

Prosthetic and Surgical Management of Congenital Anophthalmia

ABSTRACT: *A child born with congenital anophthalmos or microphthalmos provides the ophthalmologist and ocularist with their greatest challenge. The concept of conservative prosthetic "expansion therapy" combined with later stage surgical intervention is the best management choice. Specific approaches along with long-term patient follow-up are discussed.*

James H. Merritt

M.D., F.A.C.S.
Dallas, Texas

**Wm. Randall
Trawnik**

B.C.O.
Dallas Eye Prosthetics Dallas,
Texas

Congenital anophthalmos presents the ocularist and ophthalmologist team with their greatest of prosthetic and reconstructive challenges (Figure 1). The goal of the therapies used is to stimulate soft tissue and bony development. The success or failure of these treatments will affect not only the child's physical appearance, but also his or her psychological and social development.

For the ocularist/ophthalmologist team to pursue the best management and treatment of the congenital anomaly, we must first understand its dynamics. Anophthalmos results from the arrest of ocular and orbital development during organogenesis. Facial development begins in the fourth week of gestation and, by the fifth week, the eyes have made their appearance as an elevation of the lateral face. The eyes initially move medially; later, further facial growth moves them apart. Anophthalmia has its origins in this critical fourth to seventh week of development. An interesting comparison is noted in drawings of an embryo at 37 days to a photograph of an anophthalmic child with additional central facial clefting (Figures 2 and 3). The striking similarities emphasize that a developmental arrest is the basis of this anomaly.

Congenital anophthalmia and microphthalmia present the team with different challenges. The term microphthalmos includes a variety of conditions that have similar appearances. This discussion simplifies and applies microphthalmos to all cases where clinical examination finds a nonfunctional small globe or deranged ocular tissues. The clinical implications of microphthalmia observed by the authors is that the ocular adnexa (eyelids and conjunctiva) and bony orbit are of near-age appropriate size and rarely complicate the therapeutic challenge of conventional prosthetic design, supply, and therapy. Congenital anophthalmos is defined as the complete failure of outgrowth of the primary optic vesicle.

KEY WORDS:

congenital anophthalmos,
microphthalmos, clinical
anophthalmos, micro-orbitism,
expansion therapy, phimosis,
EUA (examination under
anesthesia), orbital implant



FIGURE 1. Infant with unilateral congenital anophthalmos

The differentiation between congenital anophthalmos and extreme cases of microphthalmos can be made only by histological examinations. In microphthalmia, the socket may be manageable for life with prosthetics alone with excellent function, appearance, and symmetry achieved. Reconstructive surgery is discouraged until growth is complete. Congenital anophthalmia is applied to all cases where no ocular remnants are visible. As a rule, these cases are accompanied by severely reduced conjunctival surface area, small or phimotic eyelids, and a reduced bony orbit greatly diminished when compared to age-appropriate ocular development (Figure 4). Regarding bony orbital development, the studies of Robert Kennedy, M.D., elegantly demonstrated the effects of eye loss and volume replacement or lack of volume replacement on orbital maturation. However, Dr. Kennedy's

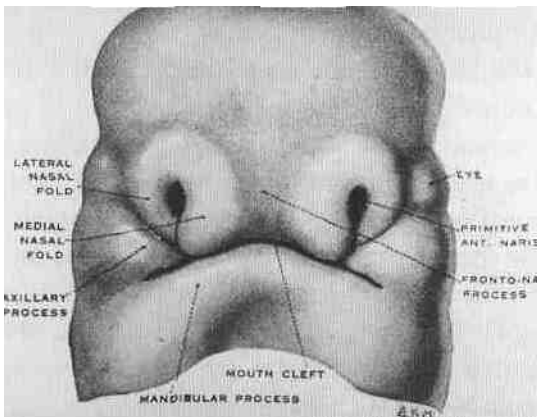


FIGURE 2. Facial development at 37 days gestation

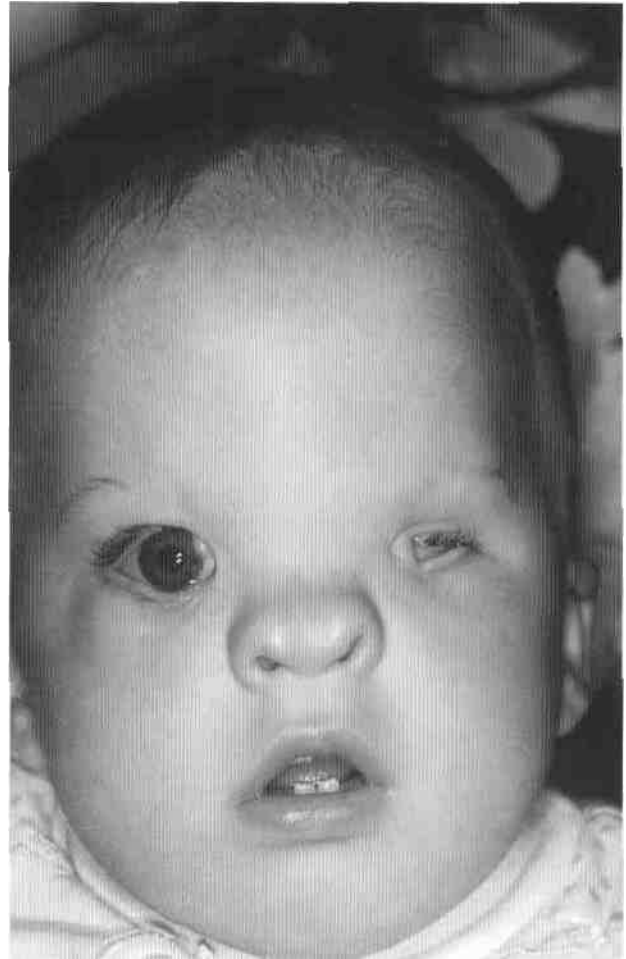


FIGURE 3. Congenital anophthalmic child with additional central facial clefting

studies removed the eyes of animals at birth; orbital development to this point was age-appropriate. Congenital anophthalmic is much more severe with arrest occurring early in the fourth to seventh week of gestation and unaided by volume replacement throughout the rest of development.

Cases of mild to moderate microphthalmia are more easily managed by the oculist (Figure 5). In such cases, the orbit, eyelids, and conjunctiva are well formed and do not present a problem in prosthetic fitting. Rapid soft tissue response is usually attained within weeks of onset of conventional prosthetic therapy. Such cases yield excellent results. The real challenge to prosthetic design and supply is presented by the severely affected cases of extreme microphthalmos and clinical anophthalmos.

In cases of congenital anophthalmos, bony orbit

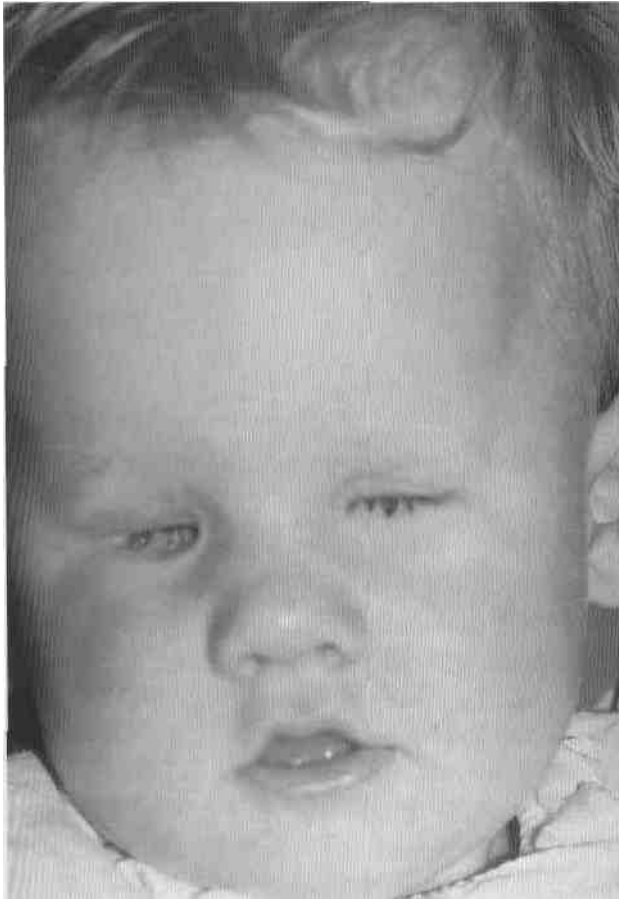


FIGURE 4. Child with bilateral congenital anophthalmos

growth can be considerably retarded. The term "micro-orbitism" (Figure 6) is used to describe this condition. In most cases, the orbital roof is thicker and flatter compared to the normal design. Overall orbital volume and conjunctival surface area are reduced relative to the degree of the defect. The resultant socket is soft and malleable without irregularities unless the lid(s) has coloboma. The congenital anophthalmic socket usually is "acorn-like" in shape (Figure 7). A depression or pit is at the apex of the socket. This pit area is a crucial feature in the fitting of the graduated expansion therapy prosthesis process. The eyelids are significantly smaller both horizontally and vertically compared to the normal size. Also, the distance between the lid lashes and the brow is proportionally reduced. The palpebral fissure is usually closed and contracted.

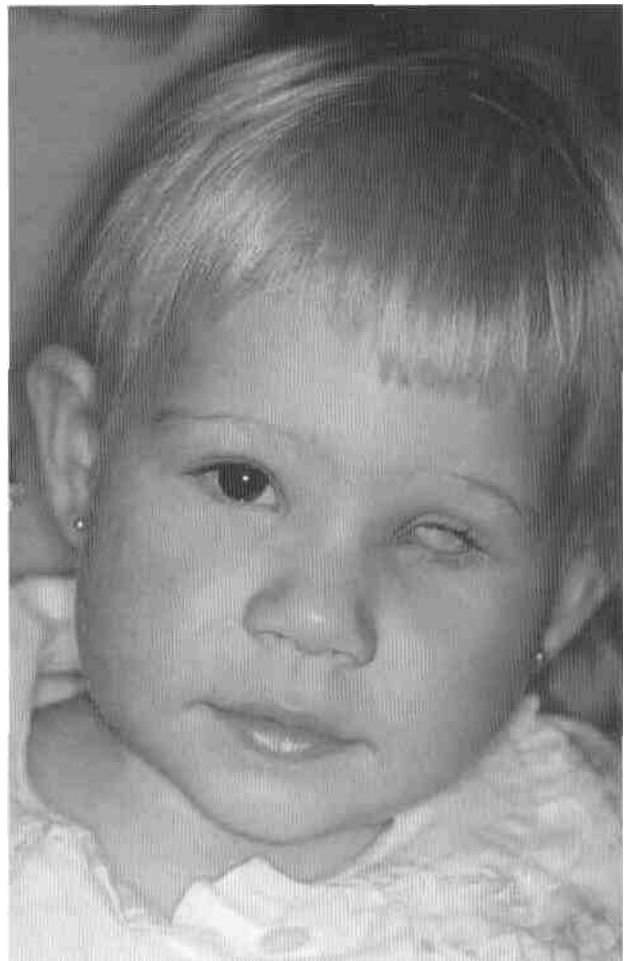


FIGURE 5. Child with unilateral microphthalmos

The diminished palpebral fissure makes the examination of the cavity very difficult (Figure 8). The management of clinical anophthalmia and extreme microphthalmia requires the close working relationship of the ophthalmologist, ocularist, patient, and family. The three traditional basic concepts of ocular prosthetic management have been: 1) mechanical or tension wire expanders, 2) pressure conformers, and 5) progressive sized conformers.

The authors believe that, in cases of overly aggressive pressure or tension expansion, the quality of dilation achieved is suboptimal and too often induces harmful scarring. Mechanical expanders and most pressure conformers expand the socket by forcing it to conform to the shape of the prosthetic appliance. While dilation is usually rapid, this technique can cause socket deformity by forcing it

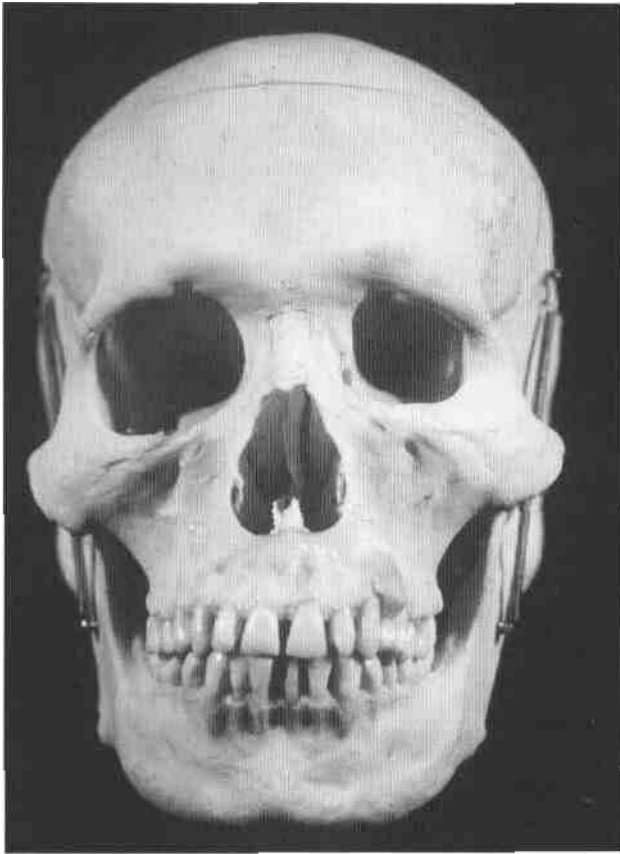


FIGURE 6. Adult skull with congenital anophthalmia



FIGURE 8. Phimoric palpebral fissure as in unilateral congenital anophthalmos

to conform to the pressure template (i.e., deformity by conformity) (Figure 9). Natural development is supplanted by forced deformation of the expander appliance. This problem was noted by Dr. Alston Callahan in his book, *Reconstructive Surgery of the Eyelids and Ocular Adnexa*. Aggressive expanders create an afunctional socket with canthal disruption and conjunctival scarring. The conjunctiva may lose elasticity and potential for future growth because of a "stretch mark" effect (Figure 10). The authors recommend the technique of anophthalmic prosthetic development based on the concept that the key to development of the anophthalmic socket is function. A similar problem is faced by infants born with dwarfed limbs. In years past, their treatments

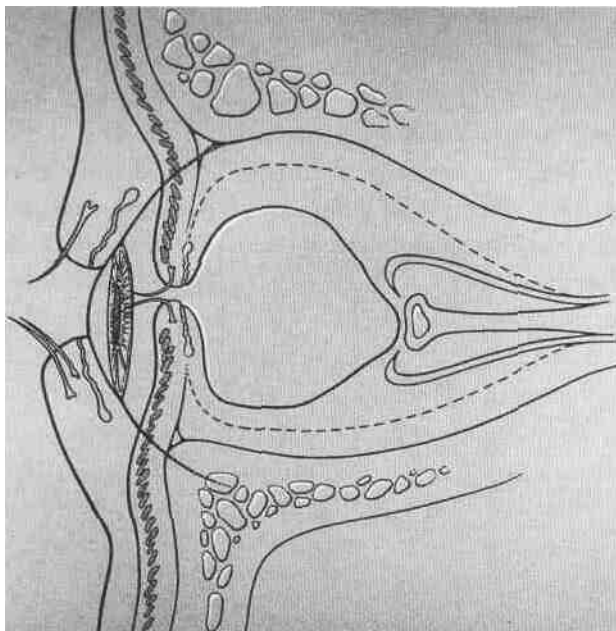


FIGURE 7. Illustration of acorn-shaped congenital anophthalmic socket



FIGURE 9. Child wearing tension wire expander

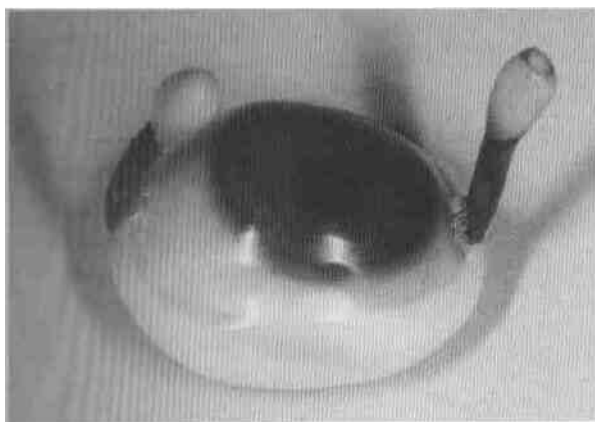


FIGURE 10. Lateral canthal erosion resultant from overly aggressive expansion

included mechanical limb stretchers and pulling weights or early radical surgery. The limbs sometimes improved in length, but the treatment negated functional development. Modern day physical therapists use a wide number of devices designed to complement function and stimulate muscle tone in the limb. Muscle function will, in turn, stimulate continued growth and development. We have applied this concept to the recommended eye expansion therapies.

In all cases of clinical anophthalmia, the expansion therapy should ideally begin as early as possible, ideally the first weeks of life. The authors have begun therapy on patients in the first week of life. The period of rapid growth in early infancy when the bony and soft tissues are mobile and adaptable lends itself best to prosthetic management. Unfortunately, not all children are seen this early, but the authors still advocate these techniques whenever first seen. The mainstay of this technique combines progressively sized appliances with the modified impression technique. Exams under anesthesia by the team are critical.

Ideally, the ophthalmologist and oculist can evaluate the first-time patient together as soon as possible. A management plan is developed and discussed with the family. Such a plan is very reassuring to the family and becomes an important mainstay in their support network dealing with their child's special situation and needs.

Although eyelid phimosis is usually found in cases of clinical anophthalmos, the authors *do not* recommend early surgery on the lids, conjunctiva, or bony orbits. The authors have also not been impressed with early implanted tissue expanders, and especially do not recommend lateral canthotomy, a common first procedure performed on many of our referred patients. The lateral canthus is an important point of capture for the prosthesis throughout growth and development. Weakening this capture point creates difficulty in retaining and stabilizing the prosthesis. The authors have observed patients where aggressive surgery on the conjunctiva, eyelids, or bony orbit has caused malignant contraction of tissue, as evidenced by a child who had 15 surgeries by age 2 years. The socket was totally contracted with eyelids fused by symblepharons (Figure 11) and now is both inoperative and unable to maintain a prosthesis.

In a typical case, the affected socket is examined and the palpebral fissure measured. In most cases, the initial fitting is done with a small stemmed



FIGURE 11. Fused eyelids and total symblepharon secondary to multiple surgeries

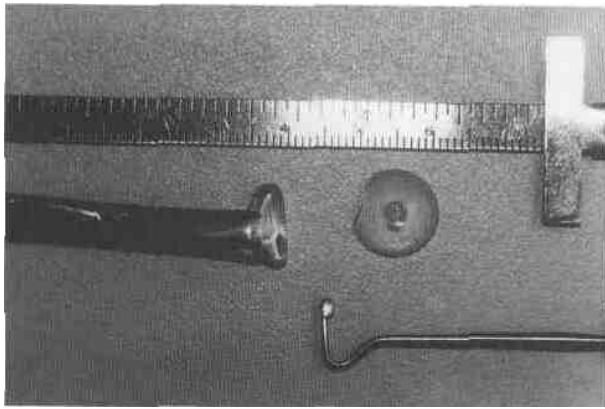


FIGURE 12a. Stemmed conformer made from a PMMA corneal button



FIGURE 12b. Child wearing bilateral stemmed conformer

conformer. The stem allows for easy insertion and removal of the appliance (Figures 12 a and b). The stemmed conformer is easily made from a prosthetic cornea button. Depending on the palpebral and socket size, the stemmed conformer is the largest that can be comfortably inserted through the eyelids. The size of this initial fit allows the ocularist to design a sequence of progressively larger con-formers for future use (Figure 1.3). A key point that cannot be overemphasized is that stretching of the socket is not the goal of this stemmed conformer. A conformer large enough to be maintained by the lids while allowing for a response is adequate (fit to function). Conformers should be changed and incrementally enlarged every 2 weeks to follow tissue expansion. As dilation occurs, the ocularist will be able to examine the socket visually and identify

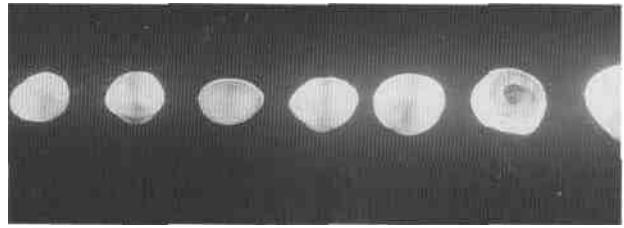


FIGURE 13. Sequence of progressively larger conformers

anatomical features. The position of the pit area is noted and incorporated in the conformer design (Figure 14). As soon as possible, the stem should be removed from the anterior surface of the con-former. To assist insertion and removal, two or three small holes are drilled in the anterior surface of the conformer. The holes should be large enough to allow the conformer to be grasped by a small hemosrat (Figure 15).

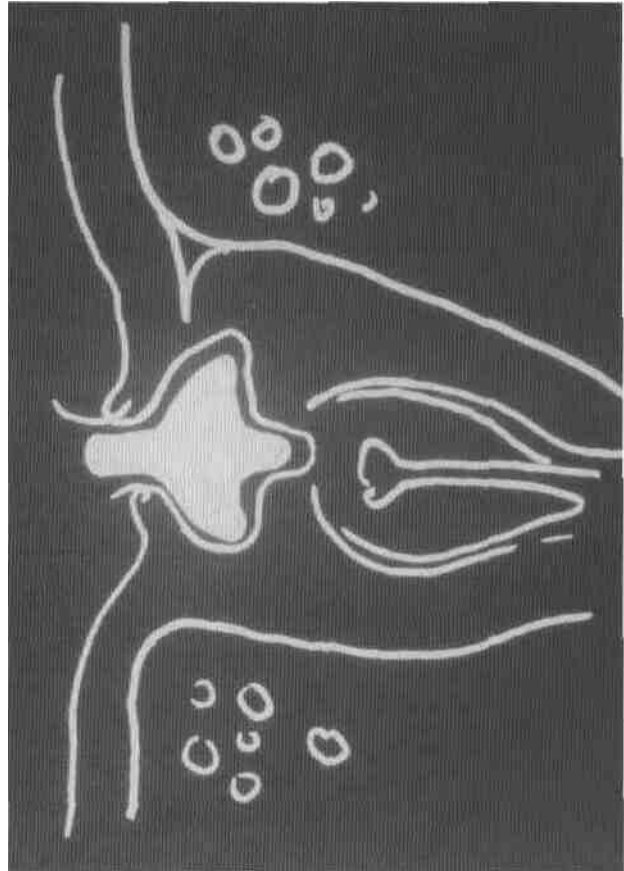


FIGURE 14. Illustration of stemmed conformer fit into pit area

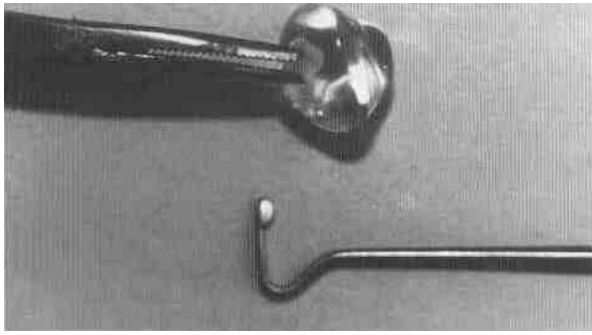


FIGURE 15. Conformer with drilled anterior surface holes for hemostat capture

The removal of the conformer stem will allow the socket and conjunctiva and eyelids greater range of function. Again, the authors emphasize that care must be taken not to overfit the socket. Progressive enlargement of conformers is soon followed by an exam under anesthesia (EUA) where an alginate impression of the socket can be made (Figure 16). Small custom-made impression trays must be used and several impressions with different viscosities of alginate taken. Information from the impression models allows the ocularist to custom-design the next stage of conformers. The pit area is identified and the custom conformers designed to fit into this area.



FIGURE 16. Alginate impression of a congenital anophthalmic socket made during EUA with custom small tray

Attention to fitting the pit area also provides an anchor to discourage the prosthesis from rotating in the socket (Figure 17). Progressive enlargement should not stress the socket conjunctiva. The anterior curve should allow the eyelids as much closure as possible. Do not expect a large palpebral fissure in the early stages of fitting. Overly rapid expansion may open the eyelids but inhibit the range of function that will retard overall development. It cannot be overemphasized that aggressive management can also cause conjunctival scarring and decrease elasticity. The authors have observed that forcefully expanded sockets rapidly contract when the appliance is removed for a day or more.

No set time schedule can be made for the sequential fitting of a congenital anophthalmic patient. Based on past experience, the authors expect the initial stage of empirical fitting to last for about 3 months. The second stage of fitting following the EUA should last an additional 6 months to 9 months. Usually during this time cosmetically designed prostheses are made, offering the parents a morale booster (Figure 18). The third stage of development occurs when the dynamic socket

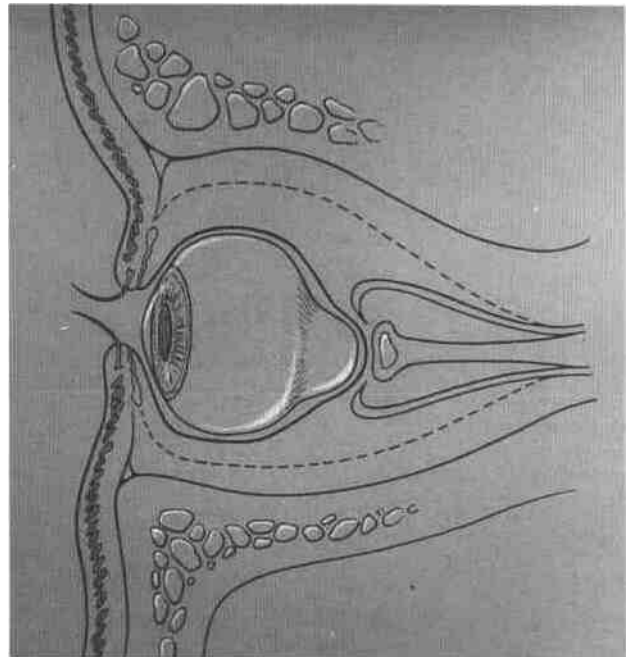


FIGURE 17. Illustration of proper prosthetic fit into pit area



FIGURE 18. E.C., 15-month-old congenital anophthalmic child with cosmetic conformer

changes end. The earlier stages represent rapid change as the socket tries to catch up on the lost development.

Empirical modifications based on the impression information and observed socket changes allow the prosthesis to keep pace with soft tissue growth and development. Close communication between oculist and ophthalmologist is extremely important at this time. This stage will see slower development consistent with the overall growth of the child. Another EUA is recommended as socket changes can no longer be accurately fit by the empirical method. Now that socket changes are less frequent, visits can usually be reduced to every 6 weeks. Adjustments are continually made to the prosthesis to ensure that the size keeps pace with the child's

overall development. The third stage usually lasts until the child is 3 years to 5 years old (Figure 19).

In many cases, socket dilation will be so great that the prosthesis becomes too large to fit through the eyelids. Two-piece appliances can be helpful with this problem (Figure 20). When the prosthesis is too large to access the orbit, an orbit implant is indicated. This step indicates the fourth stage of expansion therapy.

The authors have surgically placed orbital implants in 20 congenital anophthalmic sockets. Surgery is most often performed between 4 years and 5 years of age. Implantation in these anophthalmic sockets necessitates patience, gentle handling of tissues, critical intraoperative volume assessment, meticulous hemostasis, and wound closure. Ano-

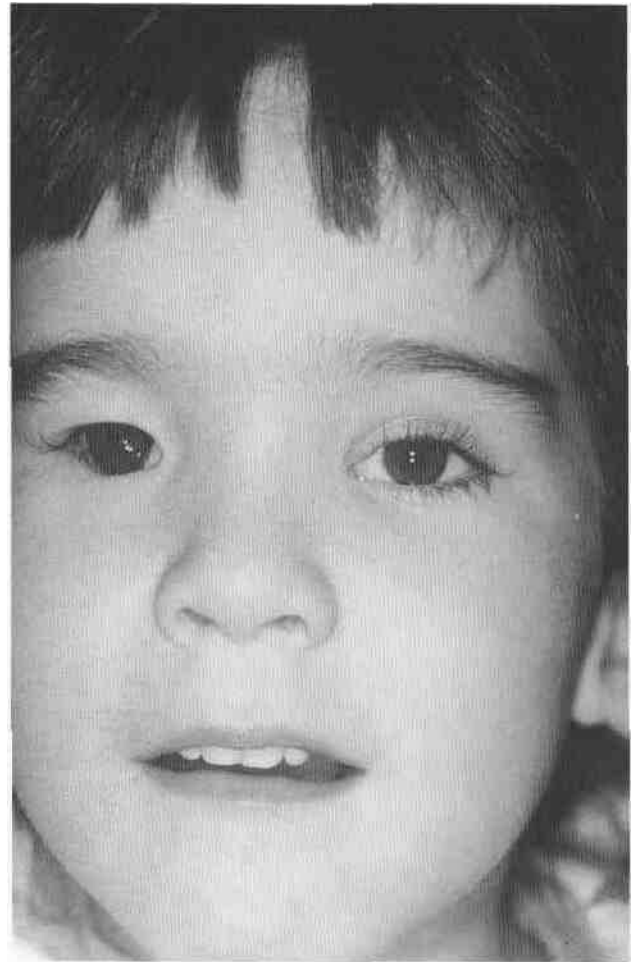


FIGURE 19. E.C., 4 1/2 years old with stage 3 prosthesis

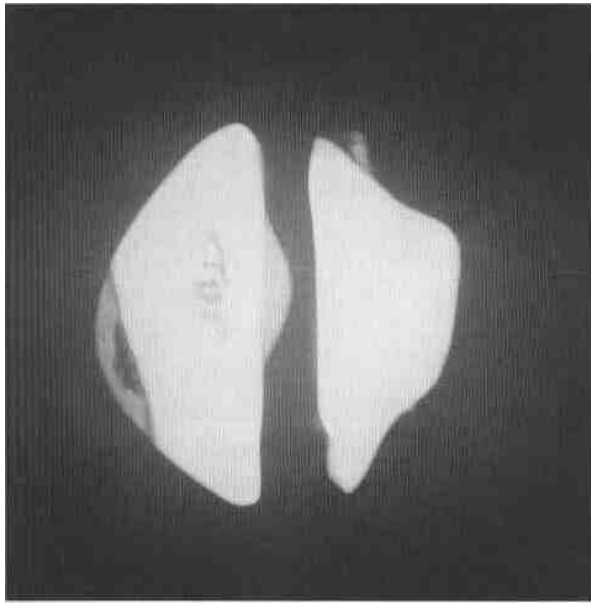


FIGURE 20. Two-piece prosthesis

ophthalmic children tend to be small in stature. Two of these patients have been growth-hormone deficient. The child's pediatrician should be consulted about this possibility. If a 4-year or 5-year-old has reached the therapeutic end point with the first three stages of prosthetic development and is ready for implantation but still quite small, surgery may be delayed for 6 months or longer until general growth increases.

Implantation allows the prosthetic design to assume a more conventional shape. Prosthetic fit and orbital access through the phimotic lids is facilitated. Lash touch of the prosthesis before implantation during prosthetic stages 1, 2, and 3 can occur and is a consequence of conjunctival shortage (Figures 21 a and b). Implantation can relieve or improve the lash position as the conjunctiva and fornices move forward with implantation (Figure 22 a and b). The authors advise against surgical lash rotation before implantation for this reason. Surgery before 4 years of age can be technically difficult. The authors recommend placement of a scleral-wrapped polymethylmethacrylate sphere behind the anterior tenon's capsule and conjunctiva in a conventional technique used with standard enucleation or secondary implantation. If ocular remnants or a

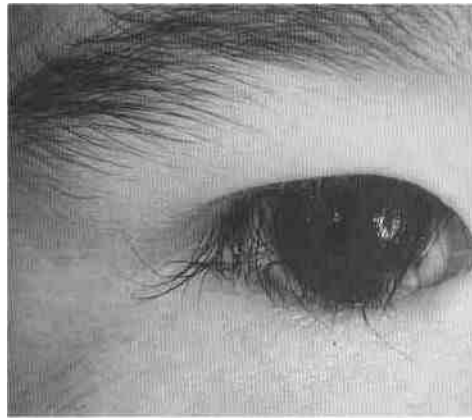


FIGURE 21a. E.G., 5 1/2 years old with lash touch of prosthesis

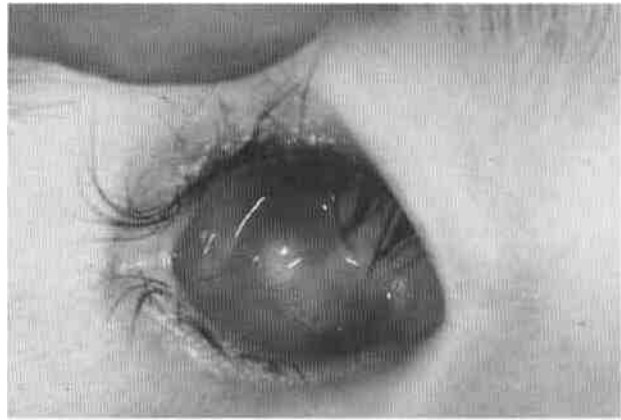


FIGURE 21b. E.C., 5 1/2 years old with prosthesis removed

cyst are present, they should be enucleated. If the ocular muscles are discernible, they can be attached to the implant in a marionette fashion. Dermis-fat grafts, hydroxyapatite, MEDPOR®, or quasi-integrated implants are not the best choices for multiple reasons. When the authors implanted tissue expanders in two anophthalmic orbits, the result was so unimpressive that we have abandoned this technique.

The orbit, eyelids, and conjunctiva may respond with future growth, development, and prosthetic management so that an implant exchange to a larger diameter can be indicated years later. The difficulty of "integrated" implant removal would be a disadvantage. Also, the tight adherence of conjunctiva and tenon's capsule to the porous implants may

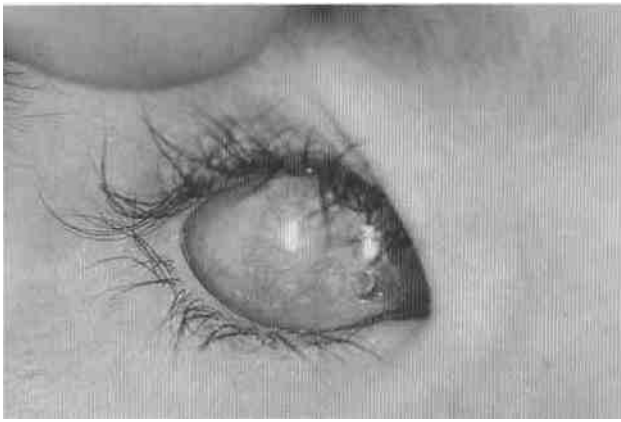


FIGURE 22a. B.C., 6¹/₂ years old, 1 month postoperative placement of 16-mm scleral-wrapped sphere



FIGURE 23a. 14-mm scleral-wrapped sphere sized to orbit for implantation in 5-year-old child

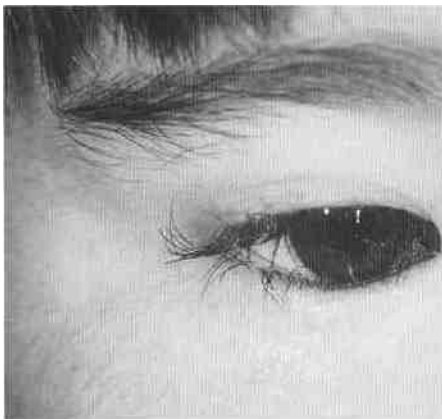


FIGURE 22b. E.G., 1 month wearing prosthesis postoperatively; note improved lash position

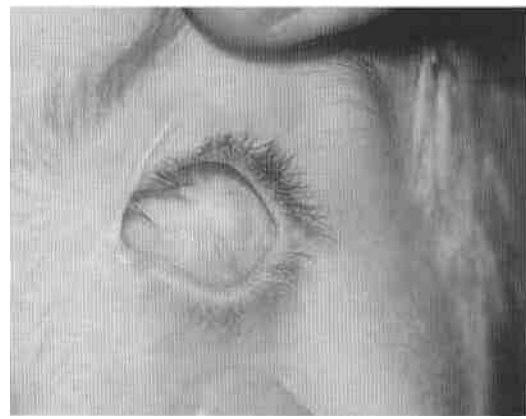


FIGURE 23b. 1 year postoperative implant placement

decrease conjunctival elasticity and growth in these immature orbits. Mucous membrane grafting is also counterproductive. These grafts and their interface with surrounding conjunctiva have poor elasticity and like dermis-fat grafts do not respond well to future prosthetic enlargement.

The diameter of the implant is critical. The authors size the implant at surgery so that it proportionately best augments the orbital volume deficiency (Figure 23). Oversizing the implant to the point of proptosis is not advantageous and will make future prosthetic fitting difficult, if not impossible, compromising cul-de-sac and lid function. Our goal is to place an implant of the largest diameter to allow space (3 mm) postoperatively for prosthetic fitting and lid function. A 16-mm sphere is most

commonly used. A total scleral-wrapping or anterior scleral-capping of the implant can refine volume augmentation and place an additional tissue barrier between prosthesis and implant.

A series of custom conformers should be prepared and critiqued before surgery by the oculist and ophthalmologist (Figure 24). Standard operating room conformers in the hospital stock will not fit these orbits postoperatively. Custom conformers are made in at least three sizes with mild variations in vertical diameter. These conformers are based in diameter and curvature on the prosthesis worn before surgery, while the curvature should reflect the spherical implant to be placed. The conformer thickness should be minimal. At surgery, the conformer that best fits the socket and least stresses

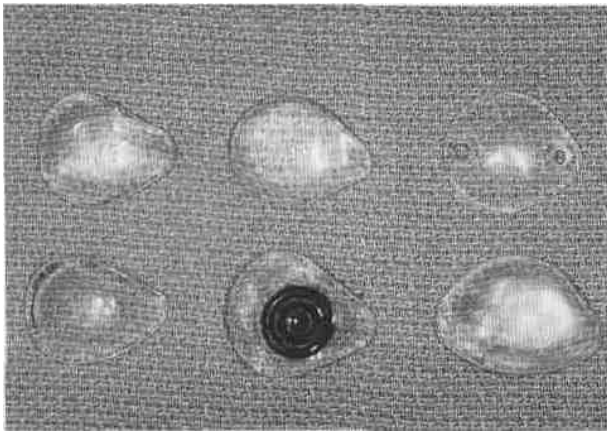


FIGURE 24. Custom conformer prepared for patient before implantation

wound closure should be used (Figure 25 a and b). The authors' first case of orbital implantation 12 years ago has recently had an implant exchange and enlargement from 14 mm to 18 mm. The second

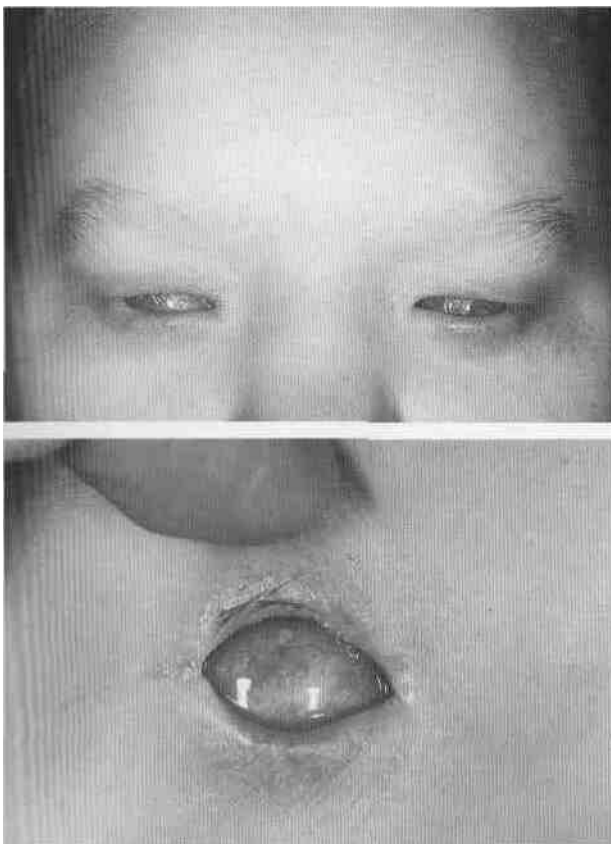


FIGURE 25a, b. Patient with bilateral congenital anophthalmia wearing custom conformers 2 weeks after placement of 16-mm scleral-wrapped implants

case of implant placement 11 years ago has continued to show tissue compliance and good health and growth. The 16-mm implant originally placed continues to provide adequate volume.

Recently, the authors have begun to apply other eyelid surgical reconstructive techniques to individual eyelid variables of congenital anophthalmia. Because of the delayed development, a judicious and conservative approach to such ancillary lid procedures is recommended. The timing of these procedures is usually several years following orbital implantation. Many of these children have the blepharophimosis syndrome and may benefit from staged Mustarde medial canthoplasty and levator resection or frontalis fascia suspension. Eyelid coloboma, which can occasionally occur with congenital anophthalmia, may require early closure before

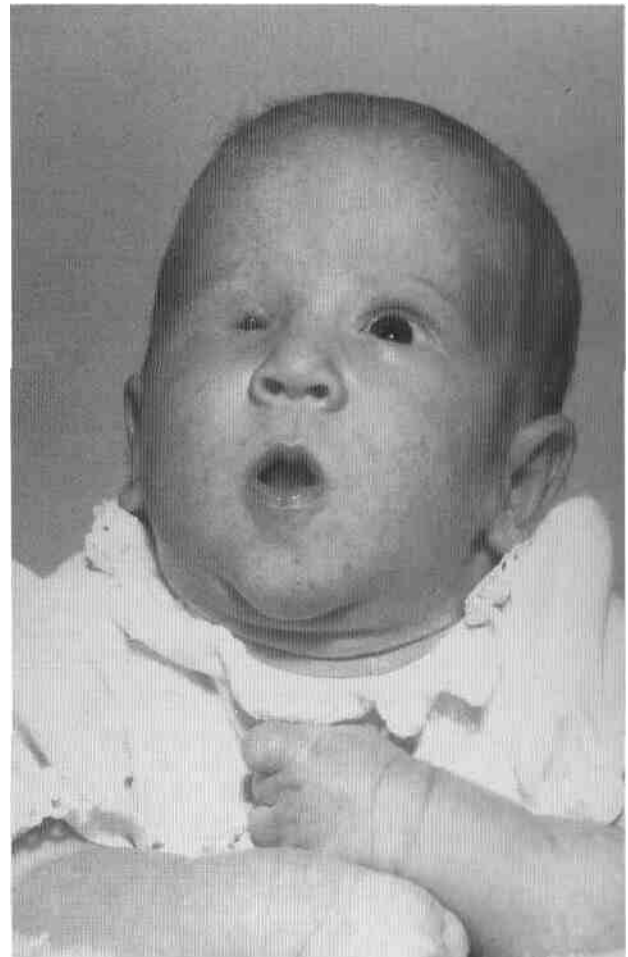


FIGURE 26. J.M., 3-month-old congenital anophthalmic patient with stemmed conformer



FIGURE 27a, b. J.M., 7-year-old patient at stage 3 prosthetic treatment

stage 1 or 2 of prosthetic therapy to facilitate prosthetic retention. Several of our patients with ptosis have had improvement in their lid elevation as they responded to orbital implantation and subsequent changes (Figures 26 through 31). Ptosis surgery is

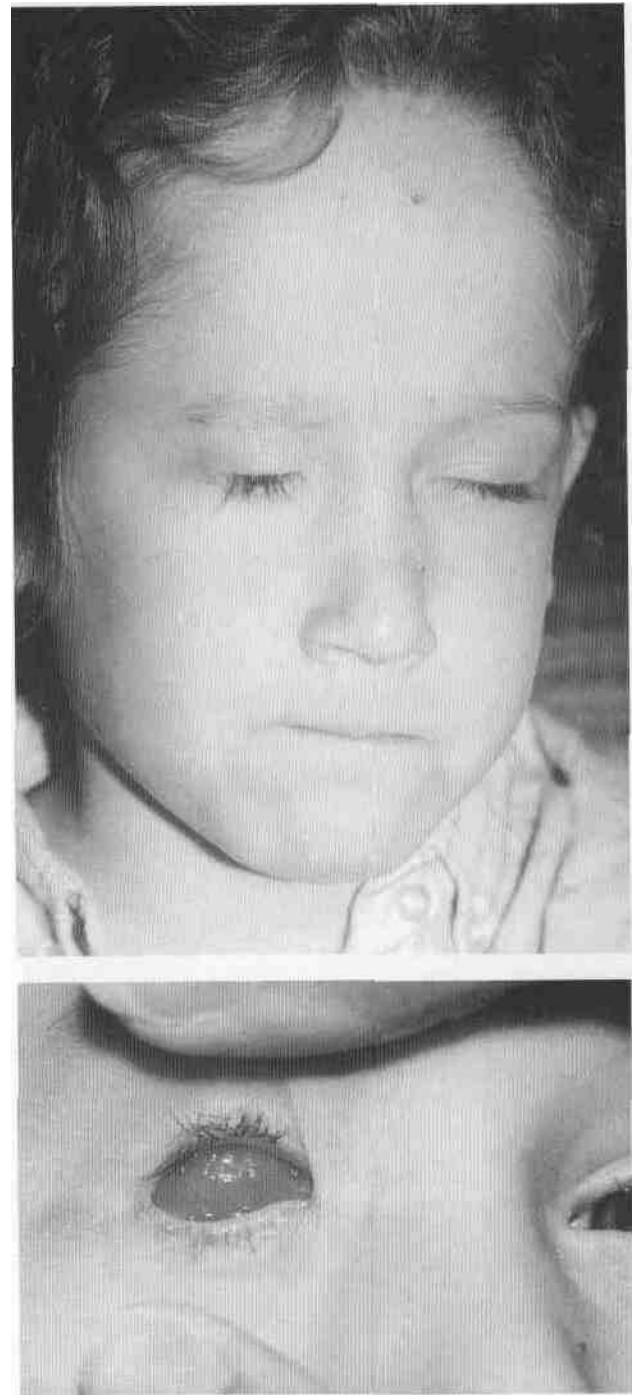


FIGURE 28a, b. J.M., at 10 days postoperative following 16-mm implant placement with lid closure

ill-advised until long-term stabilization is observed. Levator resection can also compromise the superior fornix and prosthetic retention.

The authors' oldest case managed from birth with unilateral anophthalmia is 18 years old and

has had only one surgery to date. This child was presented to us at 7 days old with clinical anophthalmia O.D. Prosthetic management began immediately (Figure 26). At 7 years of age, she had completed stage 3 (Figure 27 a and b). A 16-mm scleral-wrapped implant was surgically placed in the socket. Photographs taken postoperatively at 10 days (Figures 28 a and b), 1 year (Figure 29), 5 years (Figure 30), and 10 years (Figure 31 a and b) show that the conjunctiva orbit and eyelids are continuing to respond to gradual prosthetic expansion. Early eyelid ptosis resolved significantly without surgery. The authors stress that function is the key to development.

The management of congenital anomalies is a long-term commitment for the team. Severe microphthalmia and anophthalmia require countless

hours and teamwork of the patient, family, ocularist, and ophthalmologist. Each case will present subtle variations and challenges, but the techniques described above have been found valid and successful for our management goals. The authors emphasize that normal lid size and orbital volume are unattainable in many of these patients. Some patients with aggressive bony expansion have compromised prosthetic retention. The goal is to develop the best symmetry, eyelid function, prosthetic appearance, and orbital volume augmentation for the congenital anophthalmic patient. For the ocularist / ophthalmologist team, there is no greater satisfaction than that attained by the successful management of this complex anomaly.

Acknowledgement: The authors would like to thank our patients and their families whose stead-



FIGURE 29. J.M., at 1 year postoperative following implant placement; patient is 8 years old

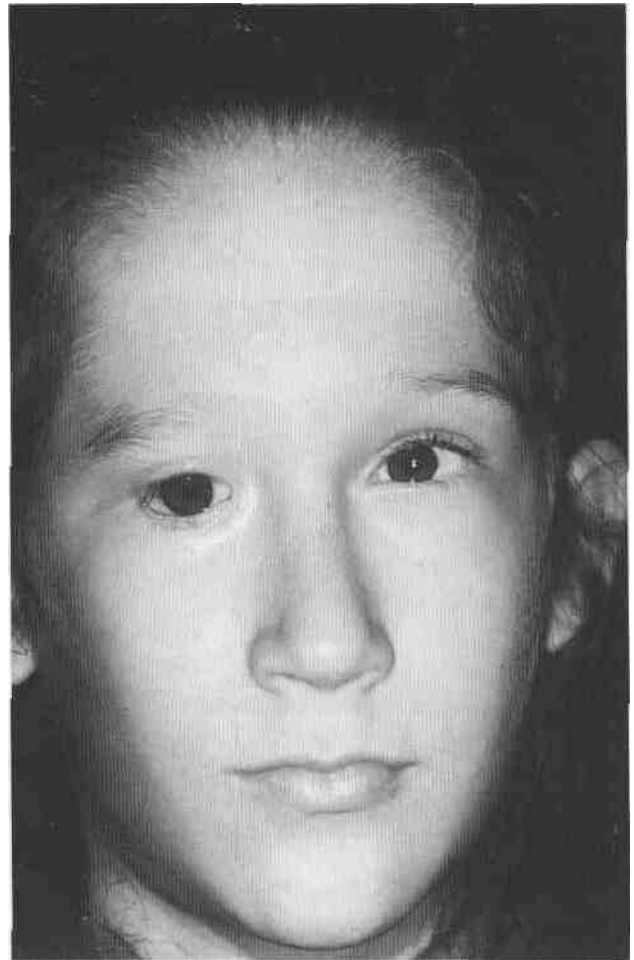


FIGURE 30. J.M., at 5 years postoperative following implant placement; patient is 12 years old

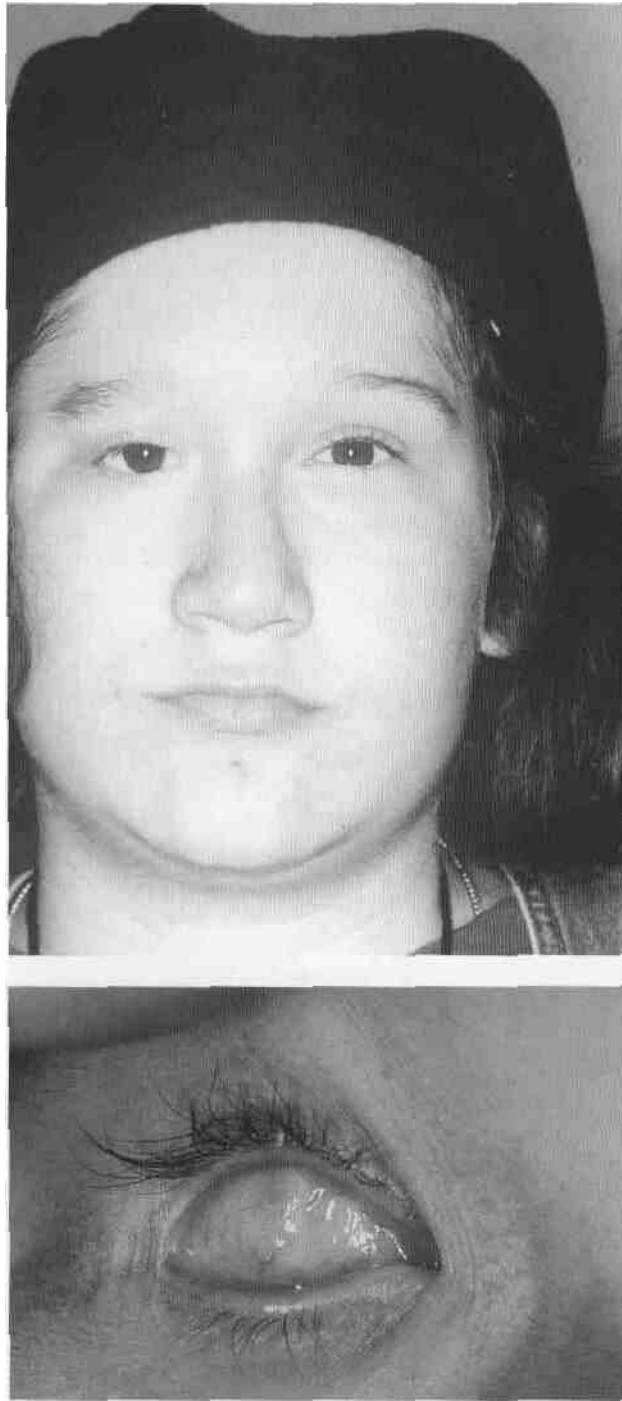


FIGURE 31. J.M., at 10 years postoperative following implant placement; patient is 17 years old

fast commitment made this work and presentation possible.

Special recognition is due our friend and mentor, John H. O'Donnell B.C.O., F.A.S.O., whose original ideas and concepts are the foundation of this work.

References

1. Callahan, A. *Reconstructive Surgery of the Eyelids and Ocular Adnexa*, Birmingham, 1977
2. Duke-Elder, S. *System of Ophthalmology; Congenital Deformities; Vol III, Part II*, London, 1964
3. Gougelmann, HP. *Problems and Treatment of Contracted Sockets*, New York, 1973
4. Pearce, WG, Nigam, S., Rootman, J. Primary Anophthalmos; Histologic and Genetic Features, *Can. Jour. of Oph.*, Vol. 9, 1974
5. Warfel, JH. A Case of Unilateral Degenerative Anophthalmia, *Am. Jour. of Oph.*, No. 151, 1968
6. Hartman, DC. Anophthalmos and microphthalmos. In *Symposium on Plastic and Reconstructive Surgery of the Eye and Adnexa*, edited by J.W. Smith and J.M. Converse, pp 514-517. C.V. Mosby Co., St. Louis, 1967
7. Kennedy, RE. The Effect of Early Enucleation on the Orbit in Animals and Humans, *Am. Jour. of Oph.*, No. 60: 277-306, 1965
8. Kennedy, RE. Growth retardation and volume determinations of the anophthalmic orbit, *Am. Jour. of Oph*
9. Mustarde, JC. Congenital absence of the eye.; *Plastic Surgery in Infancy and Childhood*. E. & S. Livingstone, Edinburgh, 1971
10. Soil, DB. Evolution and Current Concepts in the Surgical Treatment of the Anophthalmic Orbit. *Ophthalmic Plastic and Reconstructive Surgery* 1986; 2(3): 16.3-171

CORRESPONDENCE TO:

James H. Merritt, M.D.
8230 Walnut Hill Lane, Suite 518
Dallas, Texas 75230