#### REVIEW ARTICLE

# Efficacy assessment and complications of surgical management for superior semicircular canal dehiscence: a meta-analysis of published interventional studies

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Abstract Superior semicircular canal syndrome (SSCS) includes vestibular and audiological symptoms which result from the introduction of a third mobile window into the osseous cochlea. Surgical repair is considered in cases of incapacitating symptoms. The present paper aims at comparing the different surgical approaches and modes of dehiscence repair, regarding their respective efficacy and potential pitfalls. A systematic literature review and metaanalysis of pooled data were performed. Study selection included prospective- and retrospective-controlled studies, prospective- and retrospective-cohort studies, ex vivo studies, animal models, case-reports, systematic reviews and clinical guidelines. A total of 64 primary operations for SSC repair were identified; 56 ears were operated for vestibular and 7 for auditory complaints. A total of 33 ears underwent canal plugging, 16 resurfacing, and 15 capping. Success rates were 32/33, 8/16, and 14/15, respectively. The observed differences were statistically significant (P = 0.001). Resurfacing proved less effective than both plugging (P = 0.002), and capping (P = 0.01) techniques. Temporalis fascia was commonly used as sealing material and was combined with bone-pâté/bone-wax (plugging), bone-graft (resurfacing), or hydroxyapatite-cement (capping). Most operations were performed via middle-fossa approach; higher success rates were associated with plug-

were the most frequent complications encountered. Most cases were followed for 3–6 months. Precise criteria regarding follow-up duration and objective success measures are not determined. Surgical repair of SSCS is considered as a valid therapeutic option for patients with debilitating symptoms. Consensus regarding strict follow-up criteria and objective assessment of success is necessary before larger scale operations can be implemented in clinical practice.

ging and capping techniques. SNHL and disequilibrium

**Keywords** Superior canal dehiscence · Superior canal syndrome · Nystagmous · Surgery · Plugging · Resurfacing

## Introduction

Superior semicircular canal (SSC) syndrome is a recently recognized clinical condition, which was initially described by Minor et al. [1]. The syndrome usually encompasses a constellation of vestibular and audiological symptoms, such as sound- and/or pressure induced vertigo and oscillopsia [2], along with conductive hearing loss and autophony [2, 3], and typically manifests as sound- and/or pressure-induced nystagmous at the plane of the SSC (vertical direction with a torsional component) [4].

The introduction of this syndrome helped differentiating a large number of hitherto not precisely determined cases of sound- and/or pressure-induced vestibulopathy [5–9], along with undetermined cases of alleged "inner ear conductive hearing loss" [10].

The proposed underlying mechanism involves the existence of a dehiscence at the apical turn of the SSC (third mobile window), in addition to the round and oval windows of the osseous cochlea, which in effect potentiates the transmission of sudden changes in the middle and/or

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intracranial pressure, thus altering the related neural firing rates of the vestibular system, and may also alter inner ear fluid dynamics, causing dissemination of the acoustic energy [11, 12].

Even though avoidance of the precipitating stimuli may prove quite effective in the majority of cases [13, 14], surgical repair of the affected canal may be considered as a valid therapeutic option in cases of incapacitating symptoms [3]. However, the different surgical approaches to the SSC and the various modes of dehiscence repair which are reported, may lead to uncertainty, with regard to their respective efficacy and potential pitfalls.

Even though a number of studies have attempted to assess the procedures which are employed in the surgical management of SSC syndrome, the reported results were usually individualized by institution and not pooled by any means of common inclusion/exclusion criteria. The present paper represents the first meta-analysis of interventional studies with regard to the surgical treatment of the SSC syndrome, and aims at comparing the respective efficacy of the different techniques employed, taking into account the complications that may be associated with the implementation of each repair mode.

#### Materials and methods

An extensive search of the literature was performed in Medline and other available database sources, using the keywords "superior canal dehiscence", "treatment", "surgery", and "operation". The keyword "superior canal dehiscence" was considered primary and was either used individually, or combined with each of the other keywords. In addition, reference lists from the retrieved articles were manually searched.

The retrieved studies were critically appraised, according to evidence-based guidelines for the categorization of medical studies (Tables 1, 2, 3) [15]. Three methods of surgical repair of the dehiscent SSC were identified: (1) occlusion of the canal's lumen, which will be referred to as "canal plugging", (2) covering of the dehiscence in an underlay manner, which will be referred to as "canal resurfacing", and (3) shielding of the dehiscence in an overlay manner, which will be referred to as "canal capping". In addition, two surgical approaches were also identified; middle fossa craniotomy was performed in the overwhelming majority of cases, whereas a transmastoid approach was also attempted, albeit in a considerably smaller number of patients.

Tympanostomy tube placement may also prove beneficial for patients with symptoms mainly arising from pressure in the external auditory canal (EAC) [2]; however, since this intervention does not amend the underlying

Table 1 Evidence-based categorization of medical studies

Category of evidence	Origin of evidence	
Ia	Evidence from meta-analysis of randomized controlled trials	
Ib	Evidence from at least one randomized controlled trial	
IIa	Evidence from at least one controlled study without randomization	
IIb	Evidence from at least one other type of quasi-experimental study	
III	Evidence from non-experimental descriptive studies, such as comparative studies, correlation studies, and case-control studies	
IV	Evidence from expert committee reports or opinions or clinical experience of respected authorities, or both	

pathologic lesion, these operations were not included in the present study.

Using this framework of results 91 operations were initially identified. Three primary categories of outcomes were then chosen for further analysis: (1) improvement of vestibular function, or (2) improvement of auditory dysfunction, and (3) comparison of the recurrence rates, which are associated with each surgical method employed. In addition, two secondary end-points were also pursued: comparative analysis of the vestibular (1), or auditory complications (2) that may be associated with the related surgical approaches.

Inclusion criteria necessitated the existence of at least one physiologic symptom or sign indicative of a dehiscence in the SSC that was considered for surgical repair, the realization of at least one preoperative test that could assess the main complaint of the affected individual, and the radiographic demonstration of the dehiscence in thin-sliced C/T scans of the temporal bone (either in less than 0.6 mm collimation in one cut, or in 1 mm collimation in two consecutive cuts).

Exclusion criteria referred either to previous inner ear surgery, or to second operations. As a result of the first restriction, patients with previous ear surgery were included in the analysis of data only in case they had not undergone previous procedures on either the cochlea, or the labyrinth, or procedures that undoubtedly compromised audition (i.e. tympanoplasty type IV). The second restriction was mandatory in order to avoid two sources of bias: (1) double-counting of the operations, in case the initial procedure was performed at a different centre, and (2) sample heterogeneity, taking into account that second operations are associated with higher co-morbidity rates [16].

As a result of the abovementioned methodology, the final number of operated ears that was assessed in the present meta-analysis was restricted to 64. Due to the



 Table 2
 Operative management of SSC dehiscence (large series)

Authors	Type of study	Ev. lev.	No of ears	Sr	Surg. appr.	Surg. techn.	Seal. mat.	Complications	Remarks
Minor [19] Limb et al. [16] Carey et al. [13] <sup>a</sup>	Prospective Retrospective Prospective	all	26	22/26	MF	Plugging (17) Resurfacing (9)	Fascia/bone graft	(a) Vestibular hypofunction (1) (b) SNHL (2) (c) CHL (1) (d) Haematoma (1) (e) Celulitis (1) (d) Diabetes insipidus (1)	<ul> <li>(a) Mean postoperative gain value for plugged SSCs was reduced by 44%</li> <li>(b) Aggressive SSC packing may reduce PSC function</li> <li>(c) Inhibitory contribution of SSC towards the contralateral PSC is postoperatively decreased</li> </ul>
Mikulec et al. [18]	Retrospective	IIb	6	6/8	MF	Plugging (8) Resurfacing (1)	Bone wax Bone graft	(a) vestibular hypofunction (1) (b) SNHL (4) (c) MHL (1) (d) dis-equillibrium (4) (e) Tinnitus (1) (d) Seroma (2)	Extremely thin bone covering of the SSC may be difficult to be detected preoperatively
Hillman et al. [33]	Retrospective IIb	all lib	14	13/14	MF	Capping	Fascia/hydroxy-apatite cement	SNHL (1)	Atypical findings during preoperative physical examination might be indicative of negative explorations
Agrawal et al. [40]	Retrospective	IIb	2	2/2	TM	Plugging	Bone pâté	<ul><li>(a) Dis-equilibrium (2)</li><li>(b) Dural tear (1)</li></ul>	TM approach for repairing SSC dehiscence may be more familiar to ear surgeons
Brantberg et al. [14] Prospective	Prospective	IIb	7	2/2	TM	Plugging	fascia	(a) SNHL (1) (b) Vestibular hypofunction (2) (c) Dis-equillibrium (2)	VEMP testing can objectively assess surgical outcomes
Younge et al. [50]	Retrospective	IIb	2	2/2	MF	Plugging (1) Resurfacing (1)	Bone pâté/bone graft fascia/bone graft	None reported	Patients with SSC syndrome may be encountered during clinical examination by various medical specialties
Friedland et al. [11]	Retrospective	IIb	6	0/2	MF	Resurfacing	Fascia/bone graft	None reported	<ul><li>(a) The squamous portion of the temporal bone is also affected in patients with SSC syndrome</li><li>(b) Resurfacing techniques might fail due to this existing abnormality</li></ul>

<sup>a</sup> Authors belong to the same investigating team

ev. lev. Evidence level, sr success rate, surg. appr. surgical approach, surg. techn. surgical technique, seal. mat. sealing material, MF middle fossa, TM transmastoid, SNHL sensorineural hearing loss, SSC superior semicircular canal, PSC posterior semicircular canal, VEMP vestibular-evoked myogenic potential



 Table 3
 Operative management of SSC dehiscence (case studies)

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Authors	Type of study Ev. lev.	Ev. lev.	No of ears	Sr	Surg. appr.	Surg. techn.	Seal. mat.	Complications	Remarks
Wilkinson et al. [17] Case report	Case report	IIIb	1	1/1	MF	Plugging	Fascia/bone wax	None reported	Clinicians should be alert for potential SSC in patients with CHL or MHL accompanied by vertigo, dizziness or autophony
Martin et al. [51]	Case report	III	1	0/1	MF	Resurfacing	Fascia/bone graft/glue	None reported	Surgical treatment of SSC vestibulopathy represents a common field for collaboration between neurotologists and neurosurgeons
Watters et al. [20]	Retrospective IIb	IIb	1	1/1	MF	Plugging	Fascia/bone pâté/bone wax/bone graft/glue	None reported	SSC syndrome should be included in the differential diagnosis of acute post-partum vertigo or disequilibrium
Kirtane et al. [47]	Case report	IIb	1	1/1	TM	Plugging	Bone pâté/bone wax/cartilage	(a) disequilibrium (b) SNHL	TM approach avoids craniotomy-associated morbidity and can even be performed under local anaesthesia
Crovetto et al. [46]	Case report	IIb	_	0/1	MF	Resurfacing	Fascia/bone graft	None reported	TM approach may be more advantageous
Smullen et al. [36]	Case study	III	_	1/1	MF	Capping	Bone graft/hydroxy-apatite cement	None reported	SSC may be mistakenly perceived as BPPV resistant to canalith repositioning manoeuvres
Pletcher et al. [48]	Case review	III	1	1/1	MF	Resurfacing	Resurfacing Fascia/bone pâté	None reported	I

ev. lev. Evidence level, sr success rate, surg. appr. surgical approach, surg. techn. surgical technique, seal. mat. sealing material, MF middle fossa, TM transmastoid, SNHL sensorineural hearing loss, SSC superior semicircular canal, BPPV benign paroxysmal positional vertigo



qualitative nature of the evaluated data chi-square analysis was utilized; logistic regression was also used in order to determine the magnitude of the observed differences. Statistical importance was accepted at the level of 0.05.

#### Results

Three prospective controlled studies, 2 retrospective controlled studies, 9 prospective cohort studies, 13 retrospective cohort studies, 4 ex-vivo studies, 3 animal models, 7 case reports, 2 systematic reviews and 1 clinical guideline met the defined criteria and were included into the study selection.

Overall, 64 primary operations for the surgical management of SSC syndrome were identified. Among them, canal plugging was performed in 33 ears, canal resurfacing in 16 ears, whereas 15 ears underwent capping of the SSC. The respective success rates were 32/33 for the plugging technique, 8/16 for resurfacing and 14/15 for capping operations. The observed difference was found statistically significant (P = 0.001) (Table 4).

Multiple comparisons between the three repair modes demonstrated that the resurfacing technique was significantly less effective than both the plugging (P = 0.002), and the capping repair mode (P = 0.01); among the latter, success rates did not differ statistically significantly. Logistic regression determined that the plugging technique had 32 times greater relative odd for a successful repair, compared to the repair mode which exhibited the least probability of successful surgical outcome (resurfacing); a value of 14 times greater relative odd was obtained for the capping technique.

A total of 56 ears were predominantly operated for vestibular symptoms, whereas 7 were offered surgery mainly for auditory complaints; the main problem that led to surgical intervention in one instance was not possible to be identified.

Hence, the plugging technique was applied in 29 ears in order to treat balance disorders, with a success rate of 28/29, whereas 12 cases were resurfaced, with 5 of them accomplishing relief from their symptoms. All ears that underwent capping reportedly did so due to vestibular

**Table 4** Chi-square analysis of reported success rates in three SSC dehiscence repair modes

Interventional	Repair mod	le		
outcome	Plugging	Resurfacing	Capping	Total
Positive	32	8	14	54
Negative	1	8	1	10
Total	33	16	15	64

symptoms; the affected patients reached satisfying symptom resolution in the majority of cases (14/15). The observed differences were found statistically significant (P = 0.0001). Multiple comparisons again proved that the resurfacing technique was significantly less effective than both the plugging (P = 0.0002), and the capping repair mode (P = 0.005); among the latter, success rates did not differ statistically significantly.

In addition, all four of the operated ears, which reported predominantly auditory symptoms and were plugged, experienced satisfying symptom resolution [16–19], as also did their resurfacing counterparts [16] (3/3).

The overwhelming majority of operations (59 ears) were performed via a middle fossa approach; transmastoid approach was used in five ears. A rough estimation of the encountered complications with regard to these two surgical approaches revealed 22 incidences after middle fossa and 9 following a transmastoid operation. These incidences, not necessarily involving different ears are reported in Tables 2 and 3.

#### Discussion

Pathology-pathophysiology-diagnosis

Even though the pathologic lesion which is responsible for SSC syndrome has been well established, the underlying process during the development of that lesion has not yet been determined with certainty. However, the presence of an abnormally thin bony layer over the arcuate eminence of the SSC has been incriminated by many researchers as the "first event", which may later lead to the full manifestation of the syndrome [14, 19–21]. The origin of such a first event could be either developmental [21–23], or congenital [2, 24, 25], while a genetic substrate cannot be excluded in some cases [3, 14].

The syndrome typically evolves during adulthood [14, 26] and is usually triggered by a sudden change in middle ear or intracranial pressure (excessive straining, head trauma, loud low-frequency sounds etc.) which causes a disruption in the abnormally thin bone, thus acting as a "second event" [19–22]. In a large proportion of cases, however, the precipitating action of a second event could not be identified; it is possible that in these cases the syndrome has occurred as a consequence of pressure from the overlying temporal lobe [26].

Irrespective of the exact cause of the dehiscence, the absence of a bone covering in a specific area of the vestibular apparatus may allow abnormal volume displacements within the membranous labyrinth in response to stapes movements, which in effect might bring about increased susceptibility of the vestibular end organ to sound and



pressure changes, following respective deflections of the vestibular sensors [14]. Thus, a dehiscence of the SSC into the floor of the middle fossa basically introduces a "third mobile window", in addition to the round and oval windows of the osseous cochlea, namely affecting the motion of the endolymph.

This has been experimentally confirmed in chinchillas before and after fenestration of the bone that covered the uppermost portion of the SSC. Hence, SSC fenestration induced pressure sensitivity in all SSC afferents, with responses correlating with the expected changes in endolymph flow within the examined system (utriculofugal, or utriculopetal, respectively) [27]. Rigid surgical repair of the dehiscences abolished pressure sensitivity, while maintaining physiologic rotational sensitivity [27]. Thus, a mechanism not very different from the one observed in clinical conditions characterized by the existence of a labyrinthine fistula (i.e. congenital syphilis) [5, 7, 28] seems to dictate vestibular responses in SSC syndrome.

With regard to the observed auditory phenomena, experimental studies in animal models and cadaveric preparations support a three-fold effect from the existence of a pressure release point in the SSC (dissemination of the acoustic energy, decrease in the cochlear input impedance, increase of bone conduction-evoked cochlear potential) [12, 29–31]. Hence, the result of this three-fold effect seems to be the intralabyrinthine elevation of air-conducted thresholds and an apparent bone-conduction hypersensitivity [14].

The diagnosis of SSC syndrome is not solely based on the demonstration of a dehiscence in CT imaging, but requires the correlation of radiographic findings with clinical symptoms and physiologic signs, taking also into account the results from audiometric and vestibular testing [13, 32–34].

Sound, pressure, or strain-evoked vertigo and oscillopsia, and persisting imbalance represent the primary vestibular manifestations of the syndrome [1, 2, 19, 20, 33], while positional vertigo and drop-attacks have also been reported [14, 35, 36]. The corresponding signs include vertical–torsional nystagmic eye movements, with the slow-phase components directed upward and away from the affected ear in cases of positive pressure changes in the EAC, and can be explained on the grounds of an ampullofugal deflection of the cupula, which increases the discharge rates of the vestibular nerve afferents that innervate the SSC [2, 4, 19]. The converse is observed in cases of negative pressure changes in the EAC, or increased intracranial pressure [14, 19, 20]. A sound-induced tilt of the head in the plane of the SSC can be also noted in as much as 20% of patients with vestibular signs [2, 32].

Auditory manifestations usually include autophony, pulsatile tinnitus and enhanced perception of bone conducted

sound (conductive hyperacousis) [2, 13, 14, 18, 33, 37], while aural fullness [36], mild to moderate sensorineural hearing loss and variable conductive hearing loss may also be troublesome in certain cases [1, 3, 12].

Standard vestibular testing, such as caloric responses, seems to be of limited diagnostic value in the majority of patients, with the possible exception of cases demonstrating positioning vertigo [14, 36], or after modification of the employed technique [33]. Specialized three-dimensional scleral coil techniques, on the other hand, can be used to record the evoked eye movements and calculate the respective angular velocities in the Cartesian coordinate system [4, 13, 26]. The respective gain values are indicative of vestibular hypofunction of the affected canal [13]. Video-oculography may be considered as an important tool in the documentation of the nystagmic eye movements [18, 19]. Frenzel glasses should be used in clinical practice, especially when video-oculography is not available, because visual fixation can lead to suppression of the evoked eye movements [18, 19].

Auditory testing usually reveals a low-frequency conductive hearing loss [2, 12, 14, 33, 37]. The related air-bone gap usually ranges between 5 and 10 dbHL [19, 33], affecting two or more frequencies, and can exist even when air conduction thresholds are normal, as bone conduction thresholds may be lower than 0 db NHL [2, 19]. A Carhart notch can also be observed [16], though infrequently, whereas stapedial reflexes are most often preserved [12, 14, 29]. Weber testing typically lateralizes to the affected ear and may also be heard when the tuning fork is placed at a considerable distance from the head [2, 14]. VEMP responses are elicited at an abnormally low threshold in SSC dehiscence [2] and can be particularly useful for diagnosis [34, 38]. Decreased thresholds have been associated with both a sensitivity and a specificity of 80% [39]. In addition, VEMP thresholds are concordant with symptoms in bilaterally dehiscent patients and may be useful in determining the most severely affected side in cases of symptoms not readily attributed to one ear, such as chronic disequilibrium [32], although there is still some debate regarding their exact reliability [18].

The diagnosis of SSC syndrome requires the careful correlation of the above mentioned physiologic findings with the obtained CT images. Conventional CT scans of the temporal bone, performed with 1.0 mm collimation and displayed in axial and coronal planes, may fail to identify extremely thin layers of bone covering the SSC, thus leading to overdiagnosis of the syndrome, because of the partial volume averaging effect [2, 40, 41]. This inadequacy can be partially addressed by trying to identify the suspected lesion in at least two consecutive cuts, using bone-detailed CT protocols [14, 33]. Alternatively, an ultrahigh-resolution helical CT scan with 0.5 mm collimation and reformatting



of the images to the plane of the affected canal could be used, as proposed by Belden et al. [42] in order to reduce the number of false positive CT findings. Indeed, it has been demonstrated that the positive predictive value of CT imaging is improved from 50 to 93%, when 0.5 mm collimation is performed instead of conventional 1.0-mm cuts. Moreover, multi-planar reconstructions seem more specific for the diagnosis of a dehiscent SSC, compared to the more recently proposed three-dimensional surface reconstructions of the temporal bone [39].

However, there is still some debate with regard to the additional information that oblique reformatting is able to provide, although its use may undoubtedly prove useful in equivocal or confusing cases [43]. It seems that reconstruction may not be necessary if a dehiscence is not apparent in coronal images [44]. Furthermore, larger temporal bone imaging series show that the actual positive predictive value of ultra-thin CT scanning may rest quite lower than 93%, ranging between 57 and 67%. This means in effect that CT imaging should not be perceived as a screening tool for SSC dehiscence, but instead used only as a means to confirm a strong clinical suspicion [44]. In addition, prior ear surgery may be extremely important when interpreting CT scans, as both of the negative explorations reported by Mikulec et al. [18] involved operated ears, which exhibited artificially projected translucent blue-lined canals, despite the use of ultrahigh-resolution protocols. The latter may also prove unable to detect bony walls thinner than 0.1 mm [18]. The potential utilization of MRI protocols, often used in the evaluation of patients with vestibular symptoms, in the diagnosis of SSC syndrome is also controversial. Even though a recent study suggested that T2-weighted fast spin echo MRI may have as much as 96% sensitivity and 98% specificity for the identification of the syndrome [45], costeffectiveness of this method compared to CT imaging has not yet been assessed.

#### Meta-analysis of interventional studies

Following the identification of the syndrome, patients with debilitating symptoms are offered surgical repair of the existing dehiscence [2, 14, 19, 40].

Even though efficacy assessment of the different procedures which are employed in the surgical management of SSC syndrome has been previously attempted in a number of systematic reviews, no study so far had accrued the reported data by any means of common inclusion/exclusion criteria (primary-second operation, diagnostic methodology etc.). Hence, the reported results merely reflected the accumulated experience of single institutions, rather than representing an actual meta-analysis.

In the present study, a critical assessment of the pooled data regarding the surgical management of SSC syndrome has identified 64 primary operations. Temporalis fascia was the most standard used sealing material and was combined either with bone pâté (or bone wax) in cases of canal plugging, or with bone graft in the majority of resurfacing operations. When capping of the SSC was performed, fascia and hydroxyapatite cement were most commonly used (Tables 2, 3).

The respective success rates seem to favour the practice of either canal plugging, or canal capping, over resurfacing of the SSC. Chi-square analysis was utilized to compare the aforementioned discrepancy. The observed difference was found statistically significant (P = 0.001) (Table 4), and was attributed to the lower effectiveness of the resurfacing technique, both from the plugging and the capping repair mode. Logistic regression was used to determine the magnitude of the observed differences, by calculating the relative odds for the two repair modes which presented the higher success rates. The basis of relative odd calculations was the resurfacing technique, because preliminary statistics had identified this method as the one having the least likelihood for a successful outcome (Table 5). A limitation in our calculation of odds that needs to be noted is that, due to the difference in the number of observations between the plugging and capping groups, the calculated ratios refer to the specific data set and cannot be easily extrapolated; however, data selection, with regard to the type of surgical intervention, was not biased.

Categorization of the operated ears according to the main problem that led to surgical intervention revealed that when surgery was offered as a means to treat balance disorders, the respective success rates were higher both for the plugging and the capping repair mode, compared to the resurfacing technique (P = 0.0001). However, all ears that were operated predominantly for auditory symptoms experienced satisfying symptom resolution, regardless of the employed repair mode. They also demonstrated at least partial closure of the low-frequency air-bone gap, especially in the frequency of 500 Hz, resulting in the improvement, or elimination of the existing conductive hearing loss [16, 17].

It should be noted, however, that there appears to be no consensus with regard to a common measure of success after surgery for SSC syndrome. Hence, most of the studies merely report symptom resolution, as described by the

 Table 5
 Relative odd calculations regarding the most efficient repair modes for SSC dehiscence

Repair mode	r/o	z	P >  z
Plugging	32	3.06	0.002
Capping	14	2.30	0.022

95% confidence intervals did not include 1 *r/o* Relative odds



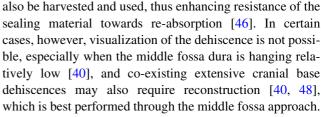
patients, however, no additional quality of life measures were employed. VEMP thresholds have been suggested as an indirect, yet technically simple and objective, physiologic measure for determining the outcome of surgical repair [32], as they seem to normalize following successful surgical intervention [14, 32]. Indeed, Welgampola et al. [32] report that two of their patients who underwent primary operation for SSC dehiscence and were tested both pre- and post-operatively demonstrated a marked increase in air-conducted VEMP thresholds after successful plugging of the canal. Despite these findings, however, a VEMP criterion has not been systematically used for efficacy assessment after surgery. Thus, even though pre-operative VEMP testing was performed in 41 out of the 64 ears which were included in the present meta-analysis, only 4 of them were also evaluated post-operatively. Further utilization of VEMP testing in the future may facilitate objective confirmation of successful surgical corrections.

Moreover, there also seems to be no consensus regarding the time of follow-up which is necessary to determine the potential success after surgery. Nevertheless, the majority of cases (40 ears) were followed for a time period of 3–6 months, whereas 12 ears were evaluated within a less than 3-month period, and only 4 ears were reportedly followed for more than 6 months. Data regarding follow up duration in eight cases were not reported (Fig. 1).

Most ears were operated via a middle fossa approach. A transmastoid approach was used in five ears, even though ear surgeons are theoretically more accustomed to it [40, 46]. Moreover, it does not necessitate craniotomy and temporal lobe retraction [40, 47], and can even be performed under local anaesthesia [47]. In addition, the canal can be occluded without first manipulating the dehiscence [40], the bone graft which is obtained from the surgical field is thicker, therefore less likely to get reabsorbed, and, once in place, its stability is substantially better [46]. Cartilage can



Fig. 1 Duration of follow-up after surgical correction of SSC dehiscence



With regard to the encountered complications by category of surgical approach, the relatively small number of ears which have been operated via the transmastoid approach does not allow robust statistical conclusions to be drawn. Respective comparisons between the three repair modes of surgical repair (plugging, resurfacing, capping) also proved impossible to perform, as most studies do not report the encountered complications in relation to the surgical mode employed.

However, an interesting notion was proposed by Agrawal and Parnes about the impact of the sealing material on post-surgical hearing. Based on their cumulative experience with posterior and superior canal occlusions and the animal studies by Kim et al., the authors proposed that bone dust (which is the basis for bone pâté formulations) may be more favourable in preserving or improving hearing, as it is potentially associated with a reduced risk for post-operative perilymphatic inflammation and serous labyrinthitis, and enhanced periosteal osteoneogenesis at the occlusion site, compared to the use of bone wax [40, 49]. Our data confirmed that bone wax was used in a total of 12 ears, all of which underwent a plugging technique; among them 5 suffered post-operative sensorineural hearing loss.

### **Conclusions**

Superior semicircular canal syndrome is a clinical condition which is caused by the introduction of a third mobile window into the osseous cochlea, and typically exhibits sound- and/or pressure-induced nystagmous at the plane of the affected canal.

The recognition of this syndrome helped differentiating a large number of hitherto undetermined cases of sound- and/ or pressure-induced vestibulopathy, as well as obscure cases of conductive hearing loss originating from the inner ear.

The presence of an abnormally thin bony layer over the arcuate eminence of the SSC has been hypothesized to represent a "first event", which may later lead to the full manifestation of the syndrome; sudden changes in middle ear or intracranial pressure ("second event"), or long-term pressure from the overlying temporal lobe may in turn be responsible for the ensuing canal dehiscence.

Accurate diagnosis is based on the correlation of ultrahigh resolution CT findings with clinical symptoms and



physiologic signs, taking also into account the results from audiometric and vestibular testing.

Even though avoidance of the precipitating stimuli may prove quite effective, patients with debilitating symptoms are offered the alternative of surgical repair of the existing dehiscence. The overwhelming majority of operations are performed via a middle fossa approach, whereas plugging and capping techniques are associated with the higher success rates. However, strict criteria for the objective assessment of a successful outcome and the appropriate duration of follow up have not been determined so far. The relatively restricted number of ears that have been operated also precludes us from drawing robust conclusions regarding the complication rates, which have been associated with the various surgical approaches and operating techniques.

A consensus regarding the aforementioned shortcomings is, therefore, necessary before larger scale operations for the treatment of SSC syndrome can be implemented in the wider clinical practice.

#### Conflicts of interest statement None declared.

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