vessels in the muscle flap. Evidence of a continuing blood supply through the nutrient artery could not be obtained in this case, but long-term vascularity could be demonstrated in the following manner. With permission of the parents, a biopsy was taken after the administration of 40 mg Vibramycin for two periods of 2 days with an interval of 5 days prior to the operation. Microscopy revealed newly formed fibrous bone with numerous osteocytes in large round lacunae adjacent to the preexisting bone. Ostoid seams covered by a layer of cylindrical osteoblasts were seen in several sites. An appositional rate of 2.6 mm per day was measured by quantitative fluorescence microscopy, providing proof of very active bone formation.

McCarthy\(^{36}\) has also used this technique for the treatment of patients with the Treacher Collins syndrome. He holds the opinion that a compound flap raised in the calvaria beyond the temporal line should include the galea for adequate vascularization. We agree that inclusion of the superficial temporal artery or its anterior branch in the osteomuscular flap should be considered when the bone flap is designed exclusively in the calvaria beyond the temporal crest. The galea is extremely well vascularized, and a somewhat more difficult dissection in a subfollicular plane is a small price to pay for better vascularity of the flap. Whether an osteoperiosteal flap nourished by the galea retains its viability is not known, but if it does, the applicability of such a flap, with its wide arc of rotation, would be extensive. Further studies are therefore indicated.

The second advantage relates to the osteogenic potential of the cambium layer of the periosteum, which is at its maximum in young animals.\(^{37,38}\) Early transfer of a viable osteomuscular flap will take full advantage of this potential and perhaps help to increase the volume of transferred bone. In addition, spontaneous closure of the donor defect can also be expected.\(^{39}\) The younger the child, therefore, the better the conditions for optimal repair.

A third advantage may eventually be formed by the fact that the cambium layer of the periosteum is a "going concern."\(^{37}\) It maintains growth and reacts to stimuli. There is evidence now that periosteum responds to surface tension\(^{40-43}\) and that detachment or transposition of the temporal muscle is followed by changes in the craniofacial morphology.\(^{43-54}\) Whether the transposition of a hypoplastic muscle has the same effect remains
questionable, but the possibility should not be neglected. The conclusion seems justified that the temporal osteoperiosteal flap may prove to be of value for the treatment of patients with the Treacher Collins syndrome. Whether the compound flap retains its viability from the beginning and whether the transfer of such a flap offers advantages over the use of an osteoperiosteal graft is, however, still uncertain. Membranous grafts are revascularized earlier than endochondral grafts and will contain a greater percentage of living bone. Grafts with periosteum are superior to those without for the same reason. It is therefore possible that the advantage of a compound flap over a membranous osteoperiosteal graft is only marginal.

The Cutaneous Flap

In the malar area of patients with the Treacher Collins syndrome, skin is not only deficient, but also atrophic. Correction of the coloboma is usually obtained by the transposition of a triangular skin flap from the upper eyelid. A composite flap may be used in cases with deficiency of the conjunctival lining. We feel that conditions in this area may benefit when the upper eyelid flap is designed as part of the temporal flap. Rotation of the palpebral part will correct the coloboma. Rotation and advancement of the temporal part will produce a dogear in the hypoplastic area and make it possible to resect atrophic skin.

SUMMARY

The clinical use of a temporal periosteal bone flap for the reconstruction of a malar bone in a patient with the Treacher Collins syndrome is presented. The temporal muscle functions as an axial carrier of the periosteum that induces osteogenesis in young children, whereas the bone segments may serve as a nucleus for further bone formation from the periosteum. Correction of the eyelid coloboma was obtained by the rotation and advancement of a temporopalpebral flap.

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