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Juvenile Menière's disease

by

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Summary

The authors report their experience in the diagnosis and treatment of Menière's disease in childhood.

They believe it to be essential to carry out a complete battery of audio-vestibular tests, with particular reference to dehydration procedures, which in this age group are considered to be more significant for a correct diagnosis than in adults.

After stressing the importance of eliminating other forms of disease which manifest themselves with vertigo, the authors conclude by asserting the efficacy of medical treatment with diuretics and they support the surgical procedure of endolymphatic-mastoid drainage as a valid means of contending with the hydrops.

Introduction

Traditionally, Menière's is considered to be a disorder which affects adults, and rightly so. It has been stated that 75 per cent of subjects suffering from this disease belong to the age range from 30 to 60 years (Harrison and Naftalin, 1968).

However, there are no specific data concerning the incidence of Menière's disease in childhood, execept for a recent report (Meyrhoff *et al.*, 1978) in which the authors claim that roughly 3 per cent of all Menière patients have a case history which can be traced back to childhood, and they quote a few case reports presented by other authors (Table I). Furthermore, Harrison (1962) described six cases which he called 'pseudo-Menière' in children, attributing such a definition to them because of the complete disappearance of symptoms after the acute phase.

A ten-year period of observation and treatment of subjects suffering from this disease has enabled us to study some cases of juvenile Menière's disease.

Report of 5 Cases

Case 1

A 9-year-old boy, V. Roberto, came to our observation in January 1982. He had a two-year history of typical symptoms, such as fluctuating hearing loss, paroxysmal vertigo, tinnitus and a sense of fullness in the ear. During that period, the boy had been admitted to hospital for recurrent episodes of vomiting and an audiogram carried out at the same time showed moderately severe, flat, unilateral sensorineural hearing loss with a type-A tympanogram and a positive Metz test; SRT at 60 db. with a tendency towards a 'plateau' function; ABR with normal I-V interpeak latency values. X-rays

TABLE I

CASES OF MENIÈRE'S DISEASE IN CHILDHOOD REFERRED TO IN THE LITERATURE

Crowe	1938	(6 yrs.)	Meyerhoff *et al.*	1978	(12 yrs.)
Simonton	1940	(4 yrs.)			(12 yrs.)
Ombredanne and Aubry	1941	(12 yrs.)			(13 yrs.)
Ford	1944	(4 yrs.)			(12 yrs.)
Fowler	1948	(8 yrs.)			(13 yrs.)
Sørenson	1959	(7 yrs.)			(17 yrs.)
Beddoe	1977	(3 yrs.)			(4 yrs.)
		(10 yrs.)			(12 yrs.)

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revealed a morphologically normal cochlea and internal auditory canal, and the glycerol test was positive.

The parents refused permission to operate and subsequently the boy was placed on diuretic therapy with clorthalidone. A pure-tone audiogram, carried out six months later, showed a further decrease in hearing threshold values.

Case 2

When admitted to hospital, a 17-year-old girl, C. Sandra, presented with a 4-year history, predominantly of vertiginous symptoms, which increased during her menstrual periods. A pure-tone audiogram showed a flat sensorineural hearing loss, with a positive Metz test, type-A tympanogram, and SRT with signs of distortion. X-ray of the internal auditory canal was normal. The glycerol test, which was repeated a number of times over a period of six months, was negative. The patient was put on diuretics, which almost completely eliminated the attacks of vertigo and stabilized the hearing loss.

Case 3

B. Fabio, 17 years of age, over a period of roughly twelve months, complained of symptoms of fluctuating hearing loss, a sense of fullness in the ear and sudden attacks of vertigo. Pure-tone audiometry revealed a flat, moderately severe sensorineural hearing loss with a positive Metz test and a dome-shaped SRT function; both ABR and X-ray of the internal auditory canal were negative; the glycerol test was positive.

We treated him with diuretics and six months later he had regained normal hearing and the vertigo had disappeared altogether.

Case 4

A 16-year-old boy, G. Bruno, was examined in our Department in January 1980. He claimed to have been suffering from typical symptoms of Menière's disease for the past year and a half. He had a mild sensorineural hearing loss at all frequencies, with a positive Metz test. ABR was negative and SRT showed signs of cochlear involvement; X-ray was negative. After taking diuretics for a period of six months, with only partial relief of vertigo, he underwent an endolymphatic-mastoid drainage procedure.

Case 5

D. P. Antonietta, 17 years of age, complained of symptoms of Menière's disease when she was 15-years-old. She had a severe sensorineural hearing loss, with a positive Metz test; ABR and X-ray of the internal auditory canal were both negative. She underwent an endolyphatic-mastoid drainage procedure, with a favourable outcome, as shown by the stablization of hearing and absence of further episodes of vertigo.

Discussion

The afore-mentioned cases allow us to make some statements about the problem of Menière's disease in childhood. Of all our patients who were diagnosed, by a complete battery of audio-vestibular test procedures, as suffering from Menière's disease, the percentage of young subjects amounted to 7 per cent, which is significantly greater than the percentage reported by Meyerhoff *et al.* (1978).

From thorough case histories of the two girls, the symptom-complex could be traced back to the menarche, and the attacks of vertigo tended to coincide with the few days prior to menstruation.

As far as the boys were concerned, in one case the symptoms first appeared very early (at 6 years of age), and in the other two they began in the post-puberal period. As regards the classic symptom-complex, a fluctuating hearing loss was present in all but one of the patients, but even in this subject the attacks of vertigo were sudden in onset and peripheral in origin.

In all the remaining cases the symptom-complex was complete. We believe that the clinical finding which helped us most in establishing the diagnosis was the unequivocally positive reaction to the dehydration test (Klockhoff and Lindblom, 1966; Snyder, 1971), which was evident in four of the five cases.

From a diagnostic point of view, this test is of more significance in the child than in the adult, as it is not always possible in children to obtain a clear case history, which is often fundamental for diagnosing Menière's disease.

In our younger patients we decided to use diuretics, which are now regarded as the most effective form of medical treatment for Menière's disease (Klokhoff *et al.*, 1974); nor do diuretics have any specific contraindications, even in children.

Two cases were treated surgically by means of endolymphatic-mastoid drainage, using a sheet of silastic shaped to fit into the isthmus of the endolymphatic duct.

Both in young subjects and in adults, we believe that the indications for surgery depend upon the following criteria:

(a) Attacks of vertigo which are considered to be

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disabling as regards their incidence, frequency and intensity;
(b) Rapid deterioration of hearing threshold, which nevertheless still shows signs of fluctuation;
(c) Failure of medical treatment to stabilize the hearing and/or to keep the vertigo under control.
Follow-up of these patients, carried out between one and five years after treatment, showed stabilization of the hearing in three patients, two of whom had undergone surgery.

Medical treatment with diuretics succeeded in stablizing the hearing in all but one of the cases.

As far as the vertigo is concerned, the intensity of the attacks was noticeably reduced, as also was their frequency; three of the five subjects, one of whom had undergone surgery, had no further attacks. The other patient who under-went a drainage procedure had a brief otolitic crisis four months later, but there has been no recurrence since then.

Conclusions

Even though we believe that Menière's disease is uncommon in childhood, we wish to point out that in these younger subjects the otologist is not always able to make an early diagnosis of this condition; for, despite the fact that more often than not the child is able to describe the vestibular symptoms quite accurately, both parents and pediatricians tend to attribute such symptoms to other pathological conditions which occur more commonly in this age group, such as vomiting of a dietary nature, metabolic acidosis, etc. They do not attribute them immediately to an inner ear disorder.

It is therefore of the utmost importance to make a thorough differential diagnosis, by doing a complete battery of audio-vestibular tests in all such patients. Table II lists the other causes of vertigo, with and without hearing loss, which must be excluded before reaching an ultimate diagnosis.

As far as the treatment of Menière's disease is concerned, we believe that in those cases where diuretics fail to control the symptoms adequately, one should consider an endolymphatic-mastoid drainage procedure; this recommendation is supported by the successful outcome we have witnessed in the two cases operated upon in our Department.

TABLE II
DIFFERENTIAL DIAGNOSIS

Vertigo	*Vertigo + Hearing Loss*
Benign paroxysmal vertigo	Physical and acoustic trauma
Vestibular neuronitis	Spontaneous perilymphatic fistula
Headache	Stenosis of the I.A.C.
Epilepsy	Mondini-type malformation
Functional vertigo	Hypothyroidism
Cerebello-pontine tumors	Adrenal and pituitary gland insufficiency
	Lues

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