INTRODUCTION

Anomalies of the external ear and hearing apparatus are seen in 66–99% of patients diagnosed with craniofacial microsomia with variabilities in severity.1,2 Treatment for patients with microtia and canal atresia requires consideration for the external ear deformity as well as rehabilitation of hearing loss.

Autologous rib cartilage is the most common material for external ear reconstruction due to complete biocompatibility and low infectivity.3–5 Although variations of surgical techniques have been reported, modern reconstruction generally requires 1 or 2 major stages.
harvest of rib cartilages, sculpting of the cartilages into a well-defined ear, and a thin, albeit well perfused, skin flap. Beyond the conceptual generalities, the details of the cartilage constructs and skin incisions are dependent on the individual deformity. Firmin classified cartilage constructs into 3 major types: I, full construct containing all anatomical components (including helical rim, antihelix, tragus, anti-tragus, lobule), II, construct without the tragus, or III, construct without the tragus or anti-tragus. Similarly, skin incisions were classified into 4 major types: I, Z plasty incision, II, transverse (transfixion) incision, IIIa, incision over the vestigial cartilage for 1-stage reconstruction, and IIIb, incision away from the vestige. Such classifications account for differences in cartilage deformities, skin availability, and location of the remnant. Drawbacks of autologous ear reconstruction include steep learning curve, disruption of the costal anatomy, revisions, and variable long-term resorption, which may affect the final aesthetic outcome. Aesthetic results in autologous ear reconstruction are, thus, challenging to achieve causing significant frustration to surgeons.

Timing of autologous ear reconstruction requires consideration of the external ear development, psychosocial development of children with craniofacial anomalies, and the availability of cartilage for reconstruction. Farkas’s seminal work in characterizing the external ear growth from birth to maturity demonstrated that the external ear width reached nearly the mature size in males at 7 years and females at 6 years, whereas length reached 86.6% of the mature size by 5 years. In terms of psychosocial ramifications, we and others have reported that significant psychosocial disturbances occur specifically in children with craniofacial anomalies between 8 and 10 years of age including increased anxiety, depression, poor peer relationships, obsessive-compulsive behavior, and aggression. For these reasons, many surgeons will aim to perform reconstruction between ages 6 and 8 years.

In patients with microtia, both conductive and sensorineural hearing loss occur in excess of the general population secondary to external auditory canal atresia and nerve abnormalities. Thus, treatment of microtia and atresia requires both reconstruction of the external ear as well as addressing the functional hearing impairment. Aural atresia is frequently found in combination with fusion of the malleus and incus and varying deformity of the ossicles. Treatment of hearing impairment can be accomplished nonsurgically, with external hearing aids, or surgically. Surgical correction includes canaloplasty, reconstruction of the tympanic membrane, and potentially implant placement in the presence of malformed ossicles. However, when the facial nerve path is anomalous, canaloplasty may not be an option. In such patients, a bone-anchored hearing aid (BAHA) is generally recommended. One limitation in the current literature on microtia reconstruction is the lack of information on outcomes with regard to treatment of the external ear and hearing impairment as a system. In this study, we review characteristics and variables in patients with craniofacial microsomia affecting the external ear and hearing apparatus that affect 3 long-term outcomes measures: wound complications, total number of surgeries, and psychosocial development.

METHODS

Patients

Patients over 13 years of age with craniofacial microsomia treated between 2008 and 2014 at the University of California Los Angeles (UCLA) Craniofacial Clinic were included. Patients were excluded if unidentified operative interventions before entry into the craniofacial clinic were performed, other craniofacial syndromes were diagnosed, and if the patients were lost to follow-up before completion of surgical plan. Operative reports, clinic evaluations, photographs, and audiograms were reviewed to assess patient characteristics, surgical algorithm, and 4 outcomes measures: complications, surgical revisions, aesthetics, and psychosocial function. For aesthetic outcomes, 3 observers evaluated frontal and lateral photographs of all patients and rated the reconstructed ear on a scale of 3 with the following criteria: 3 (excellent)—all external ear anatomy well visualized, excellent projection, no revisions necessary; 2 (moderate)—major external ear anatomy visualized, reasonable projection, minor revisions may improve outcome; 1 (poor)—abnormalities in multiple anatomical components of the ear, major revisions necessary. Psychosocial function was derived from multidisciplinary records that detailed patient or parent-reported academic performance, relationships with family and friends, depression, anxiety, and anger. Function was rated as “good” or “poor.”

Treatment Algorithm

All patients started ear reconstruction between ages 5 and 18 years. First-stage ear reconstructions were performed using a modified Nagata/Firmin technique as described previously. Six months or more after first-stage reconstruction, the cartilage construct was elevated using banked costal cartilage. In a number of patients, we used a temporoparietal fascial flap for elevation. Minor revisions such as deepening of the conchal bowl, tragal revisions, lobule revisions, and deepening of the postauricular sulci occurred frequently following the completion of the primary stages of reconstruction. For hearing loss, all patients were recommended hearing aids or frequency modulation systems on initial evaluation. Correction of aural atresia with canaloplasty occurred following the second stage of microtia reconstruction and frequently in conjunction with ear revision. Placement of osseointegrated Baha occurred in the event that a canaloplasty could not be performed, recurrent stenosis of reconstructed canals, or patient/family preference.

Statistical Analyses

Statistical analyses were performed using SPSS V.23 (SPSS, Inc., Chicago, Ill.). Descriptive statistics were reported. For total number of postoperative complications and number of surgeries, simple, robust negative binomial regression analyses were used to evaluate predictors for count
In 5 patients with grade II ears, reconstructions were performed due to the low severity of the deformity; and 75.3% of external ear abnormalities were observed. 24.2% of patients had bilateral microtia, usually of varying severity; and 75.3% occurred on the right side in unilateral cases; 63.8% of patients with microtia were male, and 91.2% had auricular abnormalities (Table 1).

Using the Firmin classification system to describe cartilage constructs and incisions in ear reconstruction, 35.7% of patients (n = 56) had exposure, 2 (3.6%) infections, and 10 (17.9%) complications resulted from ear reconstruction. When each stage was separated, first-stage ear reconstructions resulted in a 17.9% complication rate and second-stage reconstructions resulted in an 18.0% complication rate. Two of the second-stage procedures utilized porous polyethylene for elevation, and these cases were excluded from the complication rate for autologous reconstructions. Of note, both these cases resulted in extrusion, infection, and removal of the porous polyethylene. Reconstruction revisions resulted in a small complication rate of 4.3%. None of the complications required complete removal of the cartilage construct.

Revisions occurred commonly after completion of ear reconstruction with 46 patients requiring a revision of the lobule, deepening of the concha, tragus, deepening of the helical root, or projection of the construction. In virtually all cases, each surgical revision was a combination of procedures for refinement of the final result.

Hearing Loss and Atresia Reconstruction in Microtia Patients

Fifty-four of the 62 patients (87.1%) with external ear abnormalities had documented hearing loss. Forty-four

### Table 1. Patient Characteristics

| Descriptor                  | N (%)   |
|-----------------------------|---------|
| Sex                         |         |
| Male                        | 42 (67.7) |
| Female                      | 29 (32.3) |
| Laterality                  |         |
| Right only                  | 30 (48.4) |
| Left only                   | 17 (27.4) |
| Bilateral                   | 15 (24.2) |
| Microtia severity           |         |
| Grade I                     | 10 (13.0) |
| Grade II                    | 7 (9.1)  |
| Grade III                   | 58 (75.3) |
| Hearing loss                | 54 (87.1) |
| Conductive                  | 44 (81.5) |
| Mixed                       | 10 (18.5) |

### Table 2. Surgical Complications

| Stage of Ear Reconstruction | N (%)   |
|-----------------------------|---------|
| First stage (n = 56)        |         |
| Age, mean years (range)     | 8.5 (5.5–18.7) |
| Cartilage framework, n (%)  |         |
| Type I (complete)           | 20 (35.7) |
| Type II (no tragus)         | 8 (14.3)  |
| Type III (no tragus, no anti-tragus) | 28 (50.0) |
| Skin incision, n (%)        |         |
| Type I (W plasty)           | 9 (16.1)  |
| Type II (transfixion)       | 36 (64.2) |
| Type IIIa                   | 5 (8.9)  |
| Type IIIb                   | 6 (10.7)  |
| Second stage (n = 52)       |         |
| Age, mean years (range)     | 9.5 (6.3–18.5) |
| Usage of fascial flap, n (%)| 22 (46.2) |
| Aesthetic rating, mean (range) | 1.81 (1.00–3.00) |

### Table 3. Surgical Complications

| Stage of Reconstruction | N (%)   |
|-------------------------|---------|
| After autologous first stage (n = 56) | 10 (17.9) |
| Exposures               | 9 (16.1) |
| Infections              | 2 (3.6)  |
| Complete removal of cartilage | 0 (0.0)  |
| After autologous second stage (n = 50) | 9 (18.0) |
| Exposures               | 8 (16.0) |
| Infections              | 1 (2.0)  |
| Complete removal of cartilage | 0 (0.0)  |

### RESULTS

**Patient and Surgical Characteristics**

Of the 68 patients who met inclusion criteria, 62 patients (91.2%) had auricular abnormalities (Table 1). In total, 67.7% of patients with microtia were male, and 63.8% occurred on the right side in unilateral cases; 24.2% of patients had bilateral microtia, usually of varying severity; and 75.3% of external ear abnormalities were categorized as grade III microtia.

A total of 75 ears were affected, of which, 10 were not surgical candidates due to the low severity of the deformity (grade I ears). One patient had an osseointegrated implant. In 5 patients with grade II ears, reconstructions were performed with suturing, local tissue rearrangement, or auricular cartilage grafts due to the lower severity. In 3 patients, reconstruction was not undertaken for unknown reasons. A total of 56 ears underwent total ear reconstruction using a modified Nagata/Firmin technique using autologous rib cartilage (Table 2). First stage of reconstructions occurred at an average of 8.5 years of age (range, 5.5–18.7 years). Using the Firmin classification system to describe cartilage constructs and incisions in ear reconstruction, 35.7% of patients received a type I complete cartilage framework, 14.3% of patients received a type II (no tragus) cartilage framework, and 50.0% of patients received a type III (no tragus, no anti-tragus) framework. A type II transfixion, utilized in 64.2% of first-stage procedures, was the most commonly used skin incision, followed by type I (16.1%) and type IIIb (10.7%). Four ears were completed in 1 stage due to the presence of adequate skin coverage.

Second stage of reconstructions occurred at an average age of 9.5 years (range, 6.3–18.5) with 1.25 years on average elapsing between the first and second stages. The majority of second-stage procedures were completed using cartilage to elevate the cartilage construct (n = 52) with Medpor used in 2 procedures as an elevation block. A temporoparietal or occipital fascial flap was used in 46.2% of the second-stage procedures. To determine aesthetic outcomes, 3 independent observers rated reconstructed ears on frontal and lateral photographs with a scale of 1–3 with higher scores denoting better outcomes. The mean summed score for the cohort was 5.56 (range, 3–9).

All complications resultant from ear reconstruction were secondary to wound complications with no evidence of any systemic complications (Table 3). When each stage was separately analyzed, first-stage ear reconstructions resulted in a 17.9% complication rate and second-stage reconstructions resulted in an 18.0% complication rate. Two of the second-stage procedures utilized porous polyethylene for elevation, and these cases were excluded from the complication rate for autologous reconstructions. Of note, both these cases resulted in extrusion, infection, and removal of the porous polyethylene. Reconstruction revisions resulted in a small complication rate of 4.3%. None of the complications required complete removal of the cartilage construct.

Revisions occurred commonly after completion of ear reconstruction with 46 patients requiring a revision of the lobule, deepening of the concha, tragus, deepening of the helical root, or projection of the construction. In virtually all cases, each surgical revision was a combination of procedures for refinement of the final result.

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| After autologous second stage (n = 50) | 9 (18.0) |
| Exposures               | 8 (16.0) |
| Infections              | 1 (2.0)  |
| Complete removal of cartilage | 0 (0.0)  |
patients (81.5%) had conductive hearing loss, and 10 patients (18.5%) had mixed sensorineural and conductive hearing loss (Table 4). Mixed hearing loss was bilateral in 70.0% of patients, whereas conductive hearing loss was only bilateral in 29.5% of patients. Of the patients with mixed hearing loss, 8 (80.0%) had grade III microtia, 1 patient (10.0%) presented with anotia, and 1 patient (10.0%) only had grade I microtia.

Hearing deficits were treated in 38 patients (70.4%), despite clinical recommendations for all hearing loss to be addressed. Among those treated for hearing impairment, 16 patients (42.1%) opted for nonsurgical treatment using external hearing aids and 22 (57.9%) underwent surgery to correct hearing loss. Canaloplasty was performed in 16 patients. Correction of aural atresia with canaloplasty followed the second stage of microtia reconstruction and frequently in conjunction with ear revision. In total, 17 BAHAs were placed in 12 patients. Placement of osseointegrated BAHAs occurred in the event that a canaloplasty could not be performed, recurrent stenosis of reconstructed canals, or depending on the wishes of the patient and family. Five patients underwent both canaloplasty and BAHAs during treatment; 2 patients had canaloplasty in 1 ear and Baha placement in the other, whereas the other 3 received a Baha after failure of canaloplasty to adequately correct hearing loss. The differences in revisions and complications between Baha usage and canaloplasty did not reach statistical significance.

To compare the pre- and postoperative hearing results, audiograms were analyzed using paired samples t test to determine the results of surgical intervention on speech reception threshold, the lowest intensity level (in decibels hearing level, dB HL) at which the patient can correctly identify 50% of common 2-syllable words. Significant difference in the presurgery (48.1 ± 16.2 dB) and postsurgery (36.3 ± 20.0 dB) scores were found (P = 0.03).

Predictors of Total Number of Surgeries, Complications, and Aesthetic Ratings

To elucidate factors that predict surgical outcomes, patient and surgical variables were utilized in negative binomial regression analyses for 2 count outcomes: total number of surgeries (Table 5) and complications (Table 6). Additionally, patient and surgical variables were utilized in Poisson regression analyses for another count outcome: aesthetic ratings (Table 7).

The majority of patient characteristics including gender, laterality, involvement of surrounding anatomy, hearing loss, and psychosocial problems did not demonstrate any association to numbers of surgeries, complications, or aesthetic ratings. Patients with more severe microtic presentations required more surgeries to achieve optimal results than did patients with less severe presentations (IRR, 4.350; 95% CI, 2.140–8.843; P < 0.001). In addition, higher severity was associated with a lower aesthetic rating (IRR, 0.763; 95% CI, 0.658–0.886; P < 0.001). However, patients with more severe microtia did not have higher rates of exposures or infections when compared with patients with less severe deformities (P = 0.193).

In terms of surgical characteristics, age at the time of surgery, types of cartilage constructs, types of incisions,
or treatment of hearing loss did not result in statistically significant differences in total number of surgeries, complications, or aesthetic ratings. Not surprisingly, total number of complications predicted total number of surgeries (IRR, 1.304; 95% CI, 0.642–2.647; P = 0.463). In addition, increased number of surgeries predicted lower aesthetic ratings (IRR, 0.954; 95% CI, 0.927–0.982; P = 0.001), but increased complications was not associated with a statistically significant difference in aesthetic ratings.

### Treatment of Hearing Loss Predicts Improved Psychosocial Outcomes

Nineteen patients (30.7%) had documented patient or parent-reported deficits in school performance and psychosocial function, including depression, anxiety, anger, or difficulties with peer interactions, during routine annual team evaluations. Thirty-nine patients (62.9%) reported good academic standing, mood, and social interactions. Data on psychosocial function and school performance were missing for 4 patients. Patient and surgical variables were analyzed in a univariate logistic regression to determine predictors of improved psychosocial outcomes (Table 8). Of all independent variables, treatment of hearing loss was the only statistically significant predictor of good psychosocial outcomes (OR, 4.889; 95% CI, 1.459–16.381; P = 0.010). Neither surgical nor nonsurgical treatment of hearing loss alone proved to be significant independently.

### DISCUSSION

In this work, we review our long-term experience treating microtia and aural atresia in patients with craniofacial microsomia focusing on 4 outcomes measures: number of surgeries, number of postsurgical complications, aesthetic outcomes, and psychosocial outcomes. The demographics and characteristics of the patients with microtia are similar to those previously reported in the literature.1,13,18 When we reviewed the surgical characteristics of the cohort, we found that age, severity of deformity, type of incision, or size of cartilage construct were not associated with postsurgical complications, which were uniformly related to exposure and infection. All complications were salvaged with either simple closure of the exposed area or a local, small fascial flap with skin grafting. None of the cases required complete removal of the cartilage construct. Two factors were significantly associated with increased number of surgeries: severity of the microtia and number of complications. None of the factors evaluated showed a statistically significant association to increased number of complications. However, both increase in microtia severity and increase in number of total surgeries were associated with decreased aesthetic ratings. In terms of psychosocial outcomes, our experience demonstrated that treatment of hearing impairment was the only statistically significant predictor of improved psychosocial outcomes.

The total number of surgeries to achieve the optimal aesthetic result in ear reconstruction is often determined by a variety of factors. The most obvious and objective factor is addressing complications such as exposure or infection resultant from previous stages of ear reconstruction. Subjective factors such as aesthetic satisfaction from the surgeon and the patient also contribute to increased number of revisions. From both our cohort and our experience, ear reconstruction frequently entails revisions after the 2 major stages for various reasons. In our cohort, the primary reason for revision was to increase projection. All complications were salvaged with either simple closure of the exposed area or a local, small fascial flap with skin grafting. None of the cases required complete removal of the cartilage construct. Two factors were significantly associated with increased number of surgeries: severity of the microtia and number of complications. None of the factors evaluated showed a statistically significant association to increased number of complications. However, both increase in microtia severity and increase in number of total surgeries were associated with decreased aesthetic ratings.
integration. However, the Nagata/Firmin technique frequently requires a sizeable construct, which may be difficult to acquire in young children. Furthermore, changes in the cartilage from resorption may also occur with time, thereby decreasing the likelihood of an aesthetic outcome. In our cohort, we have found no increases in surgical complications, reoperations, or improved aesthetic ratings when we performed regression analyses with age of reconstruction.

In our cohort of patients followed into teenage years, hearing loss was present in 87.1% of patients with microtia, the majority of which was conductive in nature due to middle ear deformities and canal atresia. In total, 18.5% of patients had mixed sensorineural and conductive hearing loss, which is a considerably larger number than that in other reports published. The larger proportion of hearing loss that is sensorineural in nature has important consequences in treatment, as cochlear implants may have an expanding role. Our study has noted that canaloplasties have a tendency to have somewhat higher revision rates than BAHA placement, due to restenosis. However, we find correction of the abnormal anatomy via atresia reconstruction to be superior to BAHA placement in that we and others have observed that compliance with external devices in children is low and diminishes with time.20

It is now well established by many investigators that psychosocial benefits occur following reconstruction of the external ear. Steffen et al. have demonstrated a clear benefit to psychosocial functioning following microtia reconstruction using rib cartilage; however, consideration of functional hearing was not reported in this series. Similarly, many of the larger series on microtia reconstruction do not discuss the combined outcomes following treatment of functional hearing loss and external ear deformities. With the recognized importance of binaural hearing, reconstruction of the appearance of the external ear and the impairment of the hearing apparatus requires consideration as a functional unit even when the anomaly is unilateral.25-33 Our work focuses on a cohort of patients who have been offered treatment for both external ear deformities and hearing impairment. Although one may expect that severity of the ear deformity, bilaterality, deformities of other associated structures, total complications, total number of surgeries, or aesthetic ratings may affect psychosocial outcomes, we found that none of these variables predicted psychosocial outcomes in a statistically significant manner. In fact, treatment of hearing impairment, regardless of the unilaterality or bilaterality, was the only factor that had any bearing on psychosocial outcomes.

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