Advances in the treatment of microtia

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Purpose of review

Creating the fine details of the ear in a patient with a congenital absent ear is extremely challenging. Each component of the multidisciplinary team that manages the ear reconstruction, hearing restoration, and associated craniofacial anomalies of these patients has seen recent progress.

Recent findings

Population studies have provided new insights into the etiology of microtia. Novel techniques for costal cartilage harvest, implantation, and positioning add to the techniques of Brent and Nagata, which remain the gold standard for microtia repair. Advances in the use of alloplasts and tissueengineered cartilage appear promising.

Summary

Technical advances in combined aural atresia/microtia reconstruction, bone-anchored prosthetics, bone-anchored hearing aides, and use of alloplastic implants provide numerous options to patients and practitioners. Implantable, tissue-engineered auricular frameworks appear to be a promising option for the future.

Keywords

aural atresia, auricular prosthetics, congenital ear deformity, costal cartilage graft, Medpor, microtia, oculo-auriculo-vertebral spectrum

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Abbreviations

BAHA	bone-anchored hearing aid				
FTSG	ISG full-thickness skin graft				
OAVS	oculo-auriculo-vertebral spectrum				
TPF	temporoparietal fascial flap				

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Introduction

The treatment of congenital auricular deformities is one of the most difficult procedures in facial reconstructive surgery. A comprehensive understanding of the intrinsic nuances of the auricular architecture is essential to microtia repair, but is no guarantee of a final result. Even the most artistically carved costal cartilage graft or alloplastic implant can result in a mediocre appearance due to skin thickness, wound healing, and scarring. Microtia reconstruction with either autologous costal cartilage or porous polyethylene still does not equal the appearance seen with auricular prosthetics. Ultimately, tissue engineering may provide an auricular framework suitable for microtia reconstruction from human chondrocytes.

Definition and classification

Congenital malformations of the external ear can occur in a spectrum of severity. Total aplasia of the external ear is termed *anotia*. *Microtia* refers to a spectrum of deformities of the external ear that can range from a slightly smaller ear with the majority of structures present (grade I) (Fig. 1), to a greater deficiency of ear structure (i.e. absent lobule or helix, grade II) (Fig. 2) to the classic 'peanut' deformity (grade III, Fig. 3) [1]. Throughout this report, anotia will be labeled as the most severe form of microtia. The term cryptotia refers to a superior pole of the ear that is buried in the temporal skin (Fig. 4) [2]. A simplified approach classifies the microtia as lobular (presence of ear remnant and lobule) or conchal (concha, external canal, and tragus present with lobule) [3].

Etiology

The mechanism during embryologic development that induces microtia is not well understood. During the third week of gestation, the hillocks of His develop around the otic placode. The external ear can be altered by a variety of causes. Arrests in development can be correlated to a period in ear development as it relates to facial embryology. Oculo-auriculo-vertebral spectrum (OAVS) is such a condition to be considered in patients with microtia that includes hemifacial microsomia (temporal, maxillary, or mandibular hypoplasia), soft tissue abnormalities (preauricular tags or macrostomia) (Fig. 5), eyelid defects (coloboma, epibulbar dermoids), vertebral abnormalities, and congenital renal or heart defects. The craniofacial team with which the author participates would arrange a renal ultrasound and full spine radiographs on children with suspected OAVS. Cervical spine damage after microtia surgery has been reported and should reinforce the importance of proper intraoperative head positioning Figure 1 Photograph of a right grade I microtia (constricted ear)



[4[•]]. These children may be predisposed to congenital vertebral defects associated with OAVS.

Thalidomide and the acne medication Isotretinoin (Accutane, Roche, Nutley, New Jersey) are two drugs with known associations with inducing microtia in humans. Many other drugs have been used in animal models, but without clarification of the mechanism. Recently, the alkylating agent, ethane dimethanesulfonate, was given to mice during gestation and created increasingly smaller microtia (bilateral) in a dosedependent manner. Increasing the dose also inhibited the mandible ossification and may suggest that the drug targets a specific molecular event in ear development, which would be a promising site for preventive interventions [5].

Epidemiology

External ear anomalies occur as both isolated cases and as a feature of multiple congenital abnormalities (nonisolated) such as clefting, neural tube defects, and limb malformations [6]. In the United States, reported prevalence rates for microtia range from two to three births in 10000, with increased rates in Hispanics and Asians (Japanese) [7]. The occurrence of microtia in Navajo Indians has been reported to be as high as one in 1200 [8]. In a study [9] of 2.5 million live and still births in California, nonisolated cases of microtia (1.53/10000) were more common than isolated cases (0.63/10000). Shaw et al. [9] reported that isolated microtia was seen more often in foreign-born Hispanics (7:1) and Asians (3:1) than Caucasians. Consistent with previous reports [7,10[•],11[•]], microtia was found more commonly in males, unilateral (90%), and right sided. The male predisposition was attributed to gender differences in urogenital development and teratogen susceptibility.

The unique multicultural population in Hawaii was also studied for the 1986–2002 period (316 508 live births)



(a) Preoperative view of a left grade II microtia. (b) Same patient after Mustarde suture placement, and contralateral conchal cartilage graft as the first stage in creating the lobule. (c) Same patient after full thickness skin graft to the posterior lobule and cervical skin advancement to create a sulcus.

Figure 2 Left grade II microtia

Figure 3 Grade III microtia



(a) Photograph of grade III microtia. (b) Photograph of a conchal type microtia.

revealing a $3.79/10\,000$ prevalence of microtia. Filipinos, Pacific Islanders, and Far East Asians were more affected than Caucasians [11[•]]. A recent study in China [6] of 453 microtia cases in 3.2 million births (1.4/10000), however, found no male preponderance [6]. The reason for this unique difference is not clear. In fact, if one considers that the Chinese population has more males than females (~1.2:1), a greater male preponderance could be anticipated.

Figure 4 Cryptotia is represented by the superior helix being buried in the temporal skin

Castilla *et al.* [12] reported an increase in congenital craniofacial defects (microtia, cleft lip, preauricular tag, and branchial arch anomaly) at altitudes higher than 2000 m in a study of 1.6 million consecutive births in the Latin-American Collaborative Study of Congenital Malformations (ECLAMC) [12]. At above 3000 ft in Quito, Ecuador, a fivefold increase in microtia prevalence (17.4/10 000, n = 46041) compared with six South American countries (3.2/10 000, n = 553068) was reported with prenatal drug exposure, high birth order, and increased paternal age proposed as risk factors. Unlike nearly all



Figure 5 Oblique photograph illustrating left macrostomia, preauricular tags and ear anomalies seen in oculo-auricuo-vertebral spectrum (OAVS)



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other studies, most of the cases were seen to be isolated microtia [13]. Future insights into this high incidence may bring preventive advances.

Microtia reconstruction with a costal cartilage framework

In an effort to address specific aspects that affect the outcome of auricular reconstruction with autologous costal cartilage, contributions from recent publications will be categorized for direct comparison and discussion. These include framework wire extrusion; skin flap vascular compromise; loss of framework detail during wound healing; poor framework projection from the skull; framework positioning; concurrent lobular transposition and ear framework placement; prevention of complications in costal cartilage harvest; and combined hearing improvement and microtia reconstruction.

Stainless steel wire or suture fixation

The extrusion of stainless steel wires that are often used to secure the cartilage framework subunits has been reported. Nagata [3] reported use of a fine gauge wire without complication. Tai *et al.* [14^{••}] described a patient who suffered a thermal burn from the wire heating in a sauna and reported the use of 4-0 vicryl suture on a tapered needle instead of wire in 42 patients. They suggest that the braided character of vicryl reduced slippage of knots compared with smooth nylon, which has been advocated by Brent. This author prefers to use temporary needle stabilization of the subunits followed by clear, 4-0 nylon suture placement using a straight needle, taking care to bury the knots away on the posterior surface. The use of absorbable sutures may be an excellent consideration.

Skin flap vascular compromise

The two general methods for covering the costal cartilage or alloplast framework are a skin pocket or a temporoparietal fascial flap (TPF) covered by a skin graft. Traditionally, a TPF flap is used to cover an alloplast framework (e.g. high-density porous polyethylene; MedPor, Newnan, Georgia, USA). By contrast, a skin pocket that accommodates the costal cartilage (Brent or Nagata technique) provides a better color match than a skin graft. The microtia surgeon must balance the skin flap thickness and pocket size with the ability to create contours around the cartilage framework. Minimizing or completely avoiding the use of epinephrine in the flap can be helpful [15]. A simple intraoperative test is to place the framework in the pocket, activate the suction, and observe for blanching along the helical contours. To obviate vascular compromise, the pocket size should be enlarged until blanching is prevented. A tissue expander can also be placed for 2 months prior to framework placement [16[•]].

Concurrent framework placement and lobular transposition

Although the Brent technique [15] creates conchal contour and tragus projection with a contralateral composite graft at a separate stage, the Nagata technique employs simultaneous framework placement and lobular transposition [3]. Vacuum-assisted drainage around the framework can fail due to air leakage through the lobule incisions. Tai et al. [14**] counter this point by removing active suction after loose bolster sutures are placed. Using the Nagata technique, one must carefully maintain adequate vascular supply to the framework skin pocket and lobule flap. During the framework placement, the anterior-superior lobule can be added to the conchal bowl from a specially designed lobular flap. The posterior lobular remnant is used to create a lobule. The vascular supply to the anterior-superior lobular remnant is a subcutaneous pedicle from the deep lobular remnant.

Similarly, blood vessels originated in the parotid fascia behind the microtic cartilage remnant were found to supply the lobular flaps both lobular and small conchal type microtia. In conchal type microtia, Ishikura *et al.* [17] reported that the perichondrium of the conchal remnant contains vessels originating from the posterior auricular artery and suggest preservation of these vessels to enhance vascular supply to the flaps.

The vascular pedicles described in these two papers could very well be synonymous.

Loss of framework detail during wound healing

Some authors attribute the loss of detail in these reconstructions to small hematomas and infection. Tai *et al.* $[14^{\bullet\bullet}]$ suggested a 3-cm pocket surrounding the framework with an intricate drainage technique. They placed vacuum aspiration only intraoperatively so as to suture 'loose' bolsters to the concha, antihelix and helical contours that will remain for 14 days. Only passive drainage with up to nine catheters (1.2 mm) placed through the skin surrounding the framework is used. Injecting fibrin glue immediately prior to skin closure has been suggested to create stability and skin adhesion to the framework, and hematoma prevention $[18^{\bullet}]$.

Projection of the cartilage framework

Improper projection of the reconstructed framework can draw attention to even the most finely crafted ear. As early as 1952, Steffenssen [19] reported a mastoid flap to create a postauricular sulcus. Cartilage (autogenous or irradiated rib), skin grafts, and alloplasts have been suggested because soft tissue alone does not consistently provide sufficient ear elevation. Brent banked a piece of costal cartilage during the first stage and used it to elevate the framework during skin grafting of the posterior framework with scalp advancement (stage 3). A retroauricular scalp fascia flap [15,20] can be elevated off the mastoid and turned over to cover the banked cartilage in preparation for a skin graft. In this author's experience, the banked cartilage can be dissected from its temporal implantation site with the surrounding fascia and rotated into the retroauricular sulcus for ear elevation and skin grafting [21]. Another alternative is to cover a crescentic costal graft with a TPF flap to provide ear projection; however, this does preclude the TPF flap for future salvage procedures. TPF flaps can also be complicated by alopecia, additional scalp incisions, or wound contraction.

Tai *et al.* [14^{••}] introduced the use of a hydroxyapatitetricalcium phosphate ceramic (Ceratite; NGK Spark Plug, Nagoya-shi, Japan) for auricular elevation. Traumatic fracture can occur with ceramic alloplasts; however, the authors reported no infections, implant exposures or fractures (n = 42) over a 6-year period. The Ceratite was covered with a retroauricular fascial turnover flap and fullthickness skin graft (FTSG) harvested from the ilioinguinal region. The authors discouraged the use of other alloplast materials such as silicone, porous polyethylene (MedPor), and titanium mesh. In this author's experience, porous polyethylene would have better tissue integration than Ceratite (pore size 5 µm) due to a larger pore size (150 µm).

Framework positioning

Even the perfect auricular reconstruction will appear abnormal if placed aberrantly in reference to the surrounding tissues. Isolated microtia can be seen with normal adjacent structures (e.g. hairline). Patients with OAVS are challenging due to the malpositioned lobular remnant and a variety of severe mandibular and temporal bone asymmetries. Walsh et al. [22•] described a device to assist in framework positioning. They suggested creation of a three-dimensional wax model of the contralateral normal ear. A silicone mold is made attached to an acrylic positioner for intraoperative framework positioning (Fig. 6). Another method to ensure proper framework placement is to mark the lateral commissure, lateral canthus and normal ear position on a piece of X-ray paper, sterilize the template and use it on the operative field to assure positioning [10[•],23].

Prevention of complications in costal cartilage harvest

The microtia surgeon must strive toward minimizing chest wall deformity while harvesting the optimal costal cartilage for framework creation. Relatively high rates (16–63%) of chest wall deformity after rib harvest from a variety of surgeons were reviewed by Kawanabe and Nagata [24^{••}] and spawned an interesting harvest technique with special attention to maintenance of the entire Figure 6 Illustration of a novel device used to plan costal cartilage framework positioning



Source: Reproduced with permission from Walsh et al. [22*].

perichondrial sheath at the donor rib site. After carving the framework, the residual cartilage was finely minced and replaced into the perichondrial sheath. Using this technique (n = 270), no chest wall deformities were noted during follow-up (mean, 27 months). An increased occurrence of chest wall deformities in younger children was reviewed. Only patients that are at least 10 years old with a chest circumference of at least 60 cm at the xiphoid process were considered candidates for microtia reconstruction. This final point is certainly debatable, as some surgeons prefer to begin reconstruction as early as 6– 8 years of age.

During harvest of the costal cartilage, the surgeon can choose to harvest some or all of the perichondrium with the rib. Should the perichondrium be left in the donor site or on the framework? The ability of preserving perichondrium on the framework to add strength, limit resorption, and facilitate chondrocytic growth is a somewhat controversial topic. Some surgeons suggest preservation of the pericondrium on the medial side of the helical rim to prevent warping [10[•]]. Kim *et al.* [25^{••}] compared the warping of costal cartilage after carving concentrically (around the center) or eccentrically (carving only one side) and found that slow, concentric carving of the costal cartilage over a few hours may decrease warping of the graft. The applicability of this study to microtia is limited because the cadeveric rib was carved like a dorsal nasal implant without the contours of an ear framework. This author prefers to leave the perichondrium on the helical graft (floating rib), while preserving the anterior perichondrium on the graft from the synchondrosis.

The surgeon is often intent on obtaining the longest possible rib for the helix. In studying porcine rib, Snellman [26] found that 75% of longitudinal rib growth occurred at the costal chondrial junction, and therefore

Figure 7 A temporarily implantable, continuous bupivicaine infusion device





suggested that several millimeters of cartilage are preserved at the lateral harvest site. Another benefit of cutting on the cartilage side of the osseocartilaginous junction is a decreased likelihood of hematoma due to less bleeding from the bone.

Most surgeons advocate the use of intercostal nerve blocks after costal cartilage harvest to minimize pain, limit respiratory splinting, and to prevent associated atelectasis [27]. This author prefers a temporarily implantable, continuous bupivicaine infusion device (On-Q PainBuster; I-flow Corp., Lake Forrest, California, USA) (Fig. 7).

Microtia reconstruction with alloplast

Refinements in the technique of prefabricated porous polyethylene auricular implants have provided additional options. Medpor exhibits excellent biocompatibility, stability, tissue integration (due to the 150 µm pore size), and resistance to infection [28[•]]. The framework can be bent when heated to 82-100°C, carved to shape, and even affixed to additional pieces by welding with cautery. The lower two-thirds of the implant is placed into the skin pocket while the upper portion of the framework is covered with a TPF and skin graft (Fig. 8) [29^{••}]. A lobular transposition is performed after 3 months. The implant can be placed concurrent with a bone-anchored hearing aid (BAHA). Romo et al. [29**] reported a 4% complication rate in 250 cases over an 11-year period. Early implant exposure due to flap ischemia was successfully treated with soft tissue flaps if less than 1 cm of exposure and tissue integration was seen. Alloplast removal was only performed twice due to failed tissue integration.

Prosthetic versus surgical reconstruction

One of the most difficult decisions for the parents of a child with microtia and their surgeon is whether to

choose a prosthetic ear or surgical repair. Tanner and Mobley $[30^{\bullet\bullet}]$ suggested that the best results with prosthetics are seen when the ear is completely absent. The major advantage of a prosthetic ear is the capacity for remarkable anatomic replication using medical grade silicone mixed with the proper pigment to achieve a color match and translucency. A polyurethane liner is applied to enhance longevity [31]. The prosthodonist (a subspeciality of dentistry) or anaplastologist (an ocularist, medical illustrator, or orthotic specialist) can require 14 h of laboratory time to make a prosthetic ear [32].

Figure 8 Photograph of a high-density porous polyethylene framework



Framework in position with a temporoparietal fascial flap being turned over the upper third of the implant. A skin graft will be placed over the flap. Source: Reproduced with permission from Romo *et al.* [29^{••}].

Figure 9 A prosthetic ear





The prosthetic can be attached by adhesives or osseointegrated implants with the major contraindications to the latter being prior radiation therapy, which creates bone demineralization, because at least 3 mm of bone can secure at least three titanium implants (Fig. 9) [12]. Boneaugmentation techniques of the temporal bone have been reported to allow young children (1 year) to have sufficient bone for a bone-anchored prosthetic [33]. Close follow-up is imperative to address the varying degrees of inflammation around the titanium pins, mechanical trauma to the prosthetic or pins, and even tissue overgrowth [34].

A hot/humid climate, oily skin, and excessive hair are relative contraindications to adhesive application. The two options for adhesives are organic solvents and water based which are gentler on the skin and less degrading to the polyurethane prosthetic. Regardless of which adhesive is used, the lifespan of a prosthetic can be only 2-3 years due to color fading, delamination of the lining, and traumatic cracks. The global patient cost per prosthetic ranges from US\$2000 to US\$7000 [30^{••}].

The preschool age child may benefit from minimizing the social stigma of a congenital deformity by either a prosthetic ear or ear reconstruction with alloplast. The child would have to wait until age 6 (Brent) to 10 (Nagata) before sufficient costal cartilage can be harvested. Fortunately, the maturity of the external ear size approaches adult size in width at 6 (girls) to 7 (boys) and in length at 12 (girls) and 13 (boys) [35]. The size of the costal cartilage, alloplast, or prosthetic framework must be chosen taking into account the size of the contralateral normal ear growth. Creating a larger ear than the normal side with costal cartilage or alloplast in the hope of compensating for the growth of the normal ear is inadvisable. Growth of the costal cartilage framework has been reported [36]. In an effort to better understand why the auricle increases in size into advanced age, Ito *et al.* [37] measured 1958 patients of increasing age and found a rapid rate of enlargement in width and length until 15 years old with a gradual increase into the seventh decade for both men and women. Histologically, elastic fiber irregularity was found in the older ears. A smaller costal cartilage framework often has a better appearance due to the hyaline cartilage lacking elastic qualities.

Figure 10 Silastic cylinder with arranged costal cartilage grafts



Siegert uses this silastic cylinder with arranged costal cartilage grafts which will be reimplanted into the rib graft site until it is used in the second stage of a combined microtia/aural atresia repair. Source: Reproduced with permission from Siegert [41].

Combined hearing restoration and microtia reconstruction

While bilateral microtia often requires early intervention for children with either BAHAs or canaloplasty, surgical correction of unilateral atresia is controversial. Favorable features for atresia repair may include a thin atresic plate, favorable pneumatization, a normal ossicular chain with sufficient audiometric cochlear function, and a good facial nerve course. Cholesteatoma may also lead to early intervention [38]. Better development of the middle ear was found with less severe microtia in a series (n = 109) by Ishimoto *et al.* [39[•]]. Aural atresia repair can be performed at the time of framework

Figure 11 A prefabricated ear

elevation (Brent stage 3) or a BAHA can be placed when sufficient temporal bone thickness is achieved $[10^{\circ}]$.

A previously published report [40] of a novel technique using combined atresia and microtia repair to prevent meatal stenosis and reduce the number of surgeries in children with microtia has been updated [41]. After costal cartilage framework placement and lobular transposition (Brent stage 1 and 2), a silastic cylinder is wrapped in thin rib cartilage grafts to prefabricate the ear canal (Fig. 10). A tympanic membrane is created from remnants of microtic cartilage that are placed around a specially designed



(a) A skin graft is wrapped around a silastic cylinder to insert into the canalplasty during the Seigert stage 2 procedure. (b) Three results of the canalplasty and microtia repair are shown. Source: Reproduced with permission from Siegert [41].

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silastic mold. These molds are stored in the thoracic wound until the canalplasty and framework elevation.

Six months later, framework elevation with a rib graft (Brent Stage 3), TPF flap, and FTSG is followed by canalplasty with ossicular reconstruction. The prefabricated ear canal is harvested from the thorax, positioned in the new meatus, and lined with a skin graft wrapped around a smaller silastic cylinder (Fig. 11). In the last stage, a FTSG is used in a meatoplasty. Excellent microtia reconstructions were shown. Postoperatively, the majority (76%) of patients had an air-bone gap of 30 dB or smaller, which is consistent with or better than other reports (Table 1) [41–55]. In a commentary of this article, Jahrsdoerfer [56] suggested several areas of concern which include a high rate of removal of the malleus/ incus complex, use of multiple silastic implants, and a lack of hearing results that include the average preoperative bone conduction threshold of 10 dB. When adding this to the average postoperative air-bone gap (26 dB), only 59% had attained a pure tone average of 35 dB.

Cho and Lee [18[•]] also described a two-staged atresia repair and microtia reconstruction (n = 27). Audiometric improvement was reported without reference to air-bone gaps, but showed pure tone average improvement in all but five patients. Complications included chronic drainage (n = 3) and meatal stenosis (n = 6).

These reports of successful concurrent microtia and atresia repair involve novel surgical techniques, implant materials, and a thorough understanding of the complex otologic and reconstructive principles of the external and middle ear structures. Additionally, successful cochlear implantation was recently reported in a child with microtia, aural atresia, and profound, bilateral sensorineural hearing loss [57^{••}].

Table 1	Comparison	of results	of c	congenital	atresia	repair	in
special	ized centers						

Reference	No. of patients	Acoustic transmission <u> </u> 30 dB, <i>n</i> (%)
Meurman [42]	48	12 (25)
Crabtree [43]	23	12 (52)
Gill [44]	83	28 (34)
Bellucci [45]	47	24 (51)
Jahrsdoerfer [46]	20	11 (55)
Gerhardt [47]	78	23 (30)
Cremers et al. [48]	53	26 (49)
Lambert [49]	15	10 (67)
Schuknecht [50]	56	30 (54)
Molony and de la Cruz [51]	22	16 (72)
Shih and Crabtree [52]	30	3 (10)
Teunissen and Cremers [53]	4	1 (25)
Schwager and Helms [54]	70	39 (56)
Lambert [55]	46	23 (50)
Total	595	288
Average		48%
Siegert [41]	37	28 (76)

Source: Reproduced with permission from Siegert [41].

Tissue engineering

Advances in microtia reconstruction using a tissue-engineered auricle have made this application of basic science to clinical practice even more promising. Previously, a scaffold of polyglycolic acid (PGA) was used to grow the autogenous chondrocytes, but a foreign body reaction was seen after implantation into an immunocompetent rabbit [58[•]]. Kamil *et al.* [59] created a human-sized auricle by replacing the PGA with hydrogel (Pluronic F-127, BASF, Mount Olive, New Jersey, USA) in a porcine model. A perforated, pure gold ear mold was used because of its strength and inert qualities. In a subsequent article [60], chondrocytes harvested from microtic human ears were grown in a mouse model, which adds future promise.

Conclusion

Surgical creation of the fine distinctions of highlights and shadows of the external ear continues to be extremely challenging. In the nearly 50 years since Tanzer [61] introduced costal cartilage microtia reconstruction, many advances have been described. Novel insights into combined aural atresia/microtia reconstruction, boneanchored prosthetics, BAHA, and alloplast materials have provided numerous options to patients and practitioners. The future of translational research appears bright as pursuit of implantable, tissue-engineered auricular frameworks continues.

References and recommended reading

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Additional references related to this topic can also be found in the Current World Literature section in this issue (pp. 443-444).

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