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Review

Systematic Review of Round Window Operations for the Treatment of Superior Semicircular Canal Dehiscence

Waseem Ahmed, Rajini Rajagopal, Gareth Lloyd

Department of Otolaryngology, St. George's Hospital, London, United Kingdom (WA)
Department of Otolaryngology, John Radcliffe Hospital, Oxford, United Kingdom (RR)
Department of Otolaryngology, Guy's and St Thomas' NHS Foundation Trust, London, United Kingdom (GL)

ORCID IDs of the authors: W.A. 0000-0003-3832-1187; R.R. 0000-0003-0453-0845; G.L. 0000-0002-0174-2087.

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A review of the literature is presented to consider the role of round window (RW) operations in superior semicircular canal dehiscence (SSCD). Primary (PubMed) and secondary sources (TRIP, Cochrane database, Best Practice, and PubMed Clinical Queries) were used to identify relevant studies. Four original studies (three case series and one case report) were identified. All were retrospective reviews and used a number of subjective and objective outcome measures to assess the efficacy of a minimally invasive, transmeatal approach to perform RW surgery for SSCD. The current evidence suggesting that RW operations for SSCD are unlikely to replace more established surgical procedures as first-line treatment may be appropriate in a select group of patients. Further multicenter, randomized controlled trials are required to establish their efficacy in patients with SSCD.

KEYWORDS: Round window, superior semicircular canal dehiscence

INTRODUCTION

Minor et al. [1] of the Johns Hopkins University Hospital first described superior semicircular canal dehiscence (SSCD) syndrome in 1998. Prior to this, controversial theories postulated the presence of "spontaneous" perilymphatic fistulas and the supposed benefits of reinforcing oval (OW) and round windows (RW). Over the next few decades, middle ear exploration in thousands of patients to identify fistulas yielded less evidence of perilymphatic leak, and yet patients who underwent OW and RW repairs reported subjective improvements in their symptoms post-operatively [2].

Dehiscence of the otic capsule overlying the superior semicircular canal creates a direct interface between the membranous canal and the overlying dura-the third window (in addition to the physiological OW and RW). Sound or pressure changes transmitted through the OW via the stapes normally exit at the RW. However, the dehiscence creates a low-impedance pathway for this sound/pressure to dissipate through the labyrinth instead of the cochlea in SSCD. The resultant loss of energy for air-conducted sounds is illustrated as increased thresholds for hearing on pure tone audiometry. However, this mechanism permits bone-conducted sound to access the perilymph of the inner ear via the labyrinth, producing bony hyperacusis. The latter manifests as varying auditory symptoms (autophony, bone-conduction hyperacusis, pulsatile tinnitus, low-frequency hearing loss, phonophobia, and aural fullness) and vestibular phenomena (Tullio phenomenon, Hennebert's sign, oscillopsia, vertigo, and chronic disequilibrium).

SSCD is diagnostically challenging and requires multiple investigative modalities, such as pure tone audiometry, vestibular evoked myogenic potential testing, and computerized tomography of the temporal bone. Ward et al. [3] have proposed diagnostic criteria.

Surgical management of SSCD is reserved for patients in whom the symptoms are intrusive to daily living. Surgical repair requires a middle fossa, transmastoid, or more recently, an endoscopic approach. Techniques employed to repair the dehiscence include canal plugging, resurfacing, and capping. Not one repair technique (capping, plugging, resurfacing, or a combination thereof) has been shown to be statistically superior, even when surgical approach is taken into consideration [4]. The role of a transcanal approach and reinforcement of the RW or OW as a possible alternative to more established procedures has been reported. The aim of this review

Corresponding Address: Waseem Ahmed E-mail: waseem.ahmed6@nhs.net

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was to analyze the evidence for RW operations and whether they are effective in treating SSCD.

Search Strategy

Secondary Sources

An initial TRIP database search identified two primary articles that were included in the final analysis (Silverstein et al. [2] and Succar et al. [5]). Other secondary sources, such as Cochrane database and Best Practice, yielded no relevant results. PubMed Clinical Queries identified the same two articles noted in the TRIP search above, which was included in the final analysis.

Primary Sources

Primary search was conducted using EMBASE, Medline, and Cochrane Library (1950–September week 2 2018) (Figure 1).

Selection Criteria

Inclusion and exclusion criteria were pre-determined. Studies included in the review had to have the following:

Database	Time period covered	Subject headings and keywords
EMBASE on Ovid	1950-2018	Semicircular canal, round window, cochlea fenestra, superior semicircular canal, SSC, superior semicircular canal dehiscence, SSCD
Medline on Ovid	1980-2018	Semicircular canal, round window, cochlea fenestra, superior semicircular canal, SSC, superior semicircular canal dehiscence, SSCD
Cochrane Library	1990-2018	Semicircular canal, round window, fenestra cochleae, superior semicircular canal dehiscence, SSCD

Figure 1. Databases searched with the corresponding subject headings and keywords.

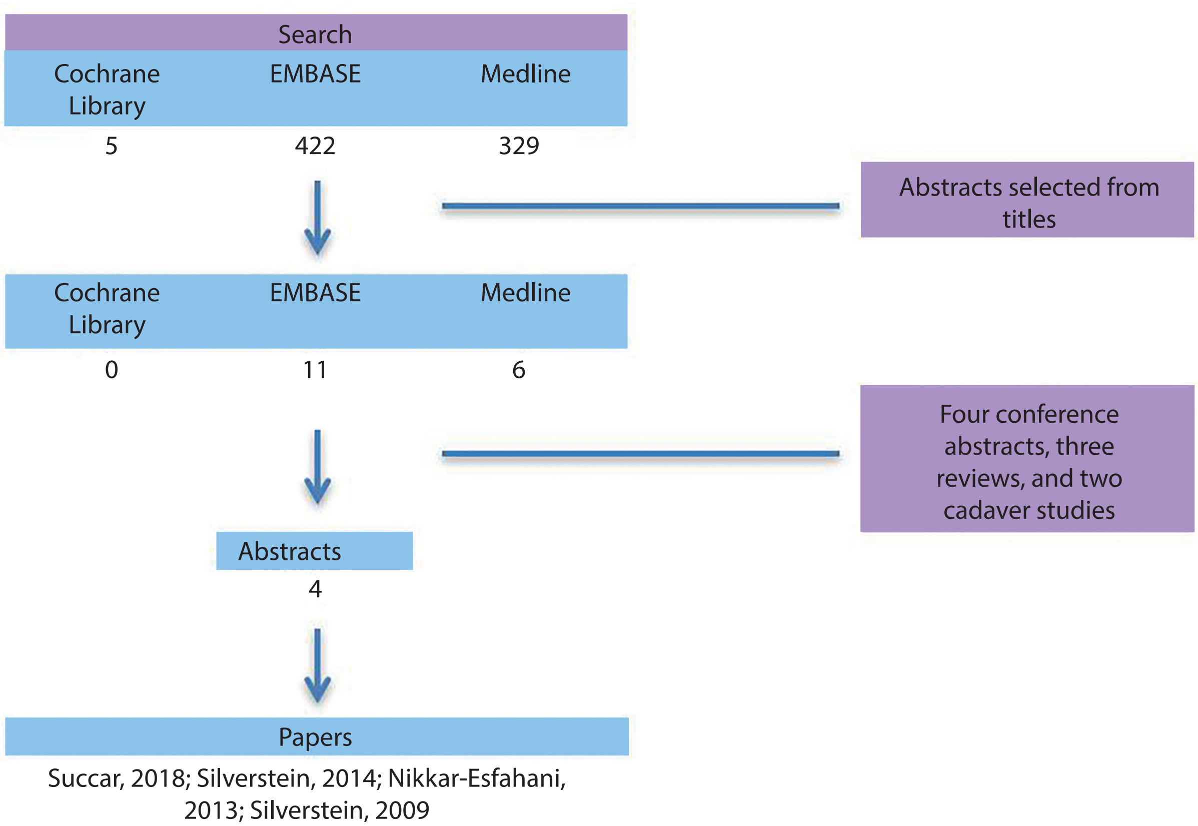

Figure 2. Flowchart showing the selection process.

1. a diagnosis of SSCD.
2. underwent surgical intervention in the form of round window obliteration (RWO) or reinforcement.

Studies were excluded for the following reasons:

1. patients were surgically managed in manners other than RW operations.
2. if written in a language other than English.

No restrictions were made on how the diagnosis was established, outcomes measures used, or study design. This enabled us to fully appreciate all the evidence published. The initial search was performed by the primary author (WA). Selection of studies included was performed by all three authors with disagreements resolved by consensus. A total of four studies were identified after the review of respective abstracts and papers (Figure 2). Three of the four studies selected were retrospective case series and one case report [2, 5-7].

Quality Assessment

We attempted to retain all the published evidence owing to the limited number of studies in this area. We attempted to qualitatively assess the merits and drawbacks of the selected studies and their relevance to the review question. We used guidance provided by Carey et al. to critique the selected studies [8].

Quality Assessment of the Evidence

Selection Bias

All the studies selected were retrospective and had defined their eligibility criteria. Although the number of participants was small in each study, they are likely to be representative of patients with SSCD.

Assessment Bias

Objective post-operative outcome measures were reported in three out of four studies (Succar, 2018; Nikkar-Esfahani, 2013; Silverstein, 2009) [5-7], with one study using a non-validated questionnaire to assess improvement (Silverstein, 2014) [7]. Assessor blinding was not relevant for any of the reported studies.

Attrition Bias

There were no missing data to account for in each of the selected studies.

DISCUSSION

A summary of the studies critiqued to assess the role of RW operations in SSCD are illustrated in Table 1. RW procedures are a surgical option for SCCD, dampening one of the three inner ear windows, leaving the OW and the dehiscence as the primary remaining windows. The major advantage is it is minimally invasive in nature. The literature search found four studies (three case series and one case report) that were relevant to the question.

The two RW procedures advocated for SSCD are RWO and RW reinforcement. Silverstein and Van Ess [7] suggested a move away from RWO despite demonstrating its success in their 2009 case report. They were unable to replicate this success in two subsequent patients and modified their technique to reinforce the RW instead [2]. However, the larger case series has significant limitations with multiple surgeons utilizing the same surgical technique but using different materials to reinforce the RW niche. Their use of parametric statistical analysis is also not explained (n=19). However, the study does discuss the role of RW and OW reinforcement in SSCD (utilized by one surgeon in the study, the number of patients was not specified), postulating that it would be safer and more effective than severely dampening the RW alone [2].

Two of the published studies (Silverstein and Van Ess [7] and Nikkar-Esfahani et al. [6]) both demonstrate the effectiveness of RWO in treating SSCD. The large case series by Succar et al. [5] is a lot more cautious in advocating this approach. It provides the most detailed analysis of pre- and post-operative outcomes, utilizing both subjective and objective outcomes. It advocates the use of RWO in select symptomatic patients who are not candidates for SSC plugging, resurfacing, or capping through a transmastoid or middle cranial fossa (MCF) approach (those with unilateral contralateral hearing loss, patients who have declined traditional surgery, or high anesthetic risk candidates).

Concerns regarding the exacerbation of conductive hearing loss have been reported in two cases [6]. These were described as mild. Cadaveric evidence suggests a modest, clinically negligible effect on conductive hearing loss secondary to RW reinforcement with perichondrium [9]. However, the results may differ clinically as scarring post-operatively may result in findings more consistent with cartilage in cadaveric experiments rather than the perichondrium. Although cartilage reinforcement resulted in a graded effect on stapes velocities (more marked at lower frequencies), the effect was still relatively small [9].

Although the evidence is limited, RW operations may be utilized as an initial lower risk, minimally invasive intervention. In some cases, patients often reported previously intolerable pre-operative symptoms. Although not eliminated post-operatively, they were sufficiently abated to not want further surgery [2]. For those who remain symptomatic, a limited number of patients who had revision surgery with plugging of the superior canal dehiscence via an MCF or transmastoid approach reported improvement in post-operative symptoms [10]. However, the follow-up period in this cohort of seven patients was short at 3 months.

The numbers of patients in the published literature undergoing RWO for SSCD are limited. Therefore, it is difficult to postulate why the procedure is successful in some and not in others. The variability in the materials and surgical techniques used may explain some of this variation. Selection criteria also varied between institutions and, therefore, are likely to be a contributing factor.

CONCLUSION

- Round window procedures may be a viable option for some patients with symptomatic SSCD.
- The procedure is low risk and may suffice in providing symptom relief without undergoing more invasive procedures.
- Oval window reinforcement in conjunction with RWO requires study to quantify any additional benefit.
- Further multicenter randomized controlled trials are needed to assess the efficacy of round window procedures and selection of appropriate surgical candidates.

Table 1. Studies describing RW operations for SSCD [2, 5-7]

		Summary			
Author (year)		**Succar et al. (2018)**	**Silverstein et al. (2014)**	**Nikkar-Esfahani et al. (2013)**	**Silverstein et al. (2009)**
Study type		Case series	Case series	Case series	Case report
No. of subjects		14 patients	22 patients	2 patients	1 patient
Clearly defined question?		Yes. Is RW plugging a suitable intervention for SSCD?	Yes. Is RW reinforcement (not obliteration) a suitable treatment for SSCD?	Yes. The role of RW occlusion for SSCD	No. Single reported case of RW occlusion
Study population	Diagnostic criteria	Clinical diagnosis of SSCD, but not defined	CT and symptoms	CT, VEMP, and symptoms	CT, VEMP, and symptoms
	Age (years)	Mean 56.9 (32-81)	Mean 48.4 (24-72)	59 and 42	37
	Gender	8 males and 6 females	12 females and 7 males	1 female and 1 male	1 male
	Laterality	13 single (10 left and 3 right) 1 bilateral	14 single (7 left and 7 right) 4 bilateral 1 unknown	2 single (1 left and 1 right)	1 single (1 left)
	Inclusion criteria	Age >18 years, SSCD diagnosis, RW plugging operation	Radiological evidence of SSCD and subjective symptoms	Diagnosis of SSCD	Diagnosis of SSCD
	Exclusion criteria	None	RWO (3)	None	None
	Post-operative follow-up	64-307 days	Not defined	2 years in one patient, undefined in the other	10 weeks
Described intervention		2 surgeons 1 institution Transcanal approach, tympanotomy, ± curettage medial EAC. RW identified, tragal cartilage graft and minced perichondrium. RW mucosal edges freshened, packed area with perichondrium and cartilage cap, with minced perichondrium on top	6 surgeons 4 institutions Transcanal approach, tympanomeatal flap, curettage to enlarge posterior auditory canal, view RW niche. One surgeon drilled RW overhang. Floor of RW niche denuded of mucosa. RW reinforcement achieved with one or more of temporalis fascia, tragal cartilage and perichondrium, fat loose connective tissue, gel foam, and silastic One surgeon reinforced both OW and RW. One surgeon used fibrinogen sealant to secure material on RW	Single surgeon 1 institution Permeatal approach. Tympanomeatal flap. RW niche identified and adjacent mucosal fold cleared. RW membrane identified and occluded with bone wax, muscle, and fascia, in three layers.	Single surgeon 1 institution Permeatal approach. Tympanomeatal flap with curettage of posterior medial bony canal. Bony overhang of RW niche removed. RW niche occluded in three layers—tragal cartilage first, bone wax, and then perichondrium

		Differentiates between RWO and RW reinforcement former excluded)		
Outcome measures	Non-validated 9-item questionnaire: 1. Pre- and post-autophony (9/14 improved) 2. Pressure-induced vertigo (8/13 improved) 3. Sound-induced vertigo (8/12 improved) 4. Hennebert's (3/12 improved) 5. Tullio's (4/10 improved) 6. Tuning fork (3/11 improved) 7. VEMP (1/6, improved, 5 no change) 7. Audiogram (14 total, 3 improved, 9 no change, 1 evident post-operative when initially negative). In addition, 6/13 increased AC thresholds by >10 dB 8. Post-operative hearing deterioration 6/13 9. DHI score (5/7 improved)	Non-validated outcome measure—9-item questionnaire designed for study. Outcome measures used (all subjective): Improvement noted in autophony, bone-conduction sensitivity, pulsatile tinnitus, dizziness with loud sound, dizziness with straining, sensitivity to increased middle ear pressure, aural fullness, and imbalance Hearing loss improvement not significant post-operatively	1 week post-operative review (1 patient), 2-year follow-up (1 patient) Post-operative VEMP (normalization in both cases) Post-operative subjective outcomes (described as patient symptoms) and VEMPs—improved	Follow-up at 6 and 10 weeks. Vertigo free (including on straining and loud noise). Pneumatic otoscopy—no vertigo or nystagmus. Aural fullness, autophony, and tinnitus resolved post-operatively. Hearing returned to pre-operative levels Post-operative VEMP remained abnormal
Statistical analysis?	Objective and subjective outcome measures. Fair assessment of success: P (subjective and at least partial objective success) (3/14) S (subjective success) (6/14) F (failure) (6/14). No statistical analysis possible	Mean and standard deviation given for each subjective outcome n=19. However, data may be non-parametric	No statistical analysis	No statistical analysis
Well-described results?	Yes. Multiple outcome measures (objective and subjective)	Outcomes were subjective with a normal distribution presumed (n=19) Not explicit as to when the post-operative questionnaire was completed-variation in post-operative duration may impact on outcome scores	Subjective outcome (patient symptoms)-improved Objective outcome: VEMPs normalized post-operative	Subjective outcomes (patient symptoms) —improved Objective outcome: VEMP no change
Appropriate discussion?	Discussed the limitations of the study and need for RCT Discuss variation in subjective/objective post-operative outcomes	Discussed limitations of the study: multiple surgeons, variations in reinforcement materials used. How much of the improvement effect	Not extensive. Limited to two cases, therefore difficult to draw major conclusions	Limited. One case only.

and significant number of failures	was placebo?
Overall: suggest that RW plugging is not the first-line treatment for SSCD	Points to advantages of less invasive technique of reinforcement compared with MCF or transmastoid techniques
	Discusses the role of RW and OW reinforcement as an area for future research.
	Advises against RWO on the lack of reproducibility of benefit noted in one case only (unsuccessful in subsequent two cases in another series)

RW: round window; EAC: external auditory canal; VEMP: vestibular evoked myogenic potential; AC: air conduction; DHI: Dizziness Handicap Inventory; RCT: randomized controlled trial; CT: computerized tomography; OW: oval window; MCF: middle cranial fossa; RWO: round window obliteration.

Peer-review: Externally peer-reviewed.

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