Expansion of the Human Microphthalmic Orbit

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Objective: To determine the effects of long-term, incremental enlargement of an orbital tissue expander on bone and eyelid growth in microphthalmia.

Design: A prospective, noncomparative case series.

Participants: Five consecutive patients with microphthalmos treated with orbital expansion were evaluated.

Intervention: A tissue expander was placed into the orbits of five children (age, 10 months–6 years) with unilateral microphthalmos and gradually enlarged by saline injections.

Main Outcome Measure: The midorbital width of each patient was determined from axial computed tomographic scans before insertion of the device. The length of the normal and abnormal eyelid fissures was measured at surgery. The postexpansion dimensions of both the normal and microphthalmic orbits and the eyelids were remeasured when the expanders were removed. The residual deficits between the normal and the microphthalmic sides were expressed in percentages.

Results: Gradual inflation of the expander to a diameter of 22 mm reduced the average preoperative orbital dimension deficit of the group from 14.6% (range, 8%–25%) to 3.8% after surgery (range, 0.5%–6.3%). The average pre-expansion eyelid length deficit for the group was 17.5% (range, 12%–26%) compared to 2.3% (range, 0.0%–5.3%) after expansion. The average expansion period was 56.8 weeks (range, 20–100 weeks). Two outpatient surgical procedures were required in each patient.

Conclusion: Incremental inflation of a tissue expander placed within the microphthalmic orbit induced sufficient osseous and eyelid growth to ameliorate the major stigmata of this syndrome in all patients treated.
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Microphthalmos is a disfiguring malformation of the eye. In a prospective study of 50,000 pregnancies, microphthalmos occurred with a frequency of 0.22/1000 live births, and there is an empiric recurrence risk of 5% to 10%.[1] Although a specific human genetic abnormality has not been identified conclusively, recent reports suggest that mutations in the PAX6 homeobox gene and chromosomal deletion in the 14q, q22q23 region of the genome may be responsible.[1,2]

The microphthalmic eye has a diameter at birth of less than 15 mm (normal range, 15–19 mm) and may exhibit other abnormalities such as coloboma.[3–5] The hallmark soft-tissue abnormalities of this disorder, minified eyelids with abbreviated palpebral and bulbar conjunctivae, often preclude wear of an ocular prosthesis. Orbital volume parallels ocular growth and its reduction in anophthalmia, and microphthalmia results in noticeable hemifacial deformity, even after treatment.[6]

Many methods have been used to improve the soft-tissue and osseous abnormalities of microphthalmos without visual potential.[3,4,7–11] Numerous surgical procedures or multiple sessions or both to fit conformers are required, and there is substantial difference of opinion regarding which of the methods offers the best outcome. One of these techniques relies on enucleation followed by serial surgical placement of orbital implants of increasing size and concurrent fitting of conjunctival conformers. Although growth of eyelids and orbit is induced by this method, a retrospective analysis of this technique in the management of congenital anophthalmia by Tucker et al[12] found that further orbital enlargement was needed after treatment.

Buchman and associates[13] showed that pressure was an effective stimulant to enlargement of the craniofacial skeleton in a neonatal feline model. The effect of different types of implants in the eviscerated orbit on growth of the cranium and contiguous facial skeleton was compared in three experimental groups: (1) fixed-dimension implants, (2) expandable implants, and (3) no implant. Both implants caused orbital enlargement, but only the expandable device resulted in normal volumes at the conclusion of the experiment. The character of the osseous growth was histologically normal. This morphometric analysis also corroborated the work of narrower studies by Lo et al,[14] Cepela et al,[15] and Heinz et al,[16] in which the effect of expandable implants on orbital dimensions was evaluated.

Orbital expansion has been used in the treatment of microphthalmos for several years (Sadov M, Epply BL, Wetherington GM. Experimental effects of early orbital expansion on orbitomaxillary growth in anophthalmos. Presented at the 47th annual meeting of the American Cleft

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Table 1. Orbital Dimension Deficit

Initials	Age at Expander Placement	Preoperative (%)	Postoperative (%)
M.C.	6 yrs	8.0	0.5
K.F.	4 yrs	12.2	7.3
J.R.	1.5 yrs	17.5	3.5
C.H.	11 mos	25.0	3.0
R.T.	10 mos	13.2	3.9

Table 2. Eyelid Fissure Deficit

Initials	Age at Expander Placement	Preoperative (%)	Postoperative (%)
M.C.	6 yrs	17.7	0.0
K.F.	4 yrs	12.0	5.0
J.R.	1.5 yrs	21.1	4.3
C.H.	11 mos	26.0	1.0
R.T.	10 mos	17.4	2.3

Palate-Craniofacial Association, St. Louis, MO, May 1990). Early implants were positioned via transcoronal incisions in subperiosteal locations within the orbital soft tissue and in one case in the anatomic position of the eye.[17] Results were encouraging but inconclusive because of the small number of patients treated. Dunaway and David[18] reported a long-term experience in seven microphthalmic patients of diverse age. Expanders were placed in either a subperiosteal or intraconal position. Patients older than 2 years of age also underwent osteotomy of the lateral orbital wall to reduce the risk of implant extrusion. The authors reported satisfactory osseous growth in most cases (intraconal expanders seemed most effective), but eyelid growth was inconsistent.

A simplified technique was formulated, based in part on the experiences of previous authors, to position an expander within the extraocular muscle cone. Five patients have been treated by this method. The influence of age, severity of the deformity, and incremental increases in orbital pressure on osseous and eyelid growth in microphthalmia were studied. The outcomes in each patient are presented. To our knowledge, this is the first long-term series reporting results of an intraconal expander in microphthalmia without supplemental osteotomy.

Materials and Methods

Patients referred for treatment of microphthalmia were evaluated by a team consisting of a craniofacial surgeon, an ocularist, and an oculoplastic surgeon. In the absence of visual potential and clear evidence of facial deformity due to underdevelopment of either the frontal bone, zygoma, or maxilla, a surgical solution was deemed appropriate. The parents were offered surgical treatment consisting of serial conjunctival expansion by serial enlarging conformers and solid sphere orbital implants, or orbital expansion. Informed consent was obtained before beginning treatment.

In the orbital expansion group, the midorbital width of both hemiorbits and the ocular diameters were evaluated by computed tomography in each patient before surgery. The orbital dimensions were obtained from an axial tomograph at a point equidistant from the superior orbital fissure and the anterior orbital rim on a plane through the lens of the normal eye. Measurements were made in millimeters, and differences between the hemiorbits were expressed as a percentage, a method similar to that used by Buchman et al.[13] Postexpansion dimensions of the two hemiorbits were measured in each patient and residual deficits were calculated (Table 1). Eyelid fissure length was determined in each patient at the time of expander placement and removal, and data obtained were expressed in the same format as the orbital measurements (Table 2).

Placement of the expander was initiated with the patient under general anesthesia by evisceration of the eye, as described in standard surgical texts. After all uveal tissue was removed, radial incisions were made in the sclera, dividing it into four sections with a rectus muscle attached to each. The sections were released from the optic nerve. Via a lateral canthotomy incision, the temporalis fascia was detached from the zygoma and the temporalis muscle separated from the lateral orbital wall. A 3-mm osteotomy was performed at the zygomaticosphenoid suture to allow the inflation tubing to pass from the orbit.

A vertical incision placed in the temporoparietal scalp, approximately 5 cm above the ear, provided access to the posterior margin of the temporalis muscle. The temporalis fascia was incised at its union with the skull, and a submuscular tunnel was created between the scalp incision and the lateral orbit for subsequent passage of the inflation tubing.

Before insertion of the expander (Micro Model 3608-10; PMT Corporation, Chanhassen, MN), its integrity was confirmed by evacuation of residual air and inflation with saline. Next, the volume of fluid required to inflate the expander to a diameter of 22 mm was determined (Table 3). Then, a silk ligature was secured to the inflation tubing and positioned in the area of the muscle cone. The suture and attached tubing were guided with the aid of a hemostat to the lateral osteotomy, passing beneath the lateral rectus muscle to avoid injury to the abducent nerve (Fig 1). The expander's position within the muscle cone was adjusted by tension on the tubing at the lateral orbit.

A hemostat, which passed through the subtemporalis tunnel to the lateral orbit, was used to grasp the inflation tubing and advance it to the scalp incision. Here it was joined to the microinjection port, and each connection was secured with silk ligatures. Residual air and saline were evacuated from the expander via the injection port, which was then positioned posterior to the incision. The scalp was closed in layers with absorbable sutures.

The scleral incisions were closed over the collapsed expander with interrupted sutures of 6–0 clear polydioxanone. The conjunctiva was approximated with the same material. An acrylic conformer was shaped to fit snugly into the conjunctival fornices. A complete tarsorrhaphy was created, using a temporary suture technique, to distribute uniform pressure to the eyelids and the cul-de-sacs.[19] Tarsorrhaphy also prevented extrusion of the conformer.

Table 3. Expander Inflation Periods (0.5 cc per session)

Initials	Duration [weeks (sessions)]	Final Volume (cc)	Follow-up (yrs)
M.C.	36 (6)	3.0	2.5
K.F.	100 (10)	5.0	2.0
J.R.	80 (8)	4.0	1.6
C.H.	20 (5)	2.5	1.5
R.T.	50 (4)	2.0	3.5

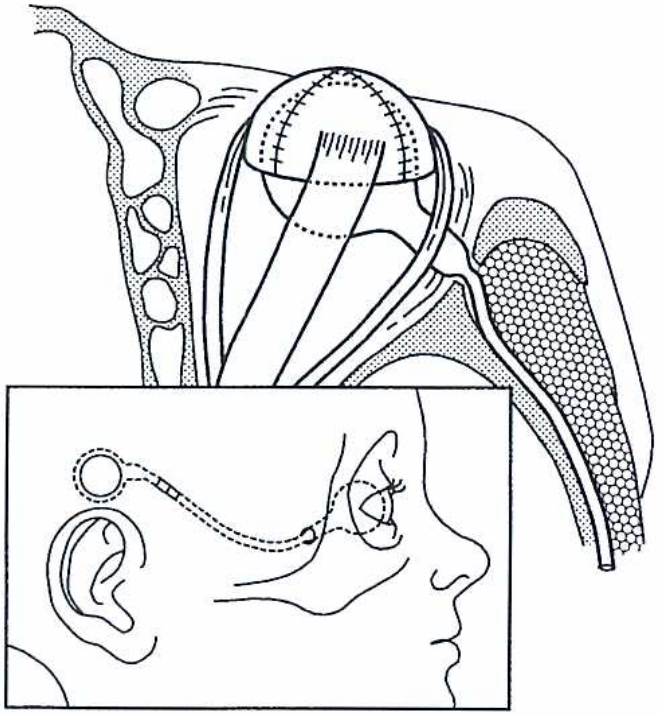

Figure 1. The expander's inflation tubing exits the lateral orbit through an incision in the periorbita via the osteotomy at the zygomaticosphenoid suture (top). The tubing is guided through the subtemporalis tunnel and joined to the microinjection port in the parietal region (inset).

Inflation of the expander was begun 3 months after the initial procedure. One-half milliliter of saline was scheduled to be injected at monthly intervals until the predetermined volume was reached (Table 3; refer to Discussion). This gradual injection protocol was chosen to minimize the risk of scleral dehiscence and extrusion of the expander. All injections were performed by the same surgeon (DMR).

The ocularist was involved when expander injections began. If the volume of the conjunctival fornices could accommodate an aesthetically satisfactory prosthesis, it was crafted at the first visit to the ocularist. Otherwise, a conformer was fashioned to fill the fornices. Once made, the prosthesis was enlarged as necessary every 3 to 4 months thereafter. All patients received a prosthesis within 10 months after initiation of expander filling.

The expander and the pseudocapsule, which it stimulates, were removed 3 months after full inflation and replaced with a solid orbital implant. The length of each eyelid fissure was determined (Table 2). Orbital measurements were obtained by subsequent computed tomography within 2 weeks of the final surgical procedure (Table 1).

Results

Five patients (age, 10 months–6 years) with severe microphthalmos were included in this study. The range of preoperative eyelid length deficit for the group (12%–26%) was reduced after surgery to 0% to 5% (Table 2). The range of preoperative orbital dimension deficit was 8% to 25% compared to a postexpansion range of 0.5% to 7.3% (Table 1). A 22-mm solid implant was placed in three patients, and in only two patients could a 20-mm implant be accommodated. Hydroxyapatite was used in four orbits and acrylic was used in one. A final diameter of 22 mm was confirmed for all PMT expanders (PMT Corporation, Chanhassen, MN) at the time of removal.

Hemifacial dysmorphism, consisting of underdevelopment of the frontal, maxillary, and zygomatic bones, was significantly improved or eliminated in all cases. The anterior orbital aperture, evaluated by coronal computed tomography, was symmetric with the normal orbit in four cases (Fig 2). Eyelid length was nearly equal to the contralateral side in three cases. In two cases, the residual eyelid deficit was 5% or less (Table 2). In the former group, there was no visible difference between the normal and treated sides (Fig 3). In the latter, a mild difference was discernible. Ptosis was present in two cases.

Two outpatient surgical procedures with the patient requiring general anesthesia were performed on each patient. The initial surgery, subsequent saline injections, and expander removal with placement of a solid implant were well-tolerated. Partial conjunctival dehiscence occurred in one case after full expansion was achieved and did not affect the outcome. This complication was attributed to the relatively large size, shape, and rigidity of the

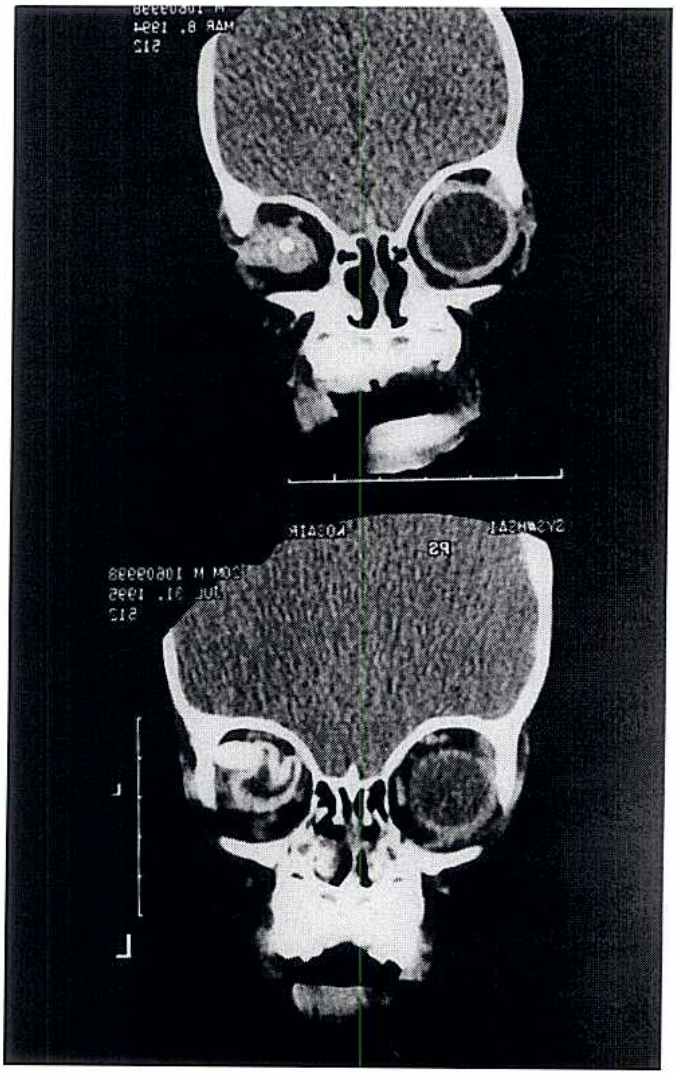

Figure 2. Pre-expansion (top) and postexpansion computed tomograph showing enlargement of the anterior orbital aperture and normalization of the position of the inferior orbital rim. The expander shown (McGhan Medical Corporation) was replaced by one manufactured by PMT Corporation (Chanhassen, MN) because of conjunctival perforation.

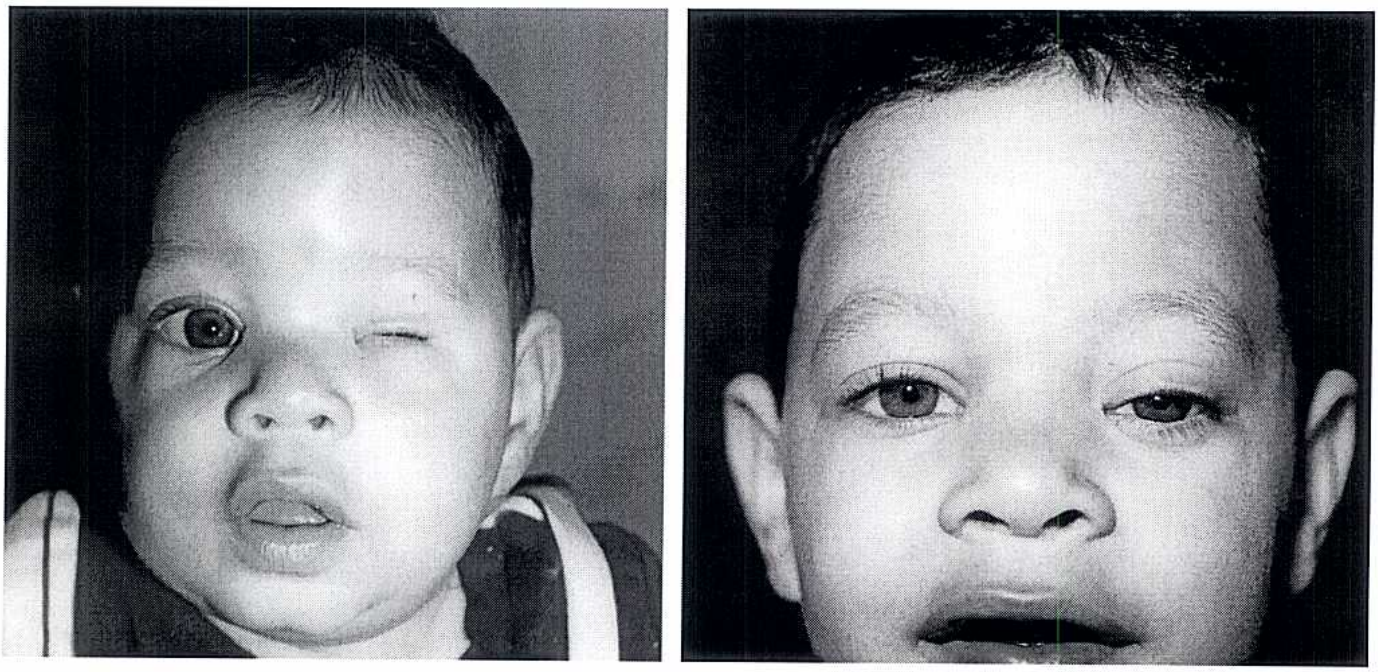

Figure 3. Preoperative appearance at 11 months of age (left). Appearance after an expansion interval of 20 weeks (right).

expander used in the first patient (McGhan Medical Corporation, Santa Barbara, CA LOT # GO 03150) (Fig 2). This problem did not occur with the spherical PMT device used in subsequent patients. There were no infections or expander leakage.

Discussion

A properly positioned tissue expander can deliver sustained, omnidirectional pressure simultaneously to the constituent bones of the orbit. Evisceration of the globe and sectioning of the sclera permit the expander to be placed in the anatomic position of the eye with minimal conjunctival trauma. A conformer that corresponds precisely to the dimensions of the conjunctival fornices allows transmission of orbital pressure to the eyelids. The results of this study show that such forces consistently produce excellent osseous and eyelid growth in microphthalmia.

Recombination of the sclera over the expander creates an adequate barrier to implant extrusion while preserving the potential for ocular motility. Implant retention is additionally favored by proper expander design coupled with its gradual inflation. Small size when collapsed and spherical form during inflation make the PMT expander design satisfactory. However, elasticity of these custom-made devices varies, precluding injection of standard saline volumes to reach a given target diameter. Thus, the number of injections required will vary among patients. The significance of this fact is discussed below.

Age of the subject at the time of implantation was not the sole determinant of the degree of eyelid and orbital growth in this series. The oldest patient, for example, achieved normal eyelid and orbital dimensions, whereas one of the youngest patients did not (Table 2). Similarly, severity of the deformity was not a dominant factor because the patient with the smallest eyelids achieved normal measurements, whereas one of the least-affected patients failed to reach normal size (Table 1).

The variable that seemed to most influence the degree of tissue growth was the time required to complete inflation of the expander (Table 3). Two patients, the oldest and one of the youngest, had expansion intervals of 20 weeks (MC) and 36 weeks (CH). Both attained normal appearance and near-normal anatomic dimensions (Tables 1–3). The expansion periods in the remainder of the group ranged between 56 and 100 weeks. None of the patients in this subset achieved completely normal orbital or eyelid measurements, although residual abnormalities of facial appearance were very slight.

The variability among the expansion intervals was because of differential compliance of the individual devices, as mentioned above, and poor adherence to injection schedules by some parents. Patients receiving less-compliant expanders required more injections to reach the target diameter, which, when combined with parental noncompliance, may have biased the final result in this group.

The important advantages of this approach include predictable growth of the orbit, contiguous facial skeleton, eyelids, and conjunctiva. Only two surgical procedures are needed, and the technique does not produce significant conjunctival scarring. All patients were able to wear a prosthesis within 10 months of expander placement.

When to begin expansion is an open question. However, because the infant eye reaches more than 70% of its adult size at birth and grows most rapidly during the first 12 months of age, initiating the process during the first year of life seems an appropriate goal.[20] A target inflation period of 20 to 36 weeks, begun 3 months after expander placement, would be expected to have a low risk of expander extrusion. The volume of saline required to reach an expander diameter of 22 mm should be apportioned equally into monthly

injections over this period. The consequences of failure to adhere to injection schedules must be made explicit to the patient's parents. In patients who do not attain normal eyelid dimensions, extension of the period of expansion (and the diameter of the expander) may be considered. The efficacy of this approach, however, can be determined only by a study of a larger patient population.

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