

Orbital expansion of the congenitally anophthalmic socket

Susan M Tucker, Nigel Sapp, Richard Collin

Abstract

Background—Congenital anophthalmos is a rare condition in which intervention at an early age can stimulate orbital expansion and maximise facial symmetry. Much is still unknown, however, regarding the degree of soft tissue and bony orbital growth achieved using the orbital expanders presently available.

Methods—A retrospective review of 59 congenitally anophthalmic orbits in 42 patients was carried out.

Results—The soft tissue and bony orbital expansion achieved using serial solid shapes is reported, and experience with hydrophilic expanders and inflatable silicone expanders is reviewed.

Conclusion—Although serially fitted solid shapes in the orbit lead to increased expansion of orbital soft tissue and bone compared with no orbital implant, further orbital tissue enlargement is required. The inflatable silicone expander may allow more rapid and extensive orbital tissue expansion, but design changes are needed to achieve this.

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Congenital anophthalmos is a rare condition, with a reported prevalence rate of 0.3/10 000.^{1 2} Social and psychological implications to the child and parents of both the visual loss and the altered physical appearance can be quite significant. It is therefore important to enhance the growth of the involved orbit to allow retention of a suitable prosthesis and to maximise facial symmetry. Although numerous clinical studies and animal model experiments have shown that deficiency of growth in the postnatally enucleated socket can be stimulated by placement of an orbital implant,³⁻¹⁰ the amount of expansion achieved by thus treating congenital anophthalmos is unknown. The goal of this study was to quantitate the degree of orbital expansion obtained with serially fitted moulded solid shapes in infants born with congenital anophthalmos, and to review our experience using hydrophilic expanders and inflatable silicone expanders in this group of patients.

Material and methods

A retrospective analysis was carried out on 42 patients (59 sockets) with congenital anophthalmos seen at Moorfields Eye Hospital in London between January 1980 and October 1993. For each of these patients, data were collected on the type of expander used, the

age at the initial insertion of the orbital expander, hospitalisation time, and surgical intervention. Three different types of expanders were used in our series of patients. Solid shapes (socket retainers, conformers, and moulded acrylic shapes) were fitted and then increased in size when they started to rotate within the orbit or when the eyelids were relatively loose around the solid shape. Hydrophilic expanders were used for very shrunken sockets which could not initially hold a solid shape, expanding in size with tear contact (and if necessary additional saline drops), and then replaced on average 72 hours later with a solid shape. The silicone expanders, periodically inflated with normal saline as required, were used in several infants in the hope that a more rapid expansion would be achieved.

For infants treated with serial solid shapes, changes in the horizontal eyelid length (HEL=the distance from the medial to lateral canthus in millimetres) over the follow up period were determined. Measurements of the vertical palpebral aperture were excluded as this measurement was considered too dependent on the size of the prosthesis. Bony orbital expansion was determined by length and volume measurements of serially fitted orbital prostheses in the anophthalmic sockets. All available prosthetic shapes used in each patient were examined to determine their horizontal width, vertical height, and anterior to posterior depth in millimetres. The greatest value for each of these dimensions was used. The volume of each prosthetic acrylic shape was then determined by measuring the volume of water displaced by the shape after submerging it in a cylinder of water calibrated to 0.1 ml.

Results were statistically analysed using split plot analysis of variance and Pearson correlation.

Results

Twenty five patients with congenital unilateral anophthalmos were reviewed: 14 male and 11 female. The right socket was anophthalmic in 17 patients, and the left socket in eight patients. An ocular remnant was present in 11/25 sockets (44%). In all patients, the involved eyelid was small. Seventeen patients (34 orbits) with congenital bilateral anophthalmos were reviewed: nine male and eight female. An ocular remnant was present in four of the 34 orbits (12%). For 31 of the 34 orbits, the involved eyelid was small with peripalpebral fibrosis.

Department of Surgery, Division of Ophthalmology, Southern Illinois University, Springfield, USA
S M Tucker

Moorfields Eye Hospital, London
N Sapp

Department of Oculoplastic and Reconstructive Surgery, Moorfields Eye Hospital, London
R Collin

Correspondence to: J R O Collin, Moorfields Eye Hospital, City Road, London EC1V 2PD.

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ORBITAL EXPANDERS

Solid shapes

Data were available for 45 orbits of 19 patients with unilateral anophthalmos and 13 patients with bilateral anophthalmos. The remaining cases were seen at Moorfields Eye Hospital for consultation, but followed up elsewhere. Serial solid shapes were used in all 45 orbits. Ocular prostheses in the 45 orbits were changed on average every 11.1 (SD 6.8) months. However, the frequency of change was greater in the earlier years, with a frequency of every 6.5 (2.4) months in patients 3 years of age or younger. Of a total of 132 mouldings/insertion of solid shapes, 64 (48%) required an anaesthetic (ketamine or general anaesthesia); 19/32 patients (59%) required hospitalisation for the insertion of the prosthesis, with an average hospital stay of 3.5 (2.6) days. Surgery was required in 11 of the 45 orbits (24%), in all cases to allow retention of a prosthesis. Bony socket reconstruction was needed in four of these orbits (all requiring additional surgery including mucous membrane socket reconstruction, lateral canthoplasty, and entropion surgery), with subsequent retention of a prosthesis.

Hydrophilic expanders

Hydrophilic expanders were used in 18 sockets of eight patients with bilateral anophthalmos and two patients with unilateral anophthalmos. Our results with hydrophilic expanders have already recently been reported in detail.¹¹ These hydrophilic expanders were subsequently replaced by a conventional conformer after an average 72 hours. In 14 orbits it was used following failed attempts to fit hard shapes, and in the remainder it was felt that the sockets could not be fitted by any other means. An ocular remnant was present in three of the 18 sockets with hydrophilic expanders; small eyelid with peripalpebral fibrosis was present in 16/18. In the two sockets with relatively normal eyelids, ocular remnants were not present. The age at insertion of the hydrophilic expander ranged from birth to 15 months, with a mean of 5.1 months. Insertion of the hydrophilic expander required a general anaesthetic in two of the 10 patients (20%). The average length of hospital stay was 3 days. All of these patients were then successfully fitted with solid shapes.

Inflatable silicone expanders

Inflatable silicone expanders were attempted in 12 orbits of eight patients. The age at insertion of the silicone expanders ranged from 8 months to 29 months, with an average of 19.5 months. In seven of the eight patients an anaesthetic (either ketamine or general anaesthesia) was needed for both the insertion and subsequent inflation of the silicone expander(s). The average length of stay in hospital for insertion or inflation of the silicone expander was 1.75 days. In seven orbits of five patients, the silicone expanders fell out within

5 weeks, and 50% of the time within the first day. All these patients were subsequently successfully fitted with solid shapes. Inflatable silicone expanders were retained in five orbits of three patients (two with bilateral anophthalmos and one with unilateral anophthalmos). In one of these patients with bilateral anophthalmos, six solid acrylic shapes were used before the silicone expanders; both silicone expanders were inflated at 2 months but then collapsed at 9 months, after which solid shapes were again fitted. In the second bilaterally anophthalmic patient, bilateral small silicone expanders followed insertion of first B, then C conformers. They were each inflated at 6 and 8 months, then exchanged for solid shapes, which fell out. A small 2 ml silicone expander was then inserted on the right and a 4 ml silicone expander on the left and both were inflated 3 weeks later. The right expander fell out within a few days. Socket reconstructive surgery was then performed, allowing subsequent retention of the solid shapes. In the patient with unilateral anophthalmos, an initial attempt at insertion of a silicone expander failed at 6.5 months and a D conformer was inserted. A re-attempt with the silicone expander at 13 months was successful, and it has been in place now for several months. This was the only patient in which the silicone expander was attempted as the first orbital shape.

SOFT TISSUE AND BONY ORBITAL EXPANSION

Solid shapes

For patients with bilateral anophthalmos, there was no statistical difference in soft tissue or bony growth between the right and left orbits, therefore only data from the right side were used for analysis. Useful data on changes in

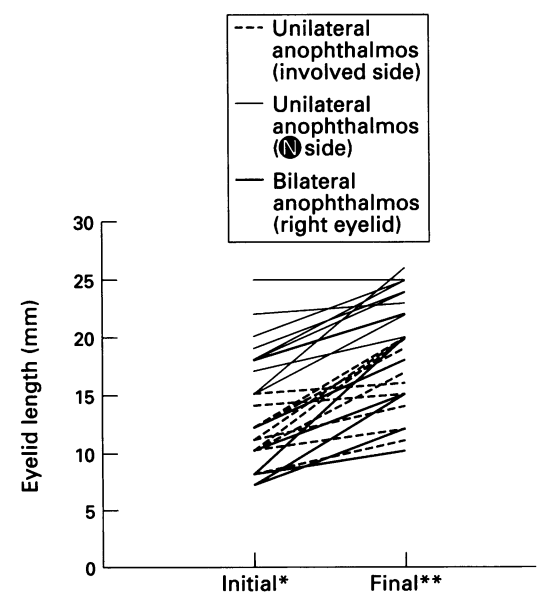


Figure 1 *Eyelid length at initial insertion of prosthesis. Mean eyelid length for the involved side of unilaterally anophthalmic sockets = 14.0 mm (normal side 21.0 mm); mean length for the right eyelid of bilaterally anophthalmic sockets = 8.8 mm. **Eyelid length at end of the follow up time (see Table 1). Mean eyelid lengths were 16.4 mm, 23.6 mm, and 13.4 mm respectively.

Table 1 Bony orbital enlargement with serial solid shapes

Patient	Age at first orbital expander (months)	Age at end of follow up (months)	Follow up time (months)	Ocular remnant	Prostheses dimensions											
					Initial				Final				Change			
					W*	H†	D‡	Vol§	W	Ht	D	Vol	W	Ht	D	Vol
1	8	36	28	+	19.0	14.0	9.0	1.00	20.0	15.0	12.0	1.10	1.0	1.0	3.0	0.10
2	2.5	48	45.5	-	12.5	12.0	8.0	0.80	15.0	12.5	14.0	1.25	2.5	0.5	6.0	0.45
3	3	45	42	+	10.5	11.5	8.5	0.40	15.0	15.0	14.5	1.50	4.5	3.5	6.0	1.10
4	3	96	93	-	13.0	12.0	12.0	1.20	17.0	15.0	14.0	1.90	4.0	3.0	2.0	0.70
5	7	46	39	-	14.0	8.5	11.0	0.80	19.0	15.0	13.5	1.80	5.0	6.5	2.5	1.00
6 RE	0.25	62	62	-	13.0	9.0	6.0	0.40	14.0	12.0	8.5	0.60	1.0	3.0	2.5	0.20
LE	0.25	62	62	-	12.0	9.0	6.0	0.35	14.0	12.0	8.5	0.70	2.0	3.0	2.5	0.35
7 RE	14	55	41	-	14.0	9.0	10.0	1.00	15.0	14.0	13.0	1.20	1.0	5.0	3.0	0.20
LE	14	55	41	-	13.0	11.5	9.5	0.60	15.0	13.0	13.0	1.40	2.0	1.5	3.5	0.80
8 RE	4	44	40	-	12.5	11.0	10.5	0.65	13.0	13.0	13.0	0.80	0.5	2.0	2.5	0.15
9 RE	1.5	29	27.5	-	9.0	7.5	3.5	0.10	15.0	12.0	9.0	0.60	6.0	4.5	5.5	0.50
LE	1.5	29	27.5	-	10.0	7.0	4.0	0.15	12.0	13.0	7.0	0.50	2.0	6.0	3.0	0.35

Patient Nos 1-5 unilateral anophthalmos, Nos 6-9 bilateral anophthalmos. *W=width in mm; †Ht=height in mm; ‡D=depth in mm; §Vol=volume in cm³. RE=right eye; LE=left eye.

HEL were available for 10 unilaterally anophthalmic and seven bilaterally anophthalmic sockets. For unilateral cases the age at insertion of the first orbital shape varied from 9 weeks to 19 months, with a mean of 13.9 months. Follow up ranged from 3 months to 13 years, with a mean of 4.5 years. For bilateral cases, the mean age for the successful retention of the initial orbital shape ranged from birth to 18 months, with a mean of 5.5 months. Follow up ranged from 28 months to 12 years with a mean of 6.5 years.

Figure 1 summarises the soft tissue expansion achieved. The mean initial and final HEL measurements for the unilateral cases (involved and normal sides), and the right sockets of the bilateral cases are listed. Although the mean initial HEL value was slightly greater for the unilateral group (11.3 mm compared with 10.0 mm), the absolute increase in the HEL achieved was statistically the same in both groups when the difference in individual follow up periods was adjusted. In the unilateral cases, the magnitude

of growth of the involved and normal sides was statistically equal; the initial length difference between the sides remained unchanged. There was no correlation between HEL growth and growth of any of the bony dimensions: width, height, depth, or volume.

Table 1 and Figure 2 summarise the bony orbital enlargement achieved based on changes in prostheses dimensions in five patients with unilateral anophthalmos and four patients with bilateral anophthalmos during their follow up period. The magnitude of growth achieved for all variables of bony expansion was the same for unilateral and bilateral anophthalmic sockets. There was no trend for expansion of the prosthesis to occur more in one dimension than in another. In this small subset of patients there was no correlation between the age at which an initial orbital expander was inserted and the subsequent enlargement of the orbit in any dimension. There was no significant difference in orbital growth between the two sockets with ocular remnants and the 10 sockets without remnants.

Discussion

Expansion of small sockets is important to maximise the potential for facial symmetry, and the resulting positive social and psychological well being that this imparts to the patient and parents. Clinical and experimental studies have shown that a deficiency of growth occurs in the anophthalmic orbit.³⁻⁹ Several animal studies, most using the cat model, have shown that the placement of a solid sphere results in partial expansion of the bony socket, and that a fully inflated serially expanded silicone implant results in growth equal to the normal side.⁷⁻⁹ Unfortunately, results following postnatal enucleation with immediate placement of an orbital expander cannot be extrapolated to the infant born with congenital anophthalmos where significant soft tissue hypoplasia (and often peripalpebral fibrosis) and lack of bony orbital growth has begun in utero. Cat orbital anatomy is also different from humans with a discontinuous orbital rim superotemporally bridged by cartilage. Although case reports and small series have suggested some expansion of microphthalmic and anophthalmic sockets in humans with orbital implants, quantitative values are lacking.¹²⁻¹⁴

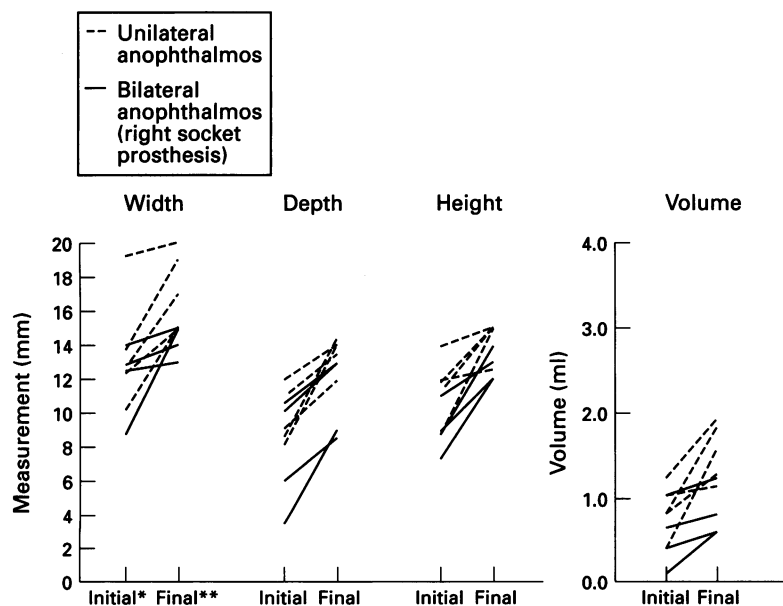


Figure 2 *Prosthetic dimensions at initial insertion of prosthesis. For unilaterally anophthalmic sockets the mean initial prosthesis width=13.8 mm, depth=9.7 mm, height=11.6 mm, and volume=0.84 cm³. For bilaterally anophthalmic sockets, the mean prosthesis width=12.0 mm, depth=7.5 mm, height=9.4 mm, volume=0.54 cm³. **Prosthetic dimensions at end of the follow up period. For unilaterally anophthalmic sockets, width=17.0 mm, depth=13.6 mm, height=14.5 mm, volume=1.5 cm³. The mean for bilaterally anophthalmic sockets: width=13.9 mm, depth=10.6 mm, height=12.8 mm, and volume=0.8 cm³.

Expansion of the horizontal eyelid length in patients with congenital anophthalmos is improved by the use of serially fitted solid shapes compared with no implant but the results are suboptimal. The magnitude of the soft tissue growth was the same for unilateral compared with bilateral anophthalmic sockets, when adjustment for differences in follow up periods were made. The mean eyelid length at insertion of the prosthesis of the affected orbit in unilateral cases was 14.0 mm (67% of the normal side which on average measured 21.0 mm); and at the end of follow up was 16.4 mm (69% of the normal side which measured 23.6 mm on average). The horizontal eyelid length of the normal uninvolved side in unilateral cases correlated well with morphometric studies performed in normal humans.¹⁵⁻²⁰ A control group of three patients with unilateral congenital anophthalmos who did not receive implants until ages 2 years, 2 years, and 37 years with resulting horizontal eyelid lengths of 10, 10, and 12 mm (on average 44% the length of their uninvolved sides) demonstrated the improved expansion achieved with orbital implants compared with that without orbital implants. A more rapid and greater expansion of the hypoplastic adnexal tissue in congenital anophthalmos is particularly important so that a small orbital entrance does not limit the size of the orbital expander which can be fitted.

The magnitude of bony orbital growth was also the same for unilaterally compared with bilaterally affected sockets when adjustment for follow up time was made. The most rapid rate of bony orbital growth in normal humans occurs between 6 months' gestation to 18 months after birth,^{16,21} with 79% of adult size achieved by 3 years of age, and full expansion by age 7.²² In this study all patients, except one who was age 29 months at end of follow up, were 3 years or older at termination of the study; a period over which the majority of bony expansion should have been achieved. The mean prosthesis width, height, depth, and volume at the end of follow up were 17.0 mm, 14.5 mm, 13.6 mm, and 1.5 cm³ respectively for unilateral cases; 13.9 mm, 12.8 mm, 10.6 mm, and 0.82 cm³ respectively for bilateral cases. This represented an average enlargement in width, height, depth, and volume of 3.2 mm, 2.9 mm, 3.9 mm, and 0.66 cm³ (19%, 20%, 29%, and 44% increases) respectively for unilateral cases and 1.9 mm, 3.4 mm, 3.1 mm, and 0.33 cm³ (14%, 26%, 29%, and 40% increases) respectively for bilateral cases. A control group for bony orbital dimensions in congenital anophthalmos not treated with implants was not available in our patient population. Kennedy reported one case of orbital dimensions from a human with untreated unilateral congenital anophthalmos with a height of 23 mm, width 26 mm, depth 45 mm, and volume of 12 cm³; a reduction of 16.7%, 20%, 12%, and 60% respectively compared with the orbital dimensions of the normal side²³; and in another case 18.4% decrease in all orbital linear dimensions with a calculated 36% decrease in orbital

volume.⁶ Hare reported two cases of untreated bilateral congenital anophthalmos, with 35% decrease in orbital linear measurements, and a calculated 58% decrease in orbital volume compared with normal children of the same age.²⁴ Because orbital dimensions in our anophthalmic sockets were initially much smaller than these reported values, these patients cannot be used as a control group. There was no correlation between changes in HEL and changes in prosthesis size, indicating that expansion of the eyelids cannot be used as an indicator of bony orbital growth.

The hydrophilic expander is extremely useful in the early management of the patient with a very small contracted socket, and can usually be inserted without anaesthesia. Our results with silicone expanders have been quite disappointing. Inflatable silicone expanders were only retained in five of 12 orbits. Seven of the eight patients required an anaesthetic for the insertion and each subsequent inflation of the silicone expander. We need larger numbers of patients to determine whether our present results are representative, and if the design of silicone expander we are now using justifies the anaesthetic risk to which we are exposing these patients. Problems with extrusion of inflatable silicone expanders and lack of good control over the direction of expansion have been experienced by other clinicians.²⁵⁻²⁷ Because the expandable orbital implants in our patients were used for only a short time in relatively few patients, we are unable directly to compare the bony enlargement with expandable implants compared with solid shapes.

In this study we have shown that with hydrophilic expanders and solid shapes we can expand the horizontal eyelid length at a rate equal to the normal side. Soft tissue enlargement does not correlate with and should not be used as a variable of bone growth. We have determined values for the magnitude of bony growth achieved by the use of serially fitted solid shapes in the orbit; the present standard of treatment for orbital expansion. Although retention of a prosthesis is usually possible following expansion with serial solid shapes, cosmesis is suboptimal. Improved clinical expansion of orbital soft tissue and bone is required. We hope the inflatable silicone expander will allow more rapid and extensive orbital tissue expansion so that the involved side can catch up with the normal side, but design changes are needed to achieve this potential. Although our results showed no correlation between age at initial insertion of the prosthesis and the degree of orbital bony growth, our numbers were small and we therefore still feel it is important to start expanding the contracted socket as early as possible.

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