

# Delayed Endolymphatic Hydrops as a Clinical Entity

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**Abstract:** Delayed endolymphatic hydrops (DEH) is a clinical entity that can be differentiated from Ménière's disease and is typically observed in patients who have been suffering from longstanding unilateral profound inner-ear hearing loss. DEH probably is caused by delayed atrophy or fibrous obliteration of the endolymphatic resorptive system of the membranous labyrinth. The time that elapses between the occurrence of hearing loss and the onset of DEH can range from 1 to 74 years. The most common cause of hearing loss preceding DEH is juvenile-onset unilateral profound deafness (early childhood unilateral profound sensorineural hearing loss of unknown etiology), followed by labyrinthitis from various causes and physical and acoustic traumas to the inner ear. Two types of DEH exist: the ipsilateral type, in which the ear with profound hearing loss suffers progressive endolymphatic hydrops, and the contralateral type, in which the formation of progressive endolymphatic hydrops takes place in the ear opposite to the previously deafened ear. The incidence of the ipsilateral type is higher than that of the contralateral type, and the contralateral type is more common in older patients. When recurrent episodic vertigo cannot be remedied through conservative treatment, labyrinthectomy and vestibular neurectomy on the deaf ear are curative for ipsilateral DEH. However, no such surgical treatment is available for the contralateral type.

**Key Words:** contralateral; endolymphatic hydrops; ipsilateral; juvenile unilateral deafness; labyrinthectomy; recurrent vertigo

No matter what its cause, a prolonged case of profound inner-ear hearing loss—called *delayed endolymphatic hydrops* (DEH)—can induce otological symptoms similar to those of Ménière's disease [1]. Two types of DEH are identified: the ipsilateral, in which vestibular symptoms arise from the profoundly deaf ear, and contralateral, in which cochlear or vestibular symptoms occur in the better-hearing ear (mostly with normal hearing). Typically, ipsilateral DEH involves repetitive violent rotational vertigo accompanied by nausea and vomiting. However, an increase in the degree of hearing loss during episodic vertigo is rarely perceived, because profound hearing loss already exists. Conversely, contralateral DEH typically

induces fluctuating hearing loss, tinnitus, and aural fullness in the ear with good hearing and, in some cases, is also complicated by episodic vertigo. DEH is a relatively new disease concept. This study provides an explanation of the way in which this concept was established and addresses several related clinical facts that have been identified.

## ESTABLISHMENT OF THE DISEASE CONCEPT

We reported the first clinical cases that became the basis for establishing the DEH disease concept [2]. We conducted statistical analyses of 44 cases of unilateral profound sensorineural hearing loss due to undetermined etiology and usually acquired in early childhood. It was called *juvenile-onset unilateral profound* (or total) *deafness*. We found that recurrent violent episodic vertigo, similar to that of Ménière's disease, developed between the ages of 16 and 60 years in nine (20%) of the cases. However, we observed no worsening of the

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degree of hearing loss and perceived no tinnitus or aural fullness in the deaf ear during episodic vertigo.

Subsequently, Nadol et al. [3] and Wolfson and Leiberman [4] simultaneously reported 12 and 5 cases, respectively, of unilateral profound sensorineural hearing loss from various causes, all of which indicated a delayed onset of recurrent episodic vertigo. Among the 17 cases of hearing loss reported, 5 were cases of hearing loss that would fall into the category of juvenile-onset unilateral profound deafness; 7 were cases of viral labyrinthitis; 2 were cases of bacterial labyrinthitis; 2 were cases of hearing loss caused by head trauma; and 1 was a case of sudden deafness. Both reports stated that labyrinthectomy of the deaf ear produced relief from episodic vertigo. Nadol et al. [3] concluded, on the basis of the following findings (which suggest dilatation of the saccule), that episodic vertigo is probably caused by endolymphatic hydrops: (1) Analysis of the inner-ear fluid aspirated by a footplate tap (diagnostic labyrinthotomy) at the time of labyrinthectomy revealed the same biochemistry as that of the normal endolymph (two cases), and (2) a histological examination of the stapes extracted during labyrinthectomy demonstrated membranous labyrinth attached to the medial surface of the footplate (one case). The former observation was later verified by LeLievre and Barber [5] in a study of five cases.

Schuknecht [6] established a hypothesis regarding the pathogenesis of delayed episodic vertigo, stating that such vertigo occurs when the following two conditions are met: (1) a labyrinthine insult of a magnitude sufficient to cause total deafness but preserving vestibular function and (2) delayed atrophy or fibrous obliteration of the endolymphatic resorptive system (the endolymphatic sac and the vestibular aqueduct). This view is widely supported today as a proper explanation of ipsilateral DEH. However, no reports of examination of temporal bone histopathology in this disorder have been made.

Furthermore, it has been made clear that patients who have suffered from unilateral profound inner-ear hearing loss for a long time also tend to suffer from otological symptoms, including episodic vertigo, in the contralateral ear with normal hearing. I confirmed that of the 27 juvenile unilateral profound deafness patients with a delayed onset of recurrent episodic vertigo, 24 cases (89%) could be attributed to ipsilateral DEH, whereas in 3 of the cases (11%), otological symptoms closely resembling those of Ménière's disease were observed in the better-hearing ear that previously had presumably normal hearing [7]. Schuknecht [1] also conducted an analysis of 18 cases of profound inner-ear hearing loss from various causes with delayed onset of otological symptoms. Symptoms consistent with ipsi-

lateral DEH were observed in 12 of the cases (67%), whereas in 6 of the cases (33%), the better-hearing ear (in most cases with normal hearing) became affected, with a later onset of fluctuating inner-ear hearing loss, recurrent episodic vertigo, or both. Thus, Schuknecht suggested that the clinical diagnosis for delayed otological symptoms resembling those of Ménière's disease and occurring in patients with profound inner-ear hearing loss should be DEH of either the ipsilateral or the contralateral type.

As for the pathogenesis of the contralateral type of DEH occurring in patients with juvenile unilateral profound deafness, we considered the following possibilities [8]: The same etiology that triggered monaural hearing loss during early childhood simultaneously induced a subclinical lesion in the contralateral inner ear, which eventually led to the delayed onset of progressive endolymphatic hydrops. Reports indicating a decreased caloric nystagmus response in the ear with normal hearing in some patients with juvenile unilateral profound deafness without complications of DEH support the foregoing hypothesis, although the percentage of such patients among the total number of such patients differs from report to report, varying from 2 percent [9] to 5 percent [8] and up to 26 percent [10]. Schuknecht et al. [11,12] held a similar view regarding the etiology of contralateral DEH. They conducted a series of histopathological studies of the temporal bone in three cases of contralateral DEH occurring in patients who had been suffering from unilateral hearing loss of unknown etiology since early childhood (which could correspond to juvenile unilateral profound deafness). Findings obtained included, in the profoundly deaf ears, pathological changes similar to those known to occur in mumps and measles labyrinthitis and, in the opposite (hearing) ears, pathological changes similar to those known to occur in Ménière's disease. Thus, Schuknecht et al. deduced that when one ear becomes deaf owing to viral infection in early childhood, the contralateral ear is also affected with subclinical viral infection, which eventually causes the ear with normal hearing to develop, over a long period, a histopathology similar to that of Ménière's disease.

## CLINICAL FACTS

### Cause of Hearing Loss Preceding DEH

The most common cause of the profound hearing loss that precedes DEH is juvenile unilateral profound deafness [12-19], found in more than one-half of all cases [12,19,20] and followed by labyrinthitis induced by otitis media and viral labyrinthitis induced by mumps, measles, or influenza [1,3-5,11,12,14,15,17,19,20]. Other

causes include acoustic trauma [20–22]; head trauma [1,3,11,12,18,21]; sudden deafness [4,12,17,19, 20,23]; bacterial labyrinthitis (induced by meningitis, scarlet fever, diphtheria, etc.) [1,12,14,18,19,24]; stapedectomy [24]; otosclerosis [25]; bilateral congenital deafness [25, 26]; cochlear implant operation [27,28]; congenital cytomegalovirus infection [29]; and migraine [30].

### Latency Period

The latency period between preceding hearing loss and the onset of DEH varies from 1 to 74 years [3,12, 18,24]. The relationship between the cause of the preceding profound sensorineural hearing loss and the latency of DEH has not been clarified. However, DEH seems to manifest itself within 10 years of the occurrence of hearing loss due to sudden deafness or acoustic trauma [20,23] and within 20 years in most cases of hearing loss caused by mumps [19]. In inner-ear hearing loss caused by head trauma, especially when the trauma does not involve a temporal bone fracture, DEH may occur even within a month of latency [21].

### Incidence of DEH

Whether the manifestation of DEH is influenced by the cause of the preceding hearing loss remains unclear. The incidence of DEH in patients with juvenile unilateral profound deafness but without episodic vertigo is 17% in an average of 15 years after the time they first consult a doctor regarding a chief complaint of hearing loss [20]. In their lifetimes, 30% of such patients could develop DEH [7]. Some reports deny any great difference between the incidence of the ipsilateral and the contralateral types [12,14,23,31,32]. However, most reports address only the ipsilateral type [3,4,18,19,24] or indicate that the incidence is higher for this type [1,5,7,13,15–17,33,34]. In our experience, 89% of DEH cases caused by juvenile unilateral profound deafness and 87% of those caused by other kinds of hearing loss are of the ipsilateral type [7,20]. Indirectly estimated, the DEH morbidity rate in the normal population could be as high as 0.05% [35], which is equivalent to the prevalence rate of Ménière's disease in the normal population (approximately 0.02% in Sagami-hara City, Japan [36], and near 0.2% in Rochester, MN [37]), and to the yearly incidence rate of Ménière's disease (approximately 0.05% in Sweden [38]).

### Age of Onset

Fewer cases of DEH occur before the age of 10 [5,7]. DEH generally occurs sometime between the ages of 11 and 20 or 41 and 60 years [20,34]. Cases that occur

between the ages of 11 and 20 years are mainly of the ipsilateral type caused by juvenile unilateral profound deafness, whereas those caused by hearing loss other than juvenile unilateral profound deafness usually occur after the age of 20 years [20]. The age at which DEH first occurs is generally higher for the contralateral type than the ipsilateral type [31,32,34], probably because residual degeneration in the inner ear with good hearing is less severe than that in the ear with profound hearing loss.

## TYPES OF DEH AND THEIR CLINICAL DIAGNOSES

### Ipsilateral DEH

Although ipsilateral DEH can occur in patients with bilateral profound inner-ear hearing loss [1,5,26], most DEH cases are observed in patients with unilateral profound hearing loss, often with normal hearing in the contralateral ear [1,5,7,14]. Hence, the guidelines for the clinical diagnosis of typical ipsilateral DEH would be as follows: (1) many years of monaural profound sensorineural hearing loss or anacusis (early phase); (2) development of recurrent vestibular symptoms resembling those of Ménière's disease, mostly rotational vertigo with vegetative symptoms lasting for less than one to several hours (late phase); (3) no perceived fluctuation in hearing loss related to episodic vertigo (although when residual hearing is observed in the ear with profound hearing loss, it is possible to prove a fluctuation in hearing loss by repeated audiometry); and (4) exclusion of central nervous system involvement.

Usually, patients with ipsilateral DEH [4,24], especially DEH caused by juvenile unilateral profound deafness [7], do not perceive tinnitus in the deaf ear, which is why they do not attribute the cause of episodic vertigo to the aural problem. However, some patients perceive tinnitus or aural fullness in the deaf ear or an increase in tinnitus during episodic vertigo, causing them to realize that their vertigo may have been induced by the deaf ear [3,4,32]. Determining the ear that is causing vertigo is usually difficult when ipsilateral DEH occurs in patients with bilateral profound inner-ear hearing loss unless it is accompanied by tinnitus or aural fullness during episodic vertigo. However, biochemical analysis of the inner-ear fluid obtained by diagnostic labyrinthotomy [3,5] or a glycerol test using vestibular evoked myogenic potentials (VEMPs) [39] may serve to reveal the presence of endolymphatic hydrops in such cases. If a decreased caloric nystagmus response is proved, the possibility that the same ear is causing the vertigo is high. However, many cases of profound inner-ear hearing loss involve a decreased

caloric nystagmus response in general, even when no DEH is present. Therefore, this observation cannot be used as a basis for a final determination of the ear responsible for vertigo.

### Contralateral DEH

The clinical diagnosis of typical contralateral DEH would be as follows: (1) many years of monaural profound sensorineural hearing loss or anacusis (early phase); (2) development of fluctuating sensorineural hearing loss in the opposite ear that previously had normal hearing (late phase); (3) occurrence of recurrent episodic vertigo similar to that of Ménière's disease in approximately 50% of all cases [12]; and (4) exclusion of central nervous system involvement.

In nearly one-half of cases of contralateral DEH, only fluctuating hearing loss takes place in the previously normal hearing ear, without the occurrence of vertigo [12]. This could be attributed to the fact that, in patients with contralateral DEH, some 39% of the better-hearing ears showed no response to caloric stimulation, whereas nearly 13% of the deaf ears of the patients with ipsilateral DEH showed no response [12]. In other words, the better-hearing ear in the contralateral cases generally suffers greater pathological changes in the vestibular labyrinth than does the deaf ear in ipsilateral cases, which means that vestibular symptoms are not frequently observed in contralateral cases [1,12]. Conversely, when episodic vertigo occurs as a complication of contralateral DEH, such cases are hardly distinguishable from Ménière's disease [1,7,12,16].

### Bilateral DEH

Later, Schuknecht [40] mentioned bilateral DEH, although it is not generally accepted as a type of DEH. His definition of the bilateral type is as follows: impairment in auditory or vestibular function (or both) in both ears due to viral labyrinthitis in the early phase and a manifestation of symptoms of progressive endolymphatic hydrops in both ears in the late phase. The problem with this definition is that the etiology of the early phase is specifically noted and the degree of hearing loss in the early phase is not limited to profound hearing loss. Consequently, this definition of bilateral DEH is not fully consistent with those of the ipsilateral and contralateral types, which could cause confusion for physicians making a clinical diagnosis of bilateral DEH. Bilateral DEH should be defined as DEH that occurs in both ears, although the time of onset could be different for each ear [35]. In other words, in bilateral DEH, both ears are affected by the ipsilateral type, or

one ear is affected by the ipsilateral type while the other ear is affected by the contralateral type.

## NEUROOTOLOGICAL EXAMINATIONS

### Audiometry

To establish a diagnosis of DEH, the presence of profound sensorineural hearing loss in one or both ears must be confirmed by pure-tone audiometry. In the case of the ipsilateral type with residual hearing, a fluctuation in hearing loss could be confirmed by running a few audiometry tests on different days. When the contralateral type is suspected, repeated audiometry is needed to confirm whether a fluctuation in hearing loss is present in the better-hearing ear. Békésy audiometry is performed for the contralateral type to prove the presence of recruitment in the better-hearing ear [1]. An increase in the SP/AP ratio observed by electrocochleography, which suggests the presence of endolymphatic hydrops, is observed in 60% of contralateral DEH cases [34,41]. The glycerol test [42] is suitable for determining the presence of endolymphatic hydrops in the contralateral type [1,32].

### Nystagmus Test

Spontaneous nystagmus during episodic vertigo observed in DEH patients is usually of the horizontal or horizontal rotatory type, the direction of which is either toward or away from the affected ear [7], as in Ménière's disease. In some cases of the ipsilateral type, paralytic spontaneous nystagmus directed toward the healthy ear occurs during episodic vertigo, followed by rotatory recovery nystagmus directed toward the deaf ear [43]. Vestibular symptoms are usually not present between vertigo attacks; however, head-shaking nystagmus can often be induced by the head-shaking test [8].

### Caloric Test

In DEH, a decreased caloric nystagmus response on one side does not necessarily indicate that the same side of the ear is responsible for vertigo, owing to the following facts: Even in juvenile unilateral profound deafness cases that do not involve DEH, 28–56% of deaf ears and 2–26% of the contralateral (hearing) ears will show a decreased caloric nystagmus response [8–10]. For all cases of DEH, 10% of the ipsilateral type [32] and 30–60% of the contralateral type [31,32] had vestibular paresis to caloric stimulation in both ears, whereas for all cases of the ipsilateral type, 82% of the deaf ears and 11% of the better-hearing ears had vestibular paresis [17].

## Vestibular Evoked Myogenic Potentials

VEMPs are an established test for exploring the sacculo-collic reflex [44,45]. Recent studies [39,46] show that the glycerol test using VEMPs may be useful to detect endolymphatic hydrops in cases of both ipsilateral and contralateral DEH.

## DISTINCTION FROM MÉNIÈRE'S DISEASE

One of the conditions for diagnosing Ménière's disease or idiopathic endolymphatic hydrops is the nonexistence of any causative diseases in the auditory and vestibular systems [1,11–13,47,48]. This and the clinical fact that profound hearing loss seldom is observed in the early phase of Ménière's disease simplify distinguishing typical ipsilateral DEH from Ménière's disease [13,14]. However, in some patients with physical and acoustic labyrinthine traumas [21,22,49,50] or subclinical chronic otitis media [51,52] involving various degrees of sensorineural hearing loss, the delayed onset of vertigo cannot be distinguished from the vertigo observed in Ménière's disease. Endolymphatic hydrops identical to that seen in Ménière's disease has been observed in postmortem histological examinations of the temporal bones of some such patients [21,51,52]. Therefore, ipsilateral DEH could occur not only in cases of profound inner-ear hearing loss but in relatively mild cases [13].

Typical contralateral DEH can be easily diagnosed because of its unique clinical features. However, when otological symptoms, including vertigo, are manifested in the normal ear of patients with unilateral profound sensorineural hearing loss, completely ruling out the possibility of the coincidental development of Ménière's disease usually is difficult. Nevertheless, as far as contralateral DEH arising from juvenile unilateral profound deafness is concerned, the incidence in patients with juvenile unilateral profound deafness is 3.4% (3 of 89 cases) [7], which is much higher than the prevalence rate (0.02–0.2%) for Ménière's disease in the general population [36–38]. Therefore, probably a justifiable consideration is that in making a clinical diagnosis of such a case, contralateral DEH should be considered first [16,35].

Conversely, the fact that clinical symptoms of contralateral DEH with vertigo in many cases closely resemble those of typical Ménière's disease indicates the possibility of past subclinical unconscious inner-ear lesion causing Ménière's disease. Schuknecht et al. [12] reasoned in their later thesis that Ménière's disease may occur as a delayed sequela of inner-ear damage sustained during an attack of subclinical viral labyrinthitis

occurring in childhood. The disease concept of DEH was previously formed on the basis of typical DEH cases, and it may have to be expanded to include less severe cases of hearing loss in the early phase.

## TINNITUS

Patients with juvenile-onset unilateral profound deafness do not usually complain of any symptoms except for unilateral hearing loss. However, our old data [7,8] showed that DEH developed in 27 (30%) of 89 patients with this deafness at and after age 9 years in all cases. They could be classified later into two groups of DEH, the ipsilateral type (24 patients) and the contralateral type (3 patients), after the concept of a contralateral type of DEH had been established by Schuknecht [1]. Among the 24 patients with ipsilateral DEH were 3 patients with mild, nonfluctuating sensorineural hearing loss unaccompanied by tinnitus due to unknown cause in the better-hearing ear and 2 patients with normal hearing in the better-hearing ear. In these, however, tinnitus had appeared in the ear with normal hearing probably during adolescence in one case and at the age of 23 in another, being not clearly related to the patients' recurrent vertigo. I think that these cochlear symptoms (mild deafness or tinnitus) in the normal or better-hearing ear in the five cases might have been caused by a latent histological DEH, although vertigo attacks in these cases were typical of the ipsilateral type of DEH (recurrent episodic vertigo unaccompanied by cochlear symptoms).

Although our data are insufficient, the idea that tinnitus of an unknown cause may occur, in general, in the ear with normal hearing based on latent (mild) DEH seems to be reasonable, because the same idea has already been expressed by Shulman [53] and Shulman et al. [54]. These researchers state that a delayed or secondary endolymphatic hydrops occurring months or years after the initial endolymphatic sac injury has clinically been found to have a significant incidence of occurrence in patients with subjective idiopathic tinnitus (SIT) of the severe disabling type and that SIT may be the initial symptom of a delayed or secondary endolymphatic hydrops and an early "soft" sign of a gradual progressive sensorineural hearing loss. We know further, from the literature, that in juvenile unilateral profound deafness cases that do not involve clinical DEH, 2–26% of the ears with normal hearing will show a decreased caloric response, as mentioned earlier. It has also been shown that the incidence of dominant negative summating potential in the better-hearing ear was 20% in the ipsilateral DEH [34]. Seemingly, therefore, clinical analytical research of the normal or better-hearing ear in patients with juvenile unilateral profound deafness with or without clinical DEH may contribute

to clarifying the pathophysiology of idiopathic cochlear tinnitus.

## TREATMENT

### Conservative Treatment and the Prognosis for Patients with Vertigo

DEH requires conservative treatment, such as that typically used for Ménière's disease, including the appropriate concomitant use of diuretics, such as isosorbide [55] or steroids for at least 3–6 months [1,5,13,16,18,19,34,35]. This disease often is resistant to conservative treatment [1,13,20,56], but sometimes episodic vertigo can subside spontaneously [1,13,15]. In approximately 65% of patients with DEH, episodic vertigo disappears within 5 years with conservative treatment [20].

### Surgical Treatment

When episodic vertigo cannot be controlled by conservative treatment, surgical treatment can be applied for the ipsilateral type. The most effective surgeries are transtympanic or transmastoid labyrinthectomy [1,3–5,12,14,18,24,25,40,55] and translabyrinthine vestibular neurectomy [13,14,19,24,57,58]. Pharmacological labyrinthectomy, especially transtympanic gentamicin therapy [15,56,59], is also effective. Some claim, however, that conservative surgeries, such as endolymphatic sac drainage [14,17], sacculotomy [5], or cochleosacculotomy [56], should be considered before a destructive surgery of the labyrinth is selected. However, the effect of such conservative surgery in controlling episodic vertigo has not been entirely satisfactory [55].

For contralateral DEH, labyrinthectomy and translabyrinthine vestibular neurectomy should not be selected, for they will completely destroy the auditory function of a patient [1]. Even selective vestibular neurectomy by the retrosigmoid or retrolabyrinthine approach could induce or aggravate hearing loss in 8% [60] to 32% [61] of all cases or in 50% in the long-term follow-up evaluation [58], with the degree of loss ranging from 10 dB to profound deafness. Hypacusis of various degrees could occur in any conservative surgery. However, endolymphatic sac drainage is relatively safe in preserving hearing [14,16], and the possibility of losing all hearing function by endolymphatic sac drainage is less than 2% [14]. Therefore, when surgical treatment is necessary for the contralateral type of DEH, endolymphatic sac drainage should be considered [14,16].

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