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Rehabilitation after Surgical Treatment for Retinoblastoma: Ocular Prosthesis for a 6-Month-Old Child

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Abstract

Maxillofacial prosthetic (MFP) rehabilitation can be especially challenging in a young, precooperative, or behaviorally compromised child presenting with an enucleated eye. Retinoblastoma is the most common intraocular malignancy in childhood and is one of the most common pediatric cancers. Treatment consists of enucleation (or removal of the entire globe) followed by placement of orbital implants. Unrestored anopthalmic sockets exhibit growth retardation and can lead to facial disfigurement. This report describes the challenges faced during rehabilitation of a 6-month-old girl with an anophthalmic socket due to enucleation for retinoblastoma. The objective of the MFP team was to provide a custom-built, acrylic ocular prosthesis in as comfortable and atraumatic manner as possible. The case was a success and underscores the value of a multidisciplinary dental approach for the treatment of children with very special needs.

Maxillofacial prosthodontists endeavor to provide care for all people with craniofacial disturbances, including children. In the pediatric population, the anophthalmic socket can be congenital or acquired. Eye removal may be necessary in recalcitrant intraocular malignancies, severe ocular trauma, intraocular and extraocular infections unresponsive to medical treatments, and blind and painful eyes, as well as for cosmetic improvement of a disfigured eye.

Retinoblastoma is the most common intraocular primary malignancy in childhood.[1] A heritable form of this malignancy is caused by a mutation in the RB1 gene, leading to intraocular tumors, and carries the risk of secondary tumors later in life, particularly in the colon.[2,3] Patients with retinoblastoma usually present with leukokoria (white pupil). Other less-common and less-specific signs and symptoms are deterioration of vision, a red and irritated eye, faltering growth, or delayed development. Some children with retinoblastoma can develop a squint,[4] commonly referred to as "cross-eyed" or "wall-eyed" (strabismus). Retinoblastoma affects males and females equally, and the mean age-adjusted incidence rate is 11.8 cases per million children aged 0 years to 4 years.[5,6] In about two-thirds of these cases,[7] only one eye is affected (unilateral retinoblastoma); in the other third, tumors develop in both eyes (bilateral retinoblastoma). Computed tomography (CT) is the study of choice in the diagnosis of retinoblastoma, but when magnetic resonance imaging (MRI) is available, it should be performed for better differentiation from lesions such as Coats' disease.[8]

The position, size, and quantity of tumors are considered when choosing the type of treatment for the disease. If left untreated, almost all patients will die of the disease; however, with early diagnosis and surgical enucleation and/or external-beam radiation, retinoblastoma patients have been shown to have a 5-year survival rate as high as 95%.[1,9]

Eye enucleation has been the method of choice in unilateral cases and often for the worst eye in many bilateral cases.[10] Radiotherapy and chemotherapy may be used in bilateral cases to preserve the possibility of vision in at least one eye.

Loss of an eye or a disfigured eye has a far-reaching impact on an individual's psyche. When it comes to children and infants, it becomes all the more important. The congenital absence or acquired loss of the ocular globe during childhood causes psychosocial and cosmetic disorders and compromises the normal development of the orbital region. The enucleated socket leads to an unsightly appearance, which may have psychological effects when the child grows up. Also the anophthalmic socket is usually reduced in size and usually develops a small orbita, hypoplastic soft tissues, and shortening of the eyelid rima. The management of the pediatric anophthalmic socket is distinguished from adult anophthalmia, primarily because normal socket and facial development is dependent on orbital growth, and secondly, behavior management in these patients is often a challenge. Installation of an eye prosthesis is essential to the rehabilitation process, so as to produce satisfactory development of the region.

Figure 1 Preoperative photograph.

Figure 3 Examination in "knee-to-knee" position.

The use of orbital implants after enucleation was first described by Frost in 1886.[11] Since then, many shapes, sizes, and materials have been used in the development of the orbital implant. Historically, many children were not fitted with an orbital ball implant after enucleation, especially in those patients whose eyes were enucleated for retinoblastoma, as the implant was believed to interfere with the detection of orbital problems and tumor recurrence. Since the advent of CT and MRI, orbital retinoblastoma and other tumors can be detected with or without an orbital implant.[12] As enucleation in a child may result in retarded orbital growth, orbital implants can provide good cosmesis, good motility, and adequate orbital volume, as well as stimulate orbital growth in the pediatric patient.[13] Porous spherical implants, such as those made from porous polyethylene and hydroxyapatite (HA), are most widely used today.[13]

An ocular prosthesis is fabricated that fits over an orbital implant and under the eyelids. The use of conventional ocular prostheses during childhood entails periodic changes with successive increases to accompany the expansion of the anophthalmic cavity, and stimulation for the growth of the tissues around the anophthalmic socket.

Clinical report

A 6-month-old girl was referred from the ophthalmology department for fabrication of an ocular prosthesis (Fig 1). Earlier, the patient had reported to the Department of Ophthalmology with a chief complaint of whitening of the pupil in the left eye. Ocular sonography, MRI, CT scan, and histopathological examination diagnosed the condition as retinoblastoma in the left eye (Fig 2). The right eye was found to be normal. The left eye was enucleated, and an HA implant was placed. The child was then referred to the Department of Prosthodontics for rehabilitation of the enucleated socket. Upon initial examination in the Department of Prosthodontics, the child demonstrated uncooperative and combative behavior, so a pediatric dentist assisted with behavior management of the child. Providing behavior management for a 6-month-old, precooperative child can be fairly challenging, even for the pediatric dentist. Careful examination of the anophthalmic socket was done in a "knee to knee" position,[14] wherein the dentist and the parent are seated face to face with their knees touching (Fig 3). Their upper legs form the "examination table" for the child. The child's legs straddle the parent's body, allowing the parent to restrain the child's legs and hands. The parents were informed before the examination that it would be necessary to gently restrain the child and that it is normal for the child to cry during the procedure. The examination showed normal depth of the upper and lower eyelids and no other abnormalities, and adequate retention and esthetics of the prosthesis could be expected (Fig 4).

A custom impression tray was fabricated by placing autopolymerizing polymethyl-methacrylate resin on the ball of the thumb and attaching a needle cap to it so the syringe could be attached to the tray for injecting the impression material (Fig 5). The needle cap also acts as a handle for easy placement and removal. The size of the tray was adjusted just smaller than the approximate size of the anophthalmic socket and was tried in the patient's socket after disinfection.

During the impression phase, the child was very uncooperative and cried obstinately at any attempt to even touch the

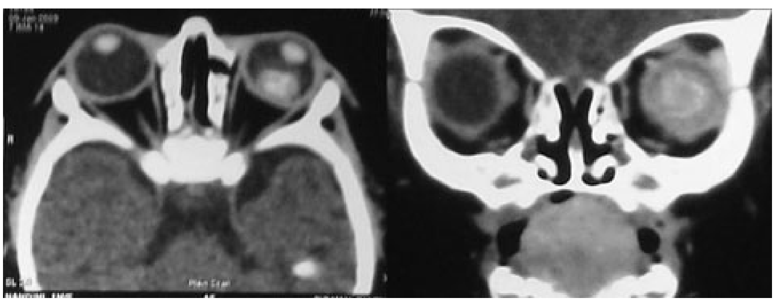

Figure 2 Presurgical MRI of the patient.

Figure 4 Examination of the socket.

Figure 5 Custom impression tray.

Figure 6 Completed impression.

Figure 7 Two-piece mold prepared.

anopthalmic socket. As it would have been almost impossible to get the 6-month-old child to cooperate for the impression procedure, it was decided to make the impression under sedation. Moreover, the eyelids tightly close during crying, and the presence of tears in the socket would have led to an inaccurate impression. The child was sedated using midozalam by an experienced anesthetist.

The impression of the anophthalmic socket was made by a modified impression technique as developed by Allen and Webster.[15] The impression tray was placed within the socket to support the eyelids and provide a more normal contour. The irreversible hydrocolloid (Alginate) was injected into the socket with a syringe through the hollow stem of the impression tray. The impression was removed from the socket and carefully evaluated. The impressions obtained depicted internal ocular structures with some herniation of surrounding tissue into the globe space. Such herniation makes the impression appear irregular, unlike the perfect rounded contours of the eyeball (Fig 6).

A two-piece stone mold was made around the impression (Fig 7). The wax conformer was prepared from this mold. The surface of the wax was smoothened, and the conformer was evaluated in the socket for extensions and soft tissue contours around the socket. Sharp ridges and undesirable irregularities were eliminated for better comfort and esthetically satisfactory results. No sedation was used during this phase or any subsequent phases to enable the evaluation of the retention, movement, and esthetics. Also, performing these steps without sedation helps in conditioning the child to the subsequent prosthesis. As it was difficult to bring the child to cooperate for these procedures, behavior management of the child was done with the help of a pediatric dentist. During evaluation it is necessary to leave the wax pattern in the socket for at least 10 minutes to allow any protective blepharospasm of the orbicular muscle to relax (Fig 8).

The wax pattern was flasked in a denture flask and processed with heat-cured acrylic resin. A custom conformer was

Figure 8 Wax conformer evaluated in the socket.

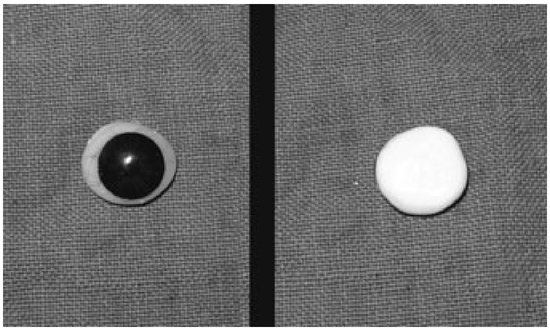

Figure 9 Definitive prosthesis anterior and posterior surfaces.

prepared. To achieve superior esthetic results, the iris and sclera were painted, and red silk fibers were used to imitate a vein pattern. The final fitting and adjusting of the custom acrylic resin eyes included careful polishing to preserve the fine details. Placement of the prosthesis was carried out by gently reflecting the eyelids and inserting the prosthesis side-to-side, much like a denture in the mouth. The musculatures in the socket immediately guide and retain the prosthesis in place (Figs 9 and 10).

The prosthesis showed adequate retention, satisfactory movements, and good esthetics. The child's parents were happy to receive the prosthesis for their child. The parents were instructed about the maintenance and care of the prosthesis and were shown how to remove and replace the prosthesis. They were also instructed that the prosthesis will need to be changed frequently according to the growth of the socket over time. Initially the child seemed to be reluctant and uncomfortable with the prosthesis and tried to remove the prosthesis with her hand, but she became more comfortable with time, and at the 1-month follow-up appointment, she did not seem to be aware of the presence of any foreign object in the socket.

Discussion

Enucleation in both children and adults is associated with a reduction of bony orbital volume, and this decrease in volume is associated with increasing time. It has been shown that adults who underwent enucleation without orbital replacement therapy experienced bony orbital collapse and impairment.[16-17]

Figure 10 Definitive prosthesis fitted in the socket.

Children with anophthalmic sockets are at high risk of craniofacial disfigurement unless timely replacements of successively larger orbital prostheses are fabricated.[9,18,19]

The Moss functional matrix hypothesis may explain this physiological response, stating that a functional relationship exists between muscles and the bones to which they are attached, whereby craniofacial growth is related to specific functional demands.[20] The interplay between the muscles and soft tissues of the eye, particularly the globe, provides the essential movement for proper osseous orbital development.[21]

In our case, a reduced functional demand in the anophthalmic socket would have caused diminished growth of the orbital walls. By acting as the functional matrix, placement of successively larger orbital prostheses during rapid craniofacial development would stimulate a more natural development of the orbital cavity by distributing pressure equally along the orbital wall, providing the tissue stimulus necessary for orbital growth. Although orbital volume varies with race and sex, imaging studies that plot orbital volumetric growth over time show that during early childhood, orbital volume increases in a linear fashion, and by the time the child has reached 5 years of age, the orbital volume for both right and left sides has reached on average 77% of the volume seen at 15 years in both sexes. Growth ends at approximately 15 years of age in boys and 11 years of age in girls.[22,23] Although no standard guidelines for an ocular prosthesis replacement schedule could be found in the literature, a 3-month periodic recall schedule may be suggested for the patient. The mean orbital volumetric growth, as described by Bentley et al,[22] may serve as a useful tool for prosthesis replacements in a growing child. Also, periodic MRI and CT scans may serve as indicators for the growth of the bony socket and may be used to determine the time at which the prosthesis must be replaced.

Conclusion

The use of conventional ocular prostheses during childhood entails periodic changes with successive increases in size to accompany the expansion of the anophthalmic cavity, and it is the only way to esthetically rebuild the anophthalmic socket. The fabrication of an ocular prosthesis for such young patients poses some challenges, which can be overcome with a team approach and effective of management techniques. The installation of an ocular prosthesis during childhood adds inestimable psychological and social contributions to the physical benefit in the patient's rehabilitation as he or she ages.

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