Acta Oto-Laryngologica, 2005; 125: 1055–1062

Taylor & Francis
Taylor & Francis

ORIGINAL ARTICLE

Canalolithiasis of the superior semicircular canal: An anomaly in benign paroxysmal vertigo

BRUNO SCHRATZENSTALLER[1], CAROLA WAGNER-MANSLAU[2], GERHARD STRASSER[1] & WOLFGANG ARNOLD[1]

[1]*Department of Otolaryngology, Klinikum rechts der Isar, Munich, Germany, and* [2]*Department of Radiology, Neuroradiology and Nuclear Medicine, Klinikum Dachau, Dachau, Germany*

Abstract
According to the canalolithiasis theory, benign paroxysmal vertigo (BPPV) is caused by gravity-dependent movements of otoconial debris that collects in the endolymph of the posterior semicircular canal. Other parts of the vestibular organ are rarely affected, and it is mainly the horizontal canal that is affected by this atypical form of BPPV. Canalolithiasis of the superior semicircular canal must be considered an anomaly because the superior semicircular canal is the highest point of the vestibular organ and debris normally cannot collect in this special location. Until now, BPPV of the superior canal has mainly been dealt with theoretically in the literature. The authors present three patients with canalolithiasis of the superior semicircular canal and offer direct proof of the condition using high-resolution 3D MRI.

Keywords: *Atypical paroxysmal vertigo, canalolithiasis, high-resolution MRI, superior semicircular canal*

Introduction

Benign paroxysmal positional vertigo (BPPV) is the most frequent peripheral vestibular disorder [1]. The patient typically suffers from attacks of vertigo with sudden onset and concomitant nystagmus of short duration, triggered by changes in head and body position. Diagnostically, the attack can be maximally provoked by employing the Hallpike maneuver, which involves moving the patient rapidly from a sitting position to a supine position with the head hanging downward to one side [2]. Dix and Hallpike [2] were the first to describe the cardinal symptoms of BPPV, as follows: (i) the attack is elicited in a critical provocative position with the affected ear pointing downwards; (ii) the nystagmus is predominantly torsional, with the fast phase beating downwards; (iii) there is a latency of onset; (iv) the nystagmus is transient; (v) the nystagmus is reversed upon repositioning the head; (vi) there is a decline in the response of nystagmus and vertigo on repetition of the provocative maneuver.

Schuknecht [3] proposed that dense basophilic particles released from the utricular macula become attached to the cupula of the posterior semicircular canal, thus making the cupula sensitive to gravity. However, this concept of a "heavy cupula" does not comply with all the typical symptoms of the disease: the nystagmus caused by a gravity-dependent cupula would persist rather than decline, and there would be no fatigability, no latency of onset and no reversal of nystagmus upon repositioning. Schuknecht's cupulolithiasis theory was subsequently replaced by the canalolithiasis theory, which is now generally accepted as the underlying pathophysiological mechanism in BPPV [4]. According to this theory, free-floating endolymph particles collect in the posterior semicircular canal and agglomerate to form "clots" with a higher density than that of the endolymph. When the Hallpike maneuver is performed these clots become mobile and cause a flow of endolymph which leads to deflection of the cupula of the posterior semicircular canal [5,6]. These "canaliths" mainly consist of otoconia which have become

Correspondence: Bruno Schratzenstaller, Department of Otolaryngology, Klinikum rechts der Isar, Ismaninger Straße 22, DE-81675 Munich, Germany. Fax: +49 89 41 40 48 53. E-mail: bruscale@t-online.de

(Received 28 June 2004; accepted 24 February 2005)

ISSN 0001-6489 print/ISSN 1651-2551 online
DOI: 10.1080/00016480510037023

detached from the utricular macula due to infection, trauma, ear surgery or the aging process [7].

Although the canalolithiasis theory provides a useful working hypothesis which can explain all of the cardinal symptoms of BPPV, there is also a small fraction of patients who present with paroxysmal positioning vertigo and concomitant nystagmus but do not fulfill all of the Dix–Hallpike criteria, i.e. vertigo attacks in untypical head positions, pure horizontal nystagmus, etc. The most common cause of this is free-floating, dense particles in an untypical location in the semicircular canals [8,9]. Epley [10] classified these variants as "atypical paroxysmal vertigo" and defined new criteria for canalolithiasis independent of the pathological site in the vestibular organ, which he divided into obligatory and facultative symptoms, as follows:

- Obligatory symptoms: (i) the nystagmus is transient; (ii) the nystagmus occurs after a latency of a few seconds; (iii) the nystagmus is accompanied by severe vertigo; and (iv) the symptoms are provoked by head positioning.
- Facultative symptoms: (i) the response declines with repetition of the provocative maneuver; (ii) the direction of the nystagmus is reversed in the same plane upon repositioning of the head; and (iii) symptoms can be cured by specific canalith repositioning maneuvers.

Atypical paroxysmal vertigo is a rare occurrence, with incidences of 1–20% having been reported in the literature [11–13]. According to the authors' experience, it represents $<1\%$ of all forms of BPPV. The best known and commonest type of BPPV is posterior canal disease but, as the underlying pathology has become more thoroughly understood, scientific research has also been carried out on other types of paroxysmal vertigo, many publications dealing with horizontal semicircular canal disease [7–9]. BPPV of the superior semicircular canal has mainly been dealt with hypothetically [10,14,15] and until now there has been no convincing proof of its existence [13]. The reason for the high frequency of posterior semicircular canal disease is the fact that the posterior semicircular canal is the most dependent part of the vestibular organ in both the upright and supine positions and endolymphatic debris is sensitive to gravity. Therefore, under normal circumstances, endolymphatic particles, having a higher density than the endolymph itself, will collect in the posterior semicircular canal and form a clot. For these physiological reasons, involvement of the superior semicircular canal must be considered an anomaly in BPPV. In >500 patients suffering from BPPV we found only 3 with involvement of this particular site and ours is the first group to offer direct proof of this by employing 3D reconstructions of the vestibular organ using high-resolution 3D MRI (HR-3D-MRI).

HR-3D-MRI of the inner ear has been performed by many groups before and offers excellent visualization of the inner ear fluid spaces. This method has mainly been used for the investigation of malformations, traumatic lesions and inflammatory processes, and for the detection of intralabyrinthine tissue formations [16–19]. Pathological processes are recognized either as an anomaly of the configuration of the inner ear or as a filling defect [20,21]. There is general agreement between published data that filling defects, especially in the semicircular canals, are always pathological [22], as HR-3D-MRI provides gapless imaging of the inner ear fluid spaces.

Material and methods

In >500 patients suffering from BPPV, we were able to prove involvement of the superior semicircular canal using HR-3D-MRI in only 3 (1 male, 2 females). All three patients had been given a diagnosis of BPPV and complied with Epley's criteria of canalolithiasis. They underwent a complete otoneurologic investigation, including audiometry, caloric irrigation and MRI of the brain to exclude concomitant neurologic diseases.

For each subject the same dataset comprising systematic series of axial MR images and 3D data was obtained using a 1.5-T Signa HS whole-body MRI system (GE, Milwaukee, WI). The manufacturer's head coil was used for all acquisitions. The major concern of the diagnostic study was the identification of anatomical and pathological sites in the semicircular canals and these data are optimally depicted on heavily T2-weighted images using a 3D dataset which was reformated afterwards. For this subject we performed fast spin echo T2 weighted images (FSE T2) with a repetition time (TR) of 4320 ms and echo time (TE) of 238 ms. The selected slice thickness was 1 mm, the matrix 512×512 mm, the field od view (FOV) 22×22 mm, the number of exaltations (NEX) 1.0.

Figure 1 shows the anatomical definitions and different views of a normal left inner ear in 3D mode in the three different planes.

Case reports

Case 1

The patient was a 39-year-old male who complained of brief attacks of rotatory vertigo when turning his head over his right shoulder in a sitting position.

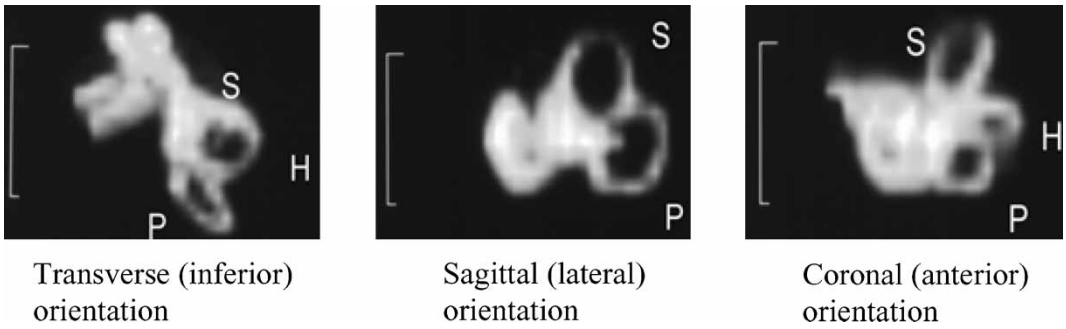

Figure 1. Anatomical definitions and views in the three different planes. HR-3D-MRI reconstructions of a normal left inner ear. S = superior semicircular canal (anterior); H = horizontal semicircular canal (lateral); P = posterior semicircular canal.

Turning the head of the patient in this position triggered a fast-beating counterclockwise pure torsional nystagmus which occurred with a latency of 5–10 s and lasted for ≈15 s. The nystagmus was accompanied by a severe spinning sensation and reversed its direction upon repositioning the head. Audiometry showed a minor high-frequency sensorineural hearing loss on both sides and caloric irrigation revealed a slightly decreased response on the left side, without the presence of head-shaking nystagmus.

This positional vertigo had occurred immediately after a skull trauma 4 years previously. Conventional X-rays, cranial CT and MRI revealed no bone fractures or brain damage and cerebral angiography and basic neurological investigations proved to be normal. HR-3D-MRI revealed an infraction or impression in the right posterior semicircular canal and a 2-mm filling defect in the right superior semicircular canal (Figure 2), while reconstruction of the left inner ear showed no abnormalities. To date, it has not been possible to cure this patient by means of any physical maneuvers.

Case 2

A 58-year-old female presented with the typical symptoms of BPPV on both sides, from which she had been suffering for many years. Ten years previously she had been diagnosed with Ménière's disease (MD) on the left side, at which time she suffered from severe pancochlear hearing loss and recurrent attacks of vertigo and vomiting lasting for hours. Local gentamicin treatment applied via a ventilation tube in the left tympanic membrane had been successful and the attacks of MD ceased; however, some years later she developed incurable positional paroxysmal vertigo on both sides. The Dix–Hallpike maneuver provoked a counterclockwise torsional nystagmus on the right side and a clockwise torsional nystagmus on the left side, with reversal on both sides when the patient was returned to a sitting position. On each side the nystagmus was accompanied by a sensation of vertigo, which lasted for ≈20 s on the right side and 25 s on the left. Caloric irrigation revealed a malfunction on the left side; audiometry showed normal hearing on the right side and pancochlear hearing loss of ≈70 dB on the left side. HR-3D-MRI revealed a 3.6-mm long occlusion of the left superior semicircular canal and a 1.7-mm filling defect in the left posterior semicircular canal (Figure 3a), whereas the right side showed no abnormalities.

Although the patient was treated with various physical maneuvers over a long period, no cure could be achieved on either side.

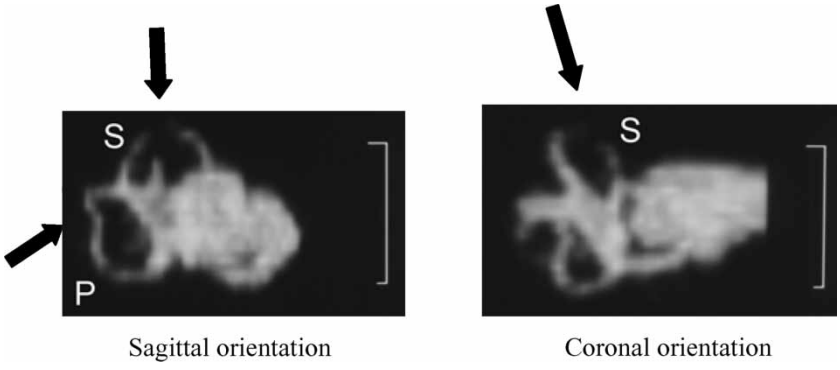

Figure 2. Case 1. HR-3D-MRI scans of the right inner ear (sagittal and coronal orientations) showing morphological abnormality of the posterior semicircular canal (traumatic impression–left arrow) and a 2-mm filling defect (upper arrows) at the top of the superior semicircular canal. For explanation of abbreviations, see Figure 1.

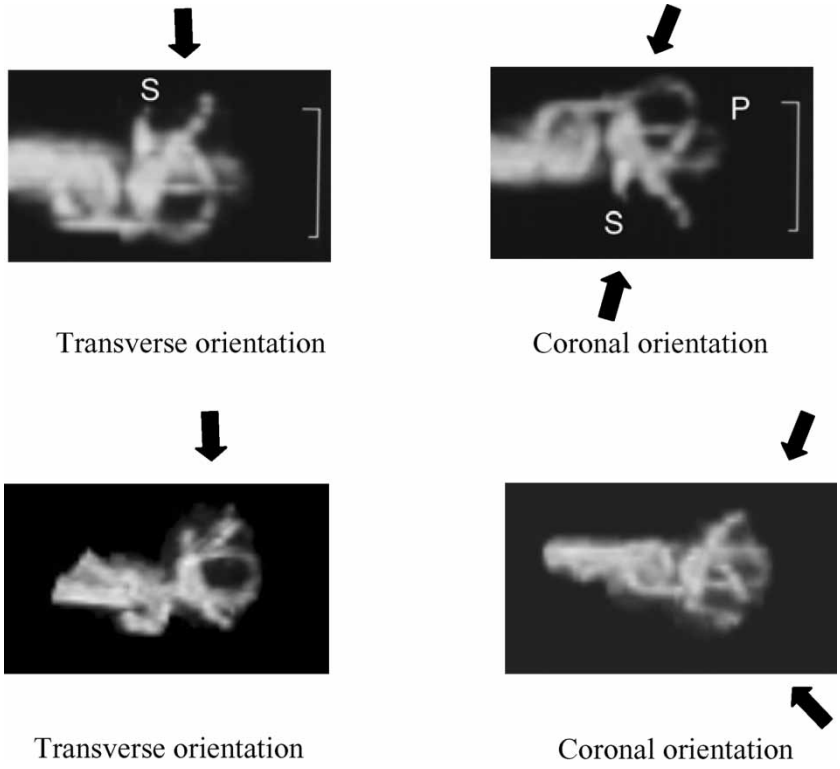

Figure 3. Case 2. HR-3D-MRI scans of the left inner ear (transverse and coronal orientations). (a) Initial MRI investigation shows a 3.6-mm long filling defect of the superior semicircular canal (upper arrow in the traverse orientation and inferior arrow in the coronal orientation) and a 1.7-mm long defect in the posterior semicircular canal (upper arrow in the coronal orientation). (b) A follow-up MRI investigation 2 years later shows no significant changes in comparison (with the arrows indicating the corresponding sites) to (a). For explanation of abbreviations, see Figure 1.

A control HR-3D-MRI scan carried out 2 years after the first scan showed no significant changes in the pathology of the semicircular canals (Figure 3b).

Case 3

This 55-year-old female presented with a history of brief attacks of rotational vertigo when turning her head to the right side while lying in bed. The vertigo had occurred for the first time 3 days before presentation. Putting the head into the right Hallpike position elicited a counterclockwise rotatory nystagmus which lasted for 15–20 s and was reversed when the head was returned to the upright position. Audiometry showed nearly normal hearing function in both ears. The patient had undergone a type I tympanoplasty 12 years previously on the contralateral (left) side without complications. For this reason we were suspicious of atypical vertigo and performed HR-3D-MRI 2 days after diagnosis without any further treatment or investigation. HR-3D-MRI showed a 2.2-mm filling defect in the superior

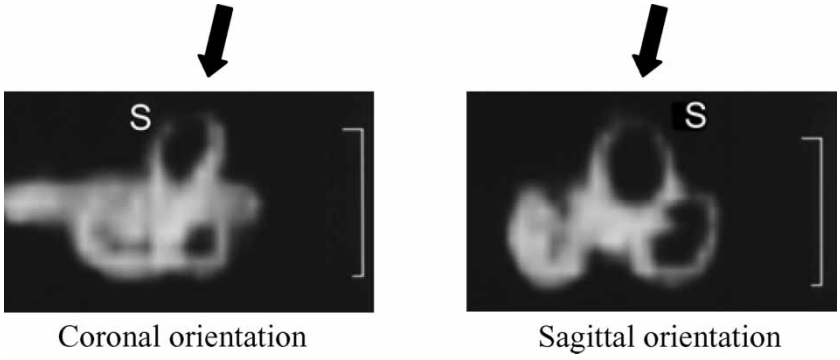

Figure 4. Case 3. HR-3D-MRI scans of the left inner ear (coronal and sagittal orientations) showing a 2.2-mm long filling defect (arrows) of the superior semicircular canal. For explanation of abbreviation, see Figure 1.

semicircular canal of the left side (Figure 4), with no pathological changes in the right labyrinth. No treatment was necessary as the patient recovered spontaneously 3 days after diagnosis. The defect in the left semicircular canal was no longer visible in a control HR-3D-MRI scan carried out 2 weeks later.

Discussion

As explained previously, due to gravitational forces endolymphatic debris is supposed to collect in the posterior semicircular canal as the most dependent part of the labyrinth, which explains the high propensity of posterior canal disease. In contrast, the superior semicircular canal is the highest point of the labyrinth in the upright position, and so particles should leave this canal on changing from a lying to an upright position, which would make the formation of canaliths in its lumen highly improbable [15]. Furthermore, sections revealed that the superior semicircular canal and its cupula had a much lower incidence of basophilic deposits than the posterior and lateral semicircular canals [23].

For all these reasons we suspect that in cases with superior canal involvement there must be structural abnormalities in the lumen of the superior canal, otherwise loose granular sediment could not collect inside this special location. Hall et al. [5] postulated different underlying pathologies for incurable and atypical forms of BPPV. It was our assumption that these abnormalities should be recognizable using HR-3D-MRI.

Schratzenstaller et al. [22] demonstrated structural changes in patients suffering from incurable and atypical variations of BPPV using HR-3D-MRI of the inner ear. While some authors [13] still doubt the existence of superior canal disease, Honrubia et al. [11] were the first to propose that canalolithiasis of the superior canal may exist. Of 292 patients suffering from BPPV, they suspected that the superior canal was affected in 4, but could not prove their assumptions using objective criteria.

Each of the three patients presented in this study showed morphological changes in the superior semicircular canal using HR-3D-MRI. This method yields highly reliable results for assessment of the vestibular organ. Following confirmation of canalolithiasis of the superior canal by means of HR-3D-MRI, interpretation of the individual cases also provides further insight into the pathophysiology of this vestibular disease.

Case 1

HR-3D-MRI showed a 2-mm filling defect at the top of the right superior semicircular canal. When the patient turned his head over his right shoulder it provoked attacks of vertigo, followed by torsional counterclockwise nystagmus. HR-3D-MRI confirms the theory of canalolithiasis because in this case the only mechanism which is able to explain BPPV attacks in the position in question is an ampullofugal deflection of the cupula of the right superior canal provoked by dislodged particles moving down from the top of the non-ampulated arm, as depicted in Figure 5.

Of all possible locations, the top of the superior semicircular canal is the most unlikely to cause BPPV [15] because this is the highest point of the inner ear and so gravity-sensitive particles cannot gather there. To date there have been no reports of similar cases.

We postulate the existence of an "adhesion point" caused by traumatization where endolymphatic particles can collect (the microfractures creating a "sticky endothelium"). This would also account for the incurability of the symptoms. Furthermore, the preceding skull trauma may have caused increased shedding of otoconia [10].

Case 2

This patient presented with recurrent BPPV on both sides preceded by MD and gentamicin therapy on the left side. BPPV following MD has long been known in the literature, and Gross et al. [24] found that it even predisposes for the intractable form of canalolithiasis. Our patient, however, suffered from incurable BPPV attacks on both sides, even though gentamicin therapy had only been applied to the left side.

HR-3D-MRI showed a normal right inner ear with filling defects in the left posterior canal and even more distinct defects in the left superior semicircular canal (Figure 3).

Repeated hydropical distension is supposed to cause damage both to the maculae, causing increased shedding of otoconia, and to the canal endothelium, resulting in stenosis, occlusions or adhesions as a consequence of loss of the resiliency of the membranous labyrinth [25]. Local application of gentamicin may have caused further damage, not only to the macula organs but also to the canal endothelium, making it easier for loose debris to adhere inside the canal walls. Vyslonzil [26] reported an increased incidence of otoconial deposits in the posterior canal in sections of patients who had been treated with streptomycin (an aminoglycoside like gentamicin). In 2002, Perez et al. [27] reported a patient suffering from intractable BPPV after gentamicin therapy for MD.

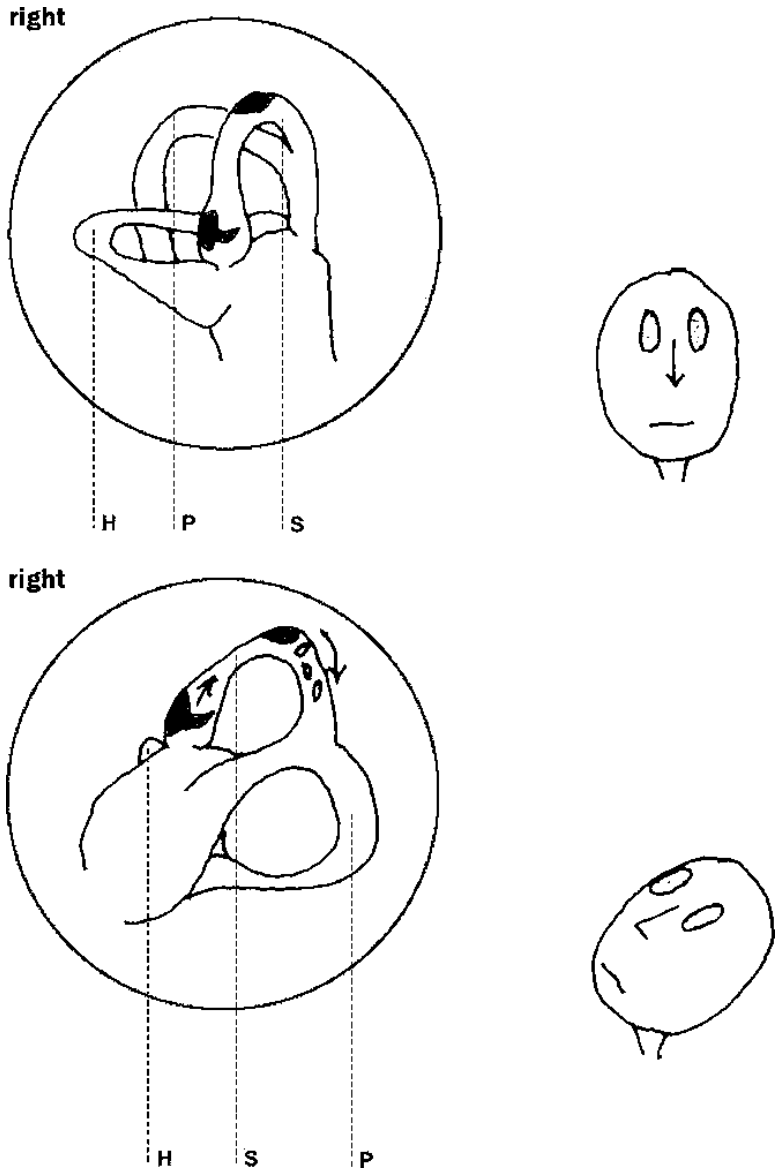

Figure 5. The pathophysiological mechanism causing endolymph flow and cupular deflection in the superior semicircular canal of Case 1. For explanation of abbreviations, see Figure 1.

Parnes and McClure [6] (for cupulolithiasis) and later Epley [10] (for canalolithiasis) postulated that, in BPPV, if the superior canal was affected on one side this would result in the same symptoms as if the posterior canal of the opposite side was affected. Epley used the term "complementary semicircular canals" to describe this [10]. As HR-3D-MRI showed no pathology in the right vestibular organ but a long filling defect in the left superior semicircular canal, we conclude that the affected left superior semicircular canal is "masking" the symptoms of typical BPPV in the right ear. The long obstruction of the canals as seen with HR-3D-MRI also accounts for the incurability of the symptoms. Figure 6 demonstrates this pathophysiological mechanism. When the head is put into the right Hallpike position a vertigo attack which fits the criteria of Dix and Hallpike [2] can be caused by canalolithiasis of both the left superior and right posterior semicircular canals.

Case 3

This patient presented with symptoms of typical BPPV on the right side but had undergone a tympanoplasty on the left side. HR-3D-MRI revealed a 2.2-mm filling defect in the left superior semicircular canal. The onset of BPPV following middle ear surgery, even years later, has frequently been reported in the literature and is thought to be due to traumatization of the inner ear [10]. The

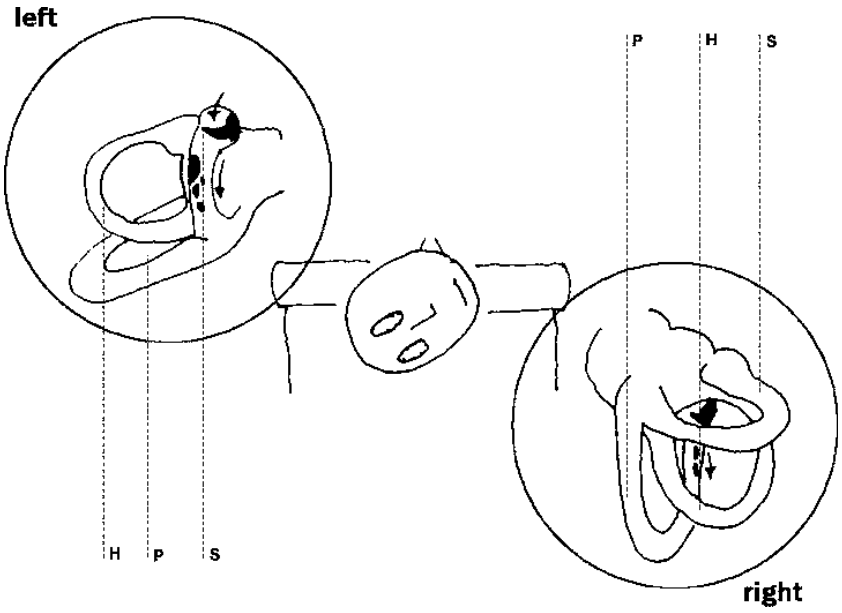

Figure 6. Epley's postulate of complementary semicircular canals causing identical symptoms when affected with canalolithiasis; shown with the patient lying in the right Hallpike position. For explanation of abbreviations, see Figure 1.

mechanism seems to be the same as that outlined above, with the symptoms of canalolithiasis of the right posterior canal "masking" an affected left superior canal. Even after increased release following macula damage, loose otoconia cannot collect in the superior semicircular canal and should leave the canal lumen as soon as the patient gets up from a lying to an upright position [15]. Therefore, as expected, HR-3D-MRI showed the formation of a solid clot. As there was no endothelial damage to make the intracanalicular endothelium adhesive, this condition must be most unstable, as confirmed by the spontaneous remission of symptoms and the absence of pathologic findings on a control MRI scan.

Conclusions

BPPV of the superior semicircular canal must be considered an anomaly in canalolithiasis, because under normal circumstances loose sediment cannot collect at this special location [15]. As the superior canal is the highest point of the vestibular labyrinth in the standing position and remains unaffected by gravitational forces in the lying position we postulate that (i) there must be structural changes in the lumen of the superior canal which makes the endothelium adhesive and (ii) there is formation of a solid structure which is recognizable as a filling defect on HR-3D-MRI.

In the following cases the authors are suspicious of canalolithiasis of the superior semicircular canal and recommend the performance of HR-3D-MRI:

- patients with previous ear surgery or ear disease (MD, vestibular neuritis, sudden sensorineural hearing loss, etc.) who present with BPPV on the contralateral side;
- patients with therapy-resistant BPPV; and
- patients who present with symptoms of BPPV when in an atypical position.

Our findings further support the canalolithiasis theory, as the pathological changes we found in the semicircular canals using HR-3D-MRI exactly caused the symptoms predicted by this hypothesis [10,11].

References

[1] Brandt Th, Steddin S. Current view of the mechanism of benign paroxysmal positioning vertigo: cupulolithiasis or canalolithiasis? J Vestib Res 1993;3:373–82.
[2] Dix R, Hallpike CS. The pathology, symptomatology and diagnosis of certain common disorders of the vestibular system. Ann Otol Rhinol Laryngol 1952;6:987–1016.
[3] Schuknecht HF. Cupulolithiasis. Arch Otolaryngol Head Neck Surg 1969;90:113–26.
[4] Epley JM. The canalith repositioning procedure: for treatment of benign paroxysmal positional vertigo. Otolaryngol Head Neck Surg 1992;107:399–404.
[5] Hall SF, Ruby RR, McClure JA. The mechanics of benign paroxysmal vertigo. J Otolaryngol 1979;8:151–8.
[6] Parnes LS, McClure JA. Free-floating endolymph particles: a new operative finding during posterior semicircular canal occlusion. Laryngoscope 1992;102:988–92.
[7] Anastasopoulos D, Lempert T, Gianna C, Gresty A, Bronstein M. Horizontal otolith-ocular responses to lateral translation in benign paroxysmal positional vertigo. Acta Otolaryngol (Stockh) 1997;117:468–71.

[8] Lempert Th, Thiel-Wilck K. A positional maneuver for treatment of horizontal-canal benign positional vertigo. Laryngoscope 1996;106:476–8.
[9] Fife TD. Recognition and management of horizontal canal benign positional vertigo. Am J Otol 1998;19:345–51.
[10] Epley JM. Positional vertigo related to semicircular canalithiasis. Otolaryngol Head Neck Surg 1995;112:154–61.
[11] Honrubia V, Baloh RW, Harris MR, Jacobson KM. Paroxysmal positional vertigo syndrome. Am J Otol 1999;20:465–70.
[12] Casani A, Vanucci G, Fattori B, Ghilardi PL. Positional vertigo and ageotropic bidirectional nystagmus. Laryngoscope 1997;107:807–13.
[13] Von Brevern M, Lempert T. Benigner paroxysmaler Lagerungsschwindel. HNO 2002;50:671–81.
[14] Brantenberg K, Bergenius J. Treatment of anterior paroxysmal positional vertigo by canal plugging. A case report. Acta Otolaryngol 2002;122:28–30.
[15] Steddin S, Brandt Th. Benigner paroxysmaler Lagerungsschwindel. Nervenarzt 1994;65:505–10.
[16] Held P, Fellner C, Fellner F, Seitz J, Graf S, Strutz J. Correlation of 3D MRI and clinical findings in the patients with sensorineural hearing loss and/or vertigo. Clin Imaging 1998;22:309–22.
[17] Albers FWJ, Demunynck KMNP, Casselman JW. Three-dimensional magnetic resonance imaging of the inner ear in idiopathic sudden sensorineural hearing loss. ORL J Otorhinolaryngol Relat Spec 1994;56:1–4.
[18] Casselman JW, Kuhweide R, Deimling M, Ampe W, Dehaene I, Meeus L. Constructive interference in steady state-3DFT MR imaging of the inner ear and cerebellopontine angle. AJNR Am J Neuroradiol 1993;14:47–57.
[19] Jäger L, Strupp M, Brandt T, Reiser M. Bildgebung von Labyrinth und Nervus vestibularis. Nervenarzt 1997;68:443–58.
[20] Casselman JW, Kuhweide R, Ampe W, Meeus L, Steyaert L. Pathology of the membranous labyrinth. Comparison of T1- and T2-weighted and gadolinium-enhanced spin-echo and 3DFT-CISS imaging. AJNR Am J Neuroradiol 1993;14:59–69.
[21] Held P, Fellner C, Fellner F, Graf S, Seitz J, Strutz J. 3D MRI of the membranous labyrinth. An age related comparison of MR findings in patients with labyrinthine fibrosis and in persons without inner ear symptoms. J Neuroradiol 1998;25:268–74.
[22] Schratzenstaller B, Wagner-Manslau C, Alexiou C, Arnold W. High-resolution three-dimensional magnetic resonance imaging of the vestibular labyrinth in patients with atypical and intractable benign positional vertigo. ORL J Otorhinolaryngol Relat Spec 2001;63:165–77.
[23] Moriarty B, Rutka J, Hawke M. The incidence and distribution of cupular deposits in the labyrinth. Laryngoscope 1992;102:56–9.
[24] Gross EM, Ress BD, Viirre ES, Nelson JR, Harris JP. Intractable benign paroxysmal positional vertigo in patients with Meniere's disease. Laryngoscope 2000;110:655–9.
[25] Hughes CA, Proctor L. Benign paroxysmal positional vertigo. Laryngoscope 1997;107:607–13.
[26] Vyslonzil E. Über eine umschriebene Ansammlung von Otokonien im hinteren häutigen Bogengang. Monatsschr Ohrenheilkd 1963;97:63–8.
[27] Perez N, Martin E, Zubieta JL, Romero MD, Garcia-Tapia R. Benign paroxysmal positional vertigo in patients with Meniere's disease treated with intratympanic gentamycin. Laryngoscope 2002;112:1104–9.