

## Guest Editorial

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### Management of Congenital Anophthalmia

*Congenital anophthalmos is a rare malformation that occurs due to developmental arrest of optic vesicle. The reported incidence varies from 0.6 to 4.2 per 100,000 births.<sup>1</sup> True anophthalmos is rare, as a rudimentary globe is mostly present within the orbital soft tissues.<sup>2</sup> It is usually associated with a small orbit, narrow palpebral fissure and shrunken fornices at birth. A delayed intervention jeopardizes the prospects of cosmetic rehabilitation as a small or absent globe hampers further development of the bony orbit.*

*The heterogeneity of presentation and association of other craniofacial anomalies such as hemifacial atrophy, cleft palate and cleft lip can be attributed to the location of defect in the Pfeifer's diencephalic region that stretches from temples to upper lip crossing the orbits and lateral parts of the nose.<sup>3</sup>*

*The extent of orbital hypoplasia, periocular and intracranial abnormalities can be determined by orbital imaging.<sup>4</sup> MRI of the orbit, is preferred over CT scan to avoid radiation exposure. It helps in quantification of the amount of asymmetry and therapy control while using orbital expanders.<sup>5</sup>*

*The management goal of these patients is retention of the prosthesis with acceptable symmetry. Interventions are therefore directed towards achieving an adequate lid contour, conjunctival fornices and orbital volume. The task is carried out by a multidisciplinary team preferably before 3 years of age, comprising of an ocularist, ophthalmologist and pediatrician with support from committed family members.*

*The guiding principle behind the urgency of treatment is based upon the rate of facial growth. The pediatric face reaches 40%, 70% and 90% of the adult size by 3 months, 2 years and 5.5 years respectively.<sup>6</sup> Thus for children presenting before 5 years of age, implants that can increase in size such as dermis fat graft and orbital tissue expanders or progressively larger conformers are utilized. Children older than 5 years are fitted with large fixed size orbital implants.*

*The treatment is initiated by using progressively enlarging static acrylic conformers soon after birth (Figure 1). These conformers can be placed with or without implant and even over a microphthalmic eye. This non-surgical conservative approach of sequentially larger sized conformers over the time is often effective.<sup>7</sup> Appropriate sizing is crucial for achieving the desired results, as a small sized conformer incites an inadequate orbital expansion whereas an inadequately large conformer causes conjunctival irritation and scarring. To enable a more accurate fit in comparison to conventional hand made conformers, 3D-printing has recently been employed with satisfactory results.<sup>8</sup> The conformer design was based on the horizontal eye lid length and visual inspection of the*



**Figure 1:** Clinical photographs showing enlargement of left orbital volume by sequential enlargement of acrylic conformers  
(Courtesy: Mr. Sachin Gupta Art Eyes)

socket. Starting from 2 months of age, a sequentially larger model was fitted over six-and-a-half- months without any sedation or anaesthesia with resultant increase in volume of the conformer from 0.449 ml to 1.69 ml and increase in horizontal palpebral fissure from 9.0mm to 19.0 mm.

In cases of severe microphthalmos, at times it becomes difficult to retain these conformers in the socket due to rotation that affects the expansion or causes unwanted expansion.

Modifications such as creation of a stem or an inverted disc, help in better retention and maintenance of pressure. Addition of a wedge over the conformer helps in localized canthal expansion, socket expansion and prevention of unwanted rotation in the socket (Figure 2).

The orbital volume deficit is addressed after the conjunctival socket has reasonably expanded using conformers. The various orbital expanders used are hard spherical implants, inflatable soft tissue expanders, and hydrogel osmotic expanders.

The expandable orbital implants cause better stimulation of bone as compared to the solid silicone spheres. They also obviate the need for multiple exchanges minimizing the trauma to the soft tissues.<sup>9</sup>

The inflatable soft tissue expanders have been reported to produce uncontrolled direction of expansion resulting in displacement of the conformer, early extrusion and atrophy of surrounding tissues.<sup>10-12</sup>

Hydrogel expander implants are made of copolymer of methylmethacrylate and N-vinyl pyrrolidone. These are placed in the orbit through a small incision and can swell up to 30 times their original volume.<sup>13</sup> Initial stimulation of the growth of conjunctival sac and eyelids is followed by growth of the orbit by serial implantation of orbital hydrogel implants. A lateral orbitotomy incision is contraindicated for implant injection in children to prevent inferolateral migration of the hydrogel implant.<sup>14</sup>

Injectable pellet expanders have also been used by implantation through an inferior skin incision deep into the orbit using a trocar. The reported disadvantage is migration of pellets.<sup>15</sup>

Traditionally, the hydrogel socket expanders are placed under general anaesthesia. A suture tarsorrhaphy is performed at the end of the procedure. Instead of tarsorrhaphy, cyanoacrylate glue can be used and the procedure can be performed under topical anaesthesia.<sup>16</sup>

Satisfactory results can also be achieved by primary dermis fat graft implantation in children with clinical anophthalmia. The disadvantages of this procedure are a second surgical site with an unpleasant scar, variable growth of the graft and prolonged healing time.<sup>17</sup>

Tse et al described an integrated orbital tissue expander in which an inflatable silicone globe was slid on a titanium T-plate secured to the lateral orbital rim. The advantage of this expander was the ease of insertion, absence of implant displacement, uniform delivery of pressure to the orbital bones without the need to change the implant and less number of procedures.

Socket and orbital reconstruction may be required in severe anophthalmia if the lids and fornix are unable to hold the conformer. However, the lid surgeries involving expansion of palpebral fissure, canthoplasty and mucous membrane grafting of the socket are to be performed after initial conformer therapy as early surgery can result in cicatricial changes.



**Figure 2:** Addition of a wedge over the conformer helps in localized canthal expansion, socket expansion and prevention of unwanted rotation in the socket. (Courtesy: Mr. Sachin Gupta Art Eyes)

*In patients with associated orbital cysts, patient is kept under observation as long as possible to ensure socket growth. For smaller cysts, surgical intervention is not required if the prosthesis fits well. Larger cysts are removed by needle aspiration or complete excision after 5 years of age. Patients presenting late may require orbito-cranial advancement surgery with osteotomies.<sup>18</sup>*

*To conclude, management of congenital clinical anophthalmia involves a long term dedicated team work that needs to be instituted as early as possible. Orbital imaging aids in assessment of the defect and monitoring the effectiveness of therapy. Progressively increasing the size of conformers and orbital expanders form the main stay of treatment. The conservative treatment is supplemented with surgical procedures involving the lid, socket and orbital bones. With the development of 3D printed conformers that can be designed, based on 3D models obtained from MRI scan, it has become possible to treat such patients even without a personal visit.*

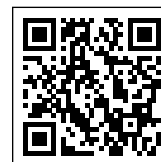
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