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Atresia Repair Before Microtia Reconstruction: Comparison of Early With Standard Surgical Timing

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Objective: To compare short-term results of atresia repair when performed before versus after microtia reconstruction.
Study Design: Retrospective case review.
Setting: Tertiary otologic referral center.
Patients: Congenital aural atresia with or without microtia: 70 cases over 24 months.
Intervention: Atresia repair before Medpor microtia reconstruction (ARM) versus atresia repair after microtia reconstruction with autogenous rib (ARR) versus atresia reconstruction without microtia (AR).
Main Outcome Measures: Surgical outcomes, short-term postoperative audiometric results (at least 4 months after surgery but within the first postoperative year), complications.
Results: Data from the 3 groups are as follows: ARM, 31 patients with median age 4.2 years (range, 2.5–9.3 yr); ARR, 28 patients with median age 12 years (range, 6.9–61); and AR, 11 patients with median age 5.9 years (range, 5.5–59 yr). Preoperative computed tomographic grading using the Jahrsdoerfer scale demonstrated an average score of 7.4 (range, 6–9) for the ARM group, 7.7 (range, 6–9) for the ARR group, and 8.5 (range, 8–9) for the AR group. For patients scoring 8 to 10 on the Jahrsdoerfer scale, postoperative pure-tone average 2 for each group were as follows: ARM, 28 dB hearing loss (HL); ARR, 32 dB HL; and AR, 29 dB HL. For patients scoring 7 or less, postoperative pure-tone average 2 were as follows: ARM, 42 dB HL; and ARR, 41 dB HL (AR, no patients). Surgical complications of infection and facial nerve injury were not seen in any group. Meatal stenosis was higher in the ARR group. One patient in the ARM group suffered a high-frequency sensorineural HL. No patient receiving Medpor microtia reconstruction suffered a complication due to the presence of the ear canal before microtia reconstruction.
Conclusion: Early results of ARM compare favorably with results achieved with atresia repair after microtia reconstruction with autogenous rib cartilage and with atresia repair without microtia repair. Hearing outcome and complications in this study are also comparable with previously reported expert results. Because restoration of binaural hearing has been shown to be advantageous for auditory development and function, timing of atresia repair can be considered before microtia reconstruction on an individual case basis, provided preoperative computed tomographic evaluation shows an adequate chance of surgical success.
Key Words: Microtia—Atresia—Congenital aural atresia—Medpor—Auricular reconstruction—Congenital anomaly.

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A parent whose child has atresia and microtia asked, "Plastic Surgeons make [microtic] ears look good while Ear Surgeons make ears work—why don't we restore hearing first and then work on the way it looks?" Good question.

Up to this point, surgeon mentors have taught hearing reconstruction in appropriately selected cases after reconstruction of the microtic outer ear. Utilizing traditional rib graft techniques, reconstruction begins at age 6 and requires several procedures (1–3). The period of time needed to reconstruct the microtic ear with a 3- to 4-stage technique most often takes 24 months and may span several years. This delay carries significant disadvantages for the hearing system in terms of function and, more importantly, in terms of its development. Surgeons and audiologists have bridged this gap with bone conduction hearing devices (implantable or nonimplantable), hearing aids or "benign" neglect of those ears affected with unilateral atresia. Still, others recommend unilateral atresia never be reconstructed if the contralateral ear is auditorily normal. With current practices, children are frequently left with suboptimal hearing during critical periods of auditory development and in demanding auditory environments such as the classroom. Conventional wisdom has implied this deficit has little impact on function or development.

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Most children who have undergone atresia repair after rib graft microtia reconstruction in our practice do so at 8 to 10 years of age at the earliest. At this point, a significant portion of the development of the auditory system has already occurred. Receptive and expressive language development proceeds rapidly in the first stages of life. Most of this development occurs by the age of 3 years and is nearly complete by the age of 5 years (4). Some complex auditory functions such as auditory processing continue to develop until the age of 10 to 12 years (5).

Previously, mentors and teachers have indicated that unilateral hearing impairment has little functional significance for patients and that late reconstruction of unilateral atresia has little impact on auditory development. Recent research (detailed in Discussion section), however, refutes this claim and has motivated us to reconsider surgical or amplification options for unilateral conductive hearing impairment associated with atresia/microtia. Reasons cited for the surgical dictum "microtia-repair first" include reluctance to disturb local blood supply in tissues used for rib graft microtia repair (B Brent, personal communication, 2007) and surgeon concern for potentially worse outcomes in terms of complications and hearing results of atresia repair in younger children. Recent techniques of microtia reconstruction utilizing the temporo-parietal fascia flap and Medpor reconstruction are independent of blood supply affected by atresia repair. This allows the potential for atresia repair to be performed before microtia repair when the techniques are coupled.

Recently, a growing number of parents and adults chose Medpor reconstruction for microtia repair (6). Atresia repair after Medpor microtia reconstruction and other synthetic auricular prostheses has been discouraged, by some surgeons because of the loss or exposure of the auricular prosthesis after creation of an ear canal in some cases (7). Requests from parents who desire for their child a functioning, hearing ear/s in addition to Medpor microtia reconstruction has fostered reconsideration of previous surgical sequencing, prompting us to move atresia reconstruction before microtia repair in selected cases. With this work, we seek to begin to understand outcomes of atresia repair surgery in the young child compared with that of older children. Given the early nature of these results, this report represents short-term follow-up. Results will need to be followed serially over time, and further reports are necessary to establish longer-term comparative analysis and results stability.

MATERIALS AND METHODS

Surgical Selection

This retrospective study was approved by the Western Institutional Review Board. The patients included in this study were diagnosed with either unilateral or bilateral atresia with or without microtia and were evaluated over a 24-month period. Only those patients graded 6 or greater on a preoperative scale (7) [via 0.3–0.5 mm coronal and axial computed tomographic (CT) scans through the temporal bone] and who went on to undergo surgery were included in the study group. Preoperative CT grading utilizing the Jahrsdoerfer scale (J-scale) demonstrated results for the 3 groups of atresia repair before Medpor microtia reconstruction (ARM), 7.4 (range, 6–9); atresia repair after microtia reconstruction with autogenous rib (ARR), 7.7 (range, 6–9); and atresia reconstruction without microtia (AR), 8.5 (range, 8–9). Preoperative and postoperative audiograms with air conduction, bone conduction, and speech discrimination scores were performed for audiometric evaluation. All preoperative audiograms were performed at the California Ear Institute, with most patients receiving at least 1 postoperative audiogram at an outside institution due to distance from our clinic.

Patients were required to be cleared for surgery from a medical perspective. After full discussion of potential risks and benefits, informed consent was obtained from the patient or the parents in the case of minors. No guardianship situation arose in the course of the study.

Surgical Technique

All surgery was ambulatory and was performed either at a community or hospital-based outpatient surgery center by a single surgeon (the senior author). After induction of general anesthesia, facial nerve monitoring utilizing the Medtronic NIM Response system was set up and tested. The postauricular skin was infiltrated with 1 to 2 mL of a mixture of 1% lidocaine with 1:100,000 epinephrine and 0.5% marcaine. The ear was prepped and draped for routine otologic surgery. Skin graft donor site (upper thigh) was prepped as well. A curvilinear incision was made 2 to 3 cm posterior to the microtic remnant. A periosteal incision mirroring the skin incision was made, and periosteum was elevated anteriorly to the level of the posterior aspect of the temporomandibular joint (TMJ). Drilling was initiated with a 4-mm cutting burr. The bone overlying the TMJ capsule was thinned, as was that of the tegmen. Gradually smaller burrs were used as the drilling proceeded medially. The level of the annulus was created slightly more medial than the lateral aspect of the ossicular mass. The atretic plate was then carefully separated from the underlying ossicular mass with sharp dissection and utilizing the potassium titanyl phosphate laser with pulse durations of 100 ms and 2 to 5 W intensity. The ossicular chain and, most critically, the incudostapedial joint were examined to ascertain anatomy and mobility and to identify areas of fibrous union. Stapes morphology and proximity to tympanic portion of the facial nerve was noted. (In the case of complete discontinuity, the lateral ossicular mass was removed and discarded; the remaining steps of atresia repair were completed, and ossicular reconstruction was scheduled as a second stage.) In those cases where the ossicular mass was fixed to the surrounding bone, the potassium titanyl phosphate laser was used to lyse this connection to allow for ossicular mobility. A temporalis fascia graft was harvested and placed onto the neoannulus as a lateral graft. A 4 $\times$ 6 cm, 0.013- to 0.015-inch split-thickness skin graft was harvested with a dermatome. The donor site was covered with gauze soaked in dilute epinephrine for hemostasis. The skin graft was folded into a longitudinal tube and placed as medially as possible. It was then smoothed onto the walls of the new canal, and the medial portion was carefully unfurled, slightly overlapping the temporalis fascia graft. A layer of chloromycetin-soaked gel foam was placed followed by a silastic ring, and then another layer of gel foam. The lateral portion of the skin graft was folded in upon itself, and the postauricular incision was closed in layers with

buried 3-0 Vicryl sutures. A meatus was created by incising a 270-degree near-circle, leaving the anterior portion of the skin intact. The skin was dissected off of the underlying soft tissue and elevated anteriorly. The intervening soft tissue was excised. The lateral portion of the skin graft was brought through the meatus and trimmed accordingly. The anteriorly based skin flap was sutured to the periosteum overlying the posterior aspect of the TMJ capsule. An Ambrus merocel pack was placed and inflated with chloromycetin. The skin graft was sutured to the F1 meatal skin with 5-0 chromic sutures (Fig. 1). A mastoid dressing was placed for 24 hours.

Postoperative Care, Examination, and Testing

The first follow-up appointment was 1 week postoperatively AQ1 F2 (Fig. 2). The Ambrus pack was trimmed by 0.5 cm to remove the hardened lateral portion and left in place. Twice daily ofloxacin drops were initiated and were used until all of the gel foam packing was out. One to two weeks later, the Ambrus pack and gel foam were removed. Two weeks later, the silastic and remainder of packing were removed. Dry ear precautions were enforced until the canal was fully epithelialized, a process that takes approximately 6 weeks. Patients were allowed to bathe and swim without restrictions thereafter. Audiometry was first performed 2 to 4 weeks after packing removal. During the first year, microscopic examination with cleaning is performed every 3 months and audiometry every 6 months. Thereafter, cleaning and audiometry are done every 6 to 12 months.

An adhesive clear dressing at the skin graft harvest site was removed after 1 week and replaced with daily nonstick dressings with antibiotic ointment for an additional week.

Data Collection

Before initial patient enrollment, data collection forms designed to begin with primary evaluation were created and used throughout the study period. Evaluation of each patient occurred at the time of initial evaluation, preoperative clinic visit, during surgery, 1 week after surgery, 3 weeks after surgery, 4 to 6 weeks after surgery, and between 6 and 12 months after surgery. Because this report focuses on short-term atresia outcomes, the last recorded audiogram at least 4 months after surgery but within the first postoperative year was used to calculate hearing outcome. For each audiogram, testing was performed at 500, 1000, 2000, 3000, 4000, and 6000 Hz for bone and air conduction. Masking as appropriate was used with standard techniques. Results are reported in compliance with the American Academy of Otolaryngology–Head and Neck Surgery Committee on Hearing and Equilibrium guidelines for evaluation of the results of treatment of conductive hearing loss (HL) (8).

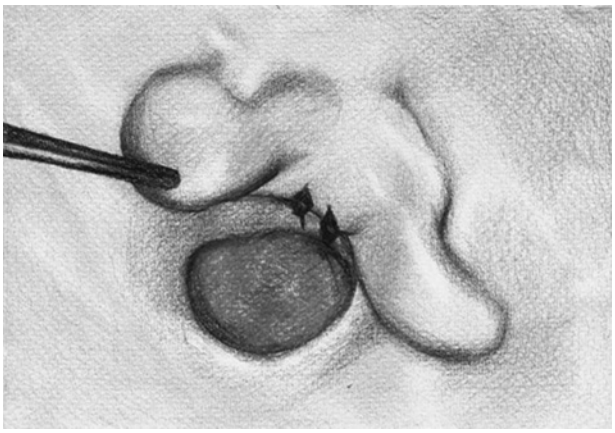

FIG. 1. Postoperative location of external auditory canal pre–Medpor microtia reconstruction.

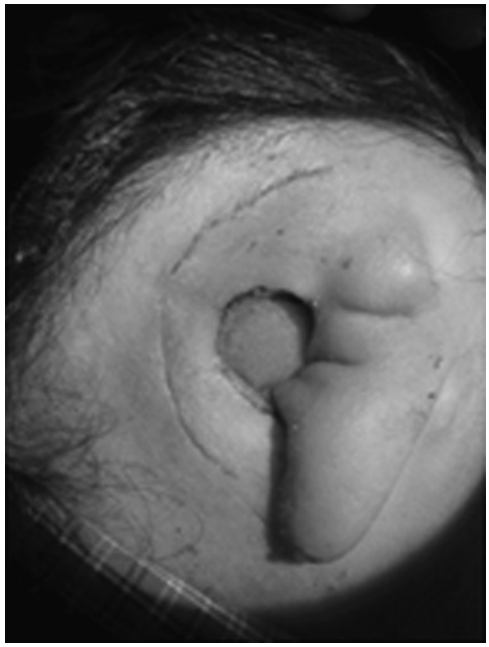

FIG. 2. Appearance of atresia reconstruction at 1 week postoperatively.

RESULTS

Study groups have been divided for analysis into 3 groups: ARM, 31 patients; ARR, 28 patients; and AR, 11 patients. Three patients (ARM-2, ARR-1) were found to have ossicular discontinuity intraoperatively. These patients were scheduled for a second-stage procedure 9 to 12 months after the initial procedure. Complete audiometric data on these patients is not yet available, and therefore they are not included in our results. Individual group ages varied with a median age in ARM of 4.2 years (mean, 4.5; range, 2.5–9.3 yr), ARR of 12 years (mean, 18.4; range, 6.9–61 yr), and AR of 5.9 years (mean, 19.8; range, 5.5–59 yr). There was no statistically significant difference in Jahrsdoerfer score between the

TABLE 1. *Short-term complications of atresia repair in the 3 study groups*

	ARM (31)	ARR (28)	AR (11)
Infection	0	0	0
Lateralized TM	1	1	1
Meatal stenosis	1	3	1
SNHL	1	0	0
Facial injury	0	0	0
TM perf	2	1	0

TABLE 2. *Audiometric outcomes for primary atresia cases: a review of the literature*

Lead author	Year	Cases	ABG criterion	Achieving ABG criterion, %	PTA/SRT criterion	Achieving PTA/SRT criterion, %
Cremers	1984	36			PTA < 35 dB HL	58
de la Cruz	1985	65	<30 dB HL	73		
Shuknecht	1989	62			PTA < 30 dB HL	50
Jahrsdoerfer	1992				SRT < 25 dB HL	70
Chang	1994	25	<30 dB HL	52		
Murphy	1997	20	<30 dB HL	65		
Lambert	1998	50			SRT < 30 dB HL	70
de la Cruz	2003	73	<30 dB HL	58.5		
Chang	2006	100	<30 dB HL	78.1		
Patel	2006	64	<30 dB HL	73	SRT < 30 dB HL	74
Digoy	2006	55	<30 dB HL	75	SRT < 30 dB HL	53
Roberson	2009	70	<30 dB HL	80 (1)/67 (2)		

(1) indicates preoperative CT grade 8 to 9 in the ARM group; (2), all patients in the ARM group with preoperative CT grade 6 to 9.

groups. Because this report focuses on short-term atresia outcomes, the last recorded audiogram at least 4 months after surgery but within the first postoperative year was used to calculate hearing outcome. Average improvement in pure-tone average 2 (PTA2) among all patients is 29.6 dB HL (SD, 11.9 dB HL). Average improvement in PTA2 across groups is as follows: ARM, 31.2 dB HL (SD, 15.74 dB HL); ARR, 28.0 dB HL (SD, 8.38 dB HL); and AR, 29.0 dB HL (SD, 1.4 dB HL). There are no statistically significant differences in postoperative PTA improvement between the groups. Patients in the ARM group with preoperative CT scores of 8 to 9 preoperative achieved ABG of <30 in 80% of cases. Across all patients in the ARM group, regardless of preoperative CT scores, an ABG of <30 was achieved in 67% of cases.

COMPLICATIONS

With the exception of meatal stenosis being higher in the ARR group, complications are very similar between the 2 groups in the short follow up period. Four patients experienced mild meatal stenosis. All of these were managed in the clinic with serial dilation and did not require return to the operating room. Table 1 lists those complications and their prevalence between the 3 study groups.

DISCUSSION

Often described as one of the most challenging otologic surgeries, aural atresia repair has benefited from general advancements in the field. The modern era of aural atresia surgery was ushered in by routine use of facial nerve electromyographic monitoring, CT grading, and the anterior canal plasty approach as pioneered by Jahrsdoerfer (7). Further modifications as suggested by Teufert and de la Cruz (9), including the use of a laser for dissection around the ossicles, split (as opposed to full)–thickness skin graft, and use of silastic and appropriately sized merocel in packing the ear canal, have further optimized outcomes. For primary cases, short-term success rates (ABG < 30 dB HL) improved from 42.2% to 66% over time with these modifications. Similar success rates have been achieved in other series, with Lambert reporting speech reception threshold (SRT) < 30 dB HL in 70% (10) and Chang et al. (11), Patel and Shelton (12), and Digoy and Cueva (13) reporting ABG < 30 dB HL in 78.1%, 73%, and 75%, respectively, whereas Jahrsdoerfer reports postoperative SRT < 25 dB HL in 75% of patients (7) (Table 2). The outcome in this series compares favorably with 80% of preoperative J-scale 8 to 10 and 67% of preoperative J-scale 6 to 9 cases achieving a postoperative ABG < 30 dB HL for those patients in the ARM group.

Short- and long-term complication rates should be considered when comparing earlier surgery to standard protocols. Short-term complication rates in this series are similar to those reported in the most recent literature and are not worse in the ARM group Table 1. The most prevalent complications resulting in revision are external auditory canal stenosis (7–18%), tympanic membrane (TM) lateralization (3.4–18%), and ossicular refixation (4%). The most concerning complications are sensorineural HL (SNHL) and facial nerve injury. Although there have been isolated cases of profound SNHL, most of the recent series' SNHL have been limited to the high frequencies (1.6–7.5%), as was the case in 1 patient in the ARM group in this series. With widespread usage of facial nerve monitoring, only several cases of temporary paresis have been reported, with facial nerve paralysis rare in experienced centers (9–13). This series had neither temporary paresis nor permanent injury of the facial nerve.

The reader will notice that the mean age of atresia repair in a subgroup of this report is significantly lower than past series. In addition to the change in parental decisions regarding microtia reconstruction previously mentioned, another impetus for this came from the mounting evidence mandating earlier intervention in children with significant unilateral and bilateral HL. In 2002, Sharma defined a "sensitive period" for the development of the central auditory system. Maximal plasticity occurs in the first 3.5 years and persists in some but not all until age 7 (14). Traditionally, atresia surgery has followed microtia surgery. With the rib graft technique, surgery is not initiated until age 5 to 6 to allow rib cartilage to reach adequate size and outer ear growth

to approach normal adult dimensions. Atresia surgery is therefore postponed until at least age 7 or 8 to allow for the multiple stages of rib graft microtia reconstruction to occur, potentially bypassing this critical period of auditory development.

Management of the patient with unilateral atresia has long been a subject of debate and can add an interesting perspective when considering timing of atresia repair. Three major schools of thought predominate. The first group advocates surgery only when the patient experiences a complication, such as facial nerve paralysis, cholesteatoma, or otorrhea. Otherwise, the decision for treatment is postponed until the patient reaches the age of consent and can fully comprehend the risks for the surgery. The second group supports intervention only with a very thin atresia plate and otherwise optimal anatomy. The third group believes in using the same timeline and candidacy criteria as with bilateral atresia patients. We support the last position for several reasons. Early in the history of aural atresia repair, results were inconsistent and morbidity rates were high. With the advent of more modern techniques, this is no longer the case. Secondly, evidence supporting the importance of binaural hearing continues to accumulate. Children with a unilateral HL have disadvantages in hearing in noisy environments and in localization (15). In addition, they are 10 times more likely than their binaurally equipped peers to fail a grade, have more behavioral problems, and have a greater need for educational assistance (16). The recommendation for nonintervention because "1 good ear is sufficient" is no longer justifiable in cases with favorable CT grading preoperatively, in our opinion. A significant number of surgeons today have been taught by mentors who considered surgery on a patient with unilateral atresia a violation of the "first, do no harm" principle. We propose re-examination of this recommendation in the light of present day knowledge of auditory development. It appears such a strategy constitutes a decision that may prevent normal auditory development in some patients.

Bone-conduction hearing aids can allow relatively normal function in quiet allowing time to pass until larger dimensions and adequate growth allow surgery at a later age. Bone anchored devices remain a very important adjuvant to auditory function in many children with atresia. In many instances where surgery is not an option based upon preoperative CT scans or medical condition and also in some patients whose surgical repair did not produce adequate results, use of a bone anchored device remains an important mechanism of hearing restoration. We recommend bone-conduction devices (usually in the form of a Bone-anchored Hearing Aid softband device) as soon as possible after diagnosis of atresia. This device is used before surgical correction of the congenital defect or until conversion to a more permanent bone-anchored solution (such as a Bone-anchored Hearing Aid). Some auditory processing functions require input from 2 independent ears (directional sound, hearing in noise, auditory processing), and bone-conduction devices would not be expected to provide the timing and loudness cues necessary for this auditory function and may hinder normal development.

Two other concerns have surfaced over the years regarding operating on a young child. First, the child's ability to cooperate with postoperative care has been questioned. In our experience, good patient rapport coupled with the insensate nature of the new ear canal results in a cooperative patient in most cases. All patients in this study have been able to complete packing removal in the clinic during the postoperative period, except for one 5 year old with significant cognitive impairment from birth. In addition, incomplete development of the mastoid cavity has also been cited as a deterrent to surgery in younger children. Studies from the cochlear implant literature indicate that the mastoid pneumatization is at 60% at age 2.5 years (17). With modern "anterior" techniques, the mastoid is not included in canal creation and properly selected cases allow enough anatomic real estate to create an adequate ear canal. This series shows an adequate sized canal may be created in younger children, allowing results similar to children and adults who have atresia surgery at older ages. More germane to the success of atresia surgery is that the middle and inner ears are essentially adult-sized at birth, as is the facial recess (18).

Atresia results have been shown to deteriorate in patients over time (10). The results in this article will in all likelihood do the same. Therefore, the conclusions of this study should be viewed as short-term only. Perhaps, no other surgical procedure performed in modern otology practice shows as much deterioration tendency. Although this may be discouraging, it also challenges the field to make further strides to improve long-term results with new and innovative techniques and practices.

Although use of rib cartilage microtia has long been the gold standard for microtia repair, the newer technique of Medpor porous polyethylene framework has become increasingly popular among our patients over the last few years. Medpor auricular reconstruction requires fewer procedures and avoids the morbidity associated with rib cartilage harvest. Use of alloplastic auricular implants is often met with caution because of historically inconsistent results with silicone. Medpor implants have been increasing in use for auricular reconstruction since the 1990s. When used with a vascularized temporoparietal fascia flap and full-thickness skin graft covering, excellent aesthetic results have been achieved with low complication rates. In 1993, Wellisz (19) reported on 41 patients receiving Medpor auricular reconstruction for traumatic and congenital auricular deformities. There were 5 implant exposures, which were all managed without removal of the implant and went on to heal uneventfully. Romo et al. (20) reported their experience with 250 ears in 2006, reporting a complication rate of 4% with total loss of framework in 2 patients. This evidence, in addition to the experience of 2 of our authors (J.R. and S.L.) and the satisfaction of our patients who have had Medpor auricular reconstruction, have impelled us to endorse this procedure as a legitimate option for microtia

repair. Long-term complication rate data is needed for this procedure, and this should be emphasized when counseling patients.

CONCLUSION

In properly selected patients, surgical outcomes of atresia repair performed at an early age before microtia reconstruction compare favorably to surgery performed in older patients with rib graft microtia reconstruction technique or to atresia repair performed alone. Atresia repair before Medpor microtia reconstruction can be performed safely with short-term results at least as good as repair after rib graft microtia reconstruction in older-age patients. Atresia repair at an early age may have developmental and functional advantages compared with traditional surgical timing and may be considered as a viable option in selected candidates. Long-term comparison of surgical intervention at an early age versus later timing is needed to investigate the stability of surgical outcomes.

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