# Advances in Oto-Rhino-Laryngology

# Radiology in Oto-Rhino-Laryngology

21

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# Radiology in Oto-Rhino-Laryngology

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# Editorial Introduction

During the last 25 years diagnostic radiology in oto-rhino-laryngology has played an increasing part in radiology as a result of advances in technology.

The last 10–15 years have brought a growing amount of papers on radiology in oto-rhino-laryngology and at the same time otoradiology has been a subspeciality in radiology.

It is the editorial committee's hope that these series will give people all over the world with interests in this field of radiology – otologists, otoradiologists as well as general radiologists – a possibility to find new things of interest and to get informations about the development in different centres of otoradiology.

I will like to thank all my friends in the editional committee, all pioneers in otoradiology for their interest in cooperating in these new series.

I also want to thank the authors for their contributions and the publisher for his encouragement and support during the preparations producing this first volume.

Spring, 1973 S. BRÜNNER

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Adv. Oto-Rhino-Laryng., vol. 21, pp. 1–8 (Karger, Basel 1974)

# Radiological Evaluation of Ménière's Disease

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## *Introduction*

Though the pathogenesis of Ménière's disease is not fully known, much evidence indicates that the endolymphatic sack plays a part in the pathophysiology of this disease.

The interest of recent years in surgical treatment of Ménière's disease [HOUSE, 1964, 1969] has also drawn attention to the endolymphatic sack.

In a number of cases, surgeons have been unable to identify the endolymphatic sack. This led VALVASSORI in 1964 to start radiological examinations of the vestibular aqueduct to assist surgeons in identifying the sack. VALVASSORI observed that, in spite of a detailed tomographic study, he was either unable to visualize the vestibular aqueduct or it appeared unusually filiformed in patients with Ménière's disease. Contrary to this, the vestibular aqueduct could be visualized in more than 90% of the patients with no history of Ménière's disease.

This possibility of finding a radiological sign in patients with Ménière's disease was the reason why we [BRÜNNER and PEDERSEN, 1971] started an investigation on this subject. It soon became evident that the vestibular aqueduct can easily be identified in a normal ear and that a very small or absent vestibular aqueduct was the rule in patients with Ménière's disease. To evaluate the diagnostic significance of the radiological examinations, 48 cases – of which 17 had a clinical verified Ménière's disease – have been examined. The results are given below.

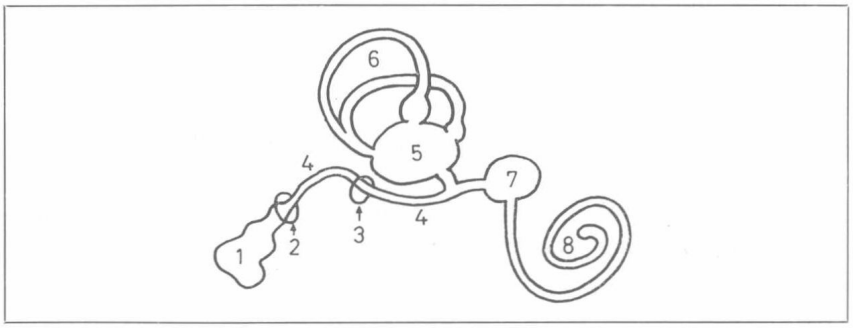


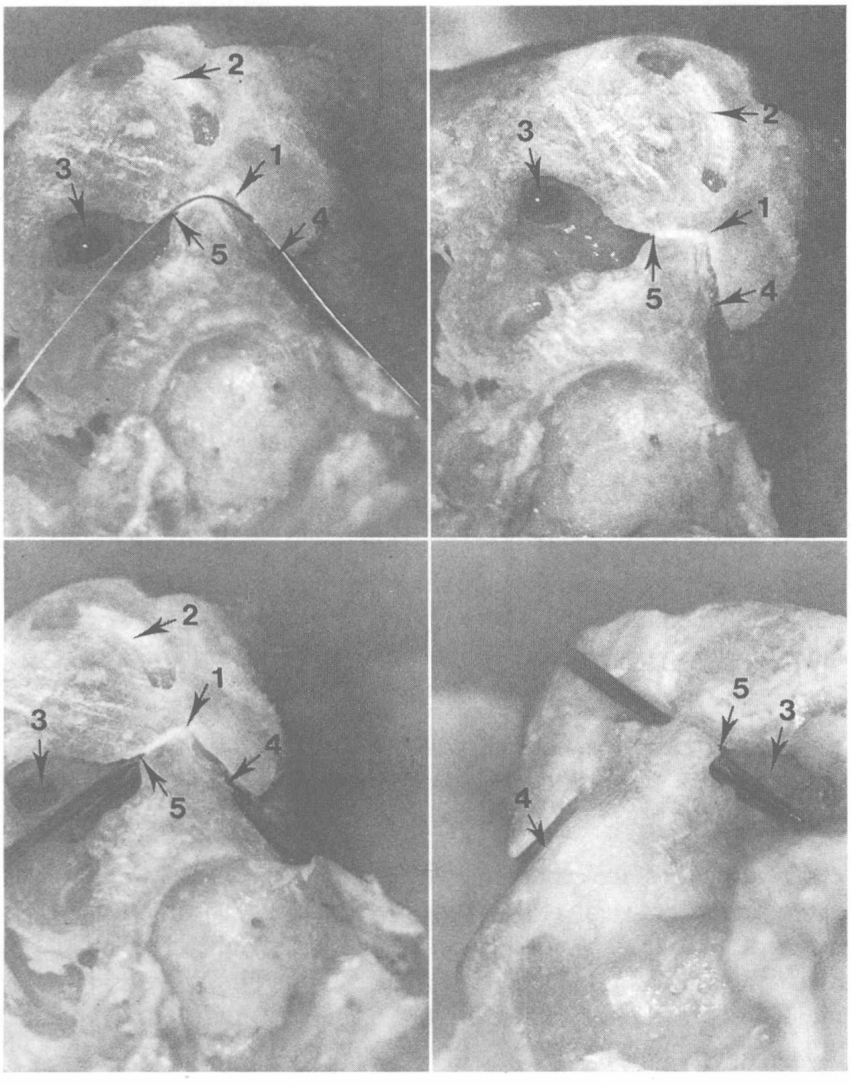

*Fig. 1.* Diagram of the endolymphatic system. 1 = Endolymphatic sack, 2 = external aperture of vestibular aqueduct, 3 = internal aperture of the vestibular aqueduct, 4 = endolymphatic duct, 5 = utricle, 6 = semicircular canals, 7 = saccule, 8 = cochlear duct.

*Anatomy*

The endolymphatic system includes the cochlear duct, the utricle, the saccule and the semicircular canals; these parts are connected with the endolymphatic duct, which continues into the endolymphatic sack (fig. 1). The endolymphatic system is located inside the petrous part of the temporal bone, but the endolymphatic sack is placed on the posterior surface of the petrosal bone and is connected with the endolymphatic system through the endolymphatic duct.

The vestibular aqueduct is the name of a small, bony canal which passes from the vestibulum to the posterior surface of the petrosal bone. The canal starts just medial to the opening of the common crus of the semicircular canals. The first part of the vestibular aqueduct is directed upwards and backwards. The canal then turns downwards and backwards and appears on the posterior surface of the petrosal bone 1 cm lateral to the internal auditory meatus. When reaching the posterior surface of the petrosal bone, the vestibular aqueduct widens so that a slit opening of 2–10 mm width appears. The diameter of the vestibular aqueduct is less than 1 mm at the beginning and increases to a few millimeters in the most posterior part of it. The external aperture of the vestibular aqueduct is subject to great variations in size and position [ANSON, 1969].

Temporal bones were dissected to visualize the anatomy of the vestibular aqueduct. Figure 2 shows a detail of the petrous part of a right




2 3

4 5

*Fig. 2–5.* The vestibular aqueduct. 1 = Vestibular aqueduct, 2 = superior semicircular canal, 3 = vestibule, 4 = external aperture of vestibular aqueduct, 5 = internal aperture of vestibular aqueduct. For further explanation, see text.

temporal bone. The specimen is seen from a medial aspect, and the section is almost sagittal. A thin, angled wire indicates the vestibular aqueduct. The wire has been removed in figure 3. Figure 4 demonstrates the close relation to the common crus of the semicircular canals; a needle

has been introduced in the common crus from the vestibulum. In figure 5, illustrating a left petrosal bone, the beginning of the vestibular aqueduct is shown situated just medial to the common crus. A needle has been introduced in the common crus from behind. The figure also shows the most posterior part of the vestibular aqueduct as it opens on the posterior surface of the petrosal bone. The part of the canal between the beginning and the posterior opening has been removed.

*Physiology*

The endolymph is believed to be produced by the stria vascularis in the periphery of the cochlear duct. DOHLMANN [1964] has stated that the endolymph is also produced from cells in plana semilunata around the vestibular sensory cells. It is believed that the endolymph floats from the cochlear and the vestibular apparatus through the endolymphatic duct to the endolymphatic sack, where a resorption takes place. A radial circulation may also occur inside the cochlea and the labyrinth.

*Pathophysiology*

Hydrops of the endolymphatic system have been described as a pathogenesis of Ménière's disease by HALLPIKE and CAIRNS [1938]. A reduced resorption of endolymph by the endolymphatic sack is believed to be the cause of the hydrops. The endolymphatic hydrops appear in some cases to be associated with ischemia of the endolymphatic sack [SHAMBAUGH, 1966].

In 4 out of 28 operations, the endolymphatic sack could not be identified [SHAMBAUGH, 1966]. CLEMIS and VALVASSORI [1968] reported a case of Ménière's disease in which a bony obliteration of the vestibular aqueduct was found at operation. SCHUKNECHT *et al.* [1968] found that loss of function of the endolymphatic sack may be the primary ethiological factor in Ménière's disease.

KOHUT and LINDSAY [1972] discuss the hypothesis that a perilymphatic, metabolic effect combined with hydrops of the endolymphatic system is responsible for the symptoms in Ménière's disease.

*Method of Investigation*

Our investigation included a total of 48 patients of whom 17 are known to have Ménière's disease. The diagnosis of Ménière's disease is based upon a history of attacks of vertigo and tinnitus, as well as loss of hearing most pronounced at lower frequencies. The cases in the control group were selected among patients who, for various reasons, were investigated by tomography of the ear. No cases with vertigo or other signs of Ménière's disease are included in this group.

The examination is performed with 1-mm cuts obtained with a Polytome. The patients lie prone with the cheek turned down 10–20°. The correct position of the posterior surface of the petrous part of the temporal bone is controlled on a TV monitor. The central rays are focussed on the external auditory meatus. Cuts are taken up to about 2.5–3.5 cm below the level of the mastoid process and about 1–1.5 cm distally; a distance of 1 mm must be maintained between the cuts [BRÜNNER and PEDERSEN, 1971].

*Results*

The results are given in table I. The vestibular aqueduct in each ear is judged and given a number 1 for normal appearance, a number 2 for a tiny canal, and a number 3 if the canal is not present. In the group

*Table I.* Results of tomographic findings of the vestibular aqueduct in patients with Ménière's disease and a control group

| Group | Hearing defect | Number of ears | Radiological evaluation of the vestibular aqueduct | | |
|---|---|---|---|---|---|
| | | | 1 | 2 | 3 |
| Ménière's disease | + | 19 | 2 | 9 | 8 |
| (n=17) | − | 13 | 3 | 5 | 5 |
| Total | | 32 | 5 | 14 | 13 |
| Control group (n=31) | | 43 | 38 | 1 | 4 |

Radiological evaluation of the vestibular aqueduct: 1=normal, 2=tiny appearance, 3=absent.

with Ménière's disease, the patients are divided into two subgroups: one in which ears are affected and another without signs of involvement.

All ears investigated without sign of Ménière's disease are included in the control group.

Table I shows that in most patients with Ménière's disease the vestibular aqueduct barely appeared or could not be identified, while in 88% of the cases in the control group a normal vestibular aqueduct could be visualized.

*Discussion*

The investigation has shown that in most cases of Ménière's disease the vestibular aqueduct cannot be visualized or barely appears under tomographic examination (fig. 6). Contrary to this, the vestibular aqueduct could be identified in 88% of the cases investigated with no history of Ménière's disease (fig. 7). Age was found not to influence the result.

Results very similar to ours have been obtained by VALVASSORI [1969]. A significant incidence of narrowing of the vestibular aqueduct evaluated by radiological examinations was found in patients with Ménière's disease by SCANLAN and GRAHAM [1971]. PORTMANN [1972] was not able to observe this finding in 12 cases investigated.

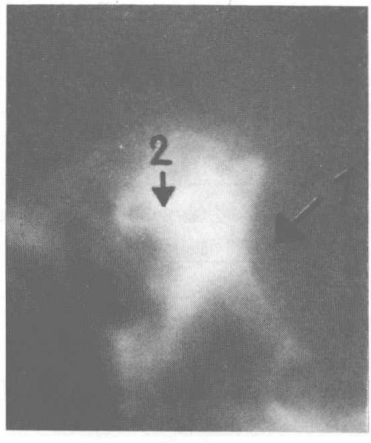



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*Fig. 6.* Tomogram of a patient with Ménière's disease. 1 = obliteration of the aqueduct, 2 = lateral semicircular canal.

*Fig. 7.* Tomogram of a normal patient in the modified lateral projection. 1 = Vestibular aqueduct, 2 = lateral semicircular canal.

The good clinical results obtained in most patients by operations on the endolymphatic sack in patients with Ménière's disease [CAWTHORNE, 1969; HOUSE, 1969; SHAMBAUGH, 1966] and from surgical findings during operation on the endolymphatic sack make it reasonable to believe that the endolymphatic sack plays a part in the pathophysiology in Ménière's disease.

The first part of the vestibular aqueduct is too small to be visualized on a tomographic examination. Therefore, the radiological examination cannot tell anything about the endolymphatic flow from the cochlea and the vestibule to the endolymphatic sack. But observations of a radiological change at the external opening of the vestibular aqueduct correlated with the fact that the endolymphatic sack plays a part in the pathophysiology of Ménière's disease indicate that location of the vestibular aqueduct by tomography may be useful as a supplement to diagnosis of Ménière's disease.

When considering operative treatment of a patient with Ménière's disease, an abnormal location of the vestibular aqueduct may be recognized before surgery by this examination. Cases of Ménière's disease with bilateral involvement may be recognized from unilateral cases by this radiological examination.

*Summary*

The results of tomographic examination of the vestibular aqueduct in 17 patients (32 ears) with Ménière's disease and a control group with 31 cases (43 ears) are given.

In patients with Ménière's disease, the vestibular aqueduct was found abnormal in 84% of the cases. Only 12% of the ears investigated in the control group showed an abnormal vestibular aqueduct.

The diagnostic significance of this finding is discussed in relation to the pathophysiology of Ménière's disease.

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# Radiology of Pendred's Syndrome

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Since the description by the English general practitioner Vaughan Pendred [28], in 1896, of two deaf-mute sisters with pronounced goiter, and the demonstration by Morgans and Trotter [25], in 1958, of a presumed specific enzymatic defect as the cause of the goiter in such deaf-mute patients, Pendred's syndrome (PS) has been defined as a triad consisting of congenital perceptive hearing loss, goiter and a pathologic perchlorate test signifying an enzymatic deficiency. Fraser [10], in 1965, collected 233 cases from the literature and estimated that the incidence in England was 8/100,000 inhabitants. The syndrome has since been described in detail by Vesterdal [37], Johnsen [23], Thould and Scowen [32], Nilsson *et al.* [27], Bax [2], Hvidberg-Hansen and Kiaer [17] and Kiaer and Hvidberg-Hansen [24], who in 1970 reviewed the clinical and thyroid aspects of our case material, consisting of 15 patients, whose genetic and clinical picture is to be further elaborated by Illum *et al.* [18].

Siebenmann [31], in 1904, mentioned incomplete development of the modiolus as an important cause of congenital deafness, and named the defect after Mondini who in 1791 described the malformation of the inner ear with hypoplasia of the modiolus, dilatation of the vestibulum and aqueductus vestibuli and coalescence of the cochlear turns to one cavity. This defect has been radiologically described more frequently in recent years, e.g. by Altman [1], Frey [11, 12], Jensen [19–21], Jensen and Rovsing [22], Gussen [14], Hvidberg-Hansen and Jörgensen [16] and Illum *et al.* [18] in cases of PS and is believed to be caused by a fault in the development of the modiolus in the 7th week of gestation, possibly resulting from a hereditary defect in the peroxidase

enzyme system, exogenous infectious, or chemical actions or radiation damage. Toxic effects on the labyrinth system before the 4th week of gestation are believed to result usually in a total aplasia while such effects at the 6th or the beginning of the 7th week are thought to result in malformations of the semicircular canals and arrest of development of the cochlea corresponding to the beginning of the basal turn, as seen in the thalidomide cases. Toxic actions at the end of the 7th week are believed to result in the fully developed Mondini defect with more or less normal semicircular canals, while a later noxious action causes smaller changes in the apex of the cochlea.

SIEBENMANN [31] proposed a classification of inner ear malformations which has proved valuable and is still widely used.

Several authors, e.g., WILDERVANCK *et al.* [39], JENSEN [19–21], PUXEDDU *et al.* [29], FREY [11, 12], VECCHI *et al.* [36], CLIFF *et al.* [7], CUMMING and MITCHELL [8], have reported malformations in cases of many different congenital anomalies. The most comprehensive review was given by JENSEN [21] who stressed the necessity of the axial-pyramidal view and thin-section tomography in the evaluation of malformations of the inner ear.

Until recent years, investigators have been unable to demonstrate radiographically anomalies of the inner ear, presumably on account of use of conventional X-ray techniques. However, since MÜNDNICH and FREY [26] in 1959 demonstrated the superior value of tomography, this has been the prefered method. In the last 5 years, PUXEDDU *et al.* [29], JENSEN [19–21], CLEMIS and VALVASSORI [6], SANSREGRET [30], BREJCHA and PIHRT [4], BRÜNNER [5], CLIFF *et al.* [7], VIGNAUD *et al.* [38], VALVASSORI [33, 34], HANAFEE *et al.* [15], BRAHE-PEDERSEN and BRÜNNER [3] and VALVASSORI and BUCKINGHAM [35] have further demonstrated the necessity of using hypocycloid thin-section tomography, primarily in the axial-pyramidal and frontal projections. The need for very careful positioning and the use of supplementary projections have been stressed by JENSEN [19] and VIGNAUD *et al.* [38].

The Mondini defect was described in detail by JENSEN [19].

### *Material*

15 cases of PS were discovered by a systematic examination of all patients at an institution for deaf-mutes, supplemented by examination