Journal articles

Perilymphatic fistula after underwater diving: a series of 11 cases

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Abstract

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Introduction: Onset of cochleovestibular symptoms (hearing loss, dizziness or instability, tinnitus) after a dive (scuba or breath-hold diving) warrants emergency transfer to an otology department. One priority is to investigate the possibility of the development of decompression sickness with a view to hyperbaric oxygen treatment of bubble-induced inner-ear damage. If this injury is ruled out, inner-ear barotrauma should be considered together with its underlying specific injury pattern, perilymphatic fistula.

Methods: We report on a series of 11 cases of perilymphatic fistula following ear barotrauma between 2003 and 2015, eight after scuba diving and three after free diving. All patients underwent a series of laboratory investigations and first-line medical treatment.

Results: Seven patients had a perilymphatic fistula in the left ear and four in the right. Eight cases underwent endaural surgical exploration. A fistula of the cochlear fenestra was visualised in seven cases with active perilymph leakage seen in six cases. After temporal fascia grafting, prompt resolution of dizziness occurred, with early, stable, subtotal recovery of hearing in seven. Of six patients in whom tinnitus occurred, this disappeared in two and improved in a further two. Two patients were not operated on because medical treatment had been successful, and one patient refused surgery despite the failure of medical treatment. Median follow-up time was 7.4 years (range 0.3 to 12).

Conclusion: The diagnosis of perilymphatic fistula is based on clinical assessments and various laboratory findings. When there was strong evidence of this condition, surgery yielded excellent functional outcomes in all patients treated early.

Key words

Inner ear; ear barotrauma; scuba diving; breath-hold diving; ENT; surgery; outcome

Introduction

Cochleovestibular symptoms (dizziness, hearing impairment and tinnitus) after scuba or breath-hold diving should steer the diagnosis towards inner-ear barotrauma (IEBt) or inner-ear decompression sickness (DCS), requiring emergency otological assessment or transfer to a hyperbaric facility in case of inner-ear DCS.[1,2] The first priority is to detect the possibility of DCS development with a view to hyperbaric oxygen treatment (HBOT) to prevent functional sequelae from the formation of tissue and intravascular bubbles in the cochlear or vestibular organs. Treating IEBt essentially relies on conservative interventions (e.g., resting prone, vestibular suppressants) in association with other medications such as corticosteroids;[1] however, it is important to be aware that there may be an underlying perilymphatic fistula, i.e., an abnormal communication between the inner ear and the middle ear due to rupture of the cochlear or vestibular fenestra, especially if medical treatment has failed. This study reports on our experience managing post-dive perilymphatic fistulae and summarises our approach to diagnosis and treatment.

MATERIAL AND METHODS

We retrospectively reviewed all the patients treated in our department for a perilymphatic fistula as a result of pressure-induced injury following an underwater dive (scuba or breath-hold) between 2003 and 2015. The local ethics committee (Sainte Anne's military hospital; approval number 40 /HIA.S.A/SMC) approved the study design.

Each injured diver was given a clinical assessment including:

- An interview to establish details of the dive and the circumstances of onset, including scuba dive, breath-hold dive, depth, duration, compliance with decompression stops, rate of ascent, yo-yo diving, difficulties with ear-clearing manoeuvres, concomitant functional symptoms (otalgia, vomiting), time to onset of symptoms after the dive and how they manifested (with or without fluctuations);
- A full physical, neurological and ENT examination;
- Laboratory examinations:
  - air and bone pure-tone liminal audiometry with, in patients with hearing loss, Fraser positional audiometry (looking for an improvement of at least 10 dB at two frequencies comparing audiometry results with the pathological ear pointed down at an angle of 90^{o} and then in the normal position). To calculate the severity of hearing loss, the average audiometric tone threshold was taken as the mean of

the air conduction thresholds at frequencies of 500, 1,000, 2,000 and 4,000 hertz;
- Videonystagmoscopy: pressure fistula test (induction of nystagmus or vertigo when pressure is exerted in the external meatus using Siegle's speculum);
- Videonystagmography: caloric and pendular tests (ipsilateral vestibular reflex inhibited or absent);
- From 2008, Tullio's sign was added to the protocol: induction of vertigo and/or nystagmus on sonic stimulation (80 dB at 250 and 500 Hz);
- CT scanning without contrast medium to rule out fracture of the petrous bone, ossicle abnormality, dehiscence of the superior canal or congenital malformation of the inner ear, as well as to investigate pathognomonic pneumolabyrinth;
- Investigation of a right-to-left shunt by transcranial Doppler ultrasonography for patients whose initial diagnosis was consistent with inner-ear DCI.

All patients were given first-line medical treatment with intravenous methylprednisolone (1 $mg \cdot kg^{-1}$ daily) for seven days. If this failed and the clinical investigations pointed to a perilymphatic fistula, the patient was offered exploratory endaural surgery on the middle ear. The surgery aimed to confirm the diagnosis of perilymphatic fistula, targeting the fenestra responsible, if possible, and then blocking the fistula if necessary. The effectiveness of the surgery was determined by follow-up clinical and audiometric evaluation.

Some patients whose initial diagnosis was one of an inner-ear DCI were first given one or more HBOT sessions (US Navy Treatment Table 5 or 6). In parallel to the findings of laboratory investigations, failure of hyperbaric treatment or worsening of the symptoms during the sessions pointed towards a diagnosis of perilymphatic fistula.

Results

Eleven patients were admitted over the study period. All of them were male with an average age of 37 (range: 22–66) years. Seven had a perilymphatic fistula in the left and four in the right ear. In eight cases, symptoms developed after scuba diving and in three after a breath-hold dive. None of the scuba divers had omitted decompression stops. Yo-yo diving with rapid descents and ascents was reported by four of the scuba divers and all of the breath-hold divers. Difficulties in pressure equalization of the ears were reported by five of the scuba divers and two of the breath-hold divers.

On average, patients had sought medical advice three days after the dive (range: within hours to 12 days). All presented with deafness, fluctuating in nature in six cases, isolated in one. Mean audiometric loss was mild (20–40 dB) in one case, severe (70–90 dB) in five and profound (> 90 dB) in the remaining five patients. Six patients complained of dizziness or gait instability with five of these reporting severe instability after a period of mild-to-moderate rotatory vertigo. In most cases, symptoms were permanent but fluctuating. One patient was asymptomatic when seen, despite a vestibular syndrome with nystagmus at initial diagnosis. Nine patients had tinnitus and five reported a feeling of fullness in their ear. The full triad – deafness/dizziness/tinnitus – was seen in five patients: deafness/tinnitus in four, deafness/dizziness in one and dizziness/tinnitus was not seen.

Positional audiometry was informative for all patients apart from one patient with profound deafness. A fistula sign was detected in one patient. Using videonystagmography, there were three cases of inhibited vestibular reflexes on the side of the affected ear. Two cases had pronounced vestibular hyperreflectivity to hot water on the side of the affected ear. Pneumolabyrinth was not seen on petrous CT scanning (without contrast). The Tullio effect (added to the protocol in 2008 for four patients) was non-contributory in all cases. Persistent foramen ovale (PFO) was investigated in three patients with negative results.

After the failure of medical treatment and strong evidence pointing to a perilymphatic fistula, eight patients were referred for endaural surgical exploration of the middle ear. A fistula of the cochlear fenestra was observed in seven cases with active perilymph leakage in six. Exploration of the vestibular fenestra was negative in all cases. The cochlear fenestra was blocked in all cases by a temporal fascia graft kept in place with biological glue. A blood patch was also preventively placed on the vestibular fenestra in six cases because we were unsure whether a small fistula could not be seen.

The eight operated patients soon recovered their hearing, within one or two days of surgery in bone conduction, with stable improvement in the long term in seven cases. Hearing loss persisted at 8,000 Hz in three patients at -80 dB, -75 dB and -35 dB respectively. One patient improved by about 50% with a mean residual deficit of -60 dB. Dizziness and instability regressed quickly after surgery in all four patients with these symptoms. Tinnitus disappeared in two cases, improved in another two but persisted in two others.

Three patients were not operated on; one refused surgery whilst two responded well to medical treatment. The first of these patients completely recovered hearing in 11 days but with persistent tinnitus as well as instability (improved by vestibular rehabilitation). The second recovered 50% of his hearing with a mean deficit of -40 dB and persistent tinnitus and instability (partly improved by vestibular rehabilitation). The third patient recovered his hearing with a residual loss at 8,000 Hz of -40 dB, without tinnitus. Median follow-up time was seven years five months, range three months to 12 years.

Discussion

There are only a limited number of studies investigating series of perilymphatic fistulae secondary to barotrauma from diving because of the low incidence of this condition.[1]

One estimate was that an ENT department detects two perilymphatic fistulae per year but this is probably underestimated.[3] In our department, we have dealt with 19 cases of perilymphatic fistulae from all causes over the 12 years covered by this report, a rare similarity to this estimate.

In a diver with persistent cochleovestibular symptoms after surfacing, two main pathological events should be considered. The differential diagnosis of inner-ear DCI or IEBt needs to be resolved urgently on presentation because DCI requires emergency recompression using one of the standard DCI treatment tables. IEBt may present with or without an underlying perilymphatic fistula. At our clinic, of 117 divers presenting between 2010 and 2015 with cochleovestibular symptoms and signs, 95 (81%) were diagnosed with inner-ear DCI and 22 (19%) with IEBt, in seven of whom a perilymphatic fistula was documented.

In the literature, perilymphatic fistula is most commonly seen in young men with an average age of 40 years. Our patients were slightly younger (average 37 years) because many of them were military divers. In seven cases, the left ear was affected and in four it was the right ear. Our series is too small to test any hypothesis, but certain anatomical features may underlie this predominance, e.g., a wider cochlear aqueduct on the left than on the right.[4] In contrast, among the inner-ear DCI seen at our hospital, a large right-to-left shunt was detected in 77% of cases, with a significantly higher incidence on the right-hand side (80% of cases, $P < 0.001$), probably owing to a paradoxical arterial gas embolism selectively going through the right common brachiocephalic artery.[2]

In our series, no fistula was visualised in one case but, because of the clinical picture coupled with strong laboratory evidence, the cochlear fenestra was occluded leading to a good functional outcome. Taking all causes together, 90% of fistulae involve the cochlear fenestra, which is more likely to be affected because it is weaker than the vestibular fenestra, which is protected by the stapes and its annular ligament.

A perilymphatic fistula has no pathognomonic sign and the final diagnosis depends on an array of signs.[5] Analysing details of the dive can rule out a DCS after a non-provocative dive.[6] Arterial gas embolism with patent foramen ovale can be investigated by identifying a right-to-left shunt. Yo-yo diving with repetitive profiles of ascent and descent is often the cause (seven cases in our series). Difficulty in equalising middle-ear pressures (e.g., Valsalva manoeuvres) is often seen (seven cases in our series). The lack of clinical improvement or exacerbation of symptoms (vomiting, dizziness) during HBOT to manage a suspected DCI case should trigger a review of the diagnosis and the consideration of IEBt and possibly a perilymphatic fistula.[7,8] The dynamics of onset (delayed onset, progressive deterioration) and how the deafness evolves (later exacerbation, fluctuation) also constitute further evidence for this diagnosis.[9]

It is important to repeat Fraser positional audiometry because, after an initially negative result, the test can turn positive later on.[10] Videonystagmoscopy can identify a fistula sign, although we only detected this in one of our patients, a similar rate to that of another series (20%).[11] Videonystagmography can detect inhibition or abolition of the vestibular reflex.[12] CT scanning without contrast medium is necessary for the detection of pneumolabyrinth, which is highly consistent with a breach through the windows.[13] CT scanning of the middle ear can also detect ossicle damage, dehiscence of the superior semicircular canal and malformation of the inner ear (abnormalities of the cochlear and vestibular aqueducts).

When inner-ear DCS is suspected, recompression should only be considered after an ENT investigation and the possibility of a perilymphatic fistula has been ruled out in a patient with ears cleared. Although experimentation has not shown that recompression is deleterious in the presence of a fistula,[14] it is important to avoid exacerbating the pressure damage by carrying out the compression phase with caution. Any exacerbation of the symptoms during HBOT points to the presence of a fistula; the sessions should be stopped and surgical investigation should be considered.

If a perilymphatic fistula is suspected, it is important to attempt medical treatment, which can afford cure in some cases.[9] The diagnosis of perilymphatic fistula should not be overestimated under the pretext of risk-free surgery without any risk of exacerbating the symptoms. The diagnosis should be backed up by the various special investigations mentioned earlier. Portmann's diagnostic scale can also be of help.[11] When this rule was followed, surgery gave an excellent outcome in every case.

In our series, hearing was improved after surgery in all eight cases and recovered fully in seven. Four patients had residual high-frequency loss. Other series have shown variable improvement with surgery, ranging from 26% to over 90%.[5,8,11,12,15] Deguine documented improvement in 63%. Pullen[8] observed a 55% cure rate, 39% improvement and a failure rate of 6% in 31 patients; Delvaux de Feffen 33% improvement,[15] 53% stable state and 15% exacerbation; Portmann 26% improvement;[11] and Black 43% improvement.[12] Disparate results are seen in the literature because the various series are not homogeneous, bringing together fistulae from different causes and, in some cases, long delays to surgery – up to years. Among the six patients with dizziness and instability, all four operated patients had excellent outcomes and improvement was obtained in both patients who received medical treatment alone. Improvement in other series has ranged from 45% to nearly 90%.[3,5,7,11,12,15] Portmann reports 33% disappearance and 12% improvement;[11] Black 89% improvement;[12] House 49% improvement;[3] Sheridan and Deguine 84% improvement;[5,7] and Delvaux de Feffen 84% improvement.[15] For tinnitus, in our series, the improvement rate was four out of six patients,

compared to about half in other reported series: Black 54% improvement;[12] Deguine 53% improvement;[5] Delvaux de Feffen 49% improvement.[15]

In a retrospective series of 50 cases of inner-ear barotrauma after scuba diving, two-thirds of divers were treated medically with improved symptoms in two-thirds of these and a cure obtained in only five.[16] Thirteen divers were operated on with improvement in hearing loss in nine cases, full recovery in two and deterioration in one. Both the tinnitus and vestibular symptoms often improved soon after the procedure.

The middle-ear compartment was investigated via an endaural approach. Examination of the two fenestrae can be facilitated by creating a Rosen bone notch[9] or using an otoendoscope. Once the fistula has been visualised – be it active or not, i.e., whether or not there is visible perilymph leakage – this fenestra has to be occluded to stop the leakage. In our series, the cochlear fenestra was occluded with a temporal fascia graft held in place with biological glue. Most experts recommend that, if no perilymphatic fistula is seen, both fenestrae ought to be occluded.[7] However, occluding the vestibular fenestra is a more delicate procedure with a mobile stapes in place. Therefore, in cases in which no fistula is seen it seems reasonable to occlude only the cochlear fenestra and to place a blood patch on the stapes footplate.

Time to treatment is important, with less likelihood of success beyond 15 days.[7,8] Some surgeons propose surgery from the outset for a broad range of different indications.[1] In our series, the timing of surgery was not a major parameter. However, rapid, ongoing hearing loss represents a relatively urgent situation. We discourage carrying on diving after surgical repair of a perilymphatic fistula.[3,7] Military divers ought to be reassigned.

Whatever the underlying mechanism suspected, a diver with persistent cochleovestibular symptoms after exiting the water ought to be put on first-aid oxygen, rehydrated and evacuated as soon as possible to a therapeutic hyperbaric facility. Once the possibility of an inner-ear DCS has been ruled out – especially if medical treatment for inner-ear barotrauma has failed – a diagnosis of perilymphatic fistula should be considered.

Conclusions

Fluctuating symptoms and sensitivity to position (positional audiometry) or pressure (fistula sign or Tullio phenomenon) are highly suggestive of a diagnosis of perilymphatic fistula. Medical treatment is always indicated. Surgical exploration of the middle ear may confirm a perilymphatic fistula, which can then be repaired. If exploration is indicated, it should be undertaken as soon as possible to ensure a good outcome. In this series, eight out of 11 divers with perilymph fistula underwent endaural surgery and had considerable sustained improvement in their symptoms and signs.

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