The Journal of Laryngology & Otology
November 2002, Vol. **116,** pp. 946–950

Scala vestibuli cochlear implantation in patients with partially ossified cochleas

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Abstract

Partial cochlear obstruction is a relatively common finding in candidates for cochlear implants and frequently involves the inferior segment of the scala tympani in the basal turn of the cochlea. In such patients, the scala vestibuli is often patent and offers an alternative site for implantation. The current report describes two patients with such partial obstruction of the inferior segment of the basal cochlear turn, caused in one case by systemic vasculitis (Takayasu's disease) and in the other by obliterative otosclerosis. A scala vestibuli implantation allowed for complete insertion of the electrode array. No problems were encountered during the surgical procedures and the good post-operative hearing and communicative outcomes achieved were similar to those reported in patients without cochlear ossification.

The importance of accurate pre-operative radiological study of the inner ear is underscored, to disclose the presence and define the features of the cochlear ossification and ultimately to properly plan the surgical approach.

Key words: Cochlear Implants; Scala Vestibuli; Ossification, Heterotopic

Introduction

Cochlear obstruction occurs in about 10–15 per cent of all candidates for cochlear implant (CI)[1,2] and in about 80 per cent of post-meningitic CI candidates.[3] The most common cause is meningitis, but obstruction has also been reported in cases of otosclerosis, autoimmune inner ear diseases, temporal bone fractures, otitis media, labyrinthectomy and other conditions.[4–10] Cochlear obstruction may be due to the presence of fibrous or bone tissue, in the form of softly calcified bony matrix as well as chalky and densely calcified new bone.[6]

For many years cochlear implantation was contra-indicated in cases of cochlear ossification, but it is now practised in both the partial and complete forms of this condition[1,2,5,9,11,12] and many surgical techniques have been proposed for both partially and totally ossified cochleas.[2,6,7,9,11,13–15] Recent clinical reports describe good, albeit variable hearing outcomes in such patients.[1,2,5,9,11,12]

This report describes the clinical features and surgical findings and results in two patients suffering from partial ossification of the basal turn of the cochlear scala tympani, who were treated by electrode array implantation in the scala vestibuli according to the surgical technique proposed by Steenerson *et al.*[11]

Case reports

Case 1 A 35-year-old female English language teacher, complained of episodes of fluctuating left-ear hearing loss with vertigo (Ménière-like symptoms) when she was 29 years old. Three years later, at age 32, her hearing function worsened bilaterally and pure-tone audiometry revealed left-ear deafness and a moderate right sensorineural hearing loss (SNHL), so she was fitted with a hearing aid on the right side, with good hearing outcomes. At age 34, she experienced worsening of her right-side hearing and vertigo, and presented left-ear deafness and profound right SNHL. Attempts at correction through a right-side hearing aid afforded little hearing or communicative improvement; she was also treated with corticosteroids and hyperbaric oxygen therapy, that were ineffective.

At age 35 the patient was referred to us and was subjected to complete medical and audiological evaluation. Pure-tone audiometry revealed left-ear deafness and profound right SNHL (Figure 1(a)). A vestibular function examination showed bilateral labyrinthine areflexia. Speech audiometry in open field with, and without, amplification showed no word discrimination. The patient also performed a speech perception test[16] and presented no open-set speech discrimination without lip-reading with the hearing aid (Figure 1(c)).

Her medical history revealed that she had been diagnosed some years earlier as suffering from ulcerative colitis and an autoimmune inner ear disease was suspected.[17,18] She was subjected to a complete blood and immunological evaluation, according to our protocol for progressive SNHL;[19] all test results were normal except for: erythrocyte sedimentation rate (ESR) 30 mm/h, C-reactive protein (CRP) 8 mg/dl, fibrinogen 130 mg/dl, and alpha-1 glucoprotein 146 mg/dl. Serum assay for antibiotics to inner-ear antigen was also conducted by Western Blot, using fresh bovine temporal-bone inner-ear antigen,[20,21] and no evidence of cross-reacting auto-antibodies was

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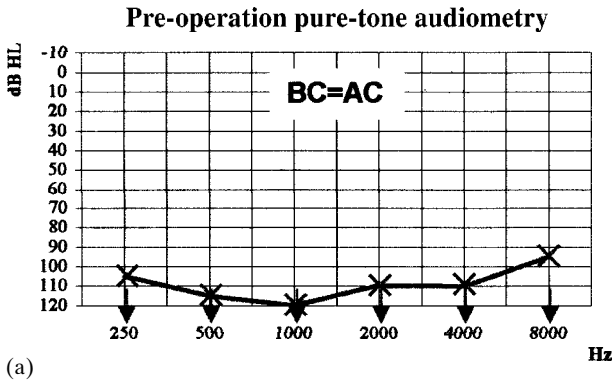
Accepted for publication: 30 April 2002.

https://doi.org/10.1258/00222150260369516 Published online by Cambridge University Press



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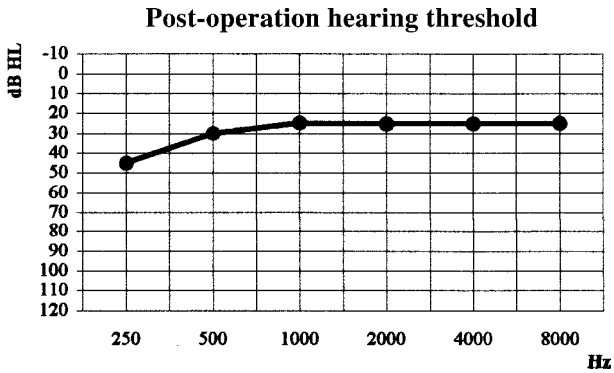

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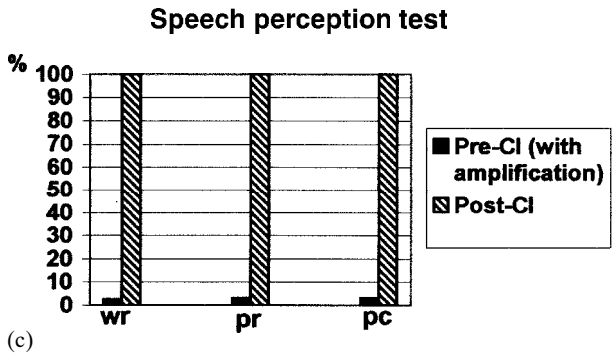

(c)

FIG. 1

(a) A pure-tone audiometry of *Case 1*, executed before cochlear implantation, shows a right deafness and a left profound hearing loss. (b) A pure-tone audiometry in open-field of the same patient, performed after cochlear implantation, shows a good hearing threshold. (c) Before cochlear implantation the patient presented a total absence of word and phrases recognition (wr, pr) and of phrases comprehension (pc) abilities, without lipreading and with amplification; after cochlear implantation the patient reached a 100 per cent word and phrases recognition and a 100 per cent phrases comprehension without lipreading.

found. Contemplating an autoimmune disorder, it was decided to have the patient undergo immunological and gastro-enterological evaluations, which both seemed to exclude systemic vasculitis, so the diagnosis of ulcerative colitis was accepted. The patient also underwent high-resolution, petrous bone spiral computed tomography (spiral HR-CT), which raised suspicions of bilateral cochlear stenosis (Figure 2(a)). High-resolution magnetic resonance imaging (HR-MRI) of the inner ear performed at high field (Figure 2(b)), with integration of three-dimensional reconstructions (Figure 2(c),(d)), confirmed symmetric bilateral partial obstruction of the inferior segment of the scala tympani of the basal turn of the cochlea, without any other middle or inner ear abnormalities. The promontory test revealed good responses bilaterally. We performed a right side cochlear implant, using a Nucleus 24 CI (CI24M-Cochlear).

As expected, during surgery we found obstruction of the round window and the scala tympani, so it was decided to insert the electrode array into the scala vestibuli. A cochleostomy was drilled anterior-superior to the round window, thereby opening the scala vestibuli, which appeared patent. The entire electrode array was introduced, including most of the stiffening rings, into the scala vestibuli without any difficulty. A skull radiograph in a modified Stenver's view was performed, two days after surgery, to confirm correct positioning of the array. Three months after activation of the CI, the patient was subjected to the same speech perception test[16] performed before implantation, with excellent outcomes: she presented a 100 per cent open-set word and phrase recognition and a 100 per cent phrase comprehension without lip-reading (Figure 1(b),(c)); she was also able to have a telephone conversation with anyone. Ten months after cochlear implantation the patient presented with a sclerotic obstruction of the coronary, femoral and renal arteries: after a complete medical evaluation, the diagnosis of Takayasu's arteritis was made, so the patient was subjected to aortic-coronary by-pass and immunosuppressive therapy with cyclosporin and high-dose corticosteroids, with complete remission of the acute pathology. Currently, three years after the CI, the patient is in good health: her left hearing threshold has remained stable, despite immunosuppressive therapy, and her communicative performance is excellent: she is able to have a telephone conversation with anyone, listen to music and has continued her career teaching English.

Case 2 A 38 year old female, complained of mild bilateral SNHL at age 17. At age 24 her hearing function began worsening bilaterally and she was fitted with bilateral hearing aids, with good communicative results. Over subsequent years, however, her hearing threshold worsened, until age 35, when she complained of profound bilateral hearing loss which hearing aids were no longer able to compensate.

At age 38 the patient was referred to us and was given a complete audiological evaluation. Pure-tone audiometry revealed profound bilateral hearing loss (Figure 3(a)); a vestibular function test showed right labyrinthine hyporeflexia. Speech audiometry in open field with, and without, hearing aids revealed a total lack of word discrimination ability. The patient moreover performed a speech perception test,[16] that showed no open-set speech discrimination ability without lip-reading despite the hearing aids (Figure 3(c)). HR-CT showed bilateral cochlear otospongiotic changes, involving the promontory, (i.e. the bony covering of the basilar turn of the cochlea) with complete obliteration of the round window. These images were suggestive of cochlear otosclerosis. HR-MRI and three-dimensional reconstructions were useful in determining the presence of stenosis of the inferior segment of cochlear basal turn bilaterally. A cochlear implant was performed in the right ear, using a Nucleus 24 CI (CI24M-Cochlear).

During surgery we found the promontory bone to have a spongiotic aspect, with complete obstruction of the oval and round windows and stapes fixation. We attempted to drill through the new bone in the round window and scala tympani region, but the scala tympani was obstructed as well, so we decided to insert the electrode array into the

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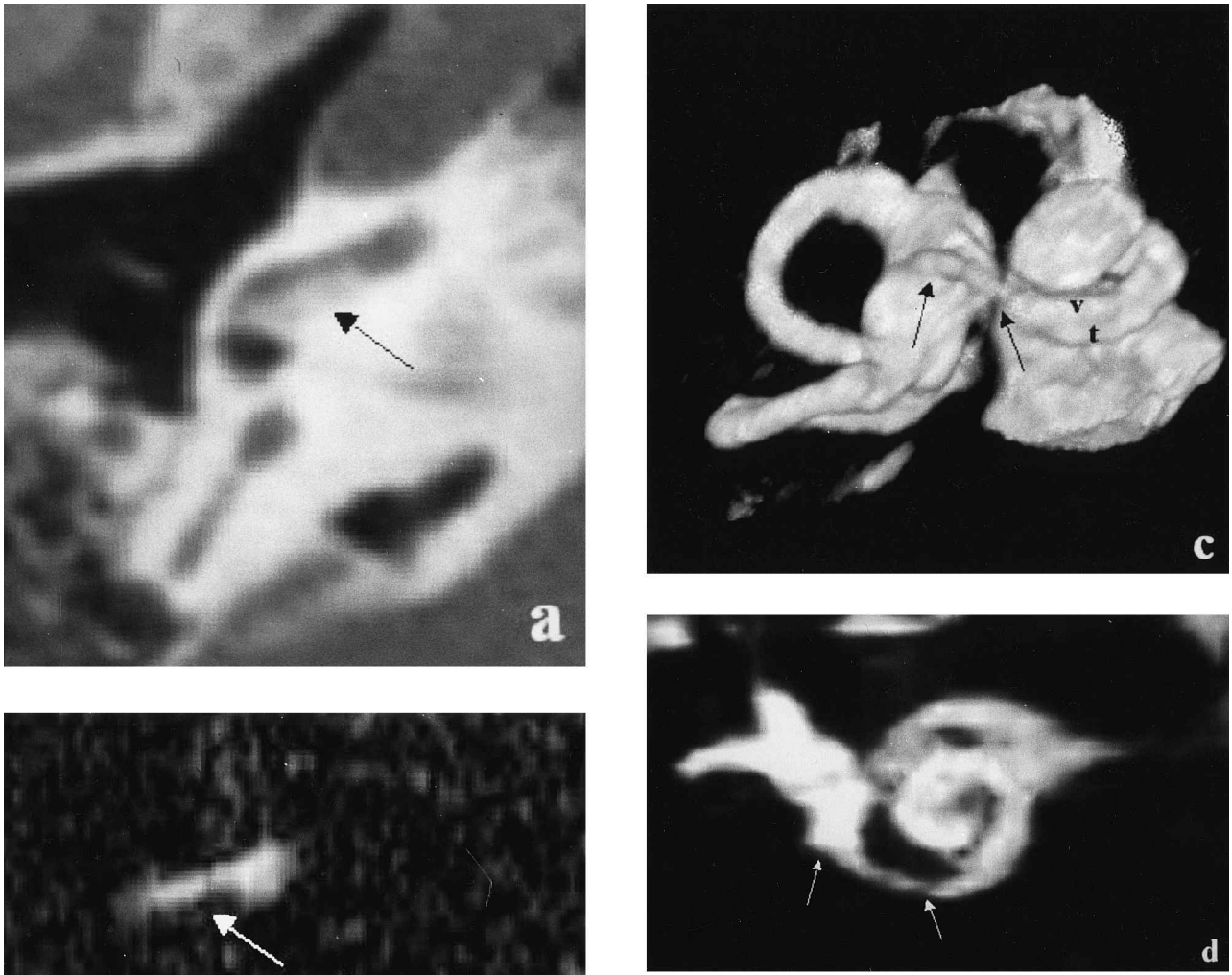

FIG. 2

Axial spiral HR-CT image of *Case 1* (a) that raised suspicions of cochlear stenosis; HR-MR (b) image confirmed the presence of a stenosis of the basal turn of the cochlea (arrows). (c) A three-dimensional reconstruction with volume rendering of the inner ear from HR-MR datasets. Scala tympani (t) and scala vestibuli (v) are visible; the obstruction of the inferior segment of the scala tympani of the basal turn of the cochlea is represented with exquisite detail (arrows). (d) Three-dimensional reconstruction with MIP (maximum intensity projection) of the inner ear shows the stenosis of the basal turn of the cochlea (arrows).

scala vestibuli. A cochleostomy was made anterior-superior to the round window, thereby exposing the scala vestibuli, which appeared patent. The entire electrode array, including all the stiffening rings, was inserted into the scala vestibuli without any difficulty.

A post-operative temporal bone radiograph in a modified Stenver's view showed good positioning of the electrode array. During the first fitting session one month after implantation, we noted a facial nerve stimulation by electrodes 17-to-12, which were therefore deactivated. One year after cochlear implantation, the patient underwent a speech perception test[16] and demonstrated a 95 per cent open-set word and 100 per cent phrase recognition and a 100 per cent phrase comprehension without lip-reading, despite deactivation of six of the electrodes (Figure 3(b),(c)).

Discussion

Cochlear obstruction or stenosis due to bone or fibrous tissue formation is a relatively common finding in candidates for cochlear implant.[4–10]

Most often cochlear ossification involves the basal turn of the cochlea, particularly its inferior segment and the scala tympani rather than the scala vestibuli, while a complete cochlear ossification is reportedly rare (< three per cent).[9,11–21] The reasons for this distribution of cochlear ossification is that in many cases such as the post-meningitic and autoimmune disease-related forms (see the first case reported above), labyrinthine ossification is a retrograde process, beginning with inflammation in the cochlear aqueduct region and spreading along the scala tympani to the rest of the cochlea; this process takes from one to two months to several years.[11] Also in otosclerosis,

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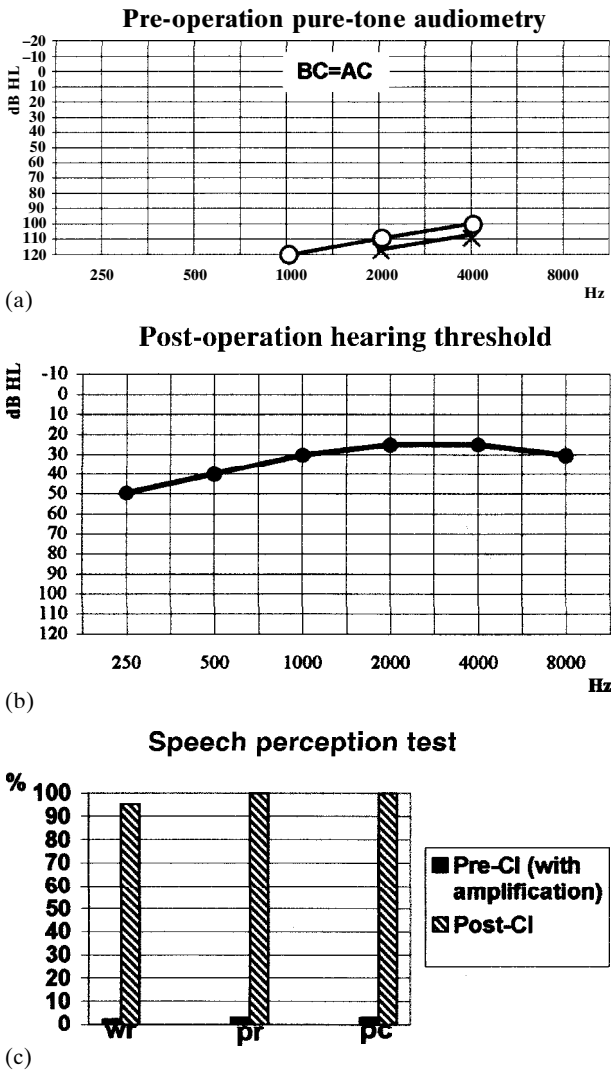

FIG. 3

(a) Pure-tone audiometry of *Case 2*, executed before cochlear implantation shows a bilateral profound hearing loss. (b) Pure-tone audiometry in open field of the same patient, performed after cochlear implantation, shows a good hearing threshold. (c) Before cochlear implantation the patient presented a total absence of word and phrases recognition (wr, pr) and of phrases comprehension (pc) abilities, without lip-reding and with amplification; after cochlear implantation the patient reached a 95 per cent word recognition and a 100 per cent phrases recognition and phrases comprehension without lip-reading.

such as the second case reported above, the most common finding is the involvement of both the round window and the inferior segment of the scala tympani of the basal turn, as the cochlear lumen may be narrowed by invasion of otosclerotic or otospongiotic bone that spreads from the round window area to the scala tympani of the cochlea.[4]

Moreover, partial obstruction of the inferior segment of the scala tympani in the basal cochlear turn is a frequent finding, although it is often misdiagnosed before surgery, especially if pre-operative assessment is made solely on the basis of petrous bone spiral HR-CT.

We wish to stress the importance of accurate pre-operative radiological evaluation, especially when cochlear obliteration is suspected. Spiral HR-CT, and HR-MRI should be performed to disclose the presence and exact extent and location of any cochlear obstruction. In the two cases reported, spiral HR-CT and HR-MR proved highly valuable, in that they revealed not only the presence of the cochlear obstruction, but provided precise indications of its localization and the involvement of the basal turn of the cochlea. The three-dimensional reconstructions moreover allowed us to better define the anatomy of the cochlea, the narrowing of the basal cochlear turn and the sole involvement of the scala tympani. The resulting accurate representation of the presence, extent and site of the cochlear obstruction proved crucial in enabling us to plan our surgical approach.[22]

The safety and efficacy of implants for partially or totally ossified cochleas have been widely demonstrated[1,2,5–7,9,11,12,23] and many surgical alternatives have been proposed for treating both partial and complete cochlear obliterations.[6,13,14,24] The reported hearing and communicative results after implant treatment of ossified cochleas are quite variable.[1,2,6,7,9,12,23]

The goal of CI surgery in treating cochlear ossification is complete insertion of the electrode array, despite cochlear obstruction. Clearly, it is to be expected that a partial insertion of the electrodes will lead to poor outcomes, although results have been quite variable.[9] Currently most authors agree that if complete electrode insertion is achieved, the results achievable in nearly all cases of partial obliteration are generally similar to those obtained in non-ossified cochleas.[1,2,6,7,9,12,23] On the other hand, some authors maintain that patients with cochlear ossification do not perform as well with CI as patients with no ossification, and that increases in ossification may result in decreased post-operative performance.[5]

In the event of obstruction of the inferior segment of the basal cochlear turn, the scala tympani is frequently obliterated, while the scala vestibuli is often patent. So in cases of difficult scala tympani insertion, the electrode array may be placed into the scala vestibuli by performing a cochleostomy anterior and superior to the round window, as suggested by Steenerson and co-workers (1990).[11] This surgical technique has been used by these and subsequently also by other authors without any surgical problem and with good communicative results.[6,11,15,23]

In the two cases reported in our study, electrode array insertion into the scala vestibuli proved to be simple and complete. Moreover, the post-operative communicative results are excellent in both patients and similar to those in patients with scala tympani implantation. The first patient reported represents a model case: after implantation she re-acquired 100 per cent open-set word and phrase recognition and phrase comprehension without lip-reading as well as the abilities to carry on telephone conversations, speak and understand foreign languages and listen to music.

Cochlear otosclerosis is a frequent cause of cochlear ossification of CI recipients, but does not seem to affect CI communicative outcomes, although one concern in cochlear implantation for otosclerosis is facial nerve stimulation.[25,26]

Our second patient suffered from obliterative otosclerosis. The surgical outcome was complicated by electrical stimulation of the facial nerve by six of the electrodes, a problem that was simply and wholly resolved by exclusion of the electrodes involved (17 to 12). No detriment to the hearing and communicative outcomes seems to have ensued: in fact, the patient now presents 95 per cent open-set word and 100 per cent phrase recognition and 100 per cent phrase comprehension without lip-reading.

Vertigo might be a problem with scala vestibuli implantation,[11] but none of our patients complained of this, even if both patients had marked pre-operative vestibular hypofunction (the first, a bilateral areflexia and

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the second, marked hypofunction on the implant side), so any vestibular damage consequent to the scale vestibuli implant is difficult to estimate.

Conclusions

Obstruction of the inferior segment of the basal turn of the cochlea, with patent scala vestibuli, is a relatively frequent condition that is often missed before CI surgery. In such cases the electrode array may be inserted into the scala vestibuli. In the two cases reported, this surgical technique allowed complete insertion of the electrode array with no surgical difficulties and good post-operative hearing and communicative outcomes, similar to those obtained in patients without cochlear ossification.

We also highlight the importance of pre-operative radiological study of the inner ear with spiral HR-CT and HR-MR to disclose the presence of cochlear ossification in CI candidates and overall the role of three-dimensional reconstructions from native MR images for precise definition of the inner-ear anatomy and the characteristics of the obstruction, the ultimate aim being proper planning of the surgical approach.

Finally, we wish to underscore the uniqueness of the two clinical cases presented. In the first patient the hearing deficit was caused by systemic vasculitis (Takayasu's disease), that presented as ulcerative colitis and was misdiagnosed before cochlear implantation. The diagnosis in the second patient, who suffered from progressive SNHL and obliterative cochlear otosclerosis, was made only during the pre-implant stages by mean of spiral HR-CT images.

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Stefano Berrettini, M.D. takes responsibility for the integrity of the content of the paper.
Competing interests: None declared

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