The Laryngoscope

Systematic Review

A Meta-analysis of Surgical Success Rates in Congenital Stapes Fixation and Juvenile Otosclerosis

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Objectives: To assess published reports in the literature on surgical success rates in patients with congenital stapes fixation (CSF) and juvenile otosclerosis (JO).

Design: Systematic review of the literature and meta-analysis of published data.

Data Sources: PubMed, SAGE, MEDLINE, and Cochrane.

Review Methods: A literature search was performed and evaluated based on established criteria. Two independent reviewers (B.A., M.B.) inspected titles and abstracts of the studies. The full texts of the studies were examined to assess their relevance to the meta-analysis. An air-bone gap (ABG) of 0 to 10 dB hearing level was described as success of surgery.

Results: A random effects model was used to analyze the data. A total of 27 studies were included in the meta-analysis, whereas 934 were excluded. The total number of operated ears was 445 (256 ears with JO and 189 ears with CSF). At the time of surgery, the age of the patients ranged from 3 to 18 years. The success rate was 80.2% for JO and 54% for CSF. The overall success rate was 69.9%. The rate of postoperative sensorineural hearing loss was 3.4% for JO and 2.1% for CSF.

Conclusions: Overall, stapes surgery in children with JO or CSF has a moderate success rate (69.9%) to provide ABG closure of 10 dB. The success rate for CSF in the setting of ossicular abnormalities is 54%. This meta-analysis suggests that if JO can be determined before surgery, a higher success rate is possible (80.2%).

Key Words: Congenital stapes fixation, juvenile otosclerosis, stapedotomy, stapedectomy, success rate.

Level of Evidence: NA

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INTRODUCTION

Osteodystrophic changes localized to the bony labyrinth and footplate of the stapes are described as otosclerosis.[1] Clinical otosclerosis usually begins in early adult life and is observed bilaterally. Otosclerotic focus begins at the fistula ante fenestram in 70% to 90% of cases. Development and etiology of otosclerosis remains unclear, but family predisposition of autosomal dominant inheritance and acceleration during the pregnancy period are the epidemiologic common features.[2] In addition, endocrine factors, immune disorders, viral involvement, and connective tissue disorders have been proposed as potential causes of otosclerosis. The age of onset of hearing loss is usually occurs after puberty.[1,2]

Congenital stapes fixation (CSF) is a rare reason for conductive hearing loss in childhood. This clinical condition usually begins in the early years, and no family history and genetic predisposition has been identified. Early onset of hearing loss and other ossicular and facial nerve anomalies have been found much more frequently than in juvenile otosclerosis (JO). In most cases, fixation of stapes footplate is the reason for stapes fixation.[3,4]

Differential diagnoses of a child with conductive hearing loss include otitis media with effusion, suppurative otitis media, CSF, congenital cholesteatoma, ossicular chain anomalies, connective tissue disorders, and JO. These entities have different treatment methods. Childhood surgical treatments for CSF and JO are controversial.

Although many case reports and small series reports are available in the literature regarding surgical results in JO and CSF, it is challenging to find large case series to compare surgical outcomes. The aim of this article was to report the results of a meta-analysis of the literature to evaluate surgical results that may aid in conducting and reporting on future research of JO and CSF, which may be of assistance when counseling patients and their families.

DATA AND METHODS

Literature Research

A literature search was independently performed by two researchers (B.A., M.B.) who searched databases

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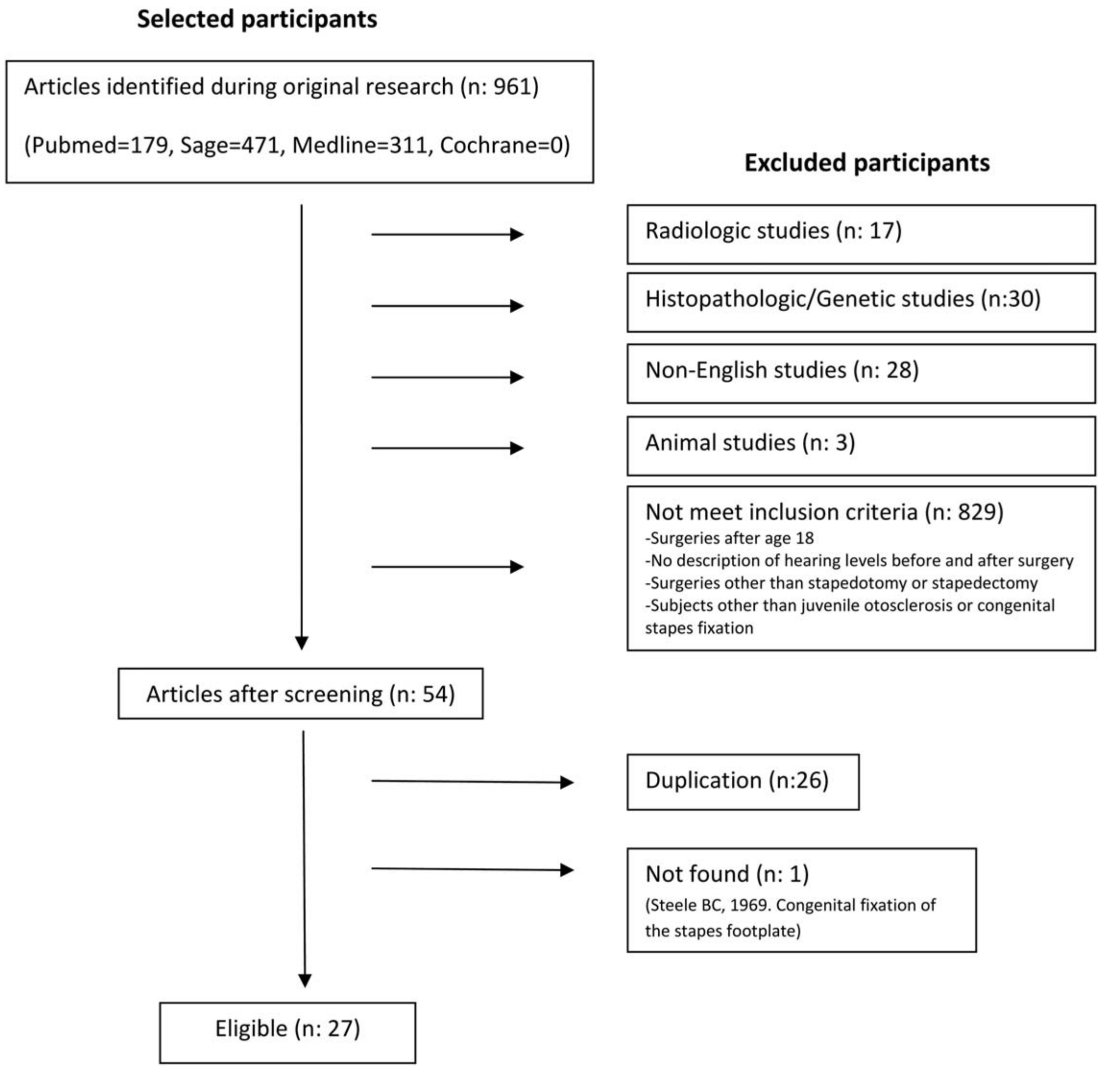

Fig. 1. Flowchart for selection of studies according to inclusion/exclusion criteria.

PubMed (January 1968 to June 2014), SAGE (January 1980 to June 2014), MEDLINE (January 1963 to June 2014), and Cochrane. The search terms used were "juvenile otosclerosis" AND "pediatric otosclerosis" [Medical Subject Heading terms] OR "congenital stapes fixation" [All Fields] OR "congenital stapes agenesis" [All Fields] AND "stapedectomy" [All Fields] OR "stapedotomy" [All Fields] AND "randomized controlled trial" AND "case report" AND "clinical retrospective trial" (ptyp). All data were evaluated by a third researcher (B.S.). Repetition of the same experiment was excluded.

Inclusion Criteria

Literature type. All published studies in international journals published in the English language addressing randomized controlled trials, case reports, and clinical retrospective trials specifying JO, CSF, and congenital stapes agenesis were included.

Subjects. JO, congenital stapedial agenesis, and CSF patients who underwent stapedotomy or stapedectomy were included. Those who underwent surgery after age 18 years were excluded.

Interventions

A literature search concerning stapedotomy and stapedectomy was performed for all age groups. Only the outcomes for patients <18 years of age were included in the meta-analysis.

Exclusion Criteria

Repeatedly published literature was excluded. Literature was also excluded if it met any of the following criteria: studies that included surgeries after 18 years of age, language other than English, no description of pre- and posthearing levels, subjects with conditions other than JO or CSF, surgeries other than stapedotomy or stapedectomy, genetic, radiologic, histopathologic studies, or animal experiments. If a study included patients both younger and older than 18 years old, only patients younger than 18 years were included; the other patients

TABLE I.
Articles on Cases With Juvenile Otosclerosis.

Study	No. (Sample Size)	No. (Postoperative ABG ≤10 dB)	Mean Preoperative ABG	Mean Postoperative ABG	Proportion (%)	95% CI
Zelenik, 2012	1	1	35	8	100.0	2.5–100.0
Huygen, 1996	2	2	30	NA	100.0	15.8–100.0
Robinson, 1983	35	35	NA	NA	100.0	90.0–100.0
Murphy, 1996	9	5	30	NA	55.6	21.2–86.3
Vincent, 2006	33	31	25.5	1.2	93.9	79.8–99.3
Schweitzer, 1984	1	1	40	10	100.0	2.5–100.0
De la Cruz, 1999	39	32	27.8	15	82.1	66.5–92.5
Hajek, 1961	2	1	37.5	NA	50.0	1.3–98.7
House, 1980	24	22	NA	NA	91.7	73.0–99.0
Raveh, 2002	2	2	30	NA	100.0	15.8–100.0
Carlson, 2013	17	12	27.5	8.8	70.6	44.0–89.7
Denoyelle, 2010	5	4	33.2	9.8	80.0	28.4–99.5
Lescanne, 2008	10	9	25.5	6.5	90.0	55.5–99.7
An, 2014	6	4	32.8	8.2	66.7	22.3–95.7
Lippy, 1998	60	55	32.6	0.4	91.7	81.6–97.2
Neilan, 2013	6	4	34.4	7.2	66.7	22.3–95.7
Welling, 2003	4	0	41	13.1	0.0	0.0–60.2
Total (fixed effects)	256	220	32.2 ± 4.8	8.0 ± 4.4	86.3	81.7–90.2
Total (random effects)	256	220			80.2	69.8–88.8

Proportion indicates the percentage of cases with postoperative hearing level improved to ABG ≤10 dB HL. Total random effects was used as a statistical model to analyze the data in this meta-analysis. Test for heterogeneity: significance level, $P < .0001$; I^2 (inconsistency) = 67.64%; 95% CI for I^2 = 46.36–80.47.

ABG = air-bone gap; CI = confidence interval; HL = hearing level.

were excluded. Thus, the n value for a given study included in this meta-analysis may have been different than the examined study's original n value. If ages of subjects were not described in a published study, the study was not included in the meta-analysis (Fig. 1).

Data Extraction

Two independent reviewers (B.A., M.B.) inspected the titles and abstracts of the studies and screened the selected literature according to inclusion and exclusion criteria. When no relevant word was found in titles and abstracts, the full text of the study was reviewed to assess its relevance to this analysis. Again, where disagreement occurred, we attempted to resolve it through discussion. The extracted data included author(s) name(s), publication year, sample size, interventions, number of cases at the time of diagnosis or surgery, outcome variables (number of reported patients, gender, postsurgical hearing level, success rates of surgeries), and follow-up time.

Main Outcome Measurements

The total number of diagnosed cases (JO, CSF) and success rates of surgeries (stapedectomy and stapedotomy) were detected. Main outcome variable assessment was a postoperative air-bone gap (ABG) of 0 to 10 dB hearing level (HL) (described as success of surgery). Mean ± standard deviation, median, and 25% to 75% quartiles (Q1–Q3) were calculated for extracted data. The outcome variables were proportion (%) and 95% confidence interval (CI) in groups with JO or CSF groups after surgery.

Statistical Analysis

Meta-analysis was performed using MedCalc version 12.7.2.0 (MedCalc Software, Ostend, Belgium). When $I^2 > 50\%$ and $P < .05$, heterogeneity was considered statistically significant, and a random effects model was used to analyze the data. To evaluate the efficacy of the surgeries for JO and CSF, 95% CIs were calculated. Sensitivity analysis was performed to identify any studies that may have influenced the results.

RESULTS

Studies Included in the Meta-analysis

Based on the inclusion criteria and exclusion criteria previously described, 27 studies were included in this meta-analysis, whereas 934 were excluded. Specifically, 10 studies related strictly to JO and 10 studies related only to CSF were identified and included in the analysis. Seven studies included patients with both entities. As a result, the meta-analysis included 17 studies for both the JO and the CSF groups. The total number of operated ears was 445. Not all of the studies provided gender information, but according to available data (24 of 27 studies), 192 patients (51.9%) were female and 178 patients (48.1%) were male. At the time of surgery, the

TABLE II.
Articles on Cases With Congenital Stapes Fixation.

Study	No. (Sample Size)	No. (Postoperative ABG ≤10 dB)	Mean Preoperative ABG	Mean Postoperative ABG	Proportion (%)	95% CI
Dornhoffer, 1995	10	5	37.4	11	50.0	18.7–81.3
Pappas, 1998	2	0	NA	NA	0.0	0.0–84.2
Wetmore, 2011	4	2	NA	NA	50.0	6.8–93.2
De la Cruz, 1999	44	34	35.2	15	77.3	62.2–88.5
Ueda, 1996	2	2	60	NA	100.0	15.8–100.0
Raveh, 2002	10	2	NA	NA	20.0	2.5–55.6
Carlson, 2013	27	9	34.1	17.2	33.333	16.5–54.0
Denoyelle, 2010	25	17	33.2	9.8	68.0	46.5–85.1
Takahashi, 1996	2	0	35	4	0.0	0.0–84.2
An, 2014	18	8	36	12.5	44.4	21.5–69.2
Kinsella, 1993	1	1	38	0	100.0	2.5–100.0
Martin, 2006	4	4	49.3	7.5	100.0	39.8–100.0
Sethi, 2005	1	1	45	10	100.0	2.5–100.0
Visvanathan, 2011	1	1	65	15	100.0	2.5–100.0
Kojima, 2014	1	1	21.3	8.8	100.0	2.5–100.0
Neilan, 2013	16	9	34.4	9.6	56.3	29.9–80.2
Welling, 2003	21	5	33.2	15.7	23.8	8.2–47.2
Total (fixed effects)	189	101	39.8 ± 11.5	10.5 ± 4.9	54.0	46.9–60.9
Total (random effects)	189	101			54.0	40.4–67.3

Test for heterogeneity: significance level, $P < .0001$; I^2 (inconsistency) = 67.14%; 95% CI for I^2 = 45.43–80.21.
ABG = air-bone gap; CI = confidence interval.

age of the patients ranged from 3 to 18 years of age. For either condition, the youngest patients were 3 years old.

Surgeries and Follow-up

We identified that 256 ears with JO and 189 ears with CSF received surgical treatment including stapedotomy, stapedectomy, and laser stapedotomy. Few studies provided information about the operation side, preventing a description of left or right preponderance. The mean follow-up time was 12.7 ± 1.2 months (median [Q1–Q3] = 3[2–14] months). The longest mean follow-up time was 79.5 months,[5] and the shortest mean follow-up time was 1 month.[6,7] In all cases, the longest follow-up period was 24 years in the case of one patient.[8]

Surgical Success Rates

The mean preoperative ABG for JO was 32.2 ± 4.8 dB and 39.8 ± 11.5 dB for CSF. After surgery, the mean postoperative ABG for JO and CSF were 8.0 ± 4.4 dB and 10.5 ± 4.9 dB respectively. For the targeted group of 256 ears, 220 (85.9%) ears in the JO group were improved to the desired ABG (≤10 dB) postsurgically (Table I). On the other hand, only 101 of 189 ears (53.4%) with CSF were improved to the desired ABG (<10 dB) after surgery (Table II). Overall, for 445 ears, 321 (72.1%) ears were improved to the desired ABG (≤10 dB) after surgery (Table III).

The success rate for the JO group was 80.2% (95% CI: 69.8–88.8) and 54% (95% CI: 40.4–67.3,) for the CSF group (Figs. 2 and 3). When both groups were combined to examine the overall effectiveness of surgery based on the postoperative ≤10 dB ABG, the success rate was 69.9% (95% CI: 58.2–80.5) (Fig. 4). The inconsistency for the JO and CSF groups was 67.6% and 67.1%, respectively.

Hearing Loss After Surgery

Postoperatively, the number of reported cases of hearing loss in the JO group was eight (3.4%) within 229 cases. Of eight with JO, four cases had poorer sensorineural hearing loss (SNHL) after surgery.[9–12] Profound hearing loss developed in the other four cases.[13–15] The number of postoperative SNHL cases after surgery for CSF was four (2.1%). No reports of cases with profound hearing loss were noted in the CSF group.

DISCUSSION

For this meta-analysis, 961 published studies were evaluated, and following the inclusion/exclusion criteria designed for this study, 27 studies were included in the review. Notably, the number of patients within each study varied considerably. The effects of sample size variation on total random effects (success rate) were recognized. Different random effects could have been calculated if the studies with small sample size were excluded; however, we thought that the most effective review would be achieved by including each study that met the aforementioned criteria for the meta-analysis. Thus, six single-case reports were included in this report

TABLE III.
All Articles on Cases With Juvenile Otosclerosis or Congenital Stapes Fixation.

Study	No. (Sample Size)	No. (Postoperative ABG ≤10 dB)	Proportion (%)	95% CI
Zelenik, 2012	1	1	100.0	2.5–100.0
Huygen, 1996	2	2	100.0	15.8–100.0
Robinson, 1983	35	35	100.0	90.0–100.0
Murphy, 1996	9	5	55.6	21.2–86.3
Vincent, 2006	33	31	93.9	79.8–99.3
Schweitzer, 1984	1	1	100.0	2.5–100.0
De la Cruz, 1999	83	66	79.5	69.2–87.6
Hajek, 1961	2	1	50.0	1.3–98.7
House, 1980	24	22	91.7	73.0–99.0
Raveh, 2002	12	4	33.3	9.9–65.1
Carlson, 2013	44	21	47.7	32.5–63.3
Denoyelle, 2010	30	21	70.0	50.6–85.3
Lescanne, 2008	10	9	90.0	55.5–99.7
An, 2014	24	12	50.0	29.1–70.9
Lippy, 1998	60	55	91.7	81.6–97.2
Neilan, 2013	22	13	59.1	36.4–79.3
Welling, 2003	25	5	20.0	6.8–40.7
Dornhoffer, 1995	10	5	50.0	18.7–81.3
Pappas, 1998	2	0	0.0	0.0–84.2
Wetmore, 2011	4	2	50.0	6.8–93.2
Ueda, 1996	2	2	100.0	15.8–100.0
Takahashi, 1996	2	0	0.0	0.0–84.2
Kinsella, 1993	1	1	100.0	2.5–100.0
Martin, 2006	4	4	100.0	39.8–100.0
Sethi, 2005	1	1	100.0	2.5–100.0
Visvanathan, 2011	1	1	100.0	2.5–100.0
Kojima, 2014	1	1	100.0	2.5–100.0
Total (fixed effects)	445	321	74.2	70.0–78.1
Total (random effects)	445	321	69.9	58.2–80.5

Test for heterogeneity: significance level, $P < .0001$; I^2 (inconsistency) = 82.09%; 95% CI for I^2 = 74.83–87.26.
ABG = air-bone gap; CI = confidence interval.

along with larger sample reports.[1,16–20] In addition to single-case reports, there were also eight studies with the total number of cases less than 10.[3,7,21–26]

Looking at the meta-analysis graph, we can see that small squares for each study are shown in different sizes depending on the sample size. Studies that reported more successful surgical results appear in the graph to be leaning to the right, whereas unsuccessful ones appear to be leaning to the left. The proportional value of individual studies reflect the size of the patient population reported within each study. In this regard, studies that appeared to most affect results of the meta-analysis include De la Cruz et al. (1999)[27] and Lippy et al.[8] (1998). On the other hand, smaller sample sizes and single-case reports result in a proportion value of zero.

The success rates of 80.2% for JO cases and 54% for CSF cases are notable. When both entities were considered together, the overall success rate was 69.9%. This value suggests the effectiveness of surgery that achieves a postoperative ABG ≤10 dB HL in children with JO or CSF. If we compare the two pathologies in terms of surgical success rates, the surgical outcomes for young patients with JO are better when compared to those pediatric patients with CSF. In the setting of JO, preoperative discussions with a patient's family regarding hearing outcomes may be made with more ease, given the results of this review. Preoperative discussion with a family regarding a child with CSF should take into account the lower success rates observed in this meta-analysis. Regardless, the diagnosis of JO versus CSF would likely depend upon the results of surgical exploration of the middle ear, and so the overall success rate of 69.9% should be included in presurgical discussions.

De la Cruz et al. (1999),[27] the largest sample in the meta-analysis, performed 83 primary and 12 revision stapedectomies. The mean age for primary cases was 12.6 ± 3.9 years. Age at onset of hearing loss was 10.2 ± 4.6 years for cases with JO ($n = 39$) and 3.0 ± 2.8 years for cases with CSF ($n = 44$). They reported that

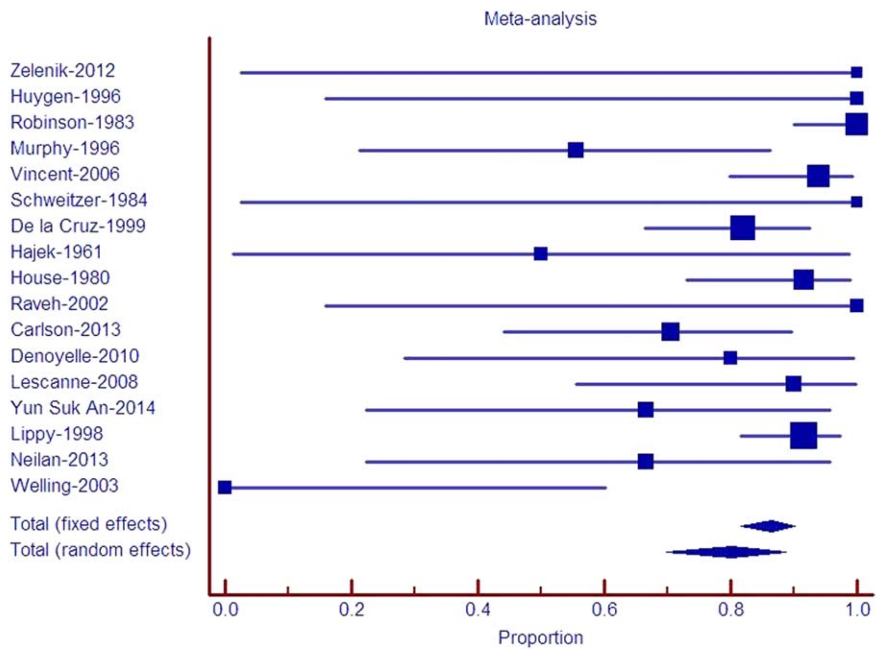

Fig. 2. Meta-analysis of articles on cases with juvenile otosclerosis. Differently sized blue squares indicate the sample size of each study. The studies including more successful surgeries in the figure are depicted over to the right (close to 1.0 or 100.0%), whereas unsuccessful surgical outcomes are depicted over to the left (close to 0.0 or 0.0%). Total random effects shows the general success rate. [Color figure can be viewed in the online issue, which is available at www.laryngoscope.com].

family history was found in only 9.8% of CSF cases and in 53% of JO cases. According to our meta-analysis, their success rate was 77.3% and 82.1% after primary surgery for CSF and JO, respectively. De la Cruz et al. concluded that primary stapedectomy in children was a safe and effective procedure, despite the fact that hearing outcomes in CSF were affected by coexisting middle ear anomalies.[27]

The next largest sample in this meta-analysis belonged to Lippy et al. (1998),[8] who reported satisfactory stapedectomy results in children with JO. They achieved an ABG of 10 dB HL in 55/60 ears (91.7%; n = 47 children). They also reported long-term results for 28 ears in their study. After 5 years or more (mean = 11.6 years), mean pure-tone averages showed deteriorations equal to 0.7 dB/year. This gradual decrease in hearing over time was reportedly similar to that in adults after stapedectomy.[8]

Lescanne et al. (2008)[11] performed laser stapedotomy in nine cases with JO and achieved an ABG <10 dB HL in 8/9 cases. These investigators stated that laser stapedotomy was technically successful, minimizing mechanical trauma to the stapes footplate while reducing postoperative complications. They stressed that stapedotomy carried out by an experienced surgeon, with or without laser, resulted in better postsurgical results.[11]

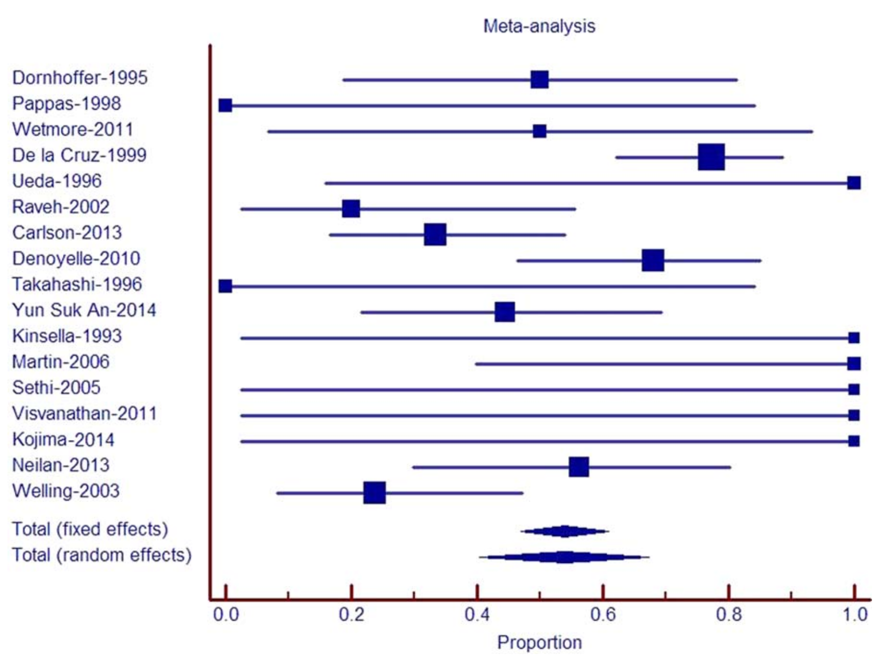

Fig. 3. Meta-analysis of articles on cases with congenital stapes fixation. [Color figure can be viewed in the online issue, which is available at www.laryngoscope.com].

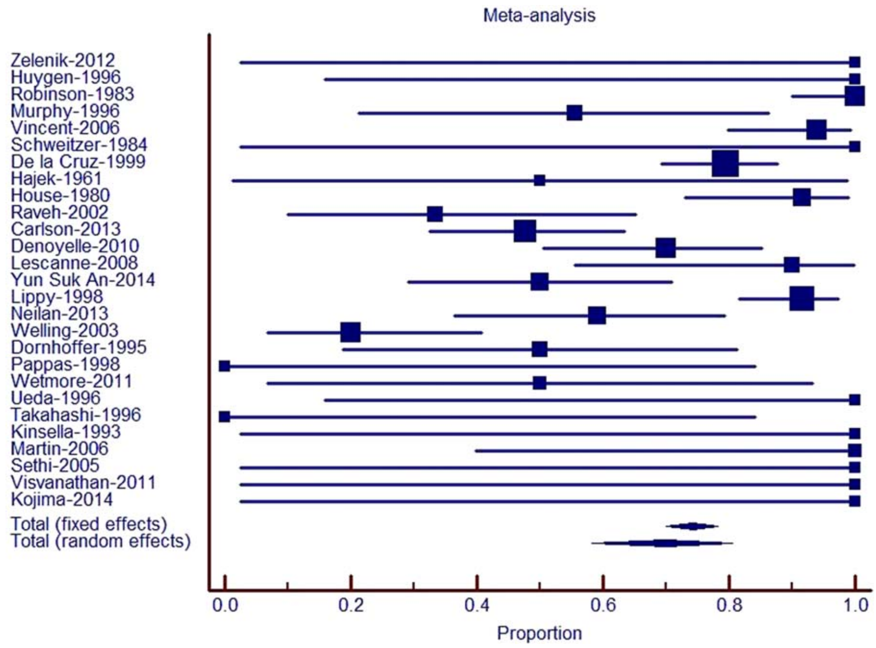

Fig. 4. Meta-analysis of all articles on cases with juvenile otosclerosis or congenital stapes fixation. [Color figure can be viewed in the online issue, which is available at www.laryngoscope.com].

Robinson (1983) performed partial or total footplate removal with vein graft used to seal the oval window in their patients with JO.[2] Criterion for success was closure of the ABG within 10 dB 6 months after the surgery. Of the patients under the age of 18 years, 77.6% had complete closure. Robinson reported no differences in hearing results between partial or total footplate removal in juvenile patients. He suggested that the degree of untreated footplate pathology may worsen with time, and that postoperative hearing results for juvenile patients might be expected to be better than for patients undergoing surgery after the age of 18 years. Robinson also reported that 80% of the juvenile patients had good cochlear reserve and no impairment of sensorineural function after stapes surgery.[2]

Several studies have demonstrated excellent results after stapes surgery. House et al. (1980) described hearing results after stapedectomy in children. Their results appear to be good as the hearing improvement seen following adult stapedectomy.[13] However, Von Haacke (1985) suggested that the use of personal hearing aids could provide satisfactory audition until a child was old enough to make a decision to undergo surgery.[28]

Early onset of otosclerosis was reported to be associated with more severe and diffuse pathology compromising stapes mobility.[29] Several studies, one of which was included in this meta-analysis, found higher prevalence of an obliterative type otosclerosis in JO.[5,30,31] However, no postoperative SNHL was reported, whereas postoperative ABG closure within 10 dB was found in 93.5% in patients with otosclerosis <18 years of age.[5] Although hearing results of the surgery for obliterative otosclerosis could be less satisfactory, Vincent et al. (2006) demonstrated hearing improvements with a four-frequency pure-tone threshold average ABG of 10 dB in their patients ($n = 5$). They also reported that long-term (mean 96.6 months) hearing outcome was also satisfactory in juvenile patients who underwent stapedotomy.[5]

CSF may be present with or without other ossicular chain or inner ear deformities. Pappas et al. (1998) reported three cases, all under the age of 13 years, who had CSF and round window atresia. Conductive hearing loss may present with round window atresia, which is difficult to distinguish from stapes ankylosis, or it could result from labyrinthine deformity.[3] House et al. (1980) also presented four conductive hearing loss cases with round window atresia.[13] Cerebrospinal fluid otorrhea is one of the major complications of stapes surgeries and has been reported in the setting of both otosclerosis and CSF. However, it is more frequent in patients with CSF.[6]

There may be concerns regarding the design and results of this meta-analysis that reflect the reported results of both larger and smaller case series. Importantly, it should be taken into account that success rates of experienced and less-experienced surgeons might differ. Postoperative stapes surgery results in children are in part related to the surgical expertise of surgeons, and pediatric patients with JO, CSF, or any other stapes pathology might be best cared for with referral to more experienced specialty care centers. Notably, the arbitrary target of ABG improved to ≤ 10 dB affected the so-called successful results. Adopting a postoperative ABG of 15 or 20 dB might have affected the results reported in this meta-analysis.

CONCLUSION

When considering the pooled data of reports in the relevant literature that met our inclusion/exclusion criteria, stapes surgery in children with JO or CSF appears to have an overall success rate (69.9%) described as an ABG closure ≤ 10 dB. If JO can be determined before surgery, families could be counseled regarding its higher postoperative rate of 80.2%, based on the results of this meta-analysis. Success rate for CSF is somewhat lower at 54%, perhaps owing to incidences of ossicular abnormalities.

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