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Pathology of endolymphatic hydrops

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THE term 'endolymphatic hydrops' means increased volume of endolymph, which is histologically shown as distention of the endolymphatic system in the inner ear. Reissner's membrane is ballooned and may contact the bony walls of the scala vestibuli. The saccule, whose membranous walls in the vestibule are distended, may contact the undersurface of the footplate of the stapes. The walls of the utricle and semicircular canals are less prone to distention due to their firm attachment to the endosteum lining the vestibule and bony semi-circular canals.

The histological diagnosis of endolymphatic hydrops is non-specific; therefore, the term should not be used as a synonym for Menière's disease. Schuknecht (1977) described two types of endolymphatic hydrops: progressive, which is due to permanent imbalance between secretion and resorption of endolymph; and non-progressive, which occurs in response to traumatic or toxic insult of limited duration to the membranous labyrinth. Table I shows the different etiologies of endolymphatic hydrops.

The present study demonstrates the incidence of endolymphatic hydrops in different otological conditions and the relationship of its severity to the clinical symptomatology.

Materials and methods

The temporal bone collection at the Ear Research Institute consists of 703 ears from 357 subjects who ranged in age from a nine-week fetus to 98 years. The collection was examined to determine the incidence of endolymphatic hydrops. This study defines endolymphatic hydrops as diffuse distention of the cochlear duct shown by displacement of Reissner's membrane toward the scala vestibuli and associated with ballooning of the saccule, with displacement of its walls towards the footplate of the stapes. The degree of hydrops was arbitrarily divided into mild, moderate, and severe according to the amount of distention of Reissner's membrane and the saccule (Fig. 1).

The relationship of histological findings to the severity of clinical symptoms was studied. Although some ears exhibited more than one pathology, a reasonable principal diagnosis was assigned on the basis of clinical history and histopathological appearance.

Findings

Endolymphatic hydrops was present in 57 ears (9 per cent), including

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TABLE I

ETIOLOGIES OF ENDOLYMPHATIC HYDROPS (SCHUKNECHT, 1977)

I. *Non-progressive hydrops*	II. *Progressive hydrops*
Acute otitis media	Menière's disease
Chronic otitis media	Congenital syphilis
Trauma	Delayed hydrops
Fenestration of lateral canal	(temporal bone fractures,
Stapedectomy	stapedectomy, sudden deafness,
Otosclerosis	mumps, diphtheria, bacterial
Paget's disease	labyrinthitis, idiopathic
Viral labyrinthitis	congenital deafness)

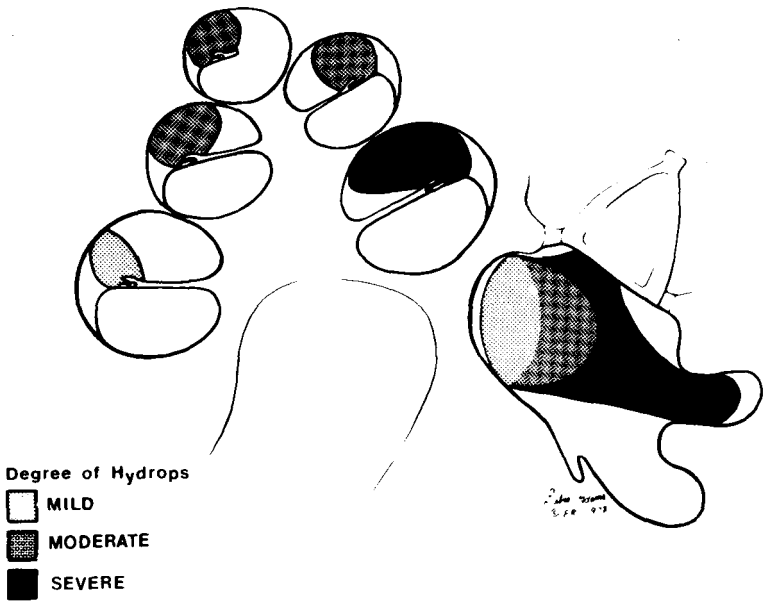


FIG. 1
Histological classification of endolymphatic hydrops.

normal ears, post-stapedectomy and post-acoustic tumor surgery ears, and ears with the primary diagnosis of Menière's disease, congenital syphilis, chronic otitis media, or other disorders (Table II). Figs. 2, 3, 4 and 5 illustrate the severity of endolymphatic hydrops in different otological disorders. Two temporal bones from two newborns who died shortly after birth showed mild endolymphatic hydrops (Fig. 6). Ten temporal bones showed hydrops localized to the cochlear duct, i.e., cochlear hydrops, associated with a normal saccule. The cochlear hydrops cases included normal ears, post-stapedectomy cases, and ears with chronic otitis media or presbycusis.

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TABLE II

SEVERITY AND DISTRIBUTION OF HYDROPS IN DIFFERENT OTOLOGICAL DISORDERS

Diagnosis	No. of persons	No. of bones	Severity of endolymphatic hydrops	
			Cochlear duct	Saccule
Otosclerosis, post-operative	14	15	5 severe 2 moderate 8 mild	3 moderate 4 mild 3 fibrosis 4 normal
Chronic otitis media	2	3	3 moderate	1 moderate 2 normal
Congenital syphilis	2	4	2 severe 2 moderate	2 severe 2 moderate
Presbycusis	3	4	2 severe 1 moderate 1 mild	1 mild 3 normal
Hereditary sensorineural hearing loss	2	3	2 severe 1 moderate	2 severe 1 moderate
Acoustic neuroma (post-operative)	2	2	1 severe 1 mild	1 mild
Paget's disease	1	1	1 mild	1 mild
Normal	2	2	2 mild	2 mild
Menière's disease	9	11	7 moderate 3 moderate 1 mild	4 severe 2 moderate 2 mild 2 fibrosis 1 collapse
Others	10	12	6 severe 4 moderate 2 mild	4 severe 2 moderate 3 mild 2 artifact 1 normal
TOTAL	47	57	25 severe 16 moderate 16 mild	12 severe 11 moderate 15 mild 10 normal 3 artifact 5 fibrosis

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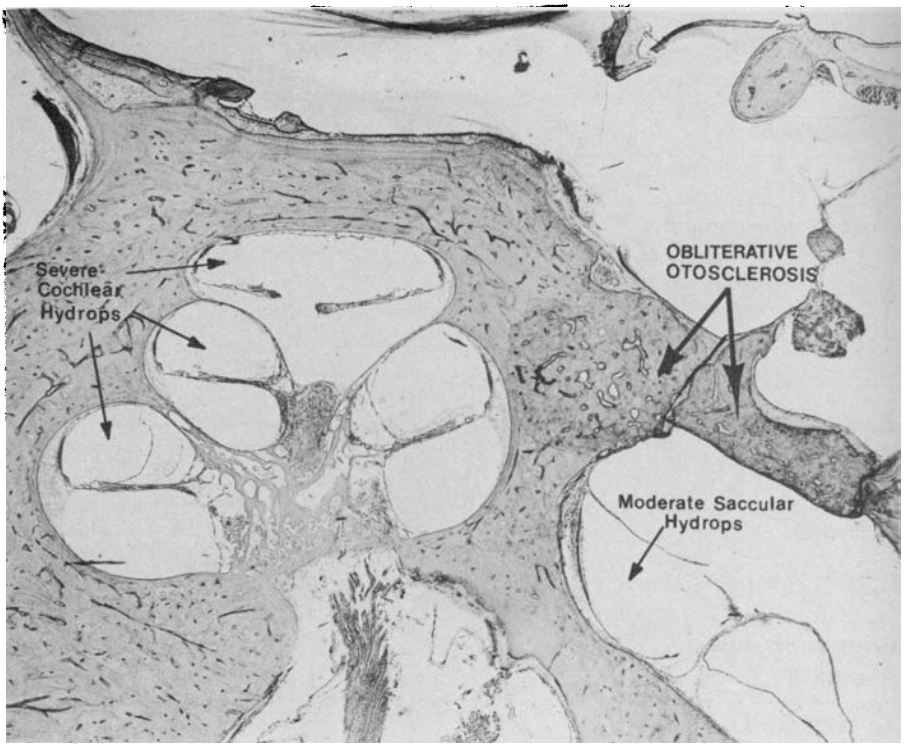


FIG. 2

M.M., a 59-year-old female with bilateral otosclerosis, underwent a right fenestration and a left fenestration-stapedectomy. She complained of intermittent dizziness aggravated by neck movements and of severe bilateral tinnitus. ENG tracings two years before death showed positional nystagmus in the head-hanging-left position. Histological examination of the right temporal bone showed obliterative otosclerosis of the footplate of the stapes. The lateral semicircular canal showed an opened fenestra. There were severe endolymphatic hydrops of the cochlear duct and moderately severe hydrops of the saccule.

TABLE III

RELATIONSHIP OF HYDROPS ETIOLOGY TO CLINICAL SYMPTOMS
(N—39)

Endolymphatic hydrops	Hearing loss		Tinnitus	Pressure	Dizziness	
	Fluctuant	Sensorineural			Episodic	Constant
Progressive (N—13)	61%	39%	100%	76%	76%	24%
Non-progressive (N—26)	4%	92%	50%	8%	20%	32%

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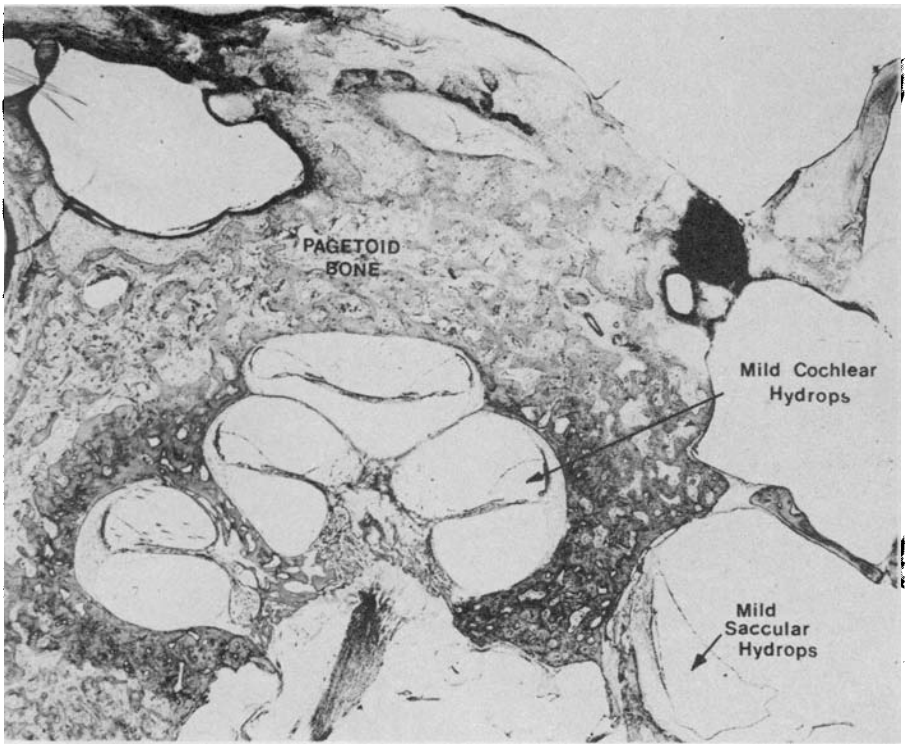


FIG. 3

K.W., a 57-year-old female, had progressive mixed bilateral sensorineural hearing loss of three years' duration. A left middle ear exploration showed no ossicular fixation. Histological examination of the temporal bones after her death at age 69 revealed involvement of the otic capsule by Paget's disease. There was mild endolymphatic hydrops in the cochlea and saccule.

TABLE IV

RELATIONSHIP OF HYDROPS SIDE TO CLINICAL SYMPTOMS
(N—39)

Endolymphatic hydrops	Hearing loss		Tinnitus	Pressure	Dizziness	
	Fluctuant	Sensorineural			Episodic	Constant
Unilateral (N—30)	16·6%	80%	70%	33·5%	37%	26·5%
Bilateral (N—9)	33·5%	66·5%	55·5%	44·5%	55·5%	33·5%

Results of complete otologic history and neurotologic evaluation were available in 39 cases of endolymphatic hydrops. The etiology, side, severity and distribution of the hydrops in these cases were correlated with the severity of clinical symptoms (Tables III, IV, V and VI).

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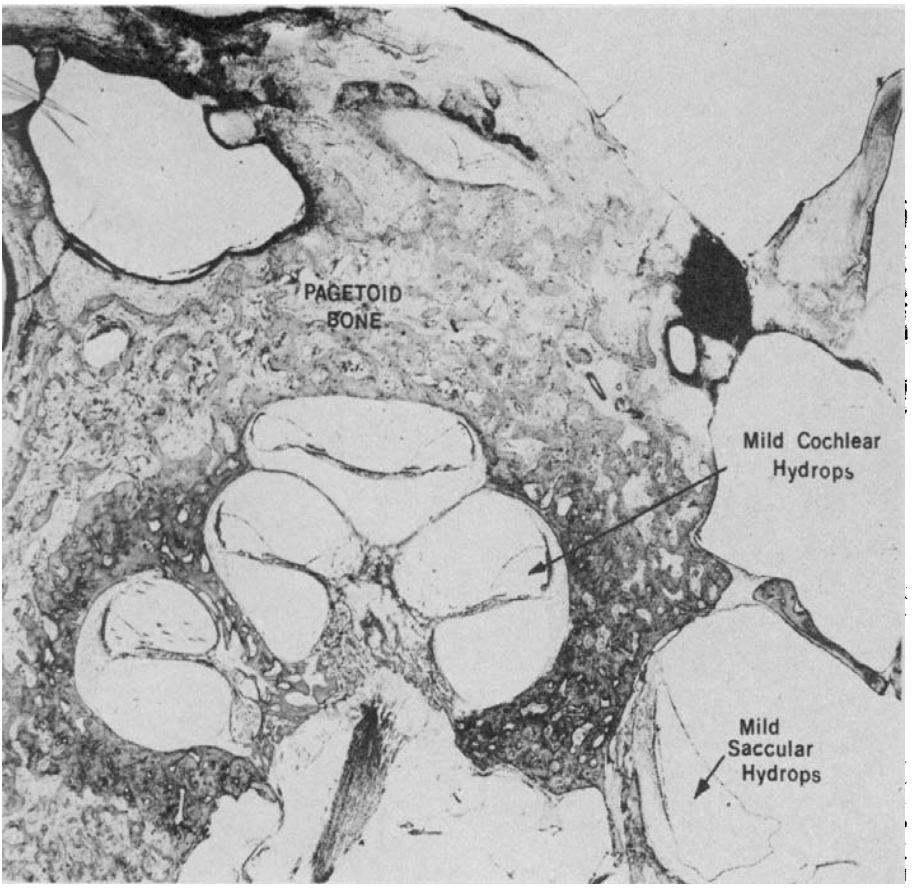


FIG. 4

This 60-year-old male had been treated intermittently for congenital syphilis since childhood. He complained of bilateral progressive sensorineural hearing loss and constant unsteadiness. ENG tracings revealed bilaterally absent caloric responses. Examination of the temporal bones following his death at age 65 showed diffuse osteitic changes in the otic capsule and severe endolymphatic hydrops in the entire membranous labyrinth.

TABLE V

RELATIONSHIP OF HYDROPS SEVERITY TO CLINICAL SYMPTOMS
(N—39)

Endolymphatic hydrops	Hearing loss		Tinnitus	Pressure	Dizziness	
	Fluctuant	Sensorineural			Episodic	Constant
Mild (N—11)		91%	45·5%	18%	27%	18%
Moderate (N—12)	41·5%	58·5%	50%	25%	33·5%	25%
Severe (N—16)	25%	75%	81%	50%	50%	25%

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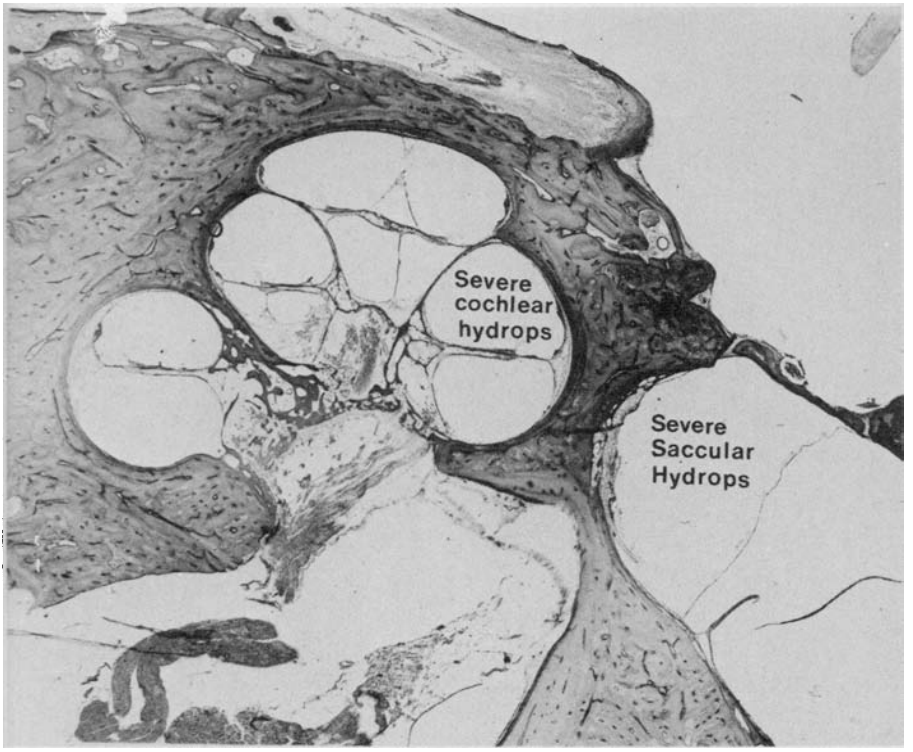


FIG. 5

M.C., a 52-year-old female, had bilateral tinnitus and intermittent dizziness for three years. She also complained of bilateral progressive hearing impairment, worse in the left ear. ENG tracings revealed spontaneous nystagmus, direction-fixed positional nystagmus and absent left caloric response. The mid-modiolar section of the left cochlea showed severe cochleosaccular endolymphatic hydrops. The right temporal bone also showed severe endolymphatic hydrops.

TABLE VI

RELATIONSHIP OF HYDROPS DISTRIBUTION TO CLINICAL SYMPTOMS
(N—39)

Endolymphatic hydrops	Hearing loss		Tinnitus	Pressure	Dizziness	
	Fluctuant	Sensorineural			Episodic	Constant
Cochlear (N—11)	9%	82%	73%	27%	18%	18%
Sacculo-cochlear (N—28)	28·5%	71·5%	60·5%	39%	46·5%	18%

Discussion

It is often difficult to obtain conclusive and objective clinical evidence for the diagnosis of endolymphatic hydrops (Berkowitz *et al.*, 1974). The deciding factor for this diagnosis is usually based on symptoms of

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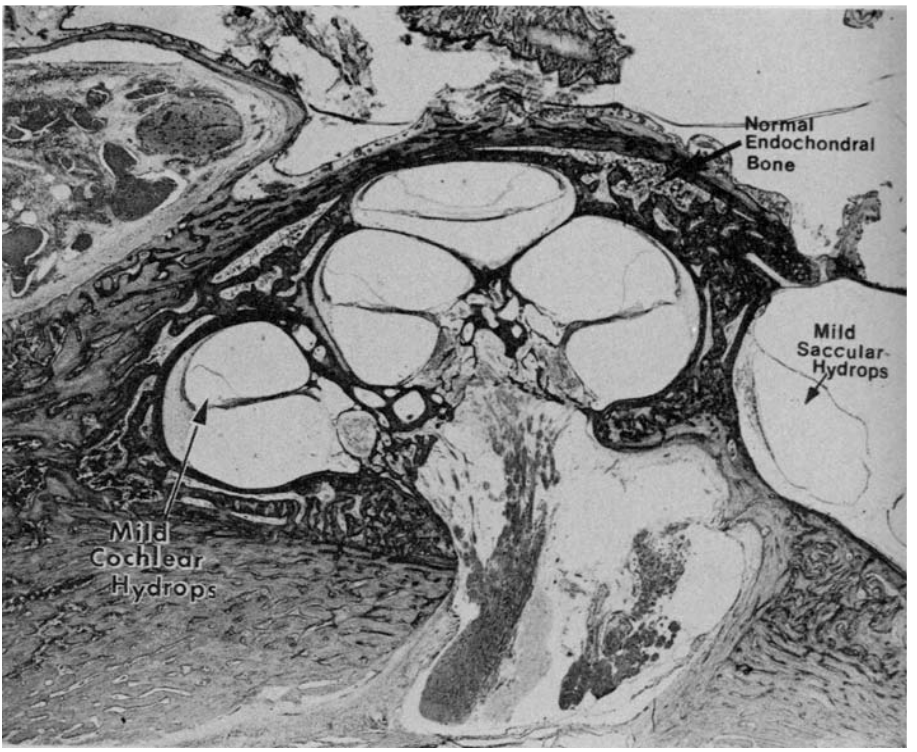


FIG. 6

Histological examination of the temporal bones of B.P., a male who died of aspiration pneumonia a few days after birth, revealed bilateral acute otitis media with mild endolymphatic hydrops.

fluctuating hearing loss, tinnitus, fullness in the ear, and vertigo. Results of audiometric and vestibular tests may support the probability of the diagnosis. However, this characteristic clinical picture is not always seen. Cochlear hydrops is characterized by fluctuant hearing loss, tinnitus, and pressure, but no vertigo. Vestibular hydrops produces vertigo, tinnitus, and pressure, but no hearing loss. Dual or multiple pathologies complicate the picture even more. For instance, presbycusis or noise-induced hearing loss is frequently associated with endolymphatic hydrops or other causes of vertigo.

In the present study, dizziness occurred in 65 per cent of cases with histologically proven endolymphatic hydrops. Characteristic episodic vertigo occurred in only 25 per cent of this total. The incidence of pressure sensation and tinnitus was 35 and 66·5 per cent respectively. All cases, except one with cochlear hydrops, had sensorineural hearing loss. Hearing levels fluctuated in only 25 per cent. Fig. 7 summarizes these clinical findings.

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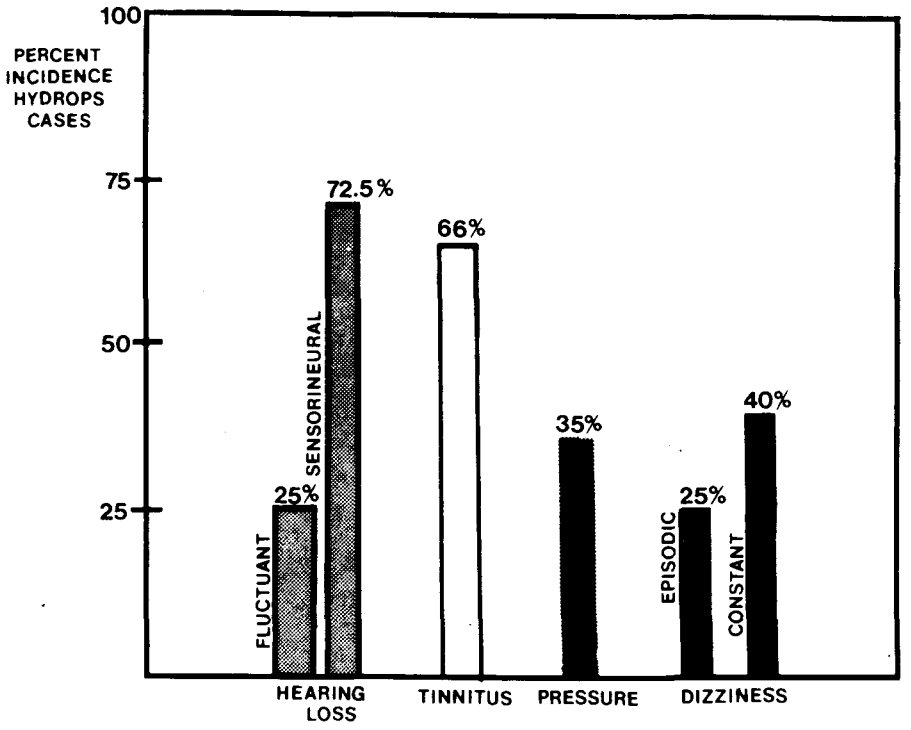


FIG. 7

Incidence of clinical symptoms in the total group of histologically proven endolymphatic hydrops.

Thus, the typical picture of endolymphatic hydrops, i.e., fluctuant hearing loss, episodic vertigo, tinnitus, and pressure, was only seen in about one-third of cases with histologically proven endolymphatic hydrops. This symptom complex commonly occurred in cases with severe bilateral cochleo-saccular endolymphatic hydrops. Progressive endolymphatic hydrops, i.e., associated with Menière's disease, and congenital syphilis were associated with the characteristic symptom complex in about 75 per cent. The clinical differentiation between progressive and non-progressive hydrops is important since the clinical symptomatology seems to depend on the disease causing progression of the hydrops, rather than on the presence of hydrops *per se*.

A reliable clinical test to determine the presence of endolymphatic hydrops is needed. Klocknoff and Lindblom (1966) reported on the glycerol test as a means of diagnosing endolymphatic hydrops. A positive result indicates hydrops, but a negative result does not exclude the diagnosis. However, psychological factors have recently been shown to play a significant role in the outcome of this test (Thomsen and Vesterhauge, 1979).

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Since endolymphatic hydrops is a non-specific diagnosis and occurs in a wide variety of otological disorders, the authors feel that neurotologic evaluation is indicated in every case suspected of having endolymphatic hydrops to determine specific causes.

Summary

The term 'endolymphatic hydrops' is not a definitive diagnosis since the condition occurs in a variety of otological disorders. Endolymphatic hydrops was found in 57 ears (9 per cent) of the 703 temporal bones in the collection of the Ear Research Institute, Los Angeles. These 57 ears included normal ears of two newborns, post-stapedectomy ears, and ears with the primary diagnosis of Menière's disease, Paget's disease, congenital syphilis, and chronic otitis media. The typical clinical picture of fluctuant hearing loss, pressure, tinnitus, and episodic vertigo occurred in about one-third of the cases with histologically documented hydrops. This symptom complex was common in cases with diffuse, bilateral, cochleosaccular, or progressive hydrops. A reliable clinical test to determine the presence of endolymphatic hydrops is needed. Complete neurotologic evaluation is indicated in every case suspected of having hydrops to determine specific causes.

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