Aggregating the symptoms of superior semicircular canal dehiscence syndrome

This is the accepted version of the following article: *Naert L, Van de Berg R, Van de Heyning P, Bisdorff A, Sharon JD, Ward BK, Van Rompaey V. Aggregating the symptoms of superior semicircular canal dehiscence syndrome. Laryngoscope. 2017 Dec 27. doi: 10.1002/lary.27062*, which has been published in final form at http://onlinelibrary.wiley.com/wol1/doi/10.1002/lary.27062/abstract

Aggregating the symptoms of superior semicircular canal dehiscence syndrome.

Review to develop evidence-based symptom set.

Naert Laura, BM[1]

Van de Berg Raymond, MD[2]

Van de Heyning Paul, MD, PhD[1,3]

Bisdorff Alexandre, MD, PhD[4]

Sharon Jeffrey D., MD[5]

Ward Bryan K., MD[6]

Van Rompaey Vincent, MD, PhD[1,3]

[1]Faculty of Medicine and Health Sciences, University of Antwerp, Belgium

[2]Maastricht University Medical Center, Department of Otorhinolaryngology and Head and Neck Surgery; Tomsk State Research University, Faculty of Physics, the Netherlands

[3]Antwerp University Hospital, Department of Otorhinolaryngology and Head & Neck Surgery; Faculty of Medicine and Health Sciences, University of Antwerp, Belgium.

[4]Department of Neurology, Centre Hospitalier Emile Mayrisch, Esch-Alzette, Luxemburg.

[5]University of California, San Francisco, Department of Otolaryngology-Head and Neck Surgery

[6]Johns Hopkins University School of Medicine, Department of Otolaryngology–Head & Neck Surgery

Corresponding Author:

Prof. dr. Vincent Van Rompaey

Antwerp University Hospital

Dept. of Otorhinolaryngology and Head & Neck Surgery

Wilrijkstraat 10, 2650 Edegem, Belgium

+32/3.821.47.30

+32/3.825.05.36

Vincent.van.rompaey@uza.be

Requests for reprints should be sent to this address.

None of the authors have financial interests in relation to this work.

No financial support was provided toward the completion of this work.

Conflicts of interest: none.

Acknowledgements

None.

This is the accepted version of the following article: *Naert L, Van de Berg R, Van de Heyning P, Bisdorff A, Sharon JD, Ward BK, Van Rompaey V. Aggregating the symptoms of superior semicircular canal dehiscence syndrome. Laryngoscope. 2017 Dec 27. doi: 10.1002/lary.27062*, which has been published in final form at http://onlinelibrary.wiley.com/wol1/doi/10.1002/lary.27062/abstract

ABSTRACT

OBJECTIVE: To aggregate symptoms reported by patients with superior canal dehiscence syndrome (SCDS) and to develop an evidence-based symptom set by performing a systematic review of the literature.

METHODS: Medline and PubMed databases were searched for articles that reported the preoperative symptoms of adult and pediatric patients with unilateral and bilateral SCDS. Articles were excluded if they reported on associated diseases or did not report symptoms.

RESULTS: Of the 397 articles retrieved, 66 were retained for quantitative analysis. Among 431 patients with SCDS, 91 symptom terms were reported. After combining synonymous terms, 22 symptoms were derived by consensus. Of the raw total number of reported symptoms 92.5% can be attributed to 5 common symptoms: spontaneous dizziness (51%), autophony (42.5%), pressure-induced vertigo (37.4%), hearing loss (39.9%) and sound-induced vertigo (42.7%).

CONCLUSIONS: This systematic review of symptoms reported by patients with SCDS identified a 22-item common symptom set. These items can be used to create an evidence-based patient-reported outcome measure to evaluate health-related quality of life in SCDS.

Key Words: Vestibular, Sensorineural hearing loss, Evidence-based medicine, Labyrinth diseases, review, patient reported outcome measures, vertigo, hearing loss

Level of evidence: NA

INTRODUCTION

Superior canal dehiscence syndrome (SCDS) was first described by Lloyd Minor in 1998. An absence of bone overlying the superior semicircular canal creates a third mobile window into the inner ear in addition to the oval and round windows[1]. This bony defect can contribute to a wide variety of auditory and vestibular symptoms, including hearing loss, autophony (hearing one's own voice louder than external sounds) [2], pulsatile and non-pulsatile tinnitus, aural fullness, bone conduction hyperacusis, imbalance, vertigo, and oscillopsia. Clinicians can also observe signs on physical examination such as Tullio phenomenon (vertigo and abnormal eye movements induced by loud sounds) [3,4] and Hennebert's sign (vertigo and nystagmus induced by pressure applied to the middle ear [5-7]). High-resolution CT imaging is important to detect a dehiscence in the otic capsule of the inner ear, but imaging alone is insufficient to diagnose SCDS [8-13]. Patients must also have symptoms consistent with SCDS as well as objective measures that the dehiscence is transmitting pressure, such as enhanced vestibular-evoked myogenic potential (VEMP) responses or negative bone conduction thresholds on pure tone audiometry. Presenting symptoms can vary and can include both auditory and vestibular symptoms or either auditory or vestibular symptoms alone[14].

When patients report their symptoms are causing significant impairments to their quality of life (QoL), surgeons have offered a variety of surgical procedures [9,15-18]. Comparing the effectiveness of these procedures, however, is challenging, as there is no patient-reported outcome measure designed specifically for evaluating SCDS-specific health-related QoL. While there are symptom-based scales like the Dizziness Handicap Inventory (DHI) that address some aspects of the disease, SCDS includes unique symptoms not included in the available measures. No scale currently exists that captures the broad variety of patient complaints in SCDS.

The aim of this study was to perform a systematic review of all symptoms reported by patients with SCDS, with the broader goal of enabling the eventual design of a condition-specific patient-reported outcome measure (PROM).

MATERIALS AND METHODS

Search strategy. The search was performed using the bibliographical databases PubMed, Medline, Cochrane Database of Systematic Reviews, Cochrane Central Register of Controlled Trials, Clinical Trials Registry, ISI Web of Knowledge and Web of Science on May 17, 2017 (LN, JS). Since no medical subject heading (MeSH) term is available for SCDS, the search was carried out using the keywords and logic: superior[All Fields] AND ("semicircular canals"[MeSH Terms] OR ("semicircular"[All Fields] AND canals"[All Fields]) OR "semicircular canals"[All Fields] OR ("semicircular"[All Fields] AND "canal"[All Fields]) OR "semicircular canal"[All Fields]) AND dehiscence[All Fields]. For a schematic overview of the search strategy see Figure 1.

Selection criteria. Articles (original studies, case series, and case reports) were included if they were written in the English language, involved humans, and reported on the presenting symptoms of adult and pediatric patients with either unilateral or bilateral SCDS. Articles were excluded if they did not report on patient symptoms associated with SCDS (n=55) or reported on symptoms associated with SCDS but in combination with other associated diseases (n=57). These associated diseases were posterior semicircular canal dehiscence, otosclerosis, migraine, vestibular schwannoma, chronic otitis media, meningo-encephalocele, lateral semicircular dysplasia, patulous Eustachian tube dysfunction, chronic ear disease, near dehiscence, cerebrospinal fluid otorrhea, otitic meningitis, enlarged vestibular aqueduct, Chiari type I malformation, meningioma, cerebral palsy, multiple semicircular canal dehiscences, glioblastoma and Pierre-Robin sequence. Articles were also excluded if classified as a systematic review (n=24), an editorial or comment on another study (n=10), or if they did not report the number of patients per symptom (n=10). Other exclusion criteria included studies focusing

primarily on radiographic techniques (n=21), VEMPs (n=23) or other diagnostic testing (n=23) or if containing a healthy subject or modeling third mobile window physiology (n=2). Figure 2 shows the flowchart for article exclusion with the number of articles excluded.

Data extraction. From the remaining 66 articles, the number of patients with SCDS and their preoperative symptoms were extracted. The risk of bias evaluation can be found in supplementary digital content e-1. Several questions were not applicable to our review because we were focusing on preoperative symptomatology and not on outcome and follow-up after interventions (incl. questions 8 and 9 on intervention and questions 11-19 on outcome and follow-up after surgery). A few patients were excluded because the authors identified comorbid diseases with symptoms that could overlap with those reported by patients with SCDS, e.g. demyelinating polyneuropathy [3], meningo-encephalocoele [19], chronic otitis media [20]. All data extracted from each study was tabulated in Microsoft Office Excel 2016 (Microsoft Redmond Campus, Redmond, Washington, USA). A committee of subject-matter experts (VVR, PVDH) reviewed the list of symptoms and deliberated on symptoms that could be merged using a common term.

Quantitative data synthesis. The primary outcome of this systematic review was a detailed overview of symptoms associated with SCDS and a classification in order of relative frequency of the symptoms reported, shown as a percentage of the total number of included patients.

RESULTS

The initial search resulted in 397 publications and was reduced to 309 by adding filters for English language and human species. After screening all abstracts against the study criteria 101 full-text articles were reviewed. Ultimately 66 articles including 431 patients were retained for quantitative analysis [1-5,8,16,19-58]. All article references can be found in supplementary digital content e-2. The number of patients included from each of these reports ranged from 1 to 35. In total, 91 symptom terms were retrieved from the 1253 patient symptoms reported. After combining synonymous terms (e.g. aural pressure – aural fullness), 22 specific symptom terms were identified: spontaneous dizziness, sound-induced vertigo, autophony, hearing loss, pressure-induced vertigo, aural pressure, nonpulsatile tinnitus, spontaneous pulsatile tinnitus, hyperacusis to environmental sounds, hyperacusis to bodily sounds, spontaneous oscillopsia,/tilting, positional vertigo, motion intolerance, gaze-evoked tinnitus, pulsatile oscillopsia, headache/ear pain, drop attack, sound distortion, head movement induced pulsatile tinnitus, anxiety, hemifacial numbness and tinnitus aggravated by Valsalva. The 91 original symptom terms and their associated common terms are provided in Table 1. The total number of patients included from each article and the number of patients reporting each symptom in the 22-item short list is provided in supplementary digital content, table e-3: Quantitative short list.

In order to determine the relative prevalence of each symptom included on the short list, the occurrence of each symptom is divided by the total number of patients. When consolidating from 91 terms to a 22-term short list, there was a risk of counting those patients who reported multiple symptoms twice, prior to our consolidating symptoms under a common term. To eliminate double counting, all articles were reevaluated and the number of reported symptoms was adjusted. The percentage of the number of patients reporting a specific symptom over the total

number of patients is shown in Figure 3. We identified eight symptoms that were most frequently reported, with spontaneous dizziness, sound induced vertigo, autophony and hearing loss being the most common.

DISCUSSION

Health services research has been promoting PROMs as indicators for quality of care, especially in areas where objective measures are lacking. [59] Many generic QoL evaluations have been developed such as the Short-Form 36 (SF36)[60] that can compare QoL among patients with different diseases and to healthy controls. Other instruments such as the Health Utility Index-Mark 3 (HUI3), which includes speech and hearing components, have been designed to assess clinical utility and to calculate cost-effectiveness[61]. Generic questionnaires, such as the SF-36, have been used to study changes of QoL after SCDS surgery and have demonstrated their responsiveness [62]. While valuable, these general QoL instruments, insufficiently evaluate disease-specific symptoms in conditions such as SCDS in which unique symptoms contribute to a patient's motivation to undergo surgery and changes in these symptoms determine whether they feel improved afterwards. Disease-specific PROMs are therefore a complementary tool that can be used to determine the effectiveness of an intervention.

When reviewing the literature, we found only one questionnaire on SCDS symptomatology, reported by Silverstein et al. [18] This questionnaire was generated in order to gather pre- and postoperative data for comparison of the efficacy of an intervention for round window reinforcement as a treatment for SCDS. The process for its development was unclear and lacks formal evaluation of reliability or validity.

Best practices in outcomes research methodology advise that development of a formal condition-specific scale should begin with a comprehensive review of the literature on all the reported symptoms for that condition. The objective of this systematic review, therefore, was to create an overview of all symptoms reported by patients with SCDS before surgery and without

concomitant pathology. From 66 articles that met inclusion criteria we obtained a list of 91 symptoms that was consolidated to a short list of 22 specific symptom terms. We subsequently identified eight symptoms that were most frequently reported, with spontaneous dizziness, sound induced vertigo, autophony and hearing loss among the most common. These symptoms will be further explored in structured interviews with patients to assist the development of a PROM for patients with SCDS.

The strength of this study is its comprehensive analysis of reported symptoms in the literature. The main limitation of this type of review, however, is the reliance on accurate assessment and reporting of symptoms by other practitioners. Patients may not mention all complaints due to their odd character, like hearing bodily sounds (e.g. eye movements, own footsteps, etc.), and it is often unclear the extent to which the physician inquired about relevant symptoms associated with SCDS. Moreover, many reports included patients that underwent surgery and those patients might present with symptoms that are more severe.

Articles that mentioned comorbid conditions with potentially overlapping symptoms were excluded; however, it is possible that in some articles included in this study the physician did not ask about associated diseases or these were not mentioned by the patient. Importantly, vestibular migraine, which is often seen in patients with SCDS can present with symptoms of spontaneous dizziness and tinnitus. Additionally, some symptoms classified as spontaneous may have triggers of which the patient may be unaware. The symptoms hemifacial numbness, anxiety and tinnitus aggravated by Valsalva were all reported only once, and should be considered in future studies.

The purpose of this study was to determine comprehensively the potential presenting symptoms in patients with SCDS as a step toward developing a novel outcome measure. Prospective studies will be used to evaluate the established symptom set before and after surgery to assess the importance of each term and its relevance for the patient. In order to develop an accurate assessment of patients presenting with SCDS and the prevalence of their presenting symptoms, however, prospective studies are also needed to review all these symptoms systematically, and provide data on symptom triggers, frequency, and duration. This future research could provide more insight into a minimum set of symptoms to enable accurate diagnosis of SCDS, in addition to imaging and electrophysiological data.

CONCLUSION

This systematic review of symptoms reported by patients with SCDS identified a 22-item common symptom set. The 5 most frequently reported symptoms were spontaneous dizziness, sound-induced vertigo, autophony, hearing loss and pressure-induced vertigo. The items derived from this study can be used as the initial steps toward creating a novel validated PROM in patients with SCDS.

REFERENCES

1. Lee GS, Zhou G, Poe Det al. Clinical experience in diagnosis and management of superior semicircular canal dehiscence in children. Laryngoscope. 2011; 121:2256-2261.

2. Crane BT, Lin FR, Minor LB, Carey JP. Improvement in autophony symptoms after superior canal dehiscence repair. Otol Neurotol. 2010; 31:140-146.

3. Younge BR, Khabie N, Brey RH, Driscoll CL. Rotatory nystagmus synchronous with heartbeat: a treatable form of nystagmus. Trans Am Ophthalmol Soc. 2003; 101:113-117; discussion 117-118.

4. Minor LB, Solomon D, Zinreich JS, Zee DS. Sound- and/or pressure-induced vertigo due to bone dehiscence of the superior semicircular canal. Arch Otolaryngol Head Neck Surg. 1998; 124:249-258.

5. Tilikete C, Krolak-Salmon P, Truy E, Vighetto A. Pulse-synchronous eye oscillations revealing bone superior canal dehiscence. Ann Neurol. 2004; 56:556-560.

6. Saliba I, Maniakas A, Benamira LZ, Nehme J, Benoit M, Montreuil-Jacques V. Superior canal dehiscence syndrome: clinical manifestations and radiologic correlations. Eur Arch Otolaryngol Head Neck Surg. 2014; 271:2905-2914.

7. Benamira LZ, Alzahrani M, Saliba I. Superior canal dehiscence: can we predict the diagnosis? Otol Neurotol. 2014; 35:338-343.

8. Beyea JA, Agrawal SK, Parnes LS. Transmastoid semicircular canal occlusion: a safe and highly effective treatment for benign paroxysmal positional vertigo and superior canal dehiscence. Laryngoscope. 2012; 122:1862-1866.

9. Van Rompaey V, Van de Heyning P. "Superior semicircular canal dehiscence"-syndroom: een nieuwe binnenooraandoening. Tijdschr voor Geneeskunde. 2015.

10. Kaski D, Davies R, Luxon L, Bronstein AM, Rudge P. The Tullio phenomenon: a neurologically neglected presentation. J Neurol. 2012; 259:4-21.

11. Saxby AJ, Gowdy C, Fandino Met al. Radiological prevalence of superior and posterior semicircular canal dehiscence in children. Int J Pediatr Otorhinolaryngol. 2015; 79:411-418.

12. Zhou G, Gopen Q, Poe DS. Clinical and diagnostic characterization of canal dehiscence syndrome: a great otologic mimicker. Otol Neurotol. 2007; 28:920-926.

13. Niesten ME, McKenna MJ, Herrmann BS, Grolman W, Lee DJ. Utility of cVEMPs in bilateral superior canal dehiscence syndrome. Laryngoscope. 2013; 123:226-232.

14. Niesten ME, Hamberg LM, Silverman JBet al. Superior canal dehiscence length and location influences clinical presentation and audiometric and cervical vestibular-evoked myogenic potential testing. Audiol Neurootol. 2014; 19:97-105.

15. Amoodi HA, Makki FM, McNeil M, Bance M. Transmastoid resurfacing of superior semicircular canal dehiscence. Laryngoscope. 2011; 121:1117-1123.

16. Brantberg K, Bergenius J, Mendel L, Witt H, Tribukait A, Ygge J. Symptoms, findings and treatment in patients with dehiscence of the superior semicircular canal. Acta Otolaryngol. 2001; 121:68-75.

17. Van Haesendonck G, Van de Heyning P, Van Rompaey V. Retrospective cohort study on hearing outcome after transmastoid plugging in superior semicircular canal dehiscence syndrome: Our Experience. Clin Otolaryngol. 2016; 41:601-606.

18. Silverstein H, Kartush JM, Parnes LSet al. Round window reinforcement for superior semicircular canal dehiscence: a retrospective multi-center case series. Am J Otolaryngol. 2014; 35:286-293.

19. Mikulec AA, Poe DS, McKenna MJ. Operative management of superior semicircular canal dehiscence. Laryngoscope. 2005; 115:501-507.

20. Modugno G, Brandolini C, Savastio G, Ceroni AR, Pirodda A. Superior semicircular canal dehiscence: a series of 13 cases. ORL J Otorhinolaryngol Relat Spec. 2005; 67:180-184.

21. Mong A, Loevner LA, Solomon D, Bigelow DC. Sound- and pressure-induced vertigo associated with dehiscence of the roof of the superior semicircular canal. AJNR Am J Neuroradiol. 1999; 20:1973-1975.

22. Smullen JL, Andrist EC, Gianoli GJ. Superior semicircular canal dehiscence: a new cause of vertigo. J La State Med Soc. 1999; 151:397-400.

23. Minor LB. Superior canal dehiscence syndrome. Am J Otol. 2000; 21:9-19.

24. Cox KM, Lee DJ, Carey JP, Minor LB. Dehiscence of bone overlying the superior semicircular canal as a cause of an air-bone gap on audiometry: a case study. Am J Audiol. 2003; 12:11-16.

25. Halmagyi GM, Aw ST, McGarvie LAet al. Superior semicircular canal dehiscence simulating otosclerosis. J Laryngol Otol. 2003; 117:553-557.

26. Minor LB, Carey JP, Cremer PD, Lustig LR, Streubel SO, Ruckenstein MJ. Dehiscence of bone overlying the superior canal as a cause of apparent conductive hearing loss. Otol Neurotol. 2003; 24:270-278.

27. Deutschlander A, Strupp M, Jahn K, Jager L, Quiring F, Brandt T. Vertical oscillopsia in bilateral superior canal dehiscence syndrome. Neurology. 2004; 62:784-787.

28. Martin JE, Neal CJ, Monacci WT, Eisenman DJ. Superior semicircular canal dehiscence: a new indication for middle fossa craniotomy. Case report. J Neurosurg. 2004; 100:125-127.

29. Mikulec AA, McKenna MJ, Ramsey MJet al. Superior semicircular canal dehiscence presenting as conductive hearing loss without vertigo. Otol Neurotol. 2004; 25:121-129.

30. Brantberg K, Ishiyama A, Baloh RW. Drop attacks secondary to superior canal dehiscence syndrome. Neurology. 2005; 64:2126-2128.

31. Pletcher SD, Oghalai JS, Reeck JB, Cheung SW. Management of superior canal dehiscence syndrome with extensive skull-base deficiency. ORL J Otorhinolaryngol Relat Spec. 2005; 67:192-195.

32. Friedland DR, Michel MA. Cranial thickness in superior canal dehiscence syndrome: implications for canal resurfacing surgery. Otol Neurotol. 2006; 27:346-354.

33. Hillman TA, Kertesz TR, Hadley K, Shelton C. Reversible peripheral vestibulopathy: the treatment of superior canal dehiscence. Otolaryngol Head Neck Surg. 2006; 134:431-436.

34. Schmuziger N, Allum J, Buitrago-Tellez C, Probst R. Incapacitating hypersensitivity to one's own body sounds due to a dehiscence of bone overlying the superior semicircular canal. A case report. Eur Arch Otolaryngol Head Neck Surg. 2006; 263:69-74.

35. Watters KF, Rosowski JJ, Sauter T, Lee DJ. Superior semicircular canal dehiscence presenting as postpartum vertigo. Otol Neurotol. 2006; 27:756-768.

36. Agrawal SK, Parnes LS. Transmastoid superior semicircular canal occlusion. Otol Neurotol. 2008; 29:363-367.

37. Hain TC, Cherchi M. Pulse-synchronous torsional pendular nystagmus in unilateral superior canal dehiscence. Neurology. 2008; 70:1217-1218.

38. Peterson EC, Lazar DA, Nemecek AN, Duckert L, Rostomily R. Superior semicircular canal dehiscence syndrome: Successful treatment with repair of the middle fossa floor: technical case report. Neurosurgery. 2008; 63:E1207-1208; discussion E1208.

39. Deschenes GR, Hsu DP, Megerian CA. Outpatient repair of superior semicircular canal dehiscence via the transmastoid approach. Laryngoscope. 2009; 119:1765-1769.

40. Kirtane MV, Sharma A, Satwalekar D. Transmastoid repair of superior semicircular canal dehiscence. J Laryngol Otol. 2009; 123:356-358.

41. Lip G, Nichols DM. Measurement of defect angle in superior semicircular canal dehiscence. Clin Radiol. 2009; 64:1210-1213.

42. Ogutha J, Page NC, Hullar TE. Postpartum vertigo and superior semicircular canal dehiscence syndrome. Obstet Gynecol. 2009; 114:434-436.

43. Silverstein H, Van Ess MJ. Complete round window niche occlusion for superior semicircular canal dehiscence syndrome: a minimally invasive approach. Ear Nose Throat J. 2009; 88:1042-1056.

44. Yagi T, Koizumi Y. 3D analysis of cough-induced nystagmus in a patient with superior semicircular canal dehiscence. Auris Nasus Larynx. 2009; 36:590-593.

45. Fiorino F, Barbieri F, Pizzini FB, Beltramello A. A dehiscent superior semicircular canal may be plugged and resurfaced via the transmastoid route. Otol Neurotol. 2010; 31:136-139.

46. Hahn Y, Zappia J. Modified resurfacing repair for superior semicircular canal dehiscence. Otolaryngol Head Neck Surg. 2010; 142:763-764.

47. Hornibrook J, O'Neill-Kerr D, Berry L, Carroll G. Bilateral superior canal dehiscence syndrome. N Z Med J. 2010; 123:64-68.

48. Koo JW, Hong SK, Kim DK, Kim JS. Superior semicircular canal dehiscence syndrome by the superior petrosal sinus. J Neurol Neurosurg Psychiatr. 2010; 81:465-467.

49. McCaslin DL, Jacobson GP, Burrows HL, Littlefield P, Haynes DS. Transforming superior canal dehiscence to chronic subjective dizziness: from SCD to CSD. J Am Acad Audiol. 2010; 21:293-300.

50. Nam EC, Lewis R, Nakajima HH, Merchant SN, Levine RA. Head rotation evoked tinnitus due to superior semicircular canal dehiscence. J Laryngol Otol. 2010; 124:333-335.

51. Phillips DJ, Souter MA, Vitkovic J, Briggs RJ. Diagnosis and outcomes of middle cranial fossa repair for patients with superior semicircular canal dehiscence syndrome. J Clin Neurosci. 2010; 17:339-341.

52. Adams ME, Levine SC. The first new otologic disorder in a century: superior canal dehiscence syndrome. Minn Med. 2011; 94:29-32.

53. Hegemann SC, Carey JP. Is superior canal dehiscence congenital or acquired? A case report and review of the literature. Otolaryngol Clin North Am. 2011; 44:377-382, ix.

54. Kanaan AA, Raad RA, Hourani RG, Zaytoun GM. Bilateral superior semicircular canal dehiscence in a child with sensorineural hearing loss and without vestibular symptoms. Int J Pediatr Otorhinolarngol. 2011; 75:877-879.

55. Li PM, Bergeron C, Monfared A, Agrawal S, Blevins NH. Superior semicircular canal dehiscence diagnosed after failed stapedotomy for conductive hearing loss. Am J Otolaryngol. 2011; 32:441-444.

56. Manzari L, Burgess AM, MacDougall HG, Curthoys IS. Enhanced otolithic function in semicircular canal dehiscence. Acta Otolaryngol. 2011; 131:107-112.

57. McCall AA, McKenna MJ, Merchant SN, Curtin HD, Lee DJ. Superior canal dehiscence syndrome associated with the superior petrosal sinus in pediatric and adult patients. Otol Neurotol. 2011; 32:1312-1319.

58. Zhang AS, Govender S, Colebatch JG. Superior canal dehiscence causes abnormal vestibular bone-conducted tuning. Neurology. 2011; 77:911-913.

59. Black N. Patient reported outcome measures could help transform healthcare. BMJ. 2013; 346:f167.

60. Fielder H, Denholm SW, Lyons RA, Fielder CP. Measurement of health status in patients with vertigo. Clinical otolaryngol Allied Sci. 1996; 21:124-126.

61. Kuthubutheen J, Mittmann N, Amoodi H, Qian W, Chen JM. The effect of different utility measures on the cost-effectiveness of bilateral cochlear implantation. Laryngoscope. 2015; 125:442-447.

62. Remenschneider AK, Owoc M, Kozin ED, McKenna MJ, Lee DJ, Jung DH. Health Utility Improves After Surgery for Superior Canal Dehiscence Syndrome. Otol Neurotol. 2015; 36:1695-1701.

Figure legends

Figure 1.

Schematic overview of the search strategy.

Figure 2.

Flowchart for article exclusion with the number of articles excluded.

Figure 3.

Proportion of symptoms reported by SCDS patients.

Supplementary digital content

Supplementary digital content e-1.

Risk of bias assessment

Supplementary digital content e-2.

Articles retained for quantitative analysis.

Supplementary digital content e-3.

Quantitative short list.