Socket expansion with conformers in congenital anophthalmia and microphthalmia

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PURPOSE To report the outcomes of acrylic conformer–assisted socket expansion in congenital anophthalmia and microphthalmia.

METHODS In this noncomparative, interventional case series, the medical records of 24 eyes of 18 consecutive patients with congenital anophthalmia (n = 3), clinical anophthalmia (n = 8), and microphthalmia (n = 13) were reviewed retrospectively. Twelve cases were unilateral; 6 were cases bilateral (3 clinical anophthalmia and 3 microphthalmia). Serial socket expansion with progressively larger acrylic conformers was managed in clinic. Horizontal palpebral fissure (HPF) width was graded as good (final HPF ≥20 mm, or interocular difference ≤2 mm for unilateral cases), fair (17–19 mm, or 3 mm interocular difference), or poor (<17 mm, or ≥4 mm difference).

RESULTS The mean initial lid lengths in anophthalmia, clinical anophthalmia, and microphthalmia were 11.0, 12.4, and 16.9, increasing to 21.0, 19.9, and 22.2, respectively, over a mean period of 51 months. Mean age at the initiation of treatment was 19 months (range, 1-78). Percentage increases in lid length were 90.9%, 61.2%, and 31.3% in anophthalmia, clinical anophthalmia, and microphthalmia, respectively, with an average 7 conformer exchanges. For unilateral cases, the mean final lid length of involved eyes was 22.3 mm, comparable to 23.5 mm in normal contralateral eyes. Good outcomes were achieved in 18 orbits (75%); fair outcomes, in 6 (25%) cases. None of the sockets had poor expansion at final follow-up. All cases had good cosmesis with acceptable prosthesis wear at last visit.

CONCLUSIONS In our patient cohort, good socket expansion was achieved with acrylic conformers alone in congenital anophthalmia and microphthalmia, with acceptable prosthesis wear. (J AAPOS 2022;26:318.e1-6)

Congenital anophthalmia and microphthalmia are rare ocular malformations, with a reported incidence of 0.18–0.4/10000 newborns.[1] This cosmetic disability requires timely intervention.[2-6] The traditional approach involves expansion of the socket either with static acrylic conformers or dynamic hydrogel tissue expanders, followed by orbital expansion when necessary.[4,5,7-12] Socket reconstruction in the initial phase is directed toward expanding the lid fissure both horizontally and vertically and expanding the conjunctival sac and fornices. There are no comparative studies for socket expansion by means of acrylic conformers versus dynamic hydrogel, and both have been used interchangeably.[2,9,12] Performing a tarsorrhaphy and suturing of hydrogel expanders to the conjunctiva requires general anesthesia, whereas acrylic conformers are easy to use but require frequent exchanges.[1,2,9]The use of acrylic conformers as a sole treatment modality is not well described in the literature. We report the outcomes of serial socket expansion alone with acrylic conformers in a cohort of children with congenital anophthalmia and microphthalmia, with long-term follow-up.

Subjects and Methods

The medical records of 18 patients (24 eyes) with congenital anophthalmia or microphthalmia who underwent serial socket expansion at the Department of Ophthalmology, Kyoto Prefectural University of Medicine, from January 2007 to December 2016, were reviewed retrospectively. Institutional ethics committee approval was obtained, and the study adhered to the tenets of Declarations of Helsinki. Informed consent was obtained from all parents.

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Reviewed data include demographic and phenotypic characteristics, horizontal palpebral fissure (HPF, measured between medial and lateral canthus with a ruler by the same observer [AW]), size of first and last conformer, frequency of conformer change, and cosmetic outcomes. Imaging details were recorded if available. Outcomes were defined on the basis of the difference in HPF between the two eyes in unilateral cases: good (difference of ≤2 mm), fair (difference of 3 mm), or poor (difference of ≥4 mm). This was a scale developed by the authors. For bilateral cases, reading of the HPF at the last visit was considered for defining outcomes as good (HPF ≥20 mm), fair (17–19 mm), or poor (<17 mm). Patients with any surgical intervention in the past, associated craniofacial anomalies, and treatment dropouts were excluded.

Anophthalmia was defined as no sign of ocular tissue, seen clinically or with the aid of imaging. Microphthalmia was defined as an abnormally small eye with axial length more than 2 standard deviations smaller than expected for that age group. Clinical anophthalmia was defined as severe microphthalmia, with a corneal diameter of <4 mm and a total axial length of <10 mm at birth or <12 mm after 1 year of age.[13] Accordingly, depending on the pathology type, the conformer type varied and was determined by the prosthesis specialist.

Treatment protocol included socket expansion with serial conformers made of clear polymethylmethacrylate (PMMA) of increasing size until the palpebral aperture was within 1 mm of the other eye or did not change with the maximum possible size. Conformers were exchanged in the outpatient clinic after instillation of proparacaine drops without the need for general anesthesia. Patients were reviewed every 2-3 months. Antibiotic and steroid drops in the form of 0.3% gatifloxacin and 0.1% fluorometholone were administered for 1 week after conformer exchange and at any time if socket congestion or discharge was observed. The acrylic conformer was increased in size when it started to rotate within the socket, and this was observed usually between 2 and 3 months. In the event of spontaneous extrusion of a conformer, a smaller size was fitted, which was replaced with a larger one at the next visit. We documented the horizontal component, vertical component, and depth of conformers at each visit. Details of ocular prosthesis wear, prosthesis fitting and follow-up data from the prosthetic clinic were also evaluated.

Results

A total of 24 eyes of 18 patients with congenital anophthalmia/microphthalmia underwent serial socket expansion over a period of 9 years. Of the 18, 6 were boys, and 6 children had bilateral involvement (3 microphthalmia and 3 clinical anophthalmia). Three orbits of 3 children had congenital anophthalmia, 8 orbits of 5 children had clinical anophthalmia, and 13 orbits of 10 children had microphthalmia. None of the patients had orbito-palpebral cysts. Follow-up data was available for 18 patients, with mean follow-up of 52 months (range, 27-95).

Treatment details are summarized in Table 1. The mean age at the initiation of socket expansion was 19 months (median, 15; range, 1-78), with a mean age of 2.7 months in the anophthalmia group and 29 months in those with microphthalmia. The average HPF at the first visit was 14.6 mm, improving to 21.3 mm at the last visit (an increase of 6.7 mm). The mean number of times conformers were changed serially was 7 (range, 3-16) over a mean treatment duration of 51 months. The final aim was to obtain almost the same lid length (within 1 mm) in unilateral cases, and a normal-sized lid length (≥20 mm) in bilateral cases. Good outcomes were achieved in 18 of 24 orbits (75%) and fair outcomes in 6 cases (25%). None of the sockets had poor expansion at final follow-up. The mean HPF of normal eyes in unilateral cases (both anophthalmia and microphthalmia) was 23.5 mm, which is comparable to 22.3 mm in the contralateral involved eyes at final follow-up. Treatment details for unilateral and bilateral cases are summarized in Tables 2-3.

Three sockets with congenital anophthalmia had a mean increase of 11 mm (increase by 90.9%) in HPF after a mean 14 conformer exchanges. The mean dimensions of the first conformer were 13 mm (horizontal), 10 mm (axial) 5.5 mm (height), increasing to 27.5 × 22.8 × 11.5 mm with the last conformer. The conformer extrusion rate was 14% (2/14), which was attributable to a conformer that was too large; in both eyes, the conformer was replaced with a smaller size and then gradually increased again. All patients were able to wear an ocular prosthesis at the final visit (mean HPF 21 mm).

Eight sockets with clinical anophthalmia had an increase of 7.5 mm (increase by 61.2%) in HPF, with good outcomes in 4 eyes and fair in 4 eyes. The mean conformer size at the first and last visit was 17 × 13.8 × 6.3 mm and 25.4 × 20.8 × 10 mm, respectively. Only 1 of 8 sockets had premature conformer extrusion. In this case, where the prosthesis was too large, the preceding conformer size was reused and the process restarted.

Thirteen sockets with microphthalmia had a mean HPF of 16.9 mm at presentation, increasing to 22.2 mm at the final visit (an increase of 31.3%). Good outcomes were achieved in 11 sockets, whereas only 2 sockets had fair outcomes. None of the cases had conformer extrusion. The mean conformer size at the first and last visit was 18.6 × 15.5 × 6.9 mm and 22.9 × 19.8 × 9.9 mm, respectively. Figures 1-3 show serial photographs of selected cases throughout the treatment course.

Eyelid abnormalities in the form of lower eyelid entropion occurred in 6 of 24 eyes (25%), of which 1 belonged to the microphthalmia group and the rest to the anophthalmia group.

Discussion

The concept of serial socket expansion using acrylic conformers is well known in the literature; however, its use as a sole treatment modality, with results from long-term follow-up, is not well described. Kiskadden and colleagues[14] reported good results with serial socket expansion with customized acrylic conformers alone in 4 cases

Table 1. Average data of all cases

	No. affected eyes	No. of cases	First date of conformer from birth, days	First conformer size, mm			First lid length, mm	Duration of conformer use, days	No. conformer changes	Last conformer size, mm			Last lid length, mm	Last/first lid length ×100 (%)
				H	V	D				H	V	D		
Anophthalmia	3	3	83.3	13.0	10.7	5.5	11.0	1685.7	14.3	27.5	22.8	11.5	21.0	190.9
Clinical anophthalmia	8	5	344.4	17.1	13.8	6.3	12.4	1573.9	7.8	25.4	20.8	10.0	19.9	160.7
Microphthalmia	13	10	888.9	18.6	15.6	7.0	16.9	1512.3	5.2	22.9	19.8	10.0	22.2	131.9
Total	24	18	606.7	17.4	14.4	6.6	14.6	1554.5	7.2	24.3	20.5	10.2	21.3	145.5

D, depth; *H*, horizontal; *V*, vertical.

Table 2. Average data of unilateral cases

Disease/normal[a]	First date of conformer from birth, days	first conformer size, mm			First lid length, mm	First rate of affected/normal lid length ×100 (%)	Duration of conformer use, days	Times of conformer change	Last conformer size, mm			Last lid length, mm	Last rate of affected/normal lid length ×100 (%)	Last/first lid length ×100 (%)
		H	V	D					H	V	D			
Anophthalmia	83.3	13.0	10.7	5.5	11.0	61.1	1685.7	14.3	27.5	22.8	11.5	21.0	90.0	190.9
Normal					18.3							23.3		
Clinical anophthalmia	187.5	17.8	14.5	6.7	13.5	73.0	2137.5	13.0	29.4	22.6	12.4	21.5	93.5	159.3
Normal					18.5							23.0		
Microphthalmia	893.7	18.6	15.6	6.7	17.6	80.4	1730.6	5.6	23.4	20.3	9.9	23.1	97.6	131.7
Normal					21.9							23.7		
Total affected	573.4	17.1	14.2	6.4	15.3	74.7	1787.2	9.0	25.4	21.3	10.7	22.3	95.0	146.4
Normal					20.4							23.5		

D, depth; *H*, horizontal; *V*, vertical.
[a]Anophthalmia, n = 3; clinical anophthalmia, n = 2; microphthalmia, n = 7; total, n = 12.

Table 3. Average data of bilateral cases

Disease[a]	First date of conformer from birth, days	first conformer size, mm			First lid length, mm	Duration of conformer use, days	Times of conformer change	Last conformer size, mm			Last lid length, mm	Last/first lid length ×100 (%)
		H	V	D				H	V	D		
Clinical anophthalmia	396.7	16.8	13.6	6.2	12.0	1386.0	6.0	24.1	20.2	9.2	19.3	161.1
Microphthalmia	883.3	18.6	15.5	7.3	16.0	2141.0	4.7	22.3	19.3	10.0	21.2	132.3
Total	640.0	17.7	14.5	6.7	14.0	1321.8	5.3	23.2	19.7	9.6	20.3	144.6

D, depth; *H*, horizontal; *V*, vertical.
[a]Clinical anophthalmia, n = 3; microphthalmia, n = 3.

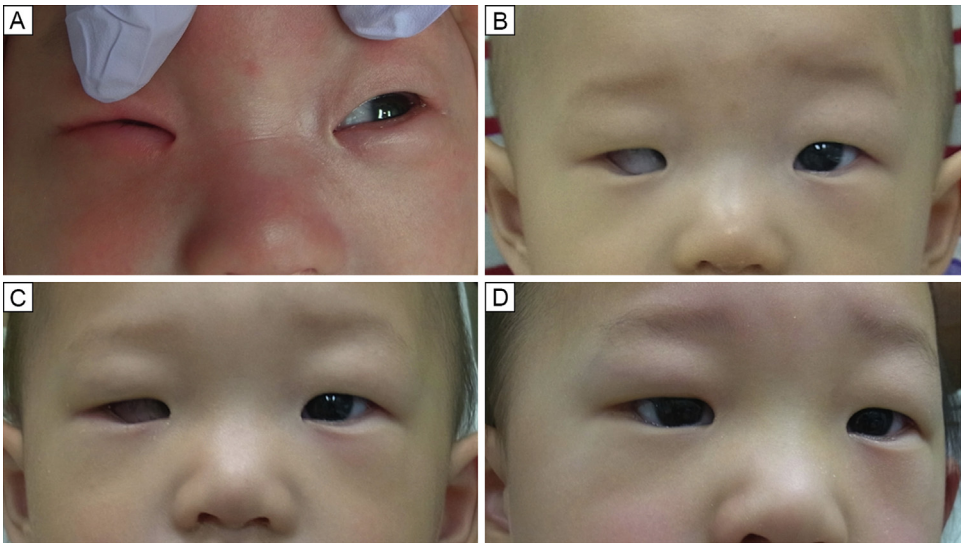

FIG 1. Girl with right clinical anophthalmia at birth (A), undergoing socket expansion using acrylic conformer; appearance at 6 months (B), 2.5 years (C), and 3.5 years (D).

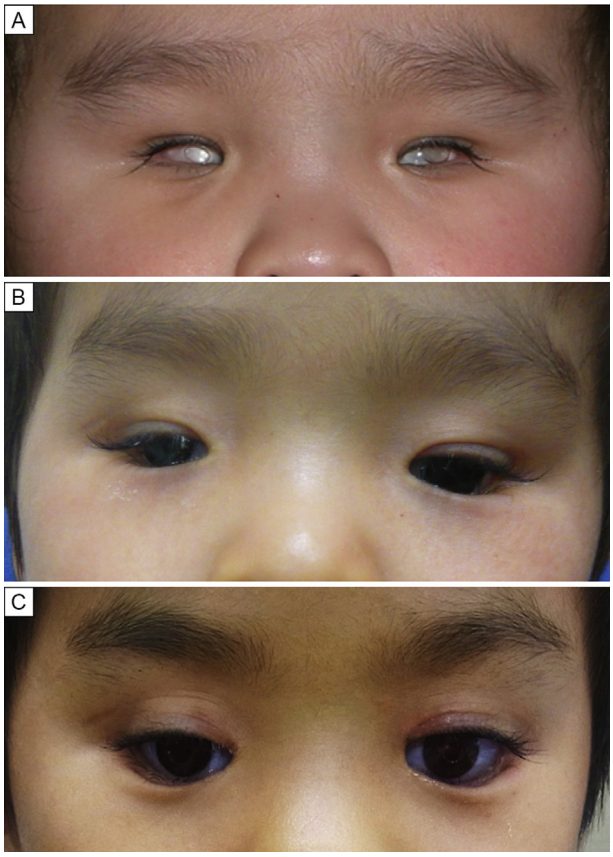

FIG 2. Boy with bilateral congenital anophthalmia at 3 months of age (A) with initiation of. therapy; appearance with painted conformer at 1 year (B) and acceptable prosthesis wear at 5 years of age (C).

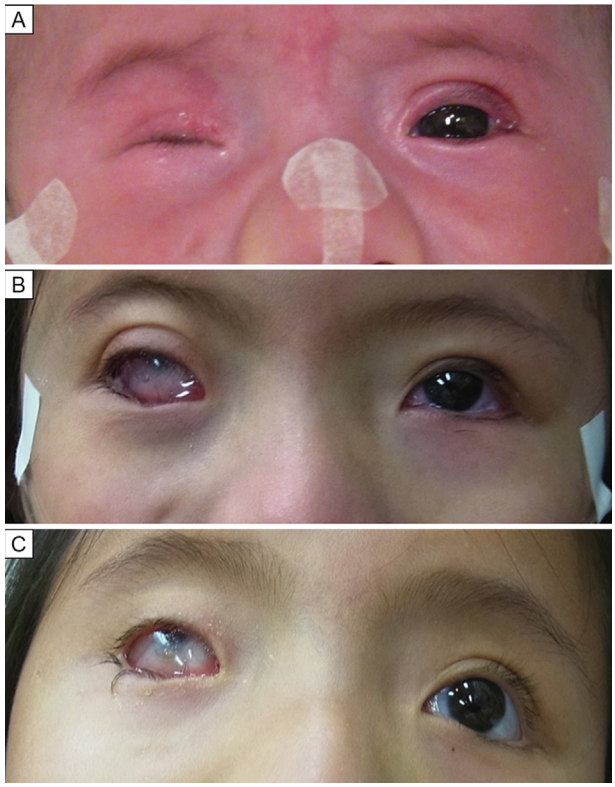

FIG 3. Girl with right microphthalmia at 1 month (A); appearance at 3 years (B) and 7 years (C) with serial socket expansion using acrylic conformer.

of clinical anophthalmia in 1949. Subsequently, similar results were reported in 2 children with clinical anophthalmia in 1968 with acrylic obturators.[4] Serial socket expansion with conformers continues to be used in tertiary hospitals.[15,16] Recently, Taha Najim and colleagues[17] described a cohort of 17 children and young adults with anophthalmia and microphthalmia, of which 16 achieved satisfactory orbital development, symmetry, and cosmesis

after 2 years of serial enlargement with custom-made conformers and prostheses.

The goal of primary treatment in congenitally contracted sockets is to achieve socket expansion, which allows suitable prosthesis wear with the best possible cosmetic appearance. Axial length, and therefore the severity of microphthalmia and anophthalmia, strongly correlates with HPF. For anophthalmic or severely microphthalmic patients, early initiation of socket expansion is paramount for normalizing eyelid and orbital development.[6] Conformers are made of clear PMMA material and can be customized. Progressively larger conformers either customized or noncustomized are used at regular intervals as socket expansion advances. Once no further change in fissure width or fornix depth is attainable, one may consider fitting an ocular prosthesis or proceed to orbital expansion if required.

Hemispherical socket expanders have also been used as an alternative for socket expansion. They are made of copolymers of PMMA and polyvinyl pyrrolidone that can be molded in any shape. They expand after absorbing tears to stretch the fornices; no further expansion occurs once the maximum size is attained, However, they require insertion under general anesthesia. Risks include infection and uncontrolled expansion, resulting in premature extrusion.[12]

El Essawy and colleagues[5] achieved a 7.8 mm increase in HPF of 15 infants with congenital anophthalmia using either solid designed acrylic conformers or hydrogel tissue expanders until the age of 2 years. They customized the conformers to be acorn-shaped to accommodate the depression which exists in the center of the socket in these cases. Hydrogel expanders were used initially in very few cases in their study and were replaced by designed prostheses. We used routinely available clear acrylic conformers, without any general anesthesia, and good expansion (mean, 6.7 mm) was achieved in all but 6 cases. These 6 eyes (3 patients) had a final HPF of 17 mm in 2 eyes, 18 mm in 1 eye, and 19 mm in 3 eyes. All these patients had bilateral involvement and treatment was started at 7, 14, and 59 months from birth. Despite the fair (rather than good) expansion achieved in these eyes, all had an ocular prosthesis with acceptable cosmesis at the final follow-up.

Wavreille and colleagues[18] reported their outcomes of socket expansion in 44 children with congenital microphthalmia. Treatment was started at 4-5 weeks of age, and conformers were changed every 8 days for 3 months. Surgery for orbital expansion was performed in 13 children for severe micro-orbitism. Socket expansion alone was sufficient for final acceptable prosthesis wear in 31 children. Unfortunately, no quantification of lid length was available in their study. However, percentage increase in lid length was 13% in their study compared with 45% in our cohort. In our study, a mean increase of 6.7 mm was obtained in HPF, which is comparable to the reported results obtained with hydrogel hemispherical expanders (7 mm).[9]

We had 3 children with congenital anophthalmia (where use of devices for orbital expansion is anticipated) who started treatment at <3 months of age. We found socket expansion alone was sufficient for achieving similarity to the contralateral eyes. None of the children required orbital expansion or orbitotomies to enable adequate prosthesis wear. The need for orbital expandable devices is debatable with no definitive guidelines. Limited volume gain and repeated surgeries for implant exchange are the limitations of orbital expansion surgeries.[2,7,9] It is possible that early treatment in congenital anophthalmia or microphthalmia with serial socket expansion might provide a limited stimulus for orbital growth as well. Orbital expansion should be performed in cases where soft tissue expansion is not possible because of a narrow orbit. Gundlach and colleagues[9] used hydrogel implants for orbital expansion, but the final volume was 40% smaller than the healthy orbit. Volumetric studies on microphthalmic orbits have shown that the involved orbits have 3 ml less volume than age-matched control groups.[19] Orbital growth slows or even stops by 3 years of age.[19,20] The translation of this deficient volume into enophthalmos may not always be cosmetically apparent—we did not observe enophthalmos in our series.

In conclusion, this study demonstrates that serial socket expansion with acrylic conformers alone may obviate the need for repeated surgical intervention in congenital anophthalmia and microphthalmia. Simultaneously, it also achieves good expansion with resultant acceptable prosthesis wear.

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