Combined Atresia Microtia (CAM) Repair

A new method of reconstruction of form and function in congential aural atresia and microtia

Abstract

A new surgical procedure to repair both the form and function deficits associated with Congential Aural Atresia & Microtia is described – Combined Atresia Microtia (CAM) repair. In properly selected patients, using the evaluation acronym HEAR MAPS, surgery can return hearing to normal ranges while providing excellent cosmetic repair. The procedure has been performed in one center on over 200 patients and is possible at 3 years of age – within the critical period of auditory development.

Introduction

I first met Dr. John Reinisch in 2004 after inviting him to a yearly conference I have organized since 1994 focused on Congenital Aural Atresia and Microtia (CAAM) and other types of hearing impairment in the pediatric population. After watching his technique of alloplastic reconstruction of the microtia defect for several years from afar, I became very intrigued by the results. Over the ensuing years, a professional relationship and friendship developed that would eventually lead into the enhancement of options available to patients with CAAM.

During the first years of my professional life, I was very fortunate to work in close association with Dr. Burt Brent - perhaps the most experienced surgeon in the history of the United States who expertly performed microtia repair via the rib graft technique. In observing the hearing function and talking to our patients about their hearing challenges in everyday life, it became obvious that patients undergoing rib graft outer ear reconstruction were experiencing similar limitations and habilitation of both form and function. We now understand these limitations are namely due to the lack of binaural hearing during the critical period of hearing development. This observation is well described by Kaplan, et al jn a recent publication,

> ""*Recent studies have shown another alarming phenomenon: Long-term hearing deficits may remain after asymmetric hearing loss has been*

> *treated if the correction was not completed within a critical time window*"[1].

The concept is further amplified in comments made after studying a population of patients who had hearing correction surgery between 11 and 20 years by Breir, et al stating

> "*Despite near-normal postoperative audiograms, all patients were found to have some lasting deficits in binaural hearing, particularly in complex processing tasks such as speech comprehension [in noise] and sound localization. Results suggest that a sensitive and critical period of development is [nearly] complete by 5 years of age*"[2].

This concept of a critical period of auditory development has been evident to those working with the hearing impaired for many years. It became more obvious to me two decades ago as I put together a group through The Let Them Hear Foundation, a foundation started by my wife and I in 2002, that established a Cochlear Implant advocacy program eventually responsible for achieving insurance coverage for bilateral cochlear implants in the United States after several years of legal action against insurance companies. Research data and empiric evidence defining the critical period of auditory development as well as critical two-ear function during this developmental time zone with hearing aids, cochlear implants or corrective surgery is very strong - and the principle applies to unilateral hearing loss seen with Atresia and Microtia as well. In short, normal human development requires independent, two-ear function within the critical period of auditory development. (***Figure 1*** *– please make this last sentence a call out text box when formatting the chapter for emphasis*)

The surgical techniques required for the rib graft microtia repair require the canal surgery to follow all stages of the rib graft microtia repair. Immediately, upon hearing John's presentation back in 2004, it became obvious the technique used for insertion of a porous polyethelene (PPE) scaffold may be able to occur after the surgery to construct an ear canal. The opportunity to restore hearing early in a child's life before reconstruction of the pinna now seemed to be a plausible possibility. I was incredibly excited thinking about the marked effect this may have in alleviating the issues patients were experiencing after undergoing hearing restoration surgery at much later ages in life.

I performed the first ear canal surgery prior to microtia repair and published my first 70 patients in 2009 [3]. **Image 1** shows an ear canal created with in this way one month after the procedure (Legend: reconstructed ear canal 1 month post-op, before PPE microtia repair). This new technique allowed for creation an ear canal to restore hearing prior to

PPE microtia repair. For the first time, binaural hearing was accomplished in patients with CAAM at three years of age - within the critical period of development of auditory development.

After several years of separate canal and microtia surgeries using following these methods, it was a patient's parent who first asked why everything could not be done in one surgery. After extensive collaboration between Dr. Reinisch, myself and our surgical teams, the first Combined Atresia Microtia (CAM) repair was performed in the California Ear Institutes's facilities in Palo Alto, California. Since that time, I have performed over 275 CAMs with three different surgeons in the California Ear Institute's facilities in Palo Alto. The vast majority of these procedures have been performed with Dr. Reinisch who kindly travels from Los Angeles to our facility for each of these surgeries so that we are able to extend this unique, comprehensive service to patients.

The average age of ear canal surgery for children following rib graft microtia repair is 12.2 years in our clinic. The average age of ear canal surgery prior to or at the same time as PPE microtia repair is 4.1 years of age. The impact on auditory development has become a strong reason for parents in choosing PPE for microtia reconstruction.

Hearing

Patients and parents now look for restoration of form and function when selecting options for treatment of Congenital Aural Atresia and Microtia. Our understanding of hearing function and auditory development has improved over the past two decades and sheds light on the inadequacies of our early strategies for treatment of the hearing component of this condition.

My medical training improperly taught me (and many others) that unilateral hearing is adequate for normal function. We now know this is wrong - *single sided hearing loss is a disability*. Perhaps the strongest data that supports this fact is the 10 times increase of having to repeat a grade for children with unilateral hearing loss and the average income levels of those with unilateral hearing loss which is one-third less than that of their two ear hearing peers (insert reference). Adults with single sided deafness will readily testify to the situations where they have difficulty - directional hearing and hearing in noise.

Parents were told (and still are by many well-intentioned providers) that one ear hearing is enough. This statement is undoubtedly not intended to harm but it is false and misleading. Directional hearing will always be impossible when hearing from only one ear. It is important to understand that an observer (such as a parent or uniformed

physician) watching a one-eared hearing child or adult in a quiet room, will see function and understanding appear to be normal. However, when the same child or adult is placed in normal, everyday situations where background noise is present, the deficits are significant.

Alarmingly, the system of complex function of the auditory system develops before its dysfunction becomes readily apparent which may be beyond the time period where intervention can improve function since it is beyond the critical period of auditory development. Since many of the deficits produced by single sided hearing loss cannot be reversed after the critical period of auditory development has passed, statements like "one ear is adequate for hearing" run the risk of withholding a child of adequate development that cannot be restored later in life.

As this becomes more widely understood and backed with even further data, I believe lack of hearing restoration with treatment of CAAM will be viewed more strongly as negligence on the side of the provider should the patient and/or his or her parent not be provided with all the options available regarding both form and function.

Critical Period of Auditory Development

Human auditory development is a very active process that occurs almost solely within the first decade of life - and mainly in the first 5 years of life. First, we develop the ability to understand spoken language (called receptive language) during the first 3 or 4 years of life. Next, expressive language ensues and continues up to about 5 years of age. Lastly, complex auditory functions begin development at birth and continue to about 10 years of age. While the development of each of these areas of hearing and communication function improve slightly with age, the vast majority of development occurs in the first years of life. Once a person is beyond those years of development, the functions cannot be altered significantly (as you know this is the definition of a critical period). Other body systems systems have similar developmental paradigms - such as amblyopia which must be corrected early in life or function of the lazy eye can never be restored.

Importantly, for the complex function to develop normally, two ear hearing MUST be achieved at an early age. Each ear's data stream must be *independent* allowing the brain to process the auditory signal needed for these functions. Three examples are illustrative:

Language Development: both the number of words and the complexity of speech composition in patients with unilateral hearing loss is reduced[4]

Directional Sound: to understand where sound is coming from, the brain takes the time the sound arrives at each year and performs a geometry calculation to determine the azimuth from which the sound originates. If only one ear has sound input, we cannot localize sound. This is very easily understood by anyone associated with unilateral CAAM patients. For the system to develop correctly, binaural sound input must be supplied during early auditory development.

Hearing in Noise: our inner ears deliver to the brain all the sound from our environment. It is the brain that focuses on spoken language filtering out unwanted noise. To perform this function - and to learn to perform this function during the critical period auditory development - the brain must have two separate and independent sound streams, one from each ear. If these two sound streams are not supplied early in life, the system does not develop normally and never will be able to process sound in noise normally. An example we all can understand is a busy restaurant. As the surrounding sounds begin to increase as dinner progresses, we have all had the experience where we 'lean in' to understand someone at our table. With two ears, our speech understanding will drop from 100% in quiet into the 90's in this situation. That can be annoying and challenging as we all have experienced. People with one ear will experience a drop in speech understanding into the 50 or 60 percent understanding range. That is a disability.

If the system does not develop normally during the critical period of development, it will not develop later in life and the patient will be left with this dysfunction in noise throughout life. Children in the classroom with single sided hearing impairment experience this phenomenon which can lead to poor performance and withdrawal, anger issues, poor concentration, and similar behavioral issues. [note single or bilateral implantable and non-implantable bone conduction devices do not provide binaural independent data streams and do not allow the brain performance or development of hearing in noise].

Table 0 lists hearing habilitation options from most to least desirable in terms of mimicry of normal hearing physiology. (Insert table based on **Table 0 Image.** Legend – rank of hearing habilitation options) It should be noted bone conduction hearing devices do not provide directional sound nor do they allow independent binaural sound input and therefore do not improve hearing in noise.

<u>Patient Selection</u>

Every patient is not a candidate for surgical creation of an ear canal. A solid understanding of 'when to say no' is critical for Surgeons to develop in advising their patients. Communication between CAAM team specialists such as Plastic Surgeons,

Craniofacial Surgeons, Orthodontic Surgeons, Audiologist, Anesthesiolgists, and Otologic Surgeons is critical to determine what practical experience has shown to be unique individualized treatment plans for each patient.

Published in 2013[5], we use a classification system we have named 'HEAR MAPS' to fully evaluate each patient. Use of this system also helps us to help to study our results and outcomes. Each letter corresponds to a characteristic of the patient of interest. Two of the letters require objective test data (H with a hearing test and A with an atresia score from a CT scan of the temporal bone) while the others are gained during physical examination.

Table 1 *- please format a table from the data I attach in the image "HEAR MAPS"*

Patients who are adequate candidates for canal creation can have an ear canal and eadrum created either as a separate procedure from PPE microtia repair (we suggest at least 4 months between atresia repair and microtia repair in this choice), or simultaneously with PPE microtia repair – combined atresia repair or CAM – the subject of this chapter.

Of note, I have performed a few dozen canal surgeries following PPE microtia repair where the implant was situated favorably but do not recommend it. Canal surgery following PPE placement puts the implant at risk of exposure and/or infection and is best avoided if possible.

Two of the eight data points in HEAR MAPS are test data (the hearing test or audiogram and the CT scan). The remainder can be determined by physical examination of the patient. The CT scan should only be read by the Otologic Surgeon and not radiology if a decision is being made for a patient with regards to potential surgery. I use the 10 point scale originally developed by Jahrsdoerfer[6] while other surgeons use different evaluation scales. As seen in Table 2, patients with a score of 6-10 on their CT scans using a 10-point scale of grading are candidates for surgical creation of an ear canal. Patients with a score of 5 with bilateral CAAM are occasionally candidates for ear canal surgery as well. Otherwise scores of 5 or less produced hearing infrequently enough that other methods of hearing provision are better options. (please format a table 2 and insert from image "Table 2 – surgical candidacy).

Nearly all patient who have an ear canal created under these guidelines enjoy improved hearing. Percent chance of successful restoration into the normal range increasing with a higher CT score (the "A" in HEAR MAPS). While hearing can be brought into normal ranges with canal surgery, we cannot yet achieve 100% function of a normally formed

ear. The hearing we do get, however, is close to normal allowing both development and function of the auditory system such as directional hearing and hearing in noise noted above.

Cholesteatoma

Every child must have a CT scan before treatment for CAAM even those who are not canal candidates or those who do not want a canal surgically created. The reason is a rare condition of canal cholesteatoma. As the ear canal develops, a small pit forms and a cell tract dissolves toward the future ear canal. In some individuals, this process never starts and CAAM results while in others the process starts and then arrests leading to a buried skin cyst within the temporal bone or soft tissue called a cholesteatoma. The condition is present in 4% of the 3,300 patients evaluated by me in my database. In multiple examples, I have encountered patients who did not have a CT and prior to microtia repair. Later, after undergoing microtia repair with either rib-graft or PPE techniques, the cyst slowly enlarges and erodes bone and may become infected. Loss and infection of both rib graft microtia repair and PPE microtia repair have occurred. In some patients, loss of inner ear function, facial nerve injury and/or meningitis result. We have found the presence of cholesteatoma cannot be excluded by physical examination alone. Importantly, patients should not have microtia repair of any type over a small ear canal as cholesteatoma can develop as a result.

*Insert **Image 2** of patient with cholesteatoma following microtia repair: image "cholesteatoma post microtia repair". Legend: red arrow - external auditory canal meatus with infected cholesteatoma. blue arrows - margin of cholesteatoma extending under microtia repair and also insert **Image 2B** "infected cholesteatoma with fistula pre-surgical repair"*

Some patients can have congenital canal cholesteatoma removed and a full ear canal and eardrum created with the intention of hearing restoration (either as separate procedures or in some instances as part of the CAM procedure). Others should have these conditions resected and treated with removal of all components of the cyst or canal by an Otologic Surgeon prior to microtia repair with either PPE or rib graft surgery. Small ear canals may also progress to canal cholesteatoma if microtia repair is performed over them. Consultation with a CT by the Otologic Surgeon on your team will determine if a cholesteatoma or an indequately small ear canal is present and how to handle it.

Age of CAM

Children should be 3 years of age and 15 kg or more in weight. Two factors lead the earliest age we perform CAM for CAAM. The first is safety. As the procedure takes 7-8 hours, anesthetic considerations for three year olds is paramount. Pediatric anesthesiologists are used and minimal inhalational agents are used due to injection of dilute solutions of lidocaine and bupivicaine both before the procedure begins and ends. Three years of age has become a dividing line for risk of general anesthesia in young children. All CAMs performed at our institution have been accomplished as an outpatient with same day discharge.

Coincidentally, 3 years of age is a good age where a significant amount of growth toward adult size of the head and ear has already occurred. Figure 1 is illustrative of this growth pattern. As can be seen, the outer ear has reached 88% of adult size at 3 years of age and 92% of adult size by 7 years of age. Both rib and PPE surgical techniques require implant size estimation based on anticipated final pinna size after growth has occurred by approximately 18 years of age.

*(Insert **Figure 1** and please redraw a figure based on data in image "Head Growth by age", reference is on the image – adapted from 'Anthropomorphic growth study of the head. Cleft Palate Craniofac J, 1992 vol 29(4) pp 303-308.[7])*

As the middle ear bones are fully formed at birth, middle ear structures can be repaired or augmented as needed without risk of future growth requiring prosthesis change for example. After performing several dozen canal reconstructions prior to PPE, a panel of surgeons at the California Ear Institute blindly viewed intra-operative video and attempted to estimate the age of the patient. No predictive ability was noted in patients pre-selected with adequate HEAR MAPS scores for the procedure.

Surgical Technique and Set-up

CAM surgery is performed in a dedicated operating room in our facility with hand selected anesthesiologists staff comfortable and adept with children and long procedures. As the procedure involved many steps, an OR team needs significant training to efficiently provide the environment and equipment and supplies necessary for successful outcomes.

Specialized equipment necessary to the performance of CAM surgery include otologic and neurotologic micro-instrumentation (some of which has been custom designed for the CEI Otology Surgery Center), high speed otologic drills, an operating microscope with attached high definition video and recording equipment, facial nerve monitoring, laser (CO2 or visible wavelength lasers such as KTP or Argon suffice), split thickness

skin graft dermatome capable of sub-millimeter thickness harvest, customs titanium ossicles reconstruction prostheses of multiple sizes and types. Each of these are needed for the ear canal creation portion of the procedure. (**Image 3:** *Legend - OR Equipment Set Up*) Plastic surgical instrumentation, eye level magnification in the form of loupes, smoke evacuation and handheld batter- operated cautery devices used to form and place the PPE implant. Anesthesia needs are discussed elsewhere in this book.

After successful induction of general anesthesia, endotrachial intubation is accomplished. The endotrachial tube is sewn in securing it to teeth in the maxilla. The patient is rotated 180 degrees on an OR bed that allows side to side rotation and for the Otologic Surgeon to sit with legs under the table with the microscope in place for his or her portion of the procedure. Facial nerve monitoring electrodes are placed in the orbicularis oris and the orbicularis occuli ipsilateral to the ear being reconstructed and covered with a sterile adhesive drape which is later prepped within the field. (**Image 4**: Facial Nerve Monitor) Continuous monitoring is performed throughout the case with the exclusion of the tempero-parietal flap elevation. A Foley catheter and temperature probe is placed. Sterile clear plastic drapes at used to drape off the face including the nose/mouth/eyes and facial nerve monitoring electrodes. The endotrachial tube position is confirmed with anesthesia and further secured with the same.

Hair is removed around the ear of subject as well as the contralateral scalp where the partial thickness skin graft is to be harvested. Using palpation and Doppler if needed, the tempero-parietal artery is located and marked with an indelible marker. (**Image 5**) Care is taken at the area of proposed ear canal to ensue the artery is adequately anterior. In some cases, the artery is directly over the proposed canal site and must be relocated anteriorly[8] (**Image 6**). Preservation of this artery is critical to PPE success.

In unilateral cases, the contralateral ear is used as a model and X-ray or clear cellophane is used to trace the ear to use as when constructing the PPE implant. Measurements in an anterior-posterior direction from the lateral canthus of the ipsilateral eye and superior-inferior positioning are calculated and the final position of the ear is marked with an indelible marker (**Image 7 and Image 8**).

Local anesthesia is infiltrated in the areas of future dissection to minimize the depth of general anesthesia needed over the ensuring hours. Additional injections may be needed during the procedure. Careful tracking of the total amount injected is recommended to remain well under potentially toxic dosing. Hair is braided if long and

potentially an impediment to the procedure. Prophylactic antibiotics (vancomycin) is administered intravenously.

Chlorhexidine prep of the entire head with sterile drapes are positioned leaving the head completely exposed as the contralateral ear will be important for positioning of the microtia repair and partial thickness skin graft and full thickness skin graft harvest on the contralateral side on unilateral cases. The endotrachial tube us placed within a sterile endoscopy sleeve and allowed to rest on top of the sterile drapes. (**Image 9 and Image 10**) The area of full thickness skin graft harvest in the groin previously marked is included in the skin preparation and draping process but covered with a drape to be cut through later in the procedure when the skin is harvested and wound closed. (Insert image)

Surgery begins with removal of the cartilagenous auricular remnant maintaining the overlying skin pedicled anteriorly maintaining its blood supply. This flap is later divided into two vascularized anteriorly based flaps. The inferior one becomes the tragus and the superior flap lines the concha as well as the superior and posterior meatus of the opening of the ear canal. The lobule is mobilized by severing it from the auricular remnant while retaining its vascularity anteriorly. It is held in a steri strip until later in the procedure inferior to the dissection field. The cartilage is retained on the back table in saline for later use.

Previous marked vessels of the tempero-parietal fascia layer are noted and the flap is elevated. Care must be taken to avoid injury to the overlying skin and hair follicles while maintaining the blood supply of the flap. Secondary incision are not needed as retractors and headlights allow work through the bed of the microtia cartilage resection. After the lateral plane of the tempero-parietal flap is dissected to an adequate size, the deep later of dissection is accomplished over the temporalis fascia completely mobilizing the flap for later use. It is left in-situ until needed later in the procedure.

Attention is turned to the ear canal where the microscope is draped and placed within the surgical field. The facial nerve monitoring is tested to ensure normal operation. A superiorly based u-shaped flap of periosteum is created with both deep and superficial layers . The base of the flap is at 1:30 with its inferior resection at 7:30 in a right ear in anatomic position. In a left ear the flap is based at 10:30 extending toward 4:30. The purpose of the flap is to support the PPE implant once inserted to prevent inferior displacement over time. The bone over the area of proposed ear canal is exposed with elevation of the flap.

Using continuous suction irrigation and progressively smaller cutting and diamond burrs, the bone is removed sculpting an ear canal while maintaining bone over the tegmen, the

temperomandibular joint, and the facial nerve inferiorly. Dissection is stopped just short of the ossicles to avoid sound transmission from drill contact. Care is made to open as few mastoid air cells as necessary for ear canal size.

The laser is then placed within the field and the remaining boney fixation of the ossicles is removed. A shelf is created for placement of the new tympanic membrane. Careful inspection of the ossicles, their attachments, and supporting ligaments are made to ensure their mobility. A careful inspection of the stapes footplate and the incudo-stapedial joint is made with endoscopes if not seen adequately through the operating microscope.

Should ossicular reconstruction be needed, measurements are take and a prosthesis is created custom for the patient's anatomy. A bed of antibiotic soaked methyl cellulose is placed in the middle ear.

Attention is turned to the temporal fascia. After moving the tempero-parietal flap, the temporal fascia is incised in a shape and size needed for transplantation and formation of an eardrum. The fascia is prepared on a block under the microscope and fashioned to the correct size for the individual patient.

The scalp is infiltrated with hydro-dissection fluid of dilute bupivicaine and flattened over the area of split thickness skin graft harvest on the opposite occipital scalp. A dermatome is used to harvest a skin graft of 0.25 mm thickness leaving the hair follicles in the scalp without damage as we have previously published[9]. **[Image 11 Legend – Split Thickness Skin Graft Donor Site]** A non-stick dressing is coated with antibiotic ointment and sewn in position as a dressing. The skin is cleaned of every hair under magnification.

The prepared fascia is placed over the the ossicular mass and placed directly on the boney ledge. The previously harvested skin graft is trimmed to size to be placed as a single graft covering the boney walls and the surface of the newly placed temporal fascia thereby creating the tympanic membrane. The medial lumen of the newly created ear canal is filled with pieces of antibiotic soaked methyl cellulose supporting the eardrum reconstruction and the skin graft against the vascularized bone of the ear canal. Lateral portions of the skin graft are folded into the canal in a 'rosebud' pattern and covered with a silastic disc to protect it from being moved until late in the procedure sequence.

Skin is harvested from the groin (or inner upper arm in some cases) in an elliptical donor site. The site is closed in layers and sealed with cyanoacrylate. The full-thickness skin graft is thinned using scissors and preserved to later surfacing of the microtia

reconstruction. In unilateral cases, a second piece of full thickness skin is harvested from the post-auricular sulcus contralateral to the reconstruction and the skin over the mastoid is advanced and the wound is closed. In bilateral cases, this second skin graft can be harvested from the upper inner arm. It is thinned as well and provides an excellent color match for any of the anterior surface of the new ear not covered by the superior retained flap originally covering the microtic remnant.

Using the previously prepared template, the PPE implant is designed and welded with a hand held cautery unit. The implant is placed in antibiotic solution for later use.

The PPE implant is placed within the field and secured with the superiorly based periostial flap. The tempers-parietal flap is removed from under the scalp and draped over the PPE implant to be secured with resorbable sutures making sure its blood supply remains intact. The posterior skin is advanced slightly from over the mastoid and sutured to the deep fascia in the retro-auricular area.

After the entire PPE implant is covered with a vascularized layer, the skin grafts are applied and trimmed to fit. Five and six chromic interrupted sutures are placed in all skin grafts to produce an air tight seal in all suture lines. The groin graft covers the posterior surface of the ear while the pedicled graft as the skin graft from the contralateral ear cover the lateral surface of the ear.

The lobule remnant is freed and dissected to fit in anatomic position where it is held in place with resorbable sutures. The cartilage remnant is used to harvest a cartilage tragal implant. The inferior flap from over the microtic remnant is sewn to the periosteum at the edge of the mandibular fossa using the operating microscope. The cartilage graft is slipped inside the skin flap and a tragus is created with through and through sutures.

The inferior edge of the superior flap which covered the microtic remnant is sewn to retained periosteum at the posterior, inferior and superior edges of the ear canal opening. The ‘rose bud’ STSG is unfurled and trimmed to meet the edge of the full thickness skin grafts laterally. The ear canal is packed with a merocel sponge which is inflated with antibiotic solution. (**Image 12** *Patient 1 - OR*, **Image 13** *Patient 1 - 4 months postop*, **Image 14** *Patient 2 - OR*, **Image 15** *Patient 2 - 8 months postop*)

Two previously inserted drains in the area of dissection are connected to suction allowing the tissue layers to be pulled together over the PPE implant. A soft material used for ear canal impressions that cures over 2 minutes - Azoft - is mixed and placed over the reconstruction and allowed to set into a firm but mildly flexible covering around the reconstruction. Prolene sutures are used to sew the Azoft mold to the surrounding

skin protecting it from inadvertent removal (**Image 16 – postop mold dressing**). After securing the mold, the drains are removed. The skin graft sites contra-laterally are dressed and a head wrap and covering applied to provide mild pressure in the areas of dissection.

All areas of dissection are injected with a mix of short and long acting anesthetic solution (including the IV site and the suture in the maxilla holding the ET in place) in order to allow the child to awake without pain from general anesthesia.

Surgical time averages around 7.5 hours All patients have been discharged the day of surgery as outpatients – a tribute to the quality of anesthetic care as described elsewhere in this work by Dr. Novak, one of our CEI Anesthesiolgists.

Ossicular Reconstruction

Previously unrecognized, we have reported a 32% incidence of malformation of the joint between the incus and stapes[10]. (**Insert Image 17**: Legend – Fibrous Incudostapedial Joint) In a significant percentage of cases, this congenital anatomic abnormality has reduced hearing results in past patients. By reconstructing the ossicular chain with customized prostheses, the impact on hearing loss can be alleviated or reduced achieving marked improvement in postoperative outcomes. Care must be use as over 50% of the time, the facial nerve is exposed congenitally just above the stapes and oval window and is at risk for injury[11]

In some patients, reconstruction of the ossicular abnormality is best accomplished with immediate reconstruction, while a small percentage of patients are best left as is to check the postoperative audiogram 5 months or more after surgery when the hearing results stabilizes. In six percent of patients, revision surgery is indicated to raise the created eardrum and performed ossicular reconstruction due to inadequate postoperative hearing results. Using this strategy as outlined in the above reference, ninety-four percent of patients require only one surgery.

In selected patients, reconstruction with an active middle ear implant is an option. Placement of a Vibrant Soundridge (MedEl Corporation) attached to either the ossicles or placed against the round window can return hearing to functionally adequate levels[12]. (**Insert Image 18**: Legend – Vibrant SoundBridge vibroplasty)

Results

Microtia repair results utilizing CAM repair are nearly identical to PPE results without ear canal creation with the following exceptions:

- Due to the mastoid periosteal flap used to suspend the PPE scaffold in CAM repair, the PPE implant is more prone to fracture due to a blow to the ear. Early results showed fracture in just over 5% of patients. After instituting more significant welding of the connection points of the PPE scaffold, this complication has been virtually eliminated

Microtia repair results utilizing CAM repair differ from separate atresia repair and microtia repair surgery as:

- CAM patients are less likely to see the PPE scaffold descent over time compared to patients who have atresia repair and microtia repair at different times. Presumably, this is due to the more vigorous suspension of the PPE with the mastoid periosteum – a significantly stronger tissue than other suspension techniques applied in microtia repair following atresia repair
- CAM patients are less likely to experience canal stenosis that patients with separate surgeries

Microtia repair results using rib graft compared to microtia repair results using PPE in the following way:

- Canal stenosis following rib graft microtia repair is more likely to occur than when PPE technique is utilized (both CAM repair and separate canal/microtia technique have lower incidence of stenosis)

Hearing Results following microtia repair using rib graft, PPE in CAM technique, and PPE in separate canal and microtia repair are identical. (**Table 3:** please create table from image Table 3 – Hearing Results. Legend – Average Hearing Results following rib graft, CAM and separate microtia repair.)

Long and short-term complications occur in less than 10% of patients requiring re-operating in less than 3% of patients. At this time, nine years after the first CAM, we see no difference in immediate or delayed complication rates compared to other techniques and ages of surgery in my practice.

Infection in the peri-operative period can occur and has been limited to PPE infection in two patients early in the series. A change to intravenous vancomycin from cefazolin has eliminated PPE infections in the last 175+ patients. No case of ear canal infection has occurred.

Eardrum movement away from the ossicles may occur late or early. Usually, a pressure from the middle ear – such as otitis media – has been responsible. In the last several

years, securing the fascia graft used for tympanic membrane reconstruction has dropped this problem to just under 3% of patients. If hearing loss accompanies eardrum lateralization, revision surgery may be indicated.

Stenosis has been the largest and most devastating complication of atresia repair and may occur in up to 30% of patients. Using minimally traumatic technique, skin graft coverage, as well as other surgical techniques we have been successful in markedly reducing this complication. Since 2012, addition of a custom-made ear canal mold made 3-4 weeks postop and worn for 4 months during sleep only (and discontinued thereafter) has reduced canal stenosis to under 2%.

Sensorineural hearing loss may occur with any ear surgery but has not been experienced in our CAM series. Similarly, facial nerve injury with resultant paresis or paralysis can occur from atresia repair but has not been experienced in CAM patients.

Skin graft loss may also mucosa to resurface the ear canal surface and may create a moist ear canal. Inadequate healing or lack of hygiene of the ear canal after healing can allow damaged skin to heal disruptively in 2% of patients. Re-surfacing of the ear canal is needed in a small percentage of this 2% as most can be managed with surface applied preparations and treatment.

Further Considerations

More than 275 patients have selected Combined Atresia Microtia Repair for reconstruction of the congenital form and function deficits associated with congenital aural atresia and microtia since first performed in February of 2008. These cases represent just under 10% of the canal reconstruction surgeries I have performed to date om patients from 49 different countries.

In the first decades of my career, children with atresia and microtia treated surgically required multiple procedures over several years. This was almost always psychologically traumatic to children. Additionally, surgery was done after school age and had effects on confidence and psychological adjustment. CAM repair performed at 3 or 4 years of age alleviates many of the ill effects of prior treatment protocols.

Undoubtedly, the future will see advanced in biomaterials and autogenous tissue growth outside the patient for future implantation impact this field wonderfully. Quite easily, a carved autogenous cartilage scaffold could be used with this technique currently. A more biocompatible and flexible scaffold than PPE will be a further advance to be enjoyed by future patients. The current CAM technique should be, with potentially small modification/s, amenable to other types of implant material.

Plastic Surgeons and Otologists both have important skills to bring when seeking to produce both form and function for patients they are privileged to treat. I have enjoyed a wonderful professional relationship and have learned much from the senior editor of this work with several advances to show for our efforts. I expect the junior editor of this work, and others, will help to achieve further advancement of the state of the art as well.

Summary

The opportunity for a 'one and done' surgical procedure with CAM repair is extremely attractive to parents, especially to those travelling a long distance for services. A cooperative effort between Pediatric Plastic Surgery and Otology and Anesthesia is necessary to achieve excellent results in this complicated and long surgical procedure. To date, surgical results and complications rates similar or better than other forms of atresia and microtia repair make Combined Atresia Microtia (CAM) repair – a new surgical procedure for treatment of congenital aural atresia and microtia – an option for properly selected patients.

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