Surgical treatment of Meniere disease via labyrinth Shunt: Past and present

Tratamento cirúrgico da doença de Ménière por meio de Shunts labirínticos: Passado e presente

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SUMMARY

Introduction: The surgeries of the endolymphatic shunt are employed in the treatment of the vertigo of Meniere disease disabling, remains a controversial topic. Portmann in 1926, was the first to perform the surgery, incising the endolymphatic sac aiming the decrease of the endolymphatic pressure of the inner ear. Planned to create a fistula that connects the endo and perilymphatic space, the sacculotomy were described by Fick in 1964, by Cody in 1969 e by Schuknecht (cochleosacculotomy) in 1982, however they were gradually abandoned because of the high incidence of hearing loss. A new and promising surgery option of shunt, still experimental is utriculostomy, performed by Lavinsky in 1999.

Objective: Make a review of literature about the main surgical techniques of endolymphatic shunt, used in the treatment of Meniere disease, its results and the advance in the understanding of these procedures.

Methods: Based on the literature data, the main surgical techniques of endolymphatic shunt are discussed and its rationality in the treatment of Meniere’s disabling disease.

Conclusion: There are a lot of controversy over the effectiveness of surgical procedures of endolymphatic shunt for the treatment of Meniere’s disabling disease, and a lot of them damage the inner ear. The experimental surgery of utriculostomy, so it seems, a promising option. Future research can give an answer to this matter.

Keywords: Meniere disease, vertigo, endolymphatic sac.

RESUMO

Introdução: As cirurgias dos shunts endolinfáticos empregadas para o tratamento da vertigem na doença de Ménière incapacitante permanece um tópico controverso. Portmann, em 1926, foi o primeiro a executar a cirurgia, incisando o saco endolinfático com o objetivo de diminuir a pressão endolinfática da orelha interna. Planejadas para criar uma fístula que conecte o espaço endo e perilinfático, as saculotomias foram descritas por Fick em 1964, por Cody em 1969 e por Schuknecht (cochleosacculotomy) em 1982, entretanto foram gradualmente abandonadas devido à alta incidência de perda auditiva. Uma nova e promissora opção cirúrgica dos shunts, em caráter experimental, é a utriculostomia, realizada por Lavinsky em 1999.

Objetivo: Realizar uma revisão da literatura sobre as principais técnicas cirúrgicas dos shunts endolinfáticos empregados no tratamento da doença de Ménière, seus resultados e o que há de avanços no entendimento desses procedimentos.

Método: Com base nos dados da literatura, são discutidas as principais técnicas cirúrgicas dos shunts endolinfáticos e sua racionalidade no tratamento da doença de Ménière incapacitante.

Conclusão: Existe muita controvérsia sobre a efetividade dos procedimentos cirúrgicos dos shunts endolinfáticos para o tratamento da doença de Ménière incapacitante, e muitos deles causam danos à orelha interna. A cirurgia experimental utriculostomia, portanto, parece-nos uma opção promissora. Futuras pesquisas poderão fornecer a resposta para essa questão.

Palavras-chave: doença de Ménière, vertigem, saco endolinfático.
INTRODUCTION

The first treatment option for Ménière Disease (MD) is pharmacological, including the use of diuretic, low sodium and low betahistine diet. A variety of other drug can be used, including antihistamines, benzodiazepines and corticosteroids. The surgical treatment is considered only when the vertigo caused by this condition evolves to such a point as to become clinically incapacitating (1).

The target of all surgical interventions is, primarily, to reduce the number and severity of the acute attacks of vertigo and, secondarily, to improve or stabilize the hearing loss and the tinnitus or aural fullness. For patients who still remain incapacitated and with imbalance attacks after three to six months of conservative therapy and unilateral development of the disease, surgical intervention can be considered (2).

Surgical interventions on MD can be divided into two groups: conservative and non-conservative of hearing (1).

The non-conservative processes include labyrinthectomies neurectomies and translabyrinthine, which remove partially or completely the sick labyrinth, and the insertion of aminoglycoside in the middle ear to perform a chemical labyrinthectomy, which decreases vertigo, but can lead to hearing loss (3).

There is another group of non-conservative of hearing techniques that act on the distal endolymphatic system, equalizing the pressures and connecting the endolymphatic and perilymphatic spaces, acting at saccule level. Some are performed through the platinum of the stapes, other through the round window. Through oval window, we have sacculotomy, the tack procedure and sacculocentesis. From the round window, there is “osmotic diuresis” with NaCl, cochleostomy, cochleosacculotomy, intracochlear shunt and cochlear dialysis. There are still other techniques, which act in other sections of the labyrinthine capsule, such as cryosurgery, determining a fistula on the semicircular side canal or acting on the promontory with a oticoperotic shunt.

Among the conservative of hearing techniques are the endolymphatic sac surgery by simple decompression, implant of sheet of Silastic® to the mastoid, through capillary tube or capillary tube with fluid chamber, shunt with unidirectional valve sensitive to pressure and, finally, shunt for the subarachnoid space. There are included, also, as conservative measures, the selective vestibular neurectomy via middle fossa, the retrolabyrinthine neurectomy and the retrossigmoid neurectomy.

The most used procedure of shunt is the endolymphatic sac. Although these surgeries are relatively complication free and technically of easy realization, their results are controversial: some authors affirm that there is no statistically significant improvement of vertigo in relation to the surgical procedures used as placebo (4); but others state the opposite (5).

The attempt to perform procedures creating endolymphatic shunts at sacule level is in disuse, for the high index of hearing loss (4).

Since this is an area about which the otorhinolaryngology has many different and of difficult application concepts, we find that the conceptual ordination about the theme and respective discussion provide a consistent contribution for readers. For this, we created the following objectives:

1) To present a bibliographic review of the main surgical techniques with endolymphatic shunts, that is, the most employed in the treatment of MD.
2) To discuss their results, highlighting a new surgical alternative in experimental phase that acts on the utricle.

LITERATURE REVIEW

Endolymphatic shunts at endolymphatic sac level

The first endolymphatic shunt procedure was performed by Georges Portmann in February 26th of 1926 (6). As a result of his own researches and the theory proposed by Guild that the longitudinal endolymphatic flow occurred towards the endolymphatic sac (7), Portmann recognized that MD is caused by an increase of pressure at the endolymphatic compartment of the inner ear. He imagined that the surgery of endolymphatic sac, designed to decrease the endolymphatic pressure on the endolymphatic compartment of inner ear through the incision of the endolymphatic sac, would improve the symptoms associated with endolymphatic hydropsis (hydrops). In this procedure, the endolymphatic sac is opened for the mastoid process. The surgery was performed with hammer and gouge instead of otologic drill, and no microscope was used. The sac was simply incised with a small knife, theoretically reserving the endolymphatic drainage under increase of pressure towards the mastoid.

With greater use of microscope and surgical drills, several modifications were made to the original surgery of Portmann with the theoretical intent to improve the drainage of endolymph. In 1954, Yamakawa and Naito removed part of the medial wall of the endolymphatic sac to direct the...
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Endolymphatic shunts endolinfáticos at saccule level

Endolymphatic shunt surgeries were planned intending to create a fistula through which the endolymphatic space can be connected to the perilymphatic space, placing the internal shunt on the otic capsule. This fistula, theoretically, would drop the pressure as it drained the flow to the perilymphatic space of low pressure.

The first procedure performed with this intent was named sacculotomy. In 1964, Pick, using histologic evidences of saccule distension in patients with MD, proposed that the rupture of the distended saccule might equalize the pressure between endolymph and perilymph. The author recommended a puncture on the platinum of the stapes with a needle to make the rupture at the base of the saccule. Cody modified Pick's procedure, placing a stainless steel tack each time there would be distension. This surgery was performed through the external auditory meatus, proceeding to tympanomeatal reattachment. Unfortunately, many patients suffered progressive hearing losses after being put through these surgeries. Schuknecht noticed that the designers of these procedures had not predicted that the distended saccular wall was usually attached to the platinum of the stapes in MD, thus causing, in long-term, a difficulty to generate an efficient and permanent fistula (18).

Cochleosacculotomy

This procedure was proposed in 1982 by Schuknecht, based on scientific observations. He observed, in animal models, that the fistulae between the perilymph and endolymph compartments on the inner ear were compatible with the preservation of hearing and demonstrated that the existence of such fistulae occurred spontaneously in temporal bones in humans. Schuknecht believed that these fistulae caused regression of the clinical symptoms of MD and that they could be permanently created via fracture and rupture of the cochlear duct (6).

Indicated as treatment for elderly patients with incapacitating symptoms, who have poor hearing, but reveal good vestibular function in electronystagmographic tests (1), and by quickness and simplicity of the procedure, the cochleosacculotomy is performed under local anesthesia. It starts with the creation of a tympanomeatal reattachment, through which the niche of the round window is exposed. A 3 mm right angle blade is introduced in all its length through the membrane of the round window, towards the oval window. At this point, the instrument advances through the bone spiral lamina and penetrates the saccule. The blade is removed, and the membrane of the round window is covered with perichondrium, fat or temporal fascia. The tympanomeatal reattachment is then reattached.

Endolymphatic shunt at utricle level

The first surgical procedure of endolymphatic shunt acting in the utricle was proposed by Lavinsky et al. in 1999 (19). The intention of this procedure was related to the expectation of a smaller auditory damage, knowing that the utricle doesn't belong to an anatomofunctional unity with the cochlea, like the saccule. In experimental stage, the method named utriculostomy consists in obtaining a permanent fistula on the membranous labyrinth, thus communicating the endolymphatic and perilymphatic spaces at utricle level. The procedure was put into practice in sheep, through the oval window, using a microcautery by radiofrequency with exposure time and temperature programming. Localized heat, applied with 0.20mm needle, generates loss of substance. As observed in histologic exams, there is regeneration in the treated area, forming a frail membrane that behaves like a valve, breaking up in moments of endolymphatic hydrops and, thus, communicating both spaces and avoiding a permanent ionic mix and its known consequences.

Experimental utriculostomy was performed in 12 sheep at the Veterinary Hospital at Veterinary Medical School of Universidade do Rio Grande do Sul, Porto Alegre, RS. After anesthesia, the endoperiauricular area of the animals was exposed and the tympanic membrane removed according to the technique describe by House et al. (20), employed in stapedectomies.

The promontory, oval and round window, facial nerve, chorda tympani, pyramidal apophysis and tensor
ligament of the stapes were exposed. Thus, the tensor ligament of the stapes was sectioned and the disjunction of the incudostapedian articulation was performed. The superstructure of the stapes was fractured, and all the platinum was removed. With a diamond drill, approximately 2mm from the posterior edge of the oval window were removed towards the facial nerve. The widening of the oval window facilitated the cautery procedure on the utricle.

The anterior wall of the utricle, partially visible during the otomicroscopy, was cauterized using a microcautery with 0.2mm tip for 0.5 seconds, at intensity of 3.5 W. the cauterization was repeated in three places close to each other. The oval window was closed with adipose tissue removed from the region close to the endoperiauricular incision and covered with Gelfoam®. The tympanum was reinserted, and the external auditory meatus tamponade with Gelfoam®. Finally, the region was covered with compressive bandage.

Three months after surgery, the animal was sacrificed, and its temporal bone was submitted to relevant histological study at the histopathology laboratory directed by Michael Paparella (University of Minnesota, Minneapolis, EUA). It was possible to demonstrate the effectiveness and safety of the proposed technique, with positive results.

**Discussion**

Mechanisms of secretion and reabsorption performed in medium scale through the heterogeneous epithelium needs to be well balanced to maintain a constant volume of fluid. Failures at the maintenance of this balance will result in dilatation of the endolymphatic compartment observed in MD. It’s also been admitted that this increase of endolymphatic pressure might break the membranous labyrinth and originate a mixture between endolymph, rich in potassium, and perilymph, poor in potassium (21). Potassium unbalance would prevent the depolarization and originate transitory loss of function until the membrane was repaired and the sodium-potassium relations were restored.

We believe that vertigo results from mechanical over-stimulation of the sensorial cells due to distension of inner ear structures. This is caused by dysfunction of the sac, damaged by fibrosis, infection or autoimmune or allergic processes. Other possible mechanism is the exaggerated production of glycoproteins, which would originate an excessive inflow of endolymph on the sac (21).

Data from literature indicate that at least 85% of the patients with MD will answer positively to clinical treatment (4). However, even after 20 years suffering from the disease, 21% of the patients remain with vertigo (22). In a prospective study involving 245 patients, there was an increase of cases with severe or very severe attacks (22), thus showing that, once vertigo becomes incapacitating despite the pharmacological treatment, surgical procedure is recommended.

The decompression of the endolymphatic sac is a surgical alternative for those patients who present resistance to the pharmacological therapy. This safe and conservative of the endolymphatic sac option can be performed with or without the use of a shunt (1).

The logic of this surgery is much debated. On the base is the understanding that the shunt drains the excess of endolymph widens the sac’s inner space, expanding the absorption surface, while the osseous decompression reduces the pressure and increases blood supply and changes (23).

On surgical procedures of the endolymphatic sac without shunt, is performed a wide osseous decompression of the sac, the sigmoid area and the area of the jugular bulb, without entering the endolymphatic sac itself. Results of long-term segments showed that the surgery is safe and efficient for the control of vertigo and the stabilization of hearing (24). Improvement indexes go from a scale of 85 to 100%, and the stabilization or improvement of hearing can reach almost 85% (25).

The shunt surgery of the endolymphatic sac is a modification of the surgery without shunt. In this case, a shunt is created for the subarachnoid space. The result of the treatment is compatible to that of the mastoid endolymphatic shunt (26). However, in long-term, the presence of fibrosis or the closing of the incision of the endolymphatic sac can prejudice the results. The application of intraoperative mitomycin C on the incised endolymphatic sac can be beneficial in these cases (27), and the instillation of steroids during surgery can help to control vertigo, to stabilize hearing and to decrease the tinnitus (28, 29). Vertigo control occurs in 60-70% of the cases in shunt surgery of the endolymphatic sac (26). Despite that, it has been suggested that the impact of endolymphatic sac shunts over the symptoms of patients with MD is very similar to the one obtained with placebo (3, 30), and some studies support the concept of placebo effect in these surgeries. The insertion of ventilation tube on the tympanic membrane, the complete extra-osseous removal of the endolymphatic sac and cortical mastoidectomy presented similar results to the procedure of shunt of the endolymphatic shunt (31-33). However, the study of Homsen et al. had a small sample and arguable statistic characteristics. Pillsbury et al. (5) repeated the study and
almost 50% of the patients will have a worsening of patient has a hearing loss in the ear with hydrops, because some authors, this surgery can be an option when the surgical procedures performed at the saccule. According to CODY’s tack operation was the most popular of the present modifications in the sacculotomy techniques, with the resulting elimination of hydrops. Despite those who hadn’t performed the surgery, and 90% long, intense and frequent vertigo crisis in comparison to patients with MD for over 20 years and that had been submitted to the endolymphatic sac surgery, all presented results also when reviewing the increase of endolymphatic control (85%) of vertigo during 2 to 5 years, with excellent less than 1% risk of paresis of the facial nerve, important than 2% index of significant sensorineural hearing loss and in cases in which MD coexists with other diseases, like fistulae, otitis media and otosclerosis. It can be performed bilaterally, but cannot be applied in vertigo conditions other than endolymphatic hydrops. The surgery can result in a 2% hearing loss, what can be attributed more to infection or inflammation than to the surgical procedure itself (36). Though the results of the endolymphatic sac surgery are conflicting, it’s been one of the most commonly performed surgical procedures in MD (35-37). A study performed at the same institution using the American Academy of Otolaryngology - Head and Neck Surgery guide showed that both the shunt surgery of the endolymphatic sac for the mastoid and the decompression of the endolymphatic sac are effective for patients with MD (38). The surgery of decompression of the endolymphatic sac, as described by PAPARELLA and SAJJADI, remains the keystone of surgical therapy for patients with MD. According to the authors, the procedure resisted to the time test, being a relatively safe procedure, with a lower than 2% index of significant sensorineural hearing loss and less than 1% risk of paresis of the facial nerve, important control (85%) of vertigo during 2 to 5 years, with excellent results also when reviewing the increase of endolymphatic sac surgery. Other studies, however, have demonstrated that the endolymphatic sac surgery does not alter, in long-term, the natural course of vertigo in MD (39, 40). In patients with MD for over 20 years and that had been submitted to the endolymphatic sac surgery, all presented long, intense and frequent vertigo crisis in comparison to those who hadn’t performed the surgery, and 90% considered their attacks to be severe or very severe (22).

Sacculotomy includes the drainage of the dilated saccule, with the resulting elimination of hydrops. Despite the present modifications in the sacculotomy techniques, CODY’s tack operation was the most popular of the surgical procedures performed at the saccule. According to some authors, this surgery can be an option when the patient has a hearing loss in the ear with hydrops, because almost 50% of the patients will have a worsening of hearing, despite the vertigo control with indexes over 60% (41). In recent histopathological study (42) of the temporal bone of patients submitted to tack operation, were observed severe endolymphatic hydrops and more severe degenerative changes in the Corti organ and cells of the spiral ganglion when compared to those of the contralateral ear, which was not operated. Severe degeneration of the saccular macula was also observed on the operated ear. According to the authors, these findings are consistent with the results of the study of hydrops in animal model, where the endoperilymphatic surgical fistulae were not enough to reduce the hydrops and still caused degenerative changes in structures of the inner ear (43). Although authors (42) didn’t have the means to detect hydrops and structural changes in the inner ear before and soon after surgery, their findings suggest that this kind of surgery was successful more by alleviating vertigo through the destruction of ciliated cells of the saccular macula, than by the resolve of the endolymphatic hydrops. Considerable damages to Corti organ also demonstrate risks of hearing loss.

SCHUNNECHT developed the cochleosacculotomy, creating a fistula between endolymph and perilymph with access via round window. Its results were analyzed by the author himself (SCHUNNECHT), designer of the procedure. According to the author, the advantage of this surgery is its technical simplicity and short realization time. In his revision of 120 cases, an extended improvement of vertigo was achieved in 70% of the cases (44). These results are better than the ones obtained with any other endolymphatic sac shunt procedure, but can be considered poor in comparison with neurectomy of the vestibular nerve, for example, that reaches 94% of vertigo control. Partial hearing losses were observed in 25% of the patients, and deep sensorineural deafness occurred in more than 10%, against less than 5% associated to vestibular neurectomy. Though other studies (45, 46) demonstrate vertigo control in more than 80% of the cases, the cochleosacculotomy is not advocated because, in long-term, this control is poor, and 30-80% of the patients will have significant hearing loss (47). Currently, cochleosacculotomy is rarely performed, but can be considered for elderly and cofotic patients with MD (48).

Utriculostomy, surgical alternative in experimental stage designed by LAVINSKY et al. (19), consists in obtaining a permanent fistula on the membranous labyrinth, communicating the endolymphatic and perilymphatic spaces on the utricle. Such objective was put to practice in sheep through the oval window, using a radiofrequency microcauter with exposure time and temperature program.

According to the author, the method is based on the fact that, when a puncture is made on the saccul, this tends to present a very fast closing, since it makes a linear rupture on the membranous labyrinth. On the other hand,
localized heat, applied with 0.20mm needle, generates the loss of substance, and was histologically observed that there is a regeneration, forming a frail membrane that behaves like a valve, disrupting itself in moments of endolymphatic hydrops and, thus, communicating the two spaces and avoiding permanent ionic mixture, with its known consequences. By the smallness of the perforation, it would tend to remain closed between episodes of MD, generating the communication only in moments of endolymphatic hydrops. Therefore, utriculostomy offers clear advantages concerning permanent endo and perilymphatic communication.

Actually, it is probable that utriculostomy might create a new "valve" in replacement of the malfunction of the utricle-endolymphatic valve, also called Bast valve (49). Although its function is still uncertain, in the opinion of some authors (50) it would close itself to prevent a larger endolymph loss on the utricular system in case of a decrease of pressure in all the endolymphatic system - or, as Zechner (51) proposed, its dysfunction would be the cause of endolymphatic hydrops.

On post-operative observations, despite the heavy manipulation of the vestibule, authors relate absence of significant losses of balance and posture. Some animals showed alterations in dynamic balance, which disappeared 24 hours after the procedure, and no nystagmus were observed.

By not acting on the saccule, that has intimate anatomical relation with the cochlea, the authors hope to be able to demonstrate the existence of a smaller cochlear repercussion in utriculostomy when compared to sacculostomies.

**Final Considerations**

Literature about endolymphatic shunt surgeries shows many controversies about its effectiveness in treatment of incapacitating MD, and many studies relate prejudicial damages to the inner ear. The experimental surgery called utriculostomy appeared as a new and promising option for the surgical treatment of MD, for presenting advantages compared to the other modalities. Additional researches must focus on the assessment of these aspects.

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