Simultaneous orbital expansion and intraoral distraction osteogenesis of upper and lower jaws in a patient with hemifacial microsomia

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In memory of Prof. Dr. med. Dr. med. dent Dr. med. h.c., Gerhard Pfeifer, a great teacher and a great scientist

SUMMARY. Background: Correction of a micro-orbit, caused by clinical anophthalmia is a very challenging task. In hemifacial microsomia a micro-orbit may be combined with hypoplasia of the malar and the ascending mandibular ramus. Material: A 5-year-old patient with hemifacial microsomia is described. Hypoplasia of the malar bone and the tilted occlusal plane were corrected by means of intraoral distraction osteogenesis in the upper jaw following precise simulation surgery on a 3D-model. At the same time, the ascending mandibular ramus was lengthened with a second distraction device and a spherical tissue expander was inserted into the hypoplastic orbit. Results: The malar as well as upper and lower jaws were lengthened and positioned symmetrically. The orbital cavity was expanded to 79% of that of the healthy side. Following removal of the distraction devices, the expander was exchanged for a larger one and orbital expansion was continued until overcorrection of the orbit up to 118 percent was achieved. Conclusion: Orbital expansion is a minimally invasive method of enlarging the volume of the eye socket three-dimensionally. Intraorally activated buried distraction devices enable “growth” of the jaws making bone transplants avoidable in many cases. By combining both methods, complex malformations can be corrected simultaneously in children. © 2003 European Association for Cranio-Maxillofacial Surgery.

Keywords: Distraction osteogenesis; Congenital malformation; Microphthalmia; Anophthalmia; Orbital expansion

INTRODUCTION

Surgical therapy of asymmetries and growth deficiencies in children, as well as three-dimensional enlargement of complex structures such as the orbit are both very difficult.

Hemifacial microsomia is a malformation of the first and second branchial arch usually associated with an underdeveloped ascending ramus and malar. It is often combined with microphthalmia or anophthalmia, the palpebral fissure is short and the conjunctival sac is underdeveloped, thus impeding ophthalmic prosthesis. Additionally, epibulbar dermoids, heart failure and spinal malformations can occur (Goldenhar, 1952).

Correction of facial asymmetry and enlargement of the orbit are the first steps to be done by the cranio-maxillofacial surgeon. After symmetrical bony structures have been achieved, eye lids and conjunctival fornices have to be constructed.

Distraction osteogenesis has proved to be a safe method for enlarging bony structures (Snyder et al., 1973; Ilizarov, 1988; Altuna et al., 1995; Sawaki et al., 1997) and for closing bony defects (Henkel et al., 2001). This procedure enables new bone formation, especially in children (Karp et al., 1990; Klein and Howaldt, 1996; Carls and Sailer, 1998; Cohen et al., 1997; Nocini et al., 2002). Not only the mandible (Pensler et al., 1995; Chin and Toth, 1996; Klesper et al., 2002). Not only the mandible (Pensler et al., 1995; Chin and Toth, 1996; Klesper et al., 2002) but also the maxilla and mid-face can be enlarged or transported by means of distraction osteogenesis (Block and Brister, 1994; Glat et al., 1994; Rachmiel et al., 1995). New subcutaneous distraction devices, submerged subperiostally and, leaving no visible facial scars, have led to this technique being frequently used in the maxillo-facial region (McCarthy et al., 1995; Wangerin and Gropp, 1995; Diner et al., 1997; Hendrickx et al., 1999; Rubio-Bueno et al., 2001; Santler et al., 2001; Van-Strijen et al., 2003).

Surgical treatments have been described on how to enlarge micro-orphbits (Wiese et al., 1999) thus avoiding major surgical correction which necessitates a cranio-facial procedure with multiple segmentation of the orbital rim and bone grafts (Marchac et al., 1977). The further growth of these enlarged orbits created by local and transplanted bone is not predictable. A purely conservative approach induces orbital growth by means of orbital conformers of increasing size (Dootz, 1992), however, the attained growth increase is limited. Rodallec et al. (1988) successfully used intraorbital expanders to induce orbital growth.

A combination of both methods in one patient is described using simultaneous distraction of the upper and lower jaws plus expansion of the orbit.

MATERIAL AND METHOD

The face of a 5-year-old boy showed deviation to the affected right side (Fig. 1). The extent of the
malformation was analysed using a CT-based three-dimensional model (Fig. 2). The right ascending mandibular ramus was too short, the occlusal plane tilted upwards to the right and the ipsilateral malar was underdeveloped. The right orbital diameters were found to be reduced from 29.2 to 15.1 mm vertically and from 29.8 to 21.3 mm horizontally when compared to the unaffected side. The orbital volumes showed corresponding differences: 5.2 ml versus 20.3 ml.

Simulation surgery on the 3D-model (Fig. 3) with submerged distraction devices (Vazquez-Diner System, Stryker-Leibinger, Freiburg, Germany) revealed location and direction of distraction osteogenesis. A vertical vector for mandibular distraction was chosen to achieve a horizontal occlusal plane. In the mid face, a maxillo-malar osteotomy on the affected side and a high LeFort I osteotomy on the contralateral side were planned. A second distraction device was responsible for moving the maxilla and malar anteroinferiorly on the right side in order to achieve symmetry of bone and teeth.

A spherical expansion device of 10 cm³ was inserted to increase the volume of the extremely small orbit (Fig. 4). The entire procedure was performed under general anaesthesia via a hemicoronal incision and two intraoral approaches. The osteotomies were performed as planned on the 3D-model. The distraction devices and the expander were placed subperiostally. The activation rods perforated the oral mucosa enabling activation from intraorally. The port of the expander, to be filled with saline, was placed subcutaneously in the temporal region enabling complete wound closure. The periosteum of the orbit was reattached over the expander and trans-osseous sutures prevented anterior dislocation during expansion.

Seven days postoperatively, following the use of antibiotics, the distraction devices were activated by 0.9 mm each day (2 turns). After 19 days the maximal distraction of upper and lower jaw (18 mm) was achieved (Figs. 5–6). A delay of 3 weeks
postoperatively before orbital expansion was performed to prevent anterior dislocation of the expander through the palpebral fissure. 0.6 ml of saline was applied transcutaneously once a week. The resistance of the expander and the protrusion of the eyelids determined the volume. Insufflation was continued until the expander was completely filled (10 ml). A new CT-based model was created 18 weeks post-operatively to check ossification of the distraction gaps and to assess orbital symmetry (Fig. 7). Osseous closure of the osteotomy and the distraction gaps was awaited before the distraction devices were removed. The orbital volume achieved was 79% of that of the healthy side.

In a second procedure, the distraction devices were removed and the orbital expander exchanged for a larger one (20 ml) to gain further orbital expansion and to achieve overcorrection. Expansion was again continued with 0.9 ml every 3 weeks. After the second expander had been filled, a further 3D-model was evaluated (Fig. 8A). In a third procedure, the port of the expander was removed. The expander itself was left in place to stabilize the orbit until growth had ceased. The palpebral fissure was widened and the conjunctival fornices required reconstruction to enable insertion of an eye prosthesis.

The measurements of the 3D models were carried out with an electronic caliper. In these the orbits were filled with soft wax and their volumes calculated using the water displacement method.

RESULTS

Activation of the distractor was painless for the patient. The parents performed the daily distraction themselves after given an initial demonstration.

A small overcorrection was planned to compensate for relapse or possibly reduced growth later. A symmetrical bony appearance was achieved (Fig. 8A) and the facial asymmetry was corrected (Fig. 8B). During removal of the distraction devices newly
formed bone was found at the original osteotomy and distraction gaps. No infection was found, despite the activation rods perforating the oral mucosa and interconnecting bone, distraction and the mouth.

A soft-tissue capsule was found enveloping the expander covering the bone. The bony orbital contour was intact and there was no evidence of any bony defect or resorption.

The spherical tissue expander induced a three-dimensional enlargement of the orbit. The orbital volume increased from 5.2 to 16.5 ml in the first step and to 25.4 ml finally, compared with 21.5 ml contralaterally side (118%). The orbital diameters were increased from 15.1 to 29 mm vertically and from 21.3 to 33.7 mm horizontally. The diameters of the unaffected orbit showed normal growth during the treatment period finally measuring 30.4 mm vertically and 31.1 mm horizontally. The clinical result remained stable during the following 3 years (Fig. 8C).

DISCUSSION
Callus distraction enables new bone formation following a minor surgical procedure. Early correction can be achieved without the need of bone grafts especially in the case of congenital malformations and other bone deficiencies in children (Santler et al., 2001). Bone grafts in children have the disadvantages of a prolonged surgical procedure and unpredictable growth (Reich, 1991). The minimal age for distraction is believed to be about 3 years (McCarthy et al., 1992). In younger children, the immature bone allows
the fixation pins to migrate through the bone, thus reducing the effective length of distraction (Hollier et al., 1999). Distraction osteogenesis lengthens not only bone but also the surrounding soft tissue (Fisher et al., 1997).

The use of submerged distraction devices has two advantages. On one hand, scars on the face can be avoided completely, as there are no fixation pins penetrating the skin. On the other hand, social interaction remains unaffected, as the distraction devices are hardly noticed.

In the treatment of complex three-dimensional malformations, as in this case, exact 3D-planning is of major importance (Kärcher, 1992; Stoker et al., 1992; Lambrecht, 1995; Roth et al., 1997; Santler et al., 1998b). The optimal choice of the individual distraction device, and determination of the distraction vector and length are possible using 3D-models. In addition, important structures like tooth germs or intrabony nerves can be taken into account on positioning the distraction device. By simulating the distraction on the 3D-model, the result of the procedure can be predicted, operation time reduced and precise results achieved (Santler et al., 1998a).

Correction of underdeveloped orbits by means of expanders has already been described by Rodallec and co-workers (1988) in 17 patients. Effectiveness has been proven in animal research, which has also shown that growth induction is reduced with increasing age (Heinz et al., 1997). Thus three-dimensional correction of bony structures with a minimum invasive procedure is possible (Tucker et al., 1995), whereas pure surgical solutions necessitate extensive craniofacial procedures and bone transplants. Achieving symmetrical orbital structures surgically is the pre-requisite for a satisfactory aesthetic and functional result. It is only after bony construction has been accomplished, that soft tissue correction of palpebral fissure and conjunctival sacs can be carried out.

CONCLUSION

Gradual expansion of the orbit by means of a spherical expander and distraction osteogenesis of both jaws can be combined successfully. The combination of these two procedures allows reduction of the extent of surgical interventions and enables symmetrical and stable bony and clinical results.

References


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