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Review Article

Scoping review of cochlear implantation in Susac's syndrome

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KEYWORDS
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Abstract *Objective:* Scoping review of published literature to establish clinical characteristics and audiologic outcomes in patients diagnosed with Susac's Syndrome(SS) who have undergone cochlear implantation (CI).
Data sources: All published studies of CI in SS and contribution of two of our own patients who have not been reported previously.
Methods: A comprehensive search of MEDLINE (via PubMed) was carried out in March 2020 using the following keywords and related entry terms: Susac's Syndrome, Cochlear Implantation.
Results: Our search identified a total of five case reports of CI in SS. With the addition of our two patients reported here, we analyzed characteristics and outcomes in seven patients. Mean age at implantation was 30 years old (range 19–46), with six women and one man implanted. Mean time from onset of hearing loss to implantation was 17 months (range three months to four years). Best reported postoperative speech understanding was reported via different metrics, with six of seven patients achieving open set speech scores of 90% or better, and one subject performing at 68%. Vestibular symptoms were present preoperatively in four of seven patients (57%), with vestibular testing reported in two patients, and showing vestibulopathy in one patient. No complications were reported following cochlear implantation.
Conclusion: Cochlear implantation is a viable option for hearing rehabilitation in patients with SS, with levels of attainment of open set speech comparable to other populations of CI candidates.

Abbreviations: SS, Susac's syndrome; CI, Cochlear implantation; SNHL, Sensorineural hearing loss.

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Introduction

Susac's Syndrome (SS) is a microangiopathy that affects the brain, retina, and cochlea, classically presenting with the clinical triad of encephalopathy, branch retinal artery occlusion, and sensorineural hearing loss (SNHL) first described by John Susac in 1979.[1] With just over 400 cases reported in the published literature and a highly variable presentation that overlaps with other more common entities, SS often presents a diagnostic quandary. Patients are commonly women aged 20—40. The underlying pathophysiology is believed to be an acquired autoimmune endotheliopathy that results in microinfarctions in the brain, retina, and inner ear.[2—4]

As part of a multidisciplinary team engaged in the evaluation and management of SS patients, Otolaryngologists play a key role in the diagnosis and treatment of vestibulocochlear insults. In its most extreme audiologic presentation, SS results in bilateral profound SNHL, for which cochlear implantation (CI) has been successfully performed to rehabilitate hearing. In this article, we review the five previous reported cases of cochlear implantation for SS and contribute our own two cases of CI in this rare disease.

Clinical case number one

Patient one's initial symptoms beganat age 26 with right-sided sudden hearing loss in June 2017, followed several days later by right hemiparesis, for which she presented to an outside hospital. MRI brain with and without contrast demonstrated numerous white matter lesions in a periventricular and subcortical distribution with involvement of the corpus callosum and infratentorium on T2-weighted sequences. She was diagnosed with multiple sclerosis and treated with three days of intravenous steroids followed by physical therapy, with near-complete recovery of her hemiparesis but no change in her right-sided hearing loss. Two months later, she developed sudden-onset left SNHL, with audiogram showing bilateral severe SNHL affecting the low and mid frequencies, upsloping to a mild hearing loss in the upper frequencies (Fig. 1A). Word recognition scores were 0% bilaterally. Despite no subjective vision changes, she was noted to have branch retinal artery occlusions on fundoscopy. A diagnosis of SS was made and she was referred to our clinic for further evaluation.

On our initial evaluation, she reported no improvement in bilateral hearing loss. She denied any vertigo but described a sense of mild imbalance since onset of right-sided hearing loss. She had a normal exam of the external and middle ear bilaterally and communicated with lip reading and written text. Cochlear implant candidacy testing demonstrated scores on HINT sentences of 0 (right) and 4% (left), and on AZBio sentences 0 (right) and 1% (left), performed in quiet. Thin-cut CT demonstrated normal temporal bone anatomy. We proceeded with right cochlear implantation in November 2017, with uncomplicated placement of a MedEl Synchrony device with full insertion of a Flex 28 electrode. The device was activated four weeks post-op, and testing at one week post-activation showed a HINT sentences score of 71% on the right in quiet. At 15 months post-activation, she had scores on HINT sentences in quiet of 100%, AZBio sentences 94% in quiet, AZBio sentences +5 dB signal-to-noise ratio of 47%, CNC words 80% and CNC phonemes 93%.

Clinical case number two

Patient two is a 43-year-old woman with a history of bilateral Eustachian tube dysfunction and bilateral post-tubal tympanic membrane perforations who developed sudden onset vision changes in October 2017, for which she presented to the ED. She had no headache or other neurological complaints and denied acute hearing change. MRI brain with and without contrast demonstrated few scattered nonspecific foci of subcortical white matter T2 signal abnormality within the frontal lobes, and mild T2 hyperintensity involving the corpus callosum. Fluorescein angiography demonstrated subtle retinal arteriole occlusion supportive of a diagnosis of SS. Rheumatology agreed with a diagnosis of SS and initiated daily PO prednisone and mycophenolate.

Five months after her initial visual symptoms, she experienced sudden hearing loss bilaterally, with audiogram showing left sensorineural and right mixed hearing loss (Fig. 1B). Exam demonstrated right larger than left tympanic membrane perforations. She received three rounds of bilateral intratympanic dexamethasone injections without change in hearing and was then referred to us. Serial audiograms over the next year demonstrated bilateral fluctuating and progressive SNHL, despite aggressive medical treatment, with diminishing benefit from bilateral amplification. We performed successful right medial graft tympanoplasty and subsequently administered cochlear implant candidacy testing. This showed scores on HINT sentences of 0 (right) and 9% (left), AZBio sentences 0 (right) and 44% (left) in quiet and 0% (right) and 0 (left) in +5 dB signal-to-noise testing.

She underwent right cochlear implantation two years after her initial sudden hearing loss with a Cochlear Corporation Nucleus CI612 device, with full insertion with normal impedance testing and good waveforms on intraoperative neural response telemetry. Two months after activation of her device, she scored 90% correct on HINT sentences, 65% correct on AzBio sentences, 42% correct on CNC words, and 70% correct on CNC phonemes.

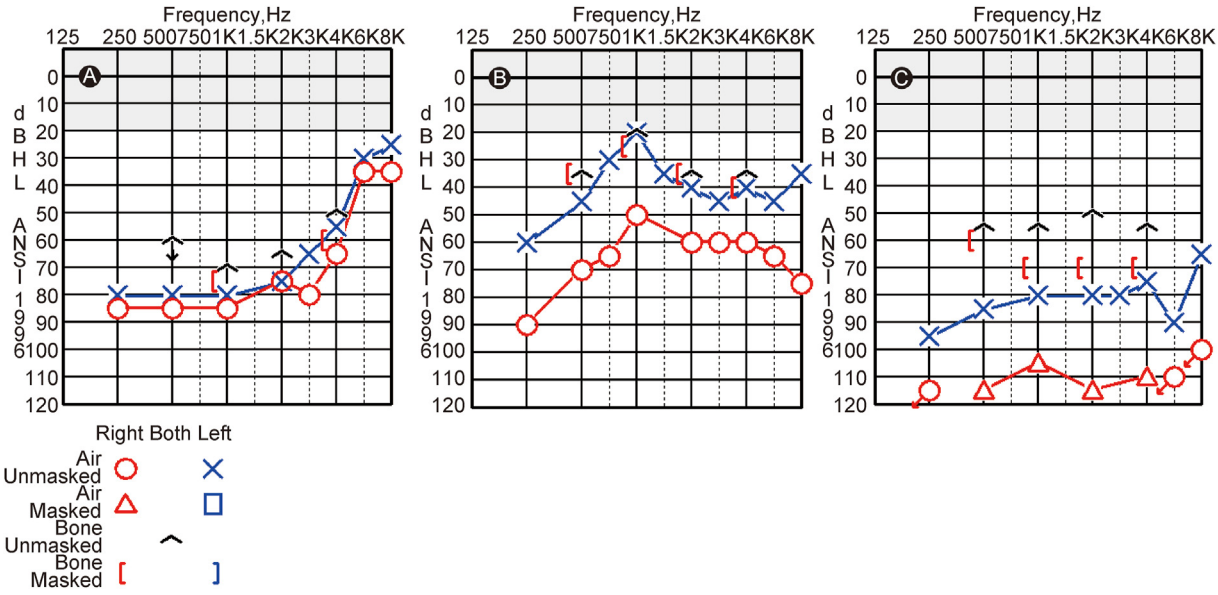

Figure 1 Preoperative audiograms for Patient one (A) showing classic upsloping sensorineural hearing loss and for Patient two (B, C), with demonstration of progressive bilateral loss despite medical therapy.

She subsequently began to experience subjective fluctuations in performance of her right CI that sometimes correlated with worsening of her migraines. She described decreased clarity of hearing on the right that could fluctuate from week to week, as well as fluctuations in hearing on her unimplanted side, with audiograms confirming changing SNHL on the left during this period. At CI programming appointments over the six months following implantation, she had testing at one visit showing shorted electrodes #11 and #20, which then returned to normal impedances at subsequent visits. Integrity testing performed with Cochlear Americas personnel did not identify any device malfunction. The patient consistently scored around 90% on HINT at 70 dB SPL on repeated testing of her implanted ear throughout this period of subjective fluctuation of CI performance. She has continued to manifest fluctuating hearing changes despite modifications of her medications.

Methods

A literature search was conducted to establish patient characteristics and outcomes in all patients, adults or children, with a diagnosis of SS who underwent cochlear implantation. All studies were considered eligible for inclusion, including abstracts of poster presentations, case reports, series, reviews, expert opinions, and randomized controlled trials. A comprehensive search of the MEDLINE database (via PubMed) was performed in March, 2020 using the following terms: (((("susac syndrome"[MeSH Terms] OR ("susac"[All Fields] AND "syndrome"[All Fields])) OR "susac syndrome"[All Fields]) OR ("susac s"[All Fields] AND "syndrome"[All Fields])) OR "susac syndrome"[All Fields]) AND (("cochlear implantation"[MeSH Terms] OR ("cochlear"[All Fields] AND "implantation"[All Fields])) OR "cochlear implantation"[All Fields]). All resulting studies were read in their entirety. Data on the clinical characteristics of patients with SS, including demographics, subjective hearing loss, tinnitus, or vestibular symptoms, time between onset of hearing loss and implantation, reported intraoperative findings and any complications, pre-operative and post-operative audiologic data, and vestibular testing were collected. The principal summary measure included attainment of open set speech following cochlear implantation. Risk of bias in reporting results of successful audiologic outcome following CI was a concern for this review.

Results of cochlear implantation in Susac's syndrome

Our search of the literature returned five studies in English and one in French, all of which were read in their entirety. Four studies in English were case reports of cochlear implantation in SS and were included in this review. One publication in English and the publication in French did not include any report of cochlear implantation in SS and both were excluded. One additional abstract from a poster presented at a national scientific conference was identified in the references of the above articles and was included in this analysis. Table 1 displays the findings of the seven patients with cochlear implantation performed for SS in the published literature, including the two cases presented in this article. Mean age at implantation was 30 years old (range 19—46), with six women and one man implanted. Mean time from onset of hearing loss to implantation was 17 months (range three months to four years). Best reported postoperative speech understanding was reported via different metrics, with six of seven patients achieving open set speech scores of 90% or better, and one subject performing at 68%. Time from implantation to reported speech perception result ranged from two to sixteen months. Vestibular symptoms were present preoperatively in four of seven patients (57%), with vestibular testing reported in

Table 1 Clinical characteristics and outcomes in patients with Susac's syndrome treated with cochlear implantation.

Patient	Publication	Age at implantation (years)	Time between hearing loss onset and CI (months)	Side of implant	Best reported speech understanding(%)	Audiologic metric	Time from surgery to testing result (months)	Vestibular symptoms
1	Connell and Brodie 2004	39	NR	NR	94	"Hearing in noise"	6	No
2	Roeser et al 2008	19	3	Bilateral	98	AZBio	2	No
3	Bittencourt et al 2011	18	48	Left	100	"Open set sentences in noise"	6	No
4	Grover et al 2011	36	9	Left	68	Bamford-Koval-Bench	6	Yes
5	Lavinsky et al 2012	29	18	Right	100	"Open field"	12	Yes
6	Perez et al 2020	26	5	Right	100	HINT in quiet	16	Yes
7	Perez et al 2020	46	21	Right	90	HINT in quiet	3	Yes

NR: not reported.

two patients, and showing vestibulopathy in one patient. No complications were reported following cochlear implantation.

Disease presentation

The diagnosis of SS is complicated by its rarity, variable presentation, and clinical overlap with other more common entities. Brain, retina, and vestibulocochlear involvement are required for definite diagnosis, but the triad is complete in only 13% of patients on presentation, requiring consideration of this diagnosis in the absence of full criteria being met.[5] In the largest review of 304 patients, the mean age of onset was 31.6 years, with a range from 8 to 65 years, and a female-to-male ratio of 3.5:1. [5]Subacute encephalopathy, focal neurological deficits, and headache are the most common CNS presentations.[3,6,7] Visual symptoms include visual field loss and central or paracentral scotoma, which are reflective of underlying retinal ischemia, although some patients are asymptomatic despite an abnormal ophthalmologic exam.[8,9] Sudden SNHL in one ear followed in short order by a similar loss in the other ear, described as "bang, bang" hearing loss, is a classic presentation, with tinnitus, vertigo, and more subacute or fluctuating SNHL being common as well.[3,10]

Diagnosis

In 2016, the European Susac Consortium published diagnostic criteria and recommended workup, including brain MRI, fluorescein angiography, fundoscopy, or optical coherence tomography, and audiogram or vestibular testing.[11] On MRI, characteristic small, round "snowball" lesions are seen affecting the central corpus callosum, which are hyperintense on T2 and FLAIR sequences and hypointense on T1 sequences. Branch retinal artery occlusion is seen on fluorescein angiography, and Gass plaques, which are yellow refractile lesions that simulate emboli along retinal arteries, may be seen on fundoscopy.[8,12]

An audiogram will commonly show asymmetric bilateral SNHL that classically affects the low and middle frequencies, with relative sparing of upper frequencies, although a flat loss may also be seen.[10,13] This pattern, along with the fluctuations that can be seen in SNHL over time, may be confused for a diagnosis of Meniere's disease. Some series have reported greater than 50% of patients presenting with only a unilateral SNHL, though limited duration of follow up may not as yet have captured those who progress to bilateral loss.[10] When performed, auditory brainstem responses have been shown to be intact and correlate with pure tone averages.[14] Normal electrocochleography and absent otoacoustic emissions have also been recorded in active SS.[14] Caloric testing, evoked myogenic responses, nystagmography, and video head impulse testing have all been used to identify peripheral vestibulopathy in patients with SS, although symptoms of dizziness may also be caused by central lesions.[13,15]

A number of laboratory tests have been considered in the workup of SS, including rheumatologic panels of antibody markers, hematologic and coagulation panels, and CSF analysis, the last of which being helpful in distinguishing similarly-presenting multiple sclerosis from SS.[9,13] To date, however, no laboratory assays have demonstrated adequate sensitivity or specificity to confirm a diagnosis of SS.[10]

Pathogenesis

SS is believed to result from an autoimmune endotheliopathy that targets the microvasculature of the brain, CNS, and inner ear. Brain biopsies have revealed microinfarcts with associated arteriolar wall proliferation, lymphocytic infiltration, and basal lamina thickening.[16,17]

Francis and colleagues have provided the only description of the histopathologic changes affecting the inner ear in active SS through temporal bone histopathology in a patient who died from a pulmonary embolus while still hospitalized for her recent diagnosis of SS.[4] Findings were notable for widespread atrophy of the inner and outer hair

cells, stria vascularis, tectorial membrane, spiral ligament, as well as capillary occlusion of the stria vascularis, with these changes restricted to the apical halves of both cochleae. The vestibular organs were unremarkable and no endolymphatic hydrops was present. These histologic changes, without any evidence of infection or inflammation, support an underlying pathophysiology of microvascular occlusion leading to SNHL in SS.

Treatment

No prospective studies of treatment for SS exist, with clinical experience and case reports guiding current recommendations. Phases of treatment can be divided into initial therapy, maintenance therapy, and therapy for relapses. Treatment directed at the CNS sequelae of this disease will usually adequately treat retinal and vestibulocochlear manifestations of SS. In general, early and aggressive treatment has been promoted to prevent the irreversible damage that can occur in SS.

The most commonly recommended initial therapy is intravenous methylprednisolone 1 g daily for 3—10 days followed by high dose prednisone, 1 mg/kg/day up to 80 mg PO daily tapered over a period of weeks, in addition to IVIG tapered over six months.[12,18] Common maintenance therapies include cyclophosphamide, mycophenolate mofetil, tacrolimus, and rituximab, with varying use depending on severity of disease, commonly continued for a period of two years.[7,9,18,19] Aspirin has also been recommended as an adjunctive agent, despite limited evidence regarding its efficacy.[19,20] Relapses may be treated with additional pulses of IV methylprednisolone and, in severe cases, plasma exchange.[18,21] Monitoring of disease is recommended with interval MRI brain, fluoroscopic angiography, and audiogram.

Intratympanic steroid injection for SNHL has been performed, with mixed results reported in case studies.[10,22] One of our presented patients had no improvement with three rounds of IT dexamethasone. Rehabilitation of hearing loss should be addressed with amplification when appropriate and an understanding of the potential for progression of SNHL. Cochlear implantation has provided significant speech perception benefit in all published cases so far.

Discussion of cochlear implantation in Susac's syndrome

In their review of 304 published cases of SS, Dorr et al[5] noted that 99% of patients had hearing loss. In its least injurious form, the SNHL associated with SS may lead to only a mild unilateral loss with minimal functional impact on the patient. Unfortunately, much more severe presentations are common, including bilateral profound loss with zero speech recognition, conferring significant morbidity on the patient. Roeser et al[10] reviewed comprehensive audiologic data for 32 ears affected by SS and identified 9 (26.5%) with word recognition scores $<$50%. In this latter scenario, cochlear implantation has been demonstrated to be a viable option for hearing rehabilitation.

Following the first report in 2004 by Connell and Brodie, a total of seven cases of CI in SS have now been published, including the two presented here.[10,23—26] Despite heterogeneity of audiologic metrics used, the open set speech understanding attained following implantation was 90% or greater for six out of seven patients, with the seventh measured at 68%, demonstrating that SS patients perform well with respect to the average CI user (Table 1). These successful hearing outcomes are similar to those published for cochlear implantation in autoimmune hearing loss, in which either improved performance or no statistical difference has been found in long term word and sentence scores when compared to results in control groups receiving CI.[27,28] In addition, despite the progressive SNHL seen in autoimmune hearing loss, long-term follow up of patients with Cogan syndrome who underwent cochlear implantation has demonstrated sustained excellent performance.[29]

The presentation of hearing loss in the SS patients discussed here was reflective of that which has been described more broadly in published cases of SS, including the classic "bang, bang" sudden SNHL affecting one followed by the other ear in some, as well as SNHL that fluctuates over more than one year's duration.[10] Published audiograms for some of these implanted patients also demonstrated the common finding of preferential loss of low and mid frequencies with relative sparing of the upper frequencies.[10,13] This pattern, along with histopathological evidence of SS preferentially affecting the apical cochlea, provides an argument for use of a full-length electrode when considering cochlear implantation for this disease. Roeser et al[10] noted intraoperative neural response telemetry with higher amplitudes in the basal and mid electrodes consistent with improved neuron survival in the basal turn of the cochlea in their report, a pattern that was not borne out in our intraoperative telemetry.

Our second patient presented here has demonstrated subjective fluctuations in her CI performance that sometimes correlated with migraine and fluctuating SNHL in her unimplanted ear, though she continued to perform well on objective testing of open set speech during this period. The loss of otoacoustic emissions and histopathological changes seen in SS demonstrate an underlying cochlear pathology that has been widely accepted in the literature. Notably, Bateman et al[14] presented a case of SS with fluctuating SNHL in which auditory brainstem responses demonstrated a prolongation of I—V wave latency (that ultimately remained within normal limits) over the course of months, suggesting the possibility of a simultaneous retrocochlear process in this disease. While no previous cases of cochlear implantation in SS have noted fluctuating CI performance, our experience prompts consideration of additional transient central pathology that compromises implant performance. Although the underlying pathophysiology is felt to differ, there are case reports of delayed CI performance degradation in patients with Cogan's syndrome and relapsing polychondritis; still, stable objective hearing outcomes are achieved in the majority of these cases.[29—31]

The unexplained subjective fluctuating cochlear implant performance in one of our patients prompted us to share this experience. We acknowledge both the small sample

size of CI in SS and the reporting bias against the publication of cochlear implantation with variable or poor audiologic outcome in this patient population. These factors somewhat limit conclusions that can be drawn from this review and reiterate the need for further study. Taken in aggregate, previous clinical experience still suggests that CI should be offered to SS patients when audiometric criteria are met.

Although approximately one third of patients who are ultimately diagnosed with SS initially present with SNHL, the average time from presentation to completion of the full clinical triad has been estimated at 21 weeks.[5] This combined with the sometimes fluctuating and progressive course of SNHL require continued audiologic follow up and patient education on the variability of the natural history of this disease. While CNS involvement can be devastating and often mandates aggressive medical treatment of SS, concurrent SNHL should also be monitored closely and amplification or implantation recommended when appropriate.

Conclusion

A rare microvasculopathy that affects the brain, retina, and inner ear, Susac's syndrome presents both a diagnostic and therapeutic challenge that requires multidisciplinary evaluation and treatment. The paucity of cases and absence of prospective studies in SS result in a reliance on clinical experience and expert opinion. The SNHL seen in SS presents with variable severity and progression. In cases of significant bilateral loss with limited benefit from hearing aid amplification, cochlear implantation has been shown to successfully improve speech understanding in all published cases so far and is a reasonable means of aural rehabilitation in the setting of SS.

Declaration of competing interest

None of the authors have any conflicts of interest related to this study to disclose.

References

1. Susac JO, Hardman JM, Selhorst JB. Microangiopathy of the brain and retina. *Neurology*. 1979;29:313—316.
2. Magro CM, Poe JC, Lubow M, Susac JO. Susac syndrome: an organ-specific autoimmune endotheliopathy syndrome associated with anti-endothelial cell antibodies. *Am J Clin Pathol*. 2011;136:903—912.
3. Susac JO, Egan RA, Rennebohm RM, Lubow M. Susac's syndrome: 1975-2005 microangiopathy/autoimmune endotheliopathy. *J Neurol Sci*. 2007;257:270—272.
4. Francis HW, Makary C, Halpin C, Crane BT, Merchant SN. Temporal bone findings in a case of Susac's syndrome. *Otol Neurotol*. 2011;32:1198—1204.
5. Dörr J, Krautwald S, Wildemann B, et al. Characteristics of Susac syndrome: a review of all reported cases. *Nat Rev Neurol*. 2013;9:307—316.
6. Zengin Karahan S, Boz C, Saip S, et al. Susac syndrome: clinical characteristics, diagnostic findings and treatment in 19 cases. *Mult Scler Relat Disord*. 2019;33:94—99.
7. Greco A, De Virgilio A, Gallo A, et al. Susac's syndrome-pathogenesis, clinical variants and treatment approaches. *Autoimmun Rev*. 2014;13:814—821.
8. Heng LZ, Bailey C, Lee R, Dick A, Ross A. A review and update on the ophthalmic implications of Susac syndrome. *Surv Ophthalmol*. 2019;64:477—485.
9. García-Carrasco M, Mendoza-Pinto C, Cervera R. Diagnosis and classification of Susac syndrome. *Autoimmun Rev*. 2014;13:347—350.
10. Roeser MM, Driscoll CL, Shallop JK, Gifford RH, Kasperbauer JL, Gluth MB. Susac syndrome-a report of cochlear implantation and review of otologic manifestations in twenty-three patients. *Otol Neurotol*. 2009;30:34—40.
11. Kleffner I, Dörr J, Ringelstein M, et al. Diagnostic criteria for Susac syndrome. *J Neurol Neurosurg Psych*. 2016;87:1287—1295.
12. Ammar MJ, Kolomeyer AM, Bhatt N, Tamhankar MA, Mullen MT, Brucker AJ. Recurrent branch retinal artery occlusion from Susac syndrome: case report and review of literature. *Retin Cases Brief Rep*. 2020;14:315—320.
13. Patel VA, Dunklebarger M, Zacharia TT, Isildak H. Otologic manifestations of Susac syndrome. *Acta Otorhinolaryngol Ital*. 2018;38:544—553.
14. Bateman ND, Johnson IJ, Gibbin KP. Susac's syndrome: a rare cause of fluctuating sensorineural hearing loss. *J Laryngol Otol*. 1997;111:1072—1074.
15. Marrodan M, Laffue A, Fiol MP, Correale J, Gualtieri F. Video head impulse test contributes to Susac syndrome diagnosis. *J Neurol*. 2019;266:2070—2072.
16. O'Halloran HS, Pearson PA, Lee WB, Susac JO, Berger JR. Microangiopathy of the brain, retina, and cochlea (Susac syndrome). A report of five cases and a review of the literature. *Ophthalmology*. 1998;105:1038—1044.
17. Fox RJ, Costello F, Judkins AR, et al. Treatment of Susac syndrome with gamma globulin and corticosteroids. *J Neurol Sci*. 2006;251:17—22.
18. Rennebohm RM, Asdaghi N, Srivastava S, Gertner E. Guidelines for treatment of Susac syndrome - an update. *Int J Stroke*. 2020;15:484—494.
19. Vodopivec I, Prasad S. Treatment of susac syndrome. *Curr Treat Options Neurol*. 2016;18:3.
20. Kleffner I, Duning T, Lohmann H, et al. A brief review of Susac syndrome. *J Neurol Sci*. 2012;322:35—40.
21. Capiluppi E, Romano L, Luccarelli SV, Macerollo A, Cislaghi G. Aggressive immunosuppression in Susac's syndrome: 10 years of follow-up. *Neurol Sci*. 2018;39:1807—1809.
22. Crawley BK, Close A, Canto C, Harris JP. Susac's syndrome: intratympanic therapy for hearing loss and a review of the literature. *Laryngoscope*. 2009;119:141—144.
23. Lavinsky L, Scarton F, Lavinsky-Wolff M, Lavinsky J, Motta LH. Successful cochlear implantation in a Susac syndrome patient. *Braz J Otorhinolaryngol*. 2012;78:123.
24. Grover N, Whiteside OJ, Ramsden JD. Susac syndrome: outcome of unilateral cochlear implantation. *J Laryngol Otol*. 2011;125:856—858.
25. Connell SS, Brodie HA. Role of cochlear implantation in Susac's syndrome. *Otolaryngol Head Neck Surg*. 2004;131, P280.
26. Bittencourt AG, Santos AF, Goffi-Gomez MV, et al. Microangiopathy of the inner ear, deafness, and cochlear implantation in a patient with Susac syndrome. *Acta Otolaryngol*. 2011;131:1123—1128.
27. Aftab S, Semaan MT, Murray GS, Megerian CA. Cochlear implantation outcomes in patients with autoimmune and immune-mediated inner ear disease. *Otol Neurotol*. 2010;31:1337—1342.
28. Wang JR, Yuen HW, Shipp DB, et al. Cochlear implantation in patients with autoimmune inner ear disease including cogan syndrome: a comparison with age- and sex-matched controls. *Laryngoscope*. 2010;120:2478—2483.

29. Bacciu A, Pasanisi E, Di Lella F, Guida M, Bacciu S, Vincenti V. Cochlear implantation in patients with Cogan syndrome: long-term results. *Eur Arch Otorhinolaryngol*. 2015;272:3201—3207.
30. Low WK, See JY, Ng WN, Fong A, Chew SH. An unusual case of post-cochlear implant performance degradation in a patient with suspected Cogan's syndrome. *Cochlear Implants Int*. 2019;20:94—99.
31. Patrizia M, Giuseppe A, Marika V, Roberto F. Deterioration of hearing in a cochlear implantee with relapsing polychondritis. *Acta Otolaryngol*. 2011;131:675—678.

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