Contemporary Management of Microtia

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Abstract

Microtia is a congenital defect in which the external ear does not develop appropriately. Microtia occurs at a rate of 1 to 10 per 10,000 births.1–3 Risk factors for microtia include low birth weight, acute maternal viral illness, and in utero exposure to teratogens, including thalidomide and retinoids.4,5 There has been evidence to show decreased risk with maternal folate ingestion.6

The majority of microtia cases are unilateral, most commonly affecting the right side. Males tend to be affected at a 30% higher rate compared with females. Specific patient populations, such as Andeans, Native Americans, Asians, and Hispanics, have a higher prevalence rate.7,8 In less than 50% of patients with microtia a syndrome, including craniofacial microsomia, Treacher Collins’ syndrome, and Goldenhar’s syndrome. Microtia is associated with aural atresia in 75% of cases.9

We discuss contemporary management of microtia, focusing on autologous rib reconstruction, and ancillary procedures, with consideration to hearing habilitation.

Patient Evaluation

At birth, it is important to examine the appearance of the ear, but more critical is assessing the patient’s audiologic status. In addition to describing the abnormal ear, the presence of aural atresia/stenosis should be noted. There are multiple systems used to describe microtia, most commonly the Marx classification. In the Marx classification, grade 1 is a small ear with subunits present, grade II contains recognizable subunits but severely underdeveloped or absent, grade III contains a cartilage rest often with a rotated lobule, and grade IV is anotia.

At birth, the patient should undergo newborn hearing screen if there is a patent ear canal. If bilateral aural atresia is present, the patient will need to undergo auditory brainstem response testing. For patients with bilateral aural atresia, it is crucial to begin hearing management and enrollment in early intervention within the first few months of life to prevent speech and language delays.

Patients with unilateral aural atresia should have regular audiological evaluations to monitor hearing in the contralateral ear.10 Band retained bone conduction systems are used for children under 5 years of age with microtia and aural atresia, either bilateral or unilateral.

Patients with microtia an aural atresia are at increased risk for congenital renal anomalies, vertebral anomalies, and congenital heart conditions. Surgeons should work with pediatricians to screen for other conditions. Patients with microtia have increased rates of depression and anxiety. These rates have been found to decrease following reconstruction.11

Conservative Management of Microtia

Conservative management options include observation or use of an adhesive-retained auricular prosthesis.
Observation may be a good choice for some, although in many cases, the microtic ear will not support glasses and face masks. A nonsurgical option is an adhesive-retained prosthesis. These can be worn at a younger age and it provides the patient with a more symmetric appearance without surgery. However, they require daily maintenance and can be insecure.

**Surgical Management of Microtia**

Surgical options include implant-retained (abutment-retained) auricular prosthesis, alloplastic reconstruction, and autologous rib reconstruction. An implant-retained prosthesis is more secure than an adhesive retained prosthesis but requires adequate bone stock within the mastoid region. It also requires daily skin maintenance of the percutaneous hardware, similar to an abutment used for retention of bone-conduction sound processors. Alloplastic reconstruction with high-density porous polyethylene (PPE) and a temporoparietal fascial flap is utilized if the patient and caregivers prefer a single-stage surgery and to avoid the morbidity of rib cartilage harvest.

Multiple techniques have been described for autologous rib reconstruction including Brent and Nagata.\textsuperscript{12–15} Autologous rib reconstruction is ideally offered at 8 years of age when the contralateral ear is of nearly adult size. Ideally, the chest circumference at the xiphoid should measure at least 60 cm to provide adequate costal cartilage. If there is any concern, an ultrasound can be used to assess costal cartilage stock.

The senior authors (K.C.Y.S., A.D. B., and R.B.) offer alloplastic and autologous auricular reconstruction. We will focus on description of autologous costochondral auricular reconstruction for patients with unilateral microtia. We use a two-stage technique and utilize a two-team approach. They have also previously described their modifications.\textsuperscript{10,16} The following sections describe the evolution of the surgical technique, role of ancillary procedures, and future directions in microtia reconstruction.

**First-Stage Autologous Rib Reconstruction**

The first stage consists of removing remnant auricular cartilage, rib cartilage harvest, creation of auricular framework from rib, and framework insertion into an elevated postauricular flap that includes lobule transposition. A template for framework construction is traced from the contralateral ear on radiographic paper. For patients with bilateral microtia, a template is created from another person’s ear with normal landmarks. The template is then used to plan incisions for positioning of the reconstruction. The framework position can be determined by replicating various distances from the contralateral normal ear to facial landmarks, such as the lateral canthus, nasal ala, or oral commissure. But many patients will have significant facial asymmetry which reduces the utility of using landmark measurements from the contralateral side. We plan framework position by standing at the head of the bed and marking points symmetric with the superior and inferior aspects of the contralateral ear. The template is then used to trace the position of the reconstructed ear. This marking is used to determine the extent of the local flap. Other considerations include the angle of the reconstructed ear and the area of nonhair bearing skin (\textsuperscript{→} Fig. 1). The template is then sterilized to be used intraoperatively during framework construction.

The lobule transposition incision is marked along the lateral aspect of the lobule remnant to an apex and then as a "W"-shaped incision along the medial aspect of the lobule extending onto the postauricular skin (\textsuperscript{→} Fig. 2A). The position of a subcutaneous pedicle planned to be maintained to the superior postauricular flap is then marked out, and often will reside in a dimple posterior to the remnant cartilage at the area of the conchal bowl.

The lobular incision is created, and the lobule is freed from the adjacent subcutaneous tissue. It is transposed to extend to the posterior aspect of previously marked framework position. The lobule isfiled to allow for insertion of the caudal aspect of the construct. The lobule is inset by creating a notch in the native postauricular skin to expose the dermis.
A dermal tab is created at the apex of the lobule by deep-epithelializing a triangular wedge. The dermal tab of the lobule is then secured to the native postauricular dermis using 5–0 plain gut suture to help prevent notching in this area. The lobule transposition is completed by insetting the medial aspect of the lobule flap into the inferior aspect of mastoid portion of the “W”-shaped incision using 5–0 polyglactin and 5–0 plain gut suture (Fig. 2B). The incisions are closed in two layers. It is critical to seal the incisions to maintain suction after closure of all the incisions.

The cartilaginous remnant is then dissected free from the surrounding soft tissue. If there is a substantial tragal component, this portion can be used to create a tragus. The postauricular skin flap is elevated to accommodate the framework. The flap is elevated in a subcutaneous plane maintaining a narrow vascular pedicle from the mastoid fascia to the superior postauricular skin flap as described above (Fig. 3).

Soft tissue is excised anterior to the subcutaneous pedicle to create increased concavity in this region resulting in definition of the conchal bowl. A 10-Fr round suction drain is placed deep to the framework exiting posterior and inferior to the reconstructed site.

Concurrently, the second team will harvest rib cartilage, typically from the contralateral side. An incision, ranging from 2 to 3 cm, is marked at the inferior aspect of the superior synchondrosis (typically sixth and seventh ribs) of cartilage planned to be harvested (Fig. 4A). The incision carried...
through the subcutaneous tissue, rectus abdominus and external abdominal oblique muscles with dissection carried much wider through the subcutaneous, and muscle tissue to allow for dissection through a small skin incision. The intercostal musculature is then freed from the superior and inferior aspects of the sixth, seventh, and eighth ribs. The floating eighth rib and its perichondrium are harvested beginning at its medial attachment and disarticulated at the bony-cartilaginous junction (►Fig. 4B). The harvested rib is typically 8 to 9 cm in length. The floating ninth rib can be harvested in a similar manner as needed.

The synchondrosis is then harvested starting with the inferior limb. The inferior limb is exposed at the bony-cartilaginous (B-C) junction, and a wedge of cartilage is excised just medial to the B-C junction to allow for direct visualization of the cartilage cut. The cartilage is cut maintaining the deep perichondrium. Dissection proceeds medially along the inferior limb between the cartilage and the deep perichondrium. A hook is used to retract the limb superficially to allow visualization of the plane of dissection (►Fig. 5). The superior limb of the synchondrosis is then freed in a similar manner. The entire synchondrosis is removed by using the curved heavy Mayo scissors with curve angled superiorly to maximize the amount of cartilage harvested (►Fig. 6). Hemostasis is achieved and the wound bed is filled with saline. A Valsalva’s maneuver is performed to evaluate for pleural injury. The wound is closed in layers, using 3–0 polyglactin to reapproximate the muscle layer. The final layers of closure are delayed until after fabrication of the framework. The unused cartilage is then sutured together and placed superficial to the rectus abdominus to be used at the second stage surgery. The deep subcutaneous tissue is then reapproximated with 3–0 polyglactin. The skin incision is closed with 4–0 poliglecaprone in a running subcuticular fashion.

The cartilaginous framework is typically constructed with four pieces as follows: (1) a base framework, (2) an antihelix projection piece, (3) antitragus–tragus complex piece, and (4) helical rim. The base framework carving is initiated by placing the template on the synchondrosis to determine the size and shape of the base (►Fig. 7). The synchondrosis is incised to create the base framework being mindful that unused segments are maintained as large as possible for possible uses as other framework pieces. The scaphoid and triangular fossa are carved with a 15-blade, exaggerating the concavities. The impression of curves is overaccentuated, especially along the scaphoid fossa (using a 5-mm skin biopsy punch) and along the anterior aspect of the antihelix. The anterior aspects of the superior and inferior helical crura are tapered significantly to allow for the helical rim to have a rolled edge appearance as the crura approach the helical rim. The antihelix projection piece is carved creating a narrower inferior crus compared with a wider superior crus. An antitragus–tragus complex piece is also carved and secured.
onto the base framework in continuity to the antihelix projection piece. The helical rim is then created from the eighth floating rib. Once all of these pieces are carved, they are secured to the base framework using 4–0 clear nylon with horizontal mattress sutures. The sutures are tied along the posterior aspect of the framework, so that the knots are not adjacent to the skin envelope. A 2-mm punch is then used to create multiple perforations within the scaphoid and triangular fossa, allowing placement of a single drain deep to the framework (► Fig. 8).

The construct is then inserted and rotated into the pocket posterior to the subcutaneous pedicle and placed superficial to the drain which is then placed on suction. Redundant skin from the microtic remnant may be present superiorly. These redundancies should be conservatively excised to avoid compromising vascularity to the skin flap. Sometimes this skin can be draped over vestigial cartilage to create the tragus. The remaining skin incisions are closed in two layers with 5–0 polyglactin for deep dermal repair and 6–0 chromic gut for the skin. The incision lines are meticulously checked for any leaks and are oversewn if necessary. Xeroform bolsters are then placed within the conchal bowl, scaphoid fossa, and triangular fossa, and are secured using 4–0 chromic suture going through the framework (► Fig. 9). Patients are admitted for two nights which allow for pain management and monitoring the status of the skin flap. The drain is kept on continuous medium wall suction, and patients are encouraged to ambulate with the drain on bulb suction. The drain is generally ready for removal by the second postoperative day. We apply topical analgesic cream 30 minutes prior to drain removal. The patient wears the Glasscock mastoid dressing for 1 week and the bolster will be removed at the 1 week follow-up.

Second-Stage Autologous Rib Reconstruction

The second stage consists of framework elevation, full thickness skin graft harvest from the thigh, retrieval of banked costal cartilage harvest, and creation of the postauricular sulcus. It is typically performed approximately 3 months

Fig. 7 Template used to guide carving of auricular framework from rib synchondrosis.

Fig. 8 (A) Helical rim, base framework, antihelix projection, and antitragus–tragus complex rib cartilage pieces separately and (B) with framework completely constructed.

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after the first stage. The skin graft harvest is started by marking out a 10 cm \(\times\) 4 cm ellipse in the nonhair bearing skin along the medial aspect of the thigh, placed in a location to avoid irritation by undergarments; typically, the incision does not cross the inguinal line. The skin graft is harvested with 20 blades in an intradermal plane. Once the graft is harvested, the dermis is thinned and the remaining dermis at the donor site is removed. The 3–0 polydioxanone is used to reapproximate the deep dermal layers and the skin is repaired with a 4–0 poliglecaprone in a running subcuticular manner (►Fig. 10).

The banked rib cartilage is harvested from the previous donor site, excising the prior scar. Dissection is performed in the subcutaneous pocket, cartilage was removed, and wound bed is then closed in a similar manner as in the first stage.

Concurrently, a second team performs the framework elevation. An incision is created at the helical rim, matching the helical attachment to the opposite side (►Fig. 11). Ideally, skin on the framework does not include any of the hair bearing skin. Elevation is performed medial to the framework, avoiding exposing cartilage. The framework is typically elevated to the level of the antihelix. Excessive elevation can destabilize the construct. The skin of the helical rim is advanced to cover more of the helical rim and secured using 5–0 plain gut suture in a modified horizontal mattress manner (►Fig. 12).

The postauricular skin is elevated in a subcutaneous plane to be advanced toward the postauricular sulcus. To provide auricular projection, the banked cartilage is fashioned into a wedge. Previously, the cartilage wedge would be fixed to the postauricular surface of the framework and then be draped by an anteriorly based mastoid fascial flap. Our technique has evolved to insert two separate cartilage wedges deep to the framework from superior and inferior pockets that span the postauricular sulcus.

The postauricular flap is then advanced and secured to the mastoid fascia using 3–0 polydioxanone. Aggressive
advancement of the postauricular flap has enabled the sulcus to be created with a single skin graft. The full-thickness skin graft is inset, using 5–0 chromic gut suture. If there is significant asymmetry in auricular projection, a contralateral otoplasty can be performed at the time of second stage surgery. The contralateral postauricular skin can be used as a skin graft to recreate the postauricular sulcus. A xeroform bolster is placed over the skin graft and secured using 4–0 prolene. The Glasscock dressing is placed and postoperative care is similar to the first stage (►Fig. 12). ►Fig. 13 demonstrates a typical postoperative result of our technique.

**Single-Stage Autologous Rib Reconstruction with Temporoparietal Fascia Flap**

In patients with deformities involving the superior portion of the auricle, such as in grade-I/II microtia, posttraumatic microtia, or microtia resulting from oncologic resection, a single-stage procedure utilizing autologous rib cartilage and temporoparietal fascial (TPF) flap is an option. In these instances, the procedure begins by templating the normal ear and planning incisions as described above. The postauricular flap is elevated in a similar manner to expose the auricular defect. The postauricular flap is elevated superiorly in a plane superficial to the temporoparietal fascia, but the extent of superior dissection is dependent on the degree of the reconstruction as the TPF flap will be reflected inferiorly. In certain cases, the dissection can be performed under endoscopic visualization. To perform full reconstruction with TPF flap, the flap typically is elevated to 13 cm above the superiorty aspect of framework position. Once the subcutaneous flap is fully elevated, incisions are planned anteriorly and posteriorly for adequate TPF width to cover the defect, and incorporate the superficial temporal artery. A superior incision is made and a uterine sound is then used to free the TPF from the deep temporalis fascia. The TPF flap is reflected inferiorly.

A second team performs the rib cartilage harvest as described above. The framework is reconstructed using the template as a guide and the pieces are secured using 4–0 clear nylon. The framework is secured to the native auricular cartilage and secured in a similar manner, and a 10-round suction drain was placed. The TPF flap is laid superficial to the reconstruction and secured to the native and reconstructed framework. The postauricular flap is then advanced into the postauricular sulcus and secured using 3–0 polydioxanone suture. A full-thickness skin graft is harvested from the thigh and secured to the superficial surface of the TPF flap (►Fig. 14). A bolster is placed over the reconstruction and remains in place for one week.

**Role of Fat Injection**

Many patients with microtia have a degree of craniofacial microsomia and associated facial asymmetry. Correcting the auricular deformity can accentuate the facial asymmetry. One technique to improve symmetry is autologous fat transfer. Subcutaneous fat is harvested from the abdomen, external oblique region, or thighs by manual liposuction. The harvested fat is centrifuged to separate the fat from oil and
blood products. The fat is injected using cannulas into volume deficient areas of the face to improve symmetry. A slight overcorrection should be considered due to a degree of fat resorption over time.

**Ear Piercing**

Following reconstruction, patients will often want their ears pierced. Piercings can be placed at the time of final-stage reconstruction. The piercings should not be placed through the cartilaginous or alloplastic framework, as this can lead to exposure and infection. Rather, the piercing should only be placed within the lobular soft tissue. As there may be persistent auricular asymmetry, the piercings should be placed at a similar level to the contralateral ear piercing, rather than the same location within the lobule (Fig. 15).

**Timing of Mandibular Reconstruction**

Syndromic microtia patients may require mandibular reconstruction if there is asymmetry due to deformities of the temporomandibular joint, condyle, and ramus. Reconstruction can be performed with distraction osteogenesis, costochondral grafts, iliac crest grafts, or fibular free flaps. Unless there is a severe deformity causing psychosocial issues, mandibular reconstruction is often not performed until the patient has permanent dentition or reaches skeletal maturity. Traditionally, microtia reconstruction would not be performed until after mandibular reconstruction, as the belief is that this would allow for better auricular symmetry. However, now microtia reconstruction is completed before patients reach skeletal maturity.

**Use of Three-Dimensional Printing and Future Directions**

The emergence of three-dimensional (3D) printing provides significant potential in facilitating preoperative surgical planning, framework reconstruction, and also with surgical training. The 3D printing has been used to create resin model templates based on anthropological analysis of the external ear which can be used intraoperatively. These templates can be placed within the soft tissue envelope to mimic skin flap contouring and guide framework alterations. Further, computer-aided design (CAD) can enhance 3D printing to create individualized templates which are ultimately used to carve PPE frameworks.

The 3D printing has significant utility in patients with craniofacial microsomia. Achieving symmetry in an auricular projection following reconstruction can be difficult due to asymmetrical landmarks and mastoid hypoplasia. Kimura and colleagues produced 3D-printed models based-off of individualized computed tomography (CT) scans in patients with craniofacial microsomia. These models were then

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**Fig. 14** A 5-year old boy with posttraumatic left ear avulsion who underwent single stage autologous rib cartilage with temporoparietal (TPF) flap. (A) Preoperative with superior ear missing. Superficial temporal artery branches marked anteriorly along with postauricular incision through TPF flap was harvested. (B) Cartilage framework. (C) TPF flap harvested prior to reflection to cover framework. (D) Skin graft in place over TPF flap covering cartilage framework.

**Fig. 15** Fifteen year old girl with grade-II left microtia who underwent two-stage autologous rib reconstruction. (A) Left microtic ear (B) Status post left ear piercing at second stage microtia reconstruction. (C) Postoperative.
used to determine a symmetric axis of projection (AOP) between the reconstructed and normal ears. A standard PPE implant was placed on the model and silicone putty was molded to achieve a satisfactory AOP. A clear plastic template was created from this model and used intraoperatively to determine the correct framework positioning.

The 3D printing has resulted in advancements in training simulations for creating auricular frameworks. Berens and colleagues used CAD and 3D printing to create a silicone and cornstarch model of costal cartilage synchondrosis and free-floating rib that was proven to have a similar texture and firmness to human cartilage. These models have been used in resident and fellow training simulations. Chang and colleagues created 3D-based models for various procedures, including microtia rib cartilage carving. Pre- and post-training surveys showed a statistically significant increase in self-rated expertise for graft design, carving, and assembly.

There has been significant progress in tissue engineering of the auricular framework. The goal of tissue engineering is to create an auricular framework with high biocompatibility that resembles elastic auricular cartilage, low infection rates, and avoid donor site morbidity. Bioengineering of an auricular framework can be difficult due to the auricular shape, balancing durability, stiffness, and flexibility, and requiring a significant number of progenitor cells. Chondrocyte and mesenchymal stem cells grown on a bioprinted scaffold shows promise to be used for auricular cartilage regeneration. Bioprinted ears have been shown to develop new chondrocytes in animal models, and there is ongoing investigations for application in a human model.

**Hearing Management in Microtia**

Hearing should be evaluated at birth and monitored closely throughout development. Patients with associated aural atresia will have a maximal conductive hearing loss. Up to 10% of these patients may have a sensorineural component to their hearing loss. The hearing management options in patients with microtia and aural atresia is shown in Fig. 16. Various options exist for hearing management, including observation, Softband-retained bone conduction devices, or osseointegrated implants. Observation is not recommended as the patient will have a persistent hearing loss that will continue to affect their development.

Nonsurgical bone conduction devices include Adhear (MED-EL USA, Durham, NC), Ponto Softband (Otonix, Somerset, NJ), and Baha Softband (Cochlear Americas, Englewood, CO). These devices do not require surgery to be placed. However, since they are not rigidly attached to the bone, there can be some artifact and reduced sound fidelity.

Osseointegrated implants can be placed with a simple procedure and can have excellent hearing results. These devices can be categorized as active or passive bone conduction devices. Passive bone conduction devices consist of an external sound processor and an osseointegrated implant. The external sound processor converts sounds into vibrations, and then the osseointegrated implant receives the vibrations, passively transmitting them to the cochlea. The Baha Attract, Baha Connect (Cochlear Americas, Englewood, CO), and Bonebridge (MED-EL, Innsbruck, Austria) are examples of passive systems. The Osia (Cochlear Americas, Englewood, CO) is an active bone conduction device with a transcutaneous sound processor and an osseointegrated implant. This system uses piezoelectricity to create an electrical charge from vibrations with the potential of increasing the high frequency gain.

The osseointegrated implant can be placed at the time of microtia surgery. The implant should be placed posterior to the incision for the microtia reconstruction. Various techniques for the skin incision can be utilized, including using a 5-mm skin biopsy punch, or making a postauricular incision. If an Osia is placed, a subcutaneous pocket is created and then the dissection transitions to a subperiosteal plane in the location in which the implant will be placed.

**Management of Aural Atresia**

If a patient has aural atresia, a CT scan of the temporal bones is obtained between the ages of 4 and 5 years. Advantages of waiting until this age allow for mastoid growth, reduce the need for sedation during imaging, and can screen for occult congenital cholesteatoma. CT findings are then used to determine if the patient is a candidate for atresiaplasty based on the Jahrsdorfer criteria. A higher score improves the patient’s candidacy for repair. Patients with a score of 7 or higher are considered candidates.

The goal of atresiaplasty is to improve the cosmesis of the ear, create a wide meatus with an epithelialized ear canal, and improve hearing. An additional goal is to create a wide meatus. Patients may have less predictable results and less hearing benefit compared with a bone implant. However, they can wear a hearing aid to enhance any residual hearing deficit. Traditionally, microtia repair was performed prior to atresiaplasty, as the vasculature would not have been altered by previous surgeries.

However, some surgeons are performing atresiaplasty before or concurrently with microtia reconstruction. If performed concurrently, autologous reconstruction is preferred over alloplastic, as alloplastic reconstruction has an increased risk of infection and framework exposure. Benefits of concurrent reconstruction include the ability to reconstruct the tragus during metaplasty, and decrease the risk of aural stenosis by utilizing the pedicled skin flap, reconstructing the tragus to also line the meatus.

The senior authors have recently started to perform atresiaplasty before microtia reconstruction. Performing the reconstruction in this order allows for the auricular framework to sit in the correct position relative to the reconstructed meatus. It also prevents the risk of unnecessary exposure of the auricular framework if atresia repair is performed before microtia reconstruction. When performing atresiaplasty in a patient with microtia, the incision is placed posterior to the vestigial cartilage. A vertical incision is created to preserve as much of the nonhair bearing skin as possible. At the time of the microtia repair, a template is created based on the contralateral ear and placed over the
Fig. 16  Overview of management of microtia and atresia. Management of microtia and atresia begins at birth with the newborn hearing screen. Based on the results, further hearing assessments will be performed in the early intervention phase, combined with Softband-retained bone conduction sound processors. During the following years, speech and language development are closely monitored. Management timeline is divided into diagnostic tests in the left column and interventions are in the right column, for both hearing loss and microtia.
neomeatus. The remainder of the microtia repair is similar to the procedure previously described (~Fig. 17).

Conclusion

Microtia can affect patients both physically and psychologically. Each microtia reconstruction is unique, and our technique has evolved to help optimize outcomes. Advancements in CAD, 3D printing, and adjunct procedures are further assisting in creating an ideal auricular contour and to improve facial symmetry. Hearing management is critical in the development of all microtia patients. There are many options for hearing rehabilitation, both nonsurgical and surgical, that can be tailored to fit the goals of the patient and their caregivers. With attention to all of these issues, microtia reconstruction can be very rewarding for patients, families, and providers alike.

Conflict of Interest

R.B. reports other from SpiWay, LLC, other from EigenHealth, Inc., and other from Wavely Diagnostics, Inc., outside the submitted work.

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