RECURRENT MENINGITIS SECONDARY TO IDIOPATHIC OVAL WINDOW CSF LEAK.*

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ABSTRACT.

Bacterial meningitis remains a life-threatening infection even in the present antibiotic era; thus, any abnormality which predisposes a patient to a recurrence of this serious disease, must be identified and corrected. This report describes the history of a 12-year-old boy with a profound neurosensory hearing loss, a related absence of vestibular function and a Mondini-type of temporal bone dysplasia who developed recurrent episodes of meningitis which were due to an idiopathic cerebrospinal fluid otorrhea. Even though the meningitis was labyrinthogenic in origin, the patient did not experience the associated symptoms of hearing loss and/or vertigo since the affected inner ear was clinically unreactive. By surgically exploring the middle ear, the presence of a cerebrospinal fluid otorrhea was confirmed. The leak was observed to be coming from a defect in the stapes footplate, and it was controlled by firmly packing the inner ear vestibule with muscle.

A remarkable similarity exists between the patient described above and the 15 previously reported cases of meningitis due to a spontaneous cerebrospinal fluid otorrhea. Generally, the problem occurred in young children, the average age being 6.4 years; male and female were equally afflicted. All 15 previously reported cases had a severe neurosensory hearing loss which was unilateral in 10 individuals and bilateral in the other five. In 11 of the case reports, the vestibular function was evaluated, and the labyrinth was noted to be unreactive in the affected ear. An associated congenital abnormality of the inner ear was described in 11 of the patients reviewed.

Anatomically, in 13 cases, the leak was observed to be coming from the oval window area. Other affected sites included one report of a fissure of the promontory and one report of a defect in the roof of the eustachian tube.

Multiple surgical procedures were required in 11 of the 15 patients in order to identify the exact source of the otorrhea and to seal it permanently. In three cases, the successful procedure was a middle ear exploration with stapedectomy and packing of the inner ear vestibule. Overall, a total of 36 operations was performed in the 15 patients reviewed.

In conclusion, when the physician is confronted by a case of meningitis in a patient with a unilateral or bilateral total loss of hearing and vestibu-

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lar function, the possible presence of an idiopathic cerebrospinal fluid leak should be considered, especially if radiographic studies demonstrate a temporal bone dysplasia. In these selected cases, if the etiology of the meningitis is obscure, a middle ear exploration should be performed both for diagnostic purposes as a means to ascertain definitely the presence of a leak and for therapeutic purposes to seal it effectively.

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INTRODUCTION.

Bacterial meningitis remains a life-threatening infection even in the present antibiotic era; thus, any abnormality which predisposes a patient to a recurrence of this serious disease, must be identified and corrected. The presence of an occult cerebrospinal fluid otorrhea is an unusual cause of bacterial meningitis. It is all the more dangerous since it may occur without producing any significant symptoms or obvious diagnostic findings.

This phenomenon has been observed in individuals having a Mondini-type of temporal bone dysplasia.[1,2,3,4] In such patients, the congenitally abnormal ear appears to be predisposed to developing a cerebrospinal fluid leak; additionally, the affected ear may be clinically unreactive, lacking both cochlear and vestibular function. If a person with this type of abnormality develops an otitis media in the abnormal ear, and it spreads retrograde to involve the labyrinth, the expected symptoms of hearing loss and vertigo will not occur; consequently, when these patients develop labyrinthitis, the infection will frequently remain undetected until meningitis supervenes. Additionally, because significant ear symptoms are absent, the underlying otitic cause of the meningitis may remain obscure and may be overlooked.

This report describes the case of a child with a unilateral congenital hearing loss associated with a Mondini-type of temporal bone dysplasia, who developed recurrent episodes of meningitis due to an idiopathic, spontaneous, cerebrospinal otorrhea. In recent years, reports of several other strikingly similar cases have been published.[1,2,3,4,5,6,7,8,9,10,11,12] The purpose of this presentation is to familiarize the clinician with this characteristic, though obscure, otologic cause of recurrent meningitis, to review the literature, and to discuss the management of these patients.

CASE REPORT.

The patient, a white male who was born in Colombia, South America, had been aware of a left-sided hearing loss since eight years of age. The impairment was called to his attention when he noticed that he could not hear friends whispering into his left ear. At about the same time, a school screening hearing test demonstrated a left-sided hearing abnormality. His family sought medical advice, and an audiometric evaluation demonstrated a profound left neurosensory hearing loss. It is important to note that at no time was the patient aware of any tinnitus or vertigo.

In January, 1973, the patient, now 12 years old, was treated for a left middle ear infection. Two weeks later, he developed headaches, fever, and a stiff neck. He was diagnosed as having meningitis and was hospitalized. On admission, the left tympanic membrane was observed to be erythematous. A lumbar puncture was performed, and the cerebrospinal fluid was found to contain many polymorphonuclear cells. Further microscopic evaluation of this fluid showed gram positive diplococci, and cultures grew out pneumococcus. The patient was treated with intravenous penicillin and sulfisoxazole* and showed immediate improvement.

*Sulfisoxazole — Gantrisin.

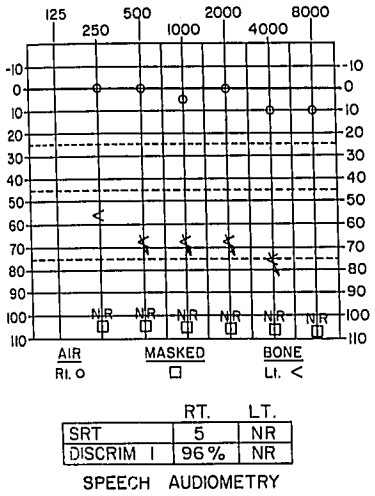

Fig. 1. Audiogram: A profound left neurosensory hearing loss. Tympanogram: No stapedial reflex is elicited when the left ear is stimulated.

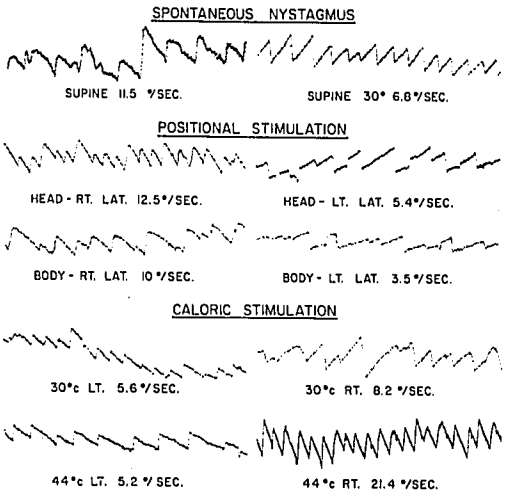

Fig. 2. Electronystagmograph: Non-reactive left labyrinth.

Following a two-week course of antibiotics, the patient became asymptomatic and was discharged.

Six weeks later, the symptoms and signs of meningitis recurred, and the patient was readmitted to the hospital. In addition to the physical findings consistent with meningeal irritation, the left tympanic membrane was dull and had normal landmarks. The remainder of the physical examination was normal. Once again, a lumbar puncture was performed and the spinal fluid contained many polymorphonuclear white cells; however, gram stains and cultures did not demonstrate any organisms. Treatment with intravenous ampicillin resulted in a dramatic improvement in the child's condition.

Considering the patient's history, it was decided to investigate all possible causes for the recurrent meningitis. The probability that the initial meningitis has been inadequately treated was evaluated, but was felt to be unlikely. The possibility that an intracranial abscess had developed as a sequel to the meningitis was also considered, but an electroencephalogram, brain scan and carotid angiogram were found to be normal.

A causal relationship between the deaf left ear and the meningeal infection was then examined. An audiometric evaluation demonstrated normal acuity in the right ear, and a profound hearing loss in the left ear. A tympanogram showed normal middle ear pressures and normal compliance. A stapedial reflex could be elicited at normal thresholds when the right ear was stimulated with the probe inserted into the left ear; however, no stapedial reflexes could be obtained when the left ear was stimulated, and the probe was inserted into the right ear (Fig. 1). Vestibular testing with electronystagmography was performed. Caloric stimulation of the left ear with 30°, 44° and ice water irrigation failed to produce nystagmus. The right ear responded normally to bi-thermal caloric stimulation (Fig. 2). Polytomographic views of the left temporal bone demonstrated a Mondini-type of dysplasia with dilatation of the vestibule and the semicircular canals. The cochlea consisted of a rudimentary basal turn (Fig. 3).

On the basis of these observations, an idiopathic leak of perilymphatic-cerebro-spinal fluid was suspected, and the left middle ear was explored.

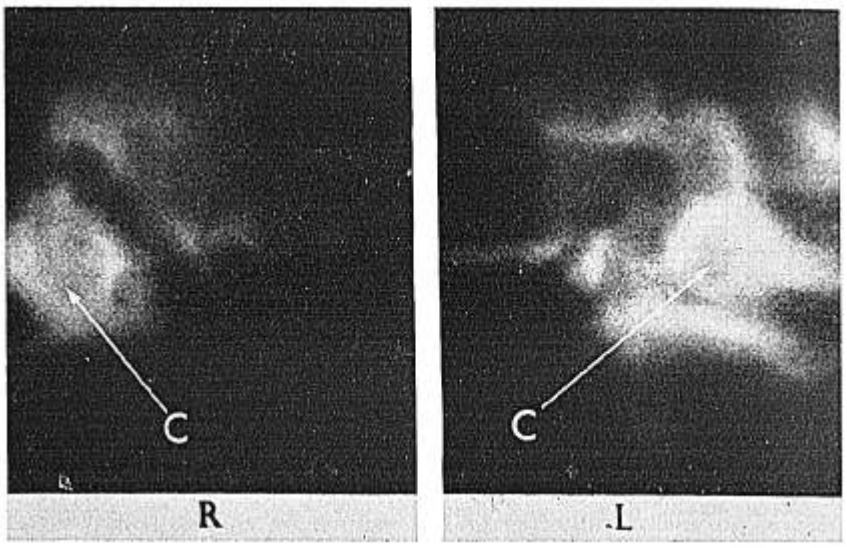

Fig. 3. Polytomographic views of temporal bone: Normal right vestibule, semicircular canals and cochlea. Left dilated vestibule and semicircular canals with a rudimentary cochlear coil consistent with a Mondini-type of temporal bone dysplasia. IAC — internal auditory canal. V — vestibule. C — cochlea. R — right temporal bone. L — left temporal bone.

After elevating a tympanomeatal flap, a normal middle ear space was entered. The malleus, incus, and stapes crura showed the usual configuration. A pulsatile flow of clear fluid was noted to be coming from a circular defect in the center of the stapes footplate. Removal of this defective stapes was accompanied by a "gush" of perilymph which welled up to fill the speculum. The cerebrospinal fluid was aspirated away until the flow slowed to a trickle. A large fat plug obtained from the lobule was used to occlude the oval window thereby sealing the leak. The tympanomeatal flap was replaced and the external canal was packed.

On the fifth postoperative day, the cerebrospinal fluid otorrhea recurred. A medical attempt to control the leak relying on acetozolamide,* bed rest with head elevation, and repeated lumbar punctures, was ineffective. Two weeks after the initial surgical procedure, the middle ear was re-explored. A defect was observed in the healing membrane through which clear fluid flowed from the anterior margin of the oval window. In order to gain better exposure of the dilated inner ear vestibule, the oval window was enlarged by drilling down the adjacent margin of the promontory. The inner ear vestibule was then tightly sealed off, using a large segment of sterno-cleido-mastoid muscle; additionally, the posterior half of the middle ear was filled with pieces of muscle, and the drum was replaced. Postoperatively, the patient did well and was discharged after 10 days.

For the next 30 months, the patient was healthy. Unknown to his doctors, he lifted weights, competed on his high school swimming team, and suffered a concussion when involved in a minibike accident.

In November, 1975, he developed an upper respiratory infection and complained of a left earache, frontal headache, and an occasional clear, watery, nasal discharge. The patient was immediately admitted for work-

*Acetozolamide — Diamox.

up to rule out a recurrent cerebrospinal fluid leak. During the hospitalization, several lumbar punctures were performed. Each time the flow of CSF fluid was observed to be very slow, and the manometric measurements revealed low CSF pressures. These findings were consistent with the presence of a CSF leak. On examination of the cerebrospinal fluid, the protein and sugar levels were normal. With appropriate stains, neither bacteria nor leukocytes were observed. Cultures of the fluid failed to grow out any organisms.

In order to detect the presence of a cerebrospinal fluid leak and ^{111} indium-DTPA* radioactive scan was performed. The material was injected through a lumbar puncture into the cerebrospinal fluid spaces. A preliminary left myringotomy was performed, but no fluid was found. A cotton ball was placed adjacent to the myringotomy opening to absorb any fluid leaking from the middle ear. Additional cotton balls were inserted intranasally to absorb any cerebrospinal fluid leaking down the eustachian tube. Serially performed scans demonstrated normal filling of the cerebral ventricles with no extravasation; moreover, the radioactivity of the individual cotton pledglets was insignificant.

While hospitalized, the patient was requested to collect any clear watery rhinorrhea for chemical analysis. Accordingly, on one occasion a small quantity of watery, nasal discharge was obtained. It was examined for sugar which was noted to be 25 mgm percent — a value suggestive of cerebrospinal fluid. At the conclusion of this work-up, the possibility still existed that this patient who had survived two episodes of meningitis might still have a cerebrospinal fluid otorrhea. It was felt that re-exploration of the left middle ear was warranted to eliminate definitely this liability.

At surgery, the previously plugged defect was seen to be covered by a smooth, durable appearing membrane. No fluid was observed to be extravasating from the area of the previous leak even when the patient was placed in the Trendelenburg position and when both internal jugular veins were compressed. Postoperatively, the patient did well and was discharged on the fourth postoperative day.

COMMENTARY.

This patient required two operations to seal his otic cerebrospinal fluid leak. In the first procedure, after the cerebrospinal fluid leak was controlled, it was relatively simple to seal the oval window with a large piece of fat. Postoperatively, however, the leak recurred, because the fat plug was not sturdy enough to withstand the fluid pressure. During the second operation, the inner ear vestibule was tightly packed with muscle to control the leak effectively; however, when the patient returned two and one half years later with a history consistent with a recurrent leak, there was great concern that the bulk of the muscle could have diminished in size as it fibrosed, and that the fluid pressure could have again eroded through the seal.

Because of the inconclusive results of the various tests to detect the presence of a cerebrospinal fluid leak, a third operative procedure was performed to examine the middle ear directly. Upon re-exploration, it was gratifying to find a strong fibrous membrane sealing the enlarged oval window area. Had a leak been present, permission had been obtained pre-

*^{111} Indium Diethylene Triamine Pentacetic Acid.

TABLE I.

Review of 15 Reported Cases of Recurrent Meningitis Due to Spontaneous CSF Otorrhea.

No. of Patients	Author	Ref.	Sex	Age at Onset (Years)	Hearing Loss		Vestibular Function	Congenital Ear Abnormality	Location of Leak
					Unilat.	Bilat.			
1	Nenzelius	1	M	1.25	X		Absent	Vestibule enlarged cochlea malformed	Fistula of promontory
2	Farrior, *et al.*	2	—	1.25		X	—	Dilated inner ear	Oval window at stapes annulus
3	Biggers, *et al.*	3	M	0.25	X		—	Dilated semicircular canals and vestibule rudimentary basal cochlear turn	Stapes footplate
4	Bottema	4	F	7	X		Absent	Dilated cochlea widened vestibule	Posterior portion of oval window niche
5	Skolnik, *et al.*	5	F	1.25	X		Absent	Possible congenital defect	Oval window at annulus
6	Barr, *et al.*	6	M	3.5		X	Absent	Promontory, stapes malformed	Stapes footplate
7	Bennett	7	F	Child		X	Absent	Klippel-Feil	Stapes footplate
8	Bennett	7	F	6.5	X		—	—	Stapes footplate
9	Stool, *et al.*	8	—	1.5	X		Absent	Widened vestibule	Stapes footplate
10	Stool, *et al.*	8	M	10	X		Absent	Widened vestibule	Mesial wall of eustachian tube
11	Crook	9	M	3	X		—	Widened vestibule	Stapes footplate
12	Rice, *et al.*	10	M	2.5		X	Absent	Possible inner ear abnormality	Stapes footplate
13	Gundersen, *et al.*	11	M	31	X		Absent	—	Oval window at stapes annulus
14	Gundersen, *et al.*	11	F	13	X		Absent	—	Stapes footplate
15	Schultz, *et al.*	12	F	2.25		X	Absent	Sagittal meningocele stapes abnormal	Stapes footplate

M — male.
F — female.
— Not reported.

TABLE II.

Meningitis in 15 Patients with CSF Otorrhea.

Patient	Author	Ref.	No. of Episodes of Meningitis	Organism
1	Nenzelius	1	Multiple	Pneumococcus
2	Farrior, *et al.*	2	4	—
3	Biggers, *et al.*	3	3	Pneumococcus
4	Bottema	4	Multiple	Pneumococcus
5	Skolnik	5	One possible episode	
6	Barr, *et al.*	6	2	Meningococcus
7	Bennett	7	2	Pneumococcus
8	Bennett	7	2	
9	Stool, *et al.*	8	3	Pneumococcus
10	Stool, *et al.*	8	20	
11	Crook	9	4	
12	Rice, *et al.*	10	3	Pneumococcus
13	Gundersen, *et al.*	11	25	
14	Gundersen, *et al.*	11	3	
15	Schultz, *et al.*	12	2	Pneumococcus

operatively, to perform a post-auricular translabyrinthine approach to the internal auditory canal and cochlear aqueduct. It was expected that the leak could have been corrected by packing the presumably widened, abnormally patent cochlear aqueduct and, if necessary, by also packing the internal auditory meatus.[2,13]

REVIEW OF THE LITERATURE.

A remarkable similarity exists between the patient described above and the 15 previously reported cases[1,2,3,4,5,6,7,8,9,10,11,12] of meningitis due to a spontaneous cerebrospinal fluid otorrhea (Table I). Generally, the problem occurred in young children, the average age being 6.4 years, and male and female were equally afflicted. All 15 previously reported cases had a severe neurosensory hearing loss which was unilateral in 10 individuals[1,3,4,5,7,8,9,11] and bilateral in the other five.[2,6,7,10,12] In 11 of the case reports, the vestibular function was evaluated, and the labyrinth was noted to be unreactive in the affected ear.[1,4,5,6,7,8,10,11,12] An associated congenital abnormality of the inner ear was described in 11 of the patients reviewed.[1,2,3,4,5,6,8,9,10,12]

Meningitis was the presenting problem that led the clinician to discover the presence of an underlying CSF leak in all but one of the case descriptions.[5] In eight cases, the bacteria responsible for the meningitis was identified.[1,3,4,6,7,8,10,12] In seven instances, the infecting organism was found to be pneumococcus. In the remaining case, meningococcus was isolated[6] (Table II).

Anatomically, in 13 cases, the leak was observed to be coming from the oval window area.[2,3,4,5,6,7,8,9,10,11,12] Other affected sites included one report[1] of a fissure of the promontory and one report[8] of a defect in the roof of the eustachian tube.

TABLE III.

Operative Procedures Performed to Seal CSF Otorrhea.

No. of Patients	Author	Ref.	Location of Leak	No. of Operations	Neurosurgical Procedures		Mastoidectomy			Middle Ear Exploration
					Post Fossa	Middle Fossa	Simple Type	Radical Type	Revision	
1	Nenzelius	1	Fistula of promontory	3				U	U	S
2	Farrior, *et al.*	2	Oval window	2			U	S*		
3	Biggers, *et al.*	3	Stapes footplate	1				S		
4	Bottema	4	Oval window	1						S
5	Skolnik, *et al.*	5	Oval window	4	a. U** b. S			U		U
6	Barr, *et al.*	6	Stapes footplate	2			U	S		
7	Bennett	7	Stapes footplate	3		U		U		S
8	Bennett	7	Stapes footplate	2			U			S
9	Stool, *et al.*	8	Stapes footplate	2		U		S		
10	Stool, *et al.*	8	Eustachian tube	3	U			U	S	
11	Crook	9	Stapes footplate	5		U	U			a. U** b. U** c. S
12	Rice, *et al.*	10	Stapes footplate	3		U	U	S		
13	Gundersen, *et al.*	11	Oval window	3	U		U			S
14	Gundersen, *et al.*	11	Stapes footplate	1						S
15	Schultz, *et al.*	12	Stapes footplate	1						S

U — Unsuccessful — leak recurred.
S — Successful.
*Vestibule packed through fenestra in lateral semicircular canal to control leak.
**Initial procedure(s) unsuccessful; repeat procedure successful.

Multiple surgical procedures were required in 11 of the 15 patients in order to identify the exact source of the otorrhea and to seal it permanently.[1,2,5,6,7,8,9,10,11] An intracranial procedure was performed in seven of the reported cases,[5,7,8,9,10,11] but it was successful in only one — a child, in whom the internal auditory meatus was packed to control the leak.[5]

Four patients required only a single operation to seal the leak.[3,4,11,12] In three cases, the successful procedure was a middle ear exploration with stapedectomy and packing of the inner ear vestibule.[4,11,12] In the fourth case, a radical mastoidectomy was performed during which the stapes was removed and the vestibule packed.[3] Eleven of the patients reveiwed underwent 15 separate mastoid procedures.[1,2,3,5,6,7,8,9,10,11] Overall, a total of 36 operations was performed in the 15 patients reviewed[1,2,3,4,5,6,7,8,9,10,11,12] (Table III).

DISCUSSION.

In treating a patient with a unilateral, or bilateral, severe hearing loss who develops meningitis, a causal relationship between these two abnormalities must be thoroughly investigated. Hearing and vestibular tests should be performed to determine whether the ear in question is reactive and capable of producing symptoms. In the individual described above, the route of infection involving the meninges was through a congenitally abnormal labyrinth. Because the affected ear was clinically unreactive, the undetected otologic complication rapidly progressed to cause meningitis, a life-threatening condition.

In reviewing the cases of recurrent meningitis due to a spontaneous oval window CSF leak, it was noted that most of these affected individuals had an underlying anatomical inner ear abnormality.[1,2,3,4,5,6,8,9,10,12] In several patients, radiographic examinations of the temporal bones demonstrated a shortened cochlear coil, a dilated semicircular canal system, and a widened inner ear vestibule, which, in extreme cases, had a cystic-like appearance.[1,3,4,8] These abnormalities are recognized as a Mondini-type of temporal bone dysplasia; therefore, in evaluating patients with both meningitis and an unreactive ear, polytomographic radiographs of the mastoids should be performed to detect the presence of a predisposing structural abnormality.

The clinical experience gained from studying these unusual cases leads one to conjecture that patients with temporal bone dysplasias are predisposed to having abnormal communications between the cerebrospinal and perilymphatic fluid spaces. The exact location of the anatomic communication is controversial;[14,15,16,17] however, there are two commonly acknowledged connections noted to exist. The first is the presence of a widened, patent cochlear aqueduct which allows the cerebrospinal fluid to pass directly from the subarachnoid space into the scala tympani of the cochlea.[18] A factor that may explain why most of the reported patients were young children is that the cochlear aqueduct is relatively wider in newborn infants than in mature adults;[19] consequently, adults may be less likely to develop a leak through this existing normal communication since the cochlear aqueduct becomes longer and relatively narrower[17] as the individual matures.

A second possible route by which cerebrospinal fluid may enter the inner ear is an abnormal communication between the fundus of the internal auditory canal and the inner ear vestibule.[5,8,9,12,13] In this area, the existing bony plate contains multiple perforations through which the nerve

fibers pass, innervating the utricular and saccular macula. In individuals having an abnormal enlargement of the inner ear vestibule, defects could exist and/or develop in this cribose plate of bone through which a leak of cerebrospinal fluid could occur.

The transmission of the pulsatile CSF pressure to the inner ear surface of the stapes footplate is probably at least partially responsible for the development of the CSF otorrhea. During embryonic development, the malleus and incus attain relative solidity, whereas the stapes sacrifices a large fraction of the mass that it attains during early growth. In the six-month fetus, resorption of the primordial stapes occurs rapidly. The progressive diminution in its bulk eventually results in the fragile mature ossicle; moreover, normal variations occur in the dimensions of the adult stapes,[20,21] and consequently, the development of the footplate fistulas in individuals with abnormal CSF-perilymphatics communication may be related to the existing thickness of their footplate.

If the footplate is thin, the pulsatile flow pressure to which it is exposed might gradually erode or displace it, resulting in a fistula with a perilymphatic leak. If the footplate is thick or even otosclerotic, it should be able to withstand the pulsatile CSF pressure. Clinically, in these later cases, the abnormal CSF communication would become manifest only during surgical manipulation of the stapes when a "gusher" would result.

Successful surgical management of these patients requires that the anatomic source of the CSF leak be definitively identified and effectively sealed. In most of the described cases, this could have been accomplished only by means of an exploratory tympanotomy. In reviewing the published reports,[1,2,3,4,5,6,7,8,9,10,11,12] it was evident that many of the neurosurgical approaches and mastoidectomies that were carried out were exploratory in nature and were performed in an attempt to locate the site of CSF fluid loss; therefore, since the leak was localized to oval window area in this select group of patients, a middle ear exploration should have been the initial operative approach. In these non-functioning ears, surgical exposure of the dilated inner ear vestibule will be enhanced by drilling away a rim of bone and enlarging the oval window. By leaving a ledge of promontory, the muscle plug, which is packed snugly into the vestibule to seal the leak, will be held in place more securely and will withstand the tendency of the CSF fluid pressure to dislodge it. If this simple procedure fails, then the surgeon can perform a trans-mastoid approach to the cochlear aqueduct and interal auditory meatus.[2,13]

In conclusion, when the physician is confronted by a case of meningitis in a patient with a unilateral or bilateral hearing loss, and the ear is nonreactive to both auditory and vestibular stimulation, the possibility of an idiopathic cerebrospinal fluid leak coming from the oval window should be strongly considered.

In cases of meningitis the observation of temporal bone dysplasia on the appropriate X-ray studies[22,23] should heighten the clinician's suspicion of an otitic etiology. The diagnosis should be substantiated by performing a myringotomy. During this procedure, the presence of a CSF leak would be confirmed by finding clear, watery fluid which, when appropriately tested, contained a significant sugar concentration.

Radiographically, the cerebrospinal fluid leak may also be demonstrated by performing a post-fossa contrast study and looking for a drop of radiopaque material in the inner ear vestibule;[8,12,24] however, if the possibility

of an undiagnosed cerebrospinal fluid leak still exists in these selected patients who have had meningitis, a middle ear exploration should be performed as a diagnostic means of directly examining the middle ear. This relatively simple procedure allows the surgeon to confirm the presence of a cerebrospinal fluid leak and, if coming from the middle ear, to seal it effectively.

SYNOPSIS.

Bacterial meningitis remains a life-threatening infection. Any abnormality which predisposes a person to this disease must be identified and corrected. This report describes the history of a 12-year-old boy with unilateral profound neurosensory hearing loss, absence of vestibular function and a Mondini-type of temporal bone dysplasia, who developed recurrent episodes of meningitis which were due to an idiopathic cerebrospinal fluid otorrhea. Even though the meningitis was labyrinthogenic in origin, the patient did not experience the expected symptoms of hearing loss and vertigo, since the affected ear was clinically unreactive. By surgically exploring the middle ear, a cerebrospinal fluid leak was observed to be coming from a defect in the stapes footplate. The leak was controlled by firmly packing the inner ear vestibule with muscle. Fifteen similar reported cases are reviewed. The possible anatomic pathways responsible for the CSF otorrhea were considered. The surgical management of these patients is discussed. This case report seeks to familiarize the clinician with this characteristic though obscure cause of recurrent meningitis.

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DISCUSSION.

G. ALEXANDER FEE, M.D., (Toronto, Ontario, Canada): It is significant that in seven of the eight cases where the organism was identified, pneumococcus was found. This bacteria has always been very insidious in the middle ear, causing a lot of damage with minimum warning symptoms. It has a tendency to develop resistance to antibiotics, and this may be one factor in the recurrence so common in these cases. The problems with recurrence may also be aggravated because the infection is in a closed space. It would be less likely to recur if drainage were used more often in conjunction with antibiotics. An early myringotomy would also make a fistula more apparent.

If a slow leak is present with an intact drum, the first evidence will be recurring meningitis; on the other hand, if a fast leak is present, this rhinorrhea might mislead the investigator to the nose as a site. A myringotomy on the first visit might obviate these occurrences.

Nearly all of the reports show that this is a life threatening condition. Except for the case of Klein in pre-antibiotic 1933, all of the cases recovered. Most of these had at least one recurrence of meningitis. One patient reported by Dr. Stool in 1967 had 20 episodes of otitic meningitis over a nine-year period before the diagnosis was established, and lived to tell the tale. Perhaps if it is a life-threatening condition, the threat is not carried out very often nowadays.

Dr. Parisier found that fat by itself will not seal a gusher. I think that if it is used, it must be held in place with a wire. I had two cases of gushers in stapedectomies, and both had a successful outcome using this procedure. This was also used in one of the Mondini cases reported in the literature, packing the vestibule, as Dr. Parisier described, was the commonest procedure used. I think that when you are looking at the windows, it is important to recognize that all that fills is not a fistula. Sometimes the local anesthetic seeps into the middle ear and fills some of the valleys, and you see fluid in the window. You aspirate it, and it refills from the neighboring crevices. This may occur three or four times and be taken for a fistula. It is important to aspirate seven or eight times to rule out this false fistula before you shove in a piece of fascia and take credit for the cure that really was due to Mother Nature.

In my opinion, sudden deafness is never due to a fistula unless there is a definite history of head trauma. Mumps is usually considered to be the commonest cause of severe monaural neuro-sensory deafness in children. As Dr. Parisier pointed out, before accepting this diagnosis, it is important to test the vestibular function, and if this is absent, do a polytome to rule out this inner ear dysplasia.

The question of whether the gusher might be a factor in a fistula in interesting. Dr. Paul Biggars, in 1973, reported a radiographically documented Mondini aplasia with no gusher and

felt that this indicated that dehiscence in the footplate was congenital and not related to perilymph pressure.

SYLVAN STOOL, M.D.: I think this is an excellent review. I feel sufficiently confident about these cases to suggest that probably one doesn't need a polytome, that one good Stenvers view that shows some abnormality is an indication that you have an abnormality of the labyrinth. Any patient who has especially a unilateral hearing loss of undetermined etiology with an acute otitis media with recurrent meningitis, then the rest of the workup, such as dye studies, is probably not going to be of value, and exploration of the middle ear with visualization of the foot-plate of the stapes is really going where the money is. The syndrome is identified well enough. There are enough cases that have been reported and not been reported, to make this an identifiable syndrome that warrants middle ear exploration after perhaps one episode of meningitis and certainly after two episodes of meningitis.

H. H. WANNAMAKER, M.D., (Syracuse, N. Y.): I saw a case referred to our office, a patient who had a myringotomy with a gusher. Apparently, testing had been done. Fortunately it healed over, When we saw it, he did have a dead ear. We worked it up and found it to be a Mondini. This child had never had meningitis and was 11 years old. As far as we could tell from a review of the literature at that time, this was about the only one that had not been diagnosed without meningitis previously. We went on to repair it prophylactically with a muscle and fat plug with a wire — and similar type findings of the enlarged vestibule also. It is 18 months now post-op.

DR. PARISIER, (closing): I thank Dr. Fee for having laboriously gone through the manuscript. In regard to meningitis, my infectious disease colleagues tell me that there is a 15 percent mortality with meningitis from pneumonic and other processes; however, Dr. Fee's observation that patients don't seem to die with meningitis from CSF otorrhea, seems to be a consistent one. It is an interesting observation. I have no good explanation, other than perhaps the flow of the fluid washes off the organism.

I agree that a wire fat prosthesis should be an ideal solution if there is a conductive type of hearing loss, and apparently, you can have a Mondini type of dysplasia with a conductive hearing loss. I think that in those cases packing the vestibule, which would necessitate a neurosensory hearing loss, would not be the proper solution.

Dr. Stool wrote two papers reviewing three patients with similar problems. In this time of medical economics being what they are, I would probably agree with him that the less expensive Stenver's view, if it is satisfactory, would supply us with the same information as the more expensive and elaborate polytomes.

In regard to Dr. Wannamaker's remarks, I thank him for pointing out that this condition which seems to be so rare may actually be one of the causes of meningitis that may be missed; and perhaps it exists more commonly in the population in children who have unilateral or bilateral hearing loss than is generally described.

FOURTEENTH CONGRESS OF THE PAN-PACIFIC SURGICAL ASSOCIATION.

The 14th Congress of the Pan-Pacific Surgical Association will be held April 1-7, 1978. Hilton Hawaiian Village Hotel, Honolulu, Hawaii, will be headquarters for this meeting. Concurrent meetings will be held in General Surgery, Neurosurgery, Obstetrics and Gynecology, Ophthalmology, Orthopedic Surgery, Otolaryngology, Plastic Surgery, Thoracic-Cardiovascular, and Urology.

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