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Bilateral loss of vestibular function: clinical findings in 53 patients

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Abstract The clinical presentations and aetiologies of a series of 53 cases of bilateral vestibular failure (BVF) seen by the authors over a decade were evaluated by retrospective review of the medical records. Thirty-nine per cent of patients had associated neurological disease; 13% had a progressive cerebellar syndrome with disabling gait ataxia, abnormal eye movements and cerebellar atrophy on neuro-imaging. BVF was usually unsuspected. Nine per cent had cranial or peripheral neuropathies and in this group there was no abnormality of brain stem/cerebellar oculomotor function, but hearing loss was common. Eleven per cent revealed BVF and hearing loss secondary to meningitis, and 6% had other neurological disorders. Idiopathic BVF was found in 21% of cases, characterised by paroxysmal vertigo and/or oscillopsia, but no abnormal clinical signs. Gentamicin ototoxicity accounted for a further 17%, while autoimmune disease was present in 9% of patients. Otological or neoplastic disease was diagnosed in the remaining 13% of patients. It was concluded that neurological, audiological and ocular motor assessments allow the probable cause of BVF to be defined in approximately 80% of cases. A group of BVF related to autoimmune pathologies is reported for the first time, indicating the need for immunological screening. Idiopathic BVF may present with only minor visual or vestibular symptoms, while in patients with cerebellar degeneration, BVF may be unsuspected and, thus, underdiagnosed.

Key words Vestibular failure · Neurological disease · Otological disorders · Ototoxicity · Autoimmine disorders · Oscillopsia

Introduction

Disequilibrium is a common symptom; in a typical family practice, 15/1000 subjects will present with such symptoms every year [33]. Many will have a vestibular cause for their disequilibrium, but few will have bilateral vestibular failure (BVF).

Patients with BVF report unsteadiness, particularly in the dark or on rough ground, reflecting the loss of vestibulospinal function, and oscillopsia (movement of the visual environment, or visual blurring, during active or passive head movements) as a result of loss of vestibulo-ocular reflexes (VOR). As a rule, these symptoms are severe in the acute stage of vestibular loss, but tend to improve with time, owing to processes generically termed "vestibular compensation". If BVF develops gradually, however, patients may complain of only minimal symptoms, as compensation occurs simultaneously and, thus, BVF may escape diagnosis.

BVF is uncommon [28, 36]. The aim of this study is to define and review the aetiologies causing BVF in patients

attending a neuro-otology clinic in a neurological institute.

Patients and methods

The medical records of all patients with a clinical diagnosis of BVF personally examined by the authors in a 10-year period between 1983 and 1993 were reviewed. These data were generated from patients referred to the hearing and balance clinics run by the Medical Research Council and the National Hospital for Neurology and Neurosurgery. Patients usually had a complaint of imbalance or oscillopsia. The cases seen in the unit are classified by neurological diagnosis and neuro-otological findings after testing. All cases with diminished caloric responses or VOR on impulsive rotation were retrieved.

Fifty-three cases were included in this study, and all had a full neurological examination. Neuro-otological examination included assessment of the Romberg test, gait, with eyes open and closed, and eye movement examination: convergence, smooth pursuit, saccades, doll's head manoeuvre and optokinetic nystagmus in response to a small motorised drum. Spontaneous nystagmus in primary gaze and during gaze deviations and positional nystagmus following the Nylen-Hallpike manoeuvre, were assessed. Otoscopy, pure tone audiometry, stapedial reflexes and, where appropriate, brain stem auditory evoked potentials (BAEP) were obtained. Normal pure tone thresholds were defined by comparison with the data from the National Survey of Hearing [25] and severity of loss determined using World Health Organisation (WHO) criteria [44]. Computed tomography (CT) and magnetic resonance imaging (MRI) and clinical neurophysiological tests were obtained as required for diagnosis.

The bithermal caloric test (30 and 44 °C) was performed according to the technique described by Fitzgerald and Hallpike [13]. The duration of the induced nystagmus, initially with and then without fixation (using an infrared viewer or Frenzel's glasses in a darkened room), was established by direct inspection of the eyes.

Horizontal eye movements were recorded with conventional direct current (DC) electro-oculography and displayed on line by an ink jet chart recorder. Bitemporal electrodes were used, unless eye movements were disconjugate or saccades appeared clinically abnormal, in which case monocular recordings were obtained. Recordings included a search for spontaneous and lateral gaze-evoked nystagmus, smooth pursuit in response to sinusoidal oscillation of a laser target at frequencies between 0.2 and 0.4 Hz (peak amplitude ± 16°) and optokinetic nystagmus, elicited by gazing passively at a full-field, encircling striped drum, rotating at ± 40°/s. Vestibulo-ocular function was investigated by whole body rotation in the dark, with velocity steps of ± 40, 60 or 80°/s.

Slow phase eye velocity was measured manually from the eye position records. The time constant of decay of the slow phase velocity of the vestibular nystagmus (defined as the time taken by the eye velocity to decay to a value of 37% of the peak velocity generated immediately after the velocity step) was determined. The two right-beating responses (one from the right start rotation and one from the left stop rotation) and the two left-beating responses were averaged.

The criteria for defining BVF were based on caloric and rotational data. Patients were considered to have bilaterally absent caloric responses, if there was total absence of a nystagmic response with and without optic fixation at both irrigation temperatures. The criterion for absence of rotational responses was based on slow phase velocity of the nystagmus during velocity steps. Mean gain (peak slow phase eye velocity/peak chair velocity; right and left combined) and time constant for normal subjects in our laboratory were 0.80 and 13 s, respectively [37]. A cut off point was established so that patients with a value of the product [gain × time constant] of less than 10% of the normal were considered to have absent rotational responses (normal product = 10.4; exclusion criterion used < 1). Patients were included in this study if they had absent caloric or rotational responses, or both, as defined above. Thirty-nine patients fulfilled both the rotational and the caloric criteria. Three patients in whom both tests were performed fulfilled only one criterion (rotational), although they showed markedly reduced caloric responses. Eleven patients underwent only one test because of a contraindication for the other (e.g. severe motor impairment, tympanic membrane perforation); 6 met the rotational and 5 the caloric criterion.

Results

The findings are summarised in the Table 1.

BVF in association with neurological diseases

Twenty-one patients (39%) with BVF had associated neurological disorders. They were referred for clarification of the mechanisms of their ataxia and for eye movement assessment.

Cerebellar degeneration

Seven patients (13%) had cerebellar degeneration, and in four there was an associated mild peripheral neuropathy (one of probable diabetic origin), while one further patient had Friedreich's ataxia. The most common symptoms were difficulty in walking, particularly in the dark, oscillopsia and vertigo. The oscillopsia was often precipitated by head movement but also occurred at rest in those with vertical nystagmus. Dysarthria and clumsiness of the hands were also reported, but only one patient reported a mild hearing loss. There was a family history of unsteadiness and oscillopsia in one case, and the parents were first cousins in another case.

Typically, patients demonstrated an ataxic gait, worse with eye closure, limb ataxia and dysarthria. Five patients had a distal sensory impairment, and four had absent ankle reflexes. Eye movement examination was abnormal in all patients, with deranged pursuit and optokinetic nystagmus, downbeat nystagmus, gaze-evoked nystagmus, rebound nystagmus and/or square waves. Two patients had the unusual combination of upbeat nystagmus on downgaze, together with downbeat nystagmus on upgaze. In five patients, the doll's head manoeuvre provoked markedly "jerky" eye movements. Saccades were dysmetric in one case.

No consistent auditory abnormality was documented. A mild sensorineural hearing loss (SNHL) on pure tone audiometry was noted in five cases, in some of which it was cochlear in origin with normal BAEP, but in others the BAEP were abnormal. Equally, normal audiometry was found in the remaining two cases, in one case with normal BAEP, but in the other case BAEP were abnormal.

Table 1 Summary of the clinical and audiological findings in 50 patients with bilateral vestibular failure (*EOM* extraocular movements, *PTA* pure tone audiogram, *BAEP* brain stem auditory evoked potentials, *GI* gastrointestinal)

Group	History					Examination				Test abnormalities		
	Unsteady	Hearing loss	Oscillopsia[c]	Other symptoms or conditions		Unsteady	Abnormal EOM	Other signs		Brain scan	PTA	BAEP
Neurological[a]												
A. Cerebellar (n = 7)	7	1[b]	4/0/0	Dysarthria Clumsiness	7 7	7	7	Dysarthria Gait ataxia Limb ataxia Peripheral neuropathy	7 7 5 5	6	5 (mild)	4
B. Neuropathy (n = 5)	5	3	0/0/0	Weakness or numbness	4	5	0	Sensory loss Reflexes	4 4	1/3	3 (severe)	3/3
C. Meningitis (n = 6)	6	6	2/1/0	Amnesia/impotence Mild dysarthria	1 1	6	3	Dysarthia Essential tremor	1 1	1/3	6 (severe)	1/2
Idiopathic (n = 11)	11	2	8/3/2	None		11	0	Essential tremor	3	0	3 (unrelated)	1/7
Ototoxic (n = 9)	9	1	7/2/0	Renal failure	5	9	1 (unrelated)	Essential tremor Old hemiplegia	2 1	None available	4 (mild)	None available
Autoimmune (n = 5)	5	5	5/0/0	Ocular Vascular GI ulceration Genital ulcers Raynaud's disease Bone marrow necrosis Bell's palsy Rheumatoid arthritis	4 3 3 2 1 1 1 1	5	0	Positive anticardiolipin antibodies	3/4	2/2	5 (severe)	1/3
Otologic (n = 4)	4	4	1/0/0	Ménière's disease Temporal bone fracture Usher's syndrome	2 1 1	4	0	Anosmia/facial palsy Retinitis pigmentosa	1 1	0/1	4 (severe)	1/2
Neoplastic (n = 3)	3	3	1/0/0	Lymphoma Leukaemia Hypernephroma	1 1 1	3	1	Ptosis + facial palsy Muscle wasting	1 1	1/2	3 (severe)	2/2

[a] Three miscellaneous neurological disorders not included

[b] Numbers represent abnormalities noted in whole group tested, except where expressed as a fraction

[c] Head movement-induced oscillopsia/oscillopsia associated with head tremor/paroxysmal oscillopsia (see text)

In six cases, CT and/or MRI findings were available and showed cerebellar atrophy.

Neuropathies

Five cases (9%), had a peripheral or cranial neuropathy, associated with vitamin B_{12} deficiency, hereditary sensory and autonomic neuropathy type IV, alcoholism and neurosarcoidosis. In a further case, a nutritional neuropathy (beriberi) was considered but not proved.

In these patients, neuro-otological examination was requested, as unsteadiness appeared to be out of proportion to the neuropathy, or there was hearing impairment, or both. In the patient with sarcoidosis, mild bilateral VI nerve palsies and benign paroxysmal positional nystagmus were observed, while progressive chronic ophthalmoplegia was found in the patient with presumed beriberi, but no patient showed evidence of a more central supranuclear oculomotor disorder.

Hearing was normal in only two of these five patients (one with an hereditary sensory and autonomic neuropathy and one with alcohol-induced neuropathy). There was an asymmetric severe neural hearing loss in the case of neurosarcoidosis and a profound bilateral SNHL, which precluded siting the lesion, in the other two cases.

Meningitis

Six patients (11%) had meningitis, although the time lapse to assessment varied from 2 weeks to 49 years. The causative agent was known in five cases: *Streptococcus suis* in three cases, *Neisseria meningitidis* and *Streptococcus pneumoniae*. During the acute phase of the illness, all patients noticed bilateral hearing loss that had persisted, and five patients felt unsteady immediately upon remobilisation.

All three patients with *Streptococcus suis* meningitis had contact with pig meat (two butchers and one farmer) and demonstrated subtle eye movement abnormalities, including slightly abnormal pursuit and optokinetic nystagmus (OKN), mildly slowed saccades, gaze-evoked and rebound nystagmus. Other neurological symptoms and signs included retrograde amnesia, seizures, impotence, dysarthria and incoordination of the limbs. One of these patients was examined initially only 2 weeks after the onset of his symptoms, and reassessment 6 months later showed substantial recovery of vestibular function [7].

Miscellaneous neurological conditions

There were three further patients with neurological disease, viz, one case each of progressive multisystem degeneration with onset in infancy, biopsy-confirmed idiopathic leptomeningitis, and arachnoiditis.

Idiopathic group

There were 11 patients (21%) in whom no associated condition or cause of BVF was found. Three different clinical presentations emerged. The most common occurred in 6 patients who developed episodes of vertigo, lasting from minutes to a few days, followed by progressive unsteadiness in the dark and oscillopsia during head movements. A second group comprised 3 patients, with sudden onset of oscillopsia during head movements, unsteadiness and brief episodes of vertigo. In both groups, the final clinical picture was similar; unsteadiness in the dark and oscillopsia with head movements. Finally, 2 patients showed a completely different picture of sudden attacks of spontaneous vertical oscillopsia lasting 5–15 s, with unsteadiness only during these attacks, while a 3rd patient with an identical presentation was seen, but not included in this series because the caloric/rotational responses were just outside the criteria set for this study.

Apart from minimal essential tremor in three patients, neurological and eye movement examinations were normal. In one case there was a symmetrical hearing loss in the low frequencies that suggested the BVF was of peripheral origin. BAEP and imaging were unhelpful in this group of patients.

Ototoxic antibiotic group

Eight of the nine cases in this group (17%) had received gentamicin. In five of the eight cases, gentamicin was combined with other potentially ototoxic antibiotics, e.g. erythromycin [1]. In one patient treated with parenteral, but unknown antibiotics, ototoxicity was presumed. Recognised risk factors [3] for ototoxicity included renal failure in five of the nine patients and old age (mean 60 years, range 33–74).

Usually, patients complained of unsteadiness, oscillopsia and vertigo upon initial mobilisation, following recovery from the acute, infectious illness, which prompted the antibiotic treatment. Only one patient noted that her hearing was impaired during the course of her illness. Patients presented from 1 month to 14 years after receiving ototoxic drugs and reported that their symptoms had gradually improved with time.

On clinical examination, all patients were unsteady, especially with eyes closed. Eye movements were normal, except those induced by the doll's head manoeuvre. Hearing was normal in five cases, but in four cases there was bilateral, usually mild, SNHL.

Autoimmune group

Five patients (9%) had autoimmune disease; in two of these the audiovestibular symptoms developed after other

symptoms of the autoimmune disease had occurred. In four cases, rotational vertigo and vomiting were accompanied by moderate to profound bilateral hearing loss. In three patients, the audiovestibular symptoms leading to BVF were of acute onset and progressive, but in two cases, the clinical presentation was that of a chronically relapsing disorder, with asymmetric audiovestibular involvement. All patients had multisystem disease, including recurrent eye symptoms (episcleritis, scleritis, uveitis, visual loss owing to retinal artery occlusion), orogenital and gastrointestinal ulceration, blistering skin lesions, synovitis and facial palsy.

Neurological and eye movement examinations were always normal. All patients had bilateral SNHL. BAEP were normal in the three cases in which they were obtained, suggesting cochlear pathology. Imaging was normal in all cases. Anticardiolipin antibodies were raised in three of the four cases tested.

The final diagnoses were Behçet's disease (three cases), vasculitis (one case) and primary antiphospolipid syndrome (one case, previously reported by Vyse et al. [43]).

Miscellaneous otological group

All of these patients (see Table 1) complained of unsteadiness and oscillopsia with head movement, and all had severe or total bilateral SNHL. General neurological examination was unremarkable.

Neoplastic group

There were three patients (6%) in whom BVF was associated with malignant disease and all reported both auditory and vestibular symptoms. In two patients (hypernephroma and chronic myeloid leukaemia), local infiltration was thought to be responsible. A third patient developed episodes of vertigo, unsteadiness, oscillopsia and hearing loss at the time when a non-Hodgkin lymphoma was diagnosed. Lymphadenopathy, BVF and a mild unilateral SNHL were the only clinical findings. It is possible that BVF was a paraneoplastic effect in this patient.

Discussion

Symptomatology and signs

Reviewing all cases, certain general observations can be made. Patients were usually referred because of a disorder of balance, but in certain instances a disorder of eye movements (cerebellar disease) or hearing loss (neuropathies, autoimmune disorders) or a combination of symptoms and/or signs (ototoxicity, meningitis, neoplasia) prompted referral. Importantly, in patients with eye movement abnormalities, BVF was often unsuspected prior to investigation. Symptoms included light-headedness, dizziness and imbalance, especially if visual input was reduced, e.g. in darkness. Rotational vertigo was common early in the course of vestibular loss, but was not a feature of the chronic state, when patients most frequently presented for investigation. Oscillopsia, induced by passive head movements, as for example travelling in a vehicle, or active movements such as walking, was common. It results from the inability of the pursuit system and cervico-ocular reflex to transduce all frequencies (>1 Hz) at an appropriate gain and phase and thereby compensate for the loss of the VOR. However, in six patients oscillopsia was reported at rest or during trivial tasks such as reading or watching television. In all six (3 previously reported [6]), "pendular pseudonystagmus" resulting from bilateral vestibular loss, with a slight essential head tremor, was demonstrated, although only two patients reported head tremor. Clinically, ophthalmoscopy revealed pendular oscillation of the fundus, which was attenuated or suppressed with rigid immobilisation of the head. There appeared no consistent aetiological relationship between the cause of the head tremor and BVF, which was due to antibiotic ototoxicity in two cases, meningitis in one case and was idiopathic in the remaining three cases.

Hearing was affected to varying degrees, but was most severely abnormal in conditions known to affect the cochlea or eighth nerve, e.g. meningitis, neuropathies and primary otological disease, whereas in the idiopathic group hearing was usually normal and in those with cerebellar disease, hearing was only mildly impaired in approximately half of the group.

Aetiology

Cases fell into seven groups, defined according to associated conditions and allowing a likely cause to be defined in approximately 80% of cases.

Neurological disease

The proportion of patients with BVF and neurological disease was, as expected, greater in our series (29%) than in previous studies [10, 28, 36, 41]. There were seven patients with cerebellar abnormalities; four were sporadic degenerations, but in two (in addition to the Friedreich's ataxia) an inherited disorder was suspected. The prominent axial ataxia and oculomotor disorder in these cases suggested that the vestibulo-cerebellum was the primary site of degeneration, although the precise site(s) of the vestibular lesion could not be determined. These patients did not complain of hearing loss, and the audiological findings were heterogeneous, indicating cochlear pathology in some cases and bilateral eighth nerve or brain stem dysfunction in others.

In this group, referral was usually for an assessment of eye movement abnormalities rather than for the investigation of auditory or vestibular symptoms. Thus, the finding of vestibular loss was unexpected and suggests that vestibular failure may be underdiagnosed in patients with cerebellar degenerations. In such patients, unsteadiness, especially in the dark, in excess of that which would be expected from cerebellar pathology, should signal the need for vestibular investigation. In addition, a clinical sign that should prompt vestibular investigation is the presence of "jerky" eye movements when the patient is asked to fixate on the examiner's nose and slow head movements (ca. 0.1 Hz) are made (doll's head manoeuvre). This phenomenon results from the combined loss of slow pursuit and vestibular eye movements and leads to extreme instability of the optic fundus during slow head movements [5].

In five further cases, the cause of the underlying cranial or peripheral neuropathy was felt to be associated with BVF, as such cases had been documented in the literature: vitamin B_{12} deficiency [30], alcohol [45], hereditary sensory and autonomic neuropathy type IV (HSAN IV) [31, 40], neurosarcoidosis [21] and beriberi [11, 16]. In two of our cases, auditory brain stem evoked responses suggested brain stem or VIII nerve involvement.

BVF and auditory loss are common sequelae of bacterial meningitis [4, 14]. *Streptococcus suis* type II causes meningitis and sepsis in pigs [24] and meningitis in humans. Three of our patients acquired this infection, and all reported contact with pigs or pork. The most common recognised complication is hearing loss [4, 12], and all three patients demonstrated a bilateral SNHL, which was severe in two. In our patients, minor eye movement abnormalities, which have not been reported previously, were present, including saccadic pursuit, gaze-evoked and rebound nystagmus and slight saccadic slowing, suggestive of subtle central nervous system (CNS) involvement.

Most interestingly, in one case there was a significant improvement in both auditory and vestibular function [7]. It has been suggested that the presence or absence of recovery of hearing function following meningitis may depend on the underlying pathological process. Cases with permanent loss may be owing to suppurative destructive labyrinthitis, whereas a serous labyrinthitis may resolve and explain the auditory improvement [14]. Presumably, a similar mechanism may operate with respect to vestibular function and explain the recovery in our patient. The fact that he was the only patient to suffer a moderate SNHL during the infection – which subsequently improved – would support this view.

Idiopathic BVF

As in previous studies, the proportion of patients with idiopathic BVF is significant (21%) [2, 10, 17, 29, 36, 41]. Three different clinical presentations emerged in this study: (a) episodes of vertigo followed by progressive unsteadiness, (b) episodes of head movement-induced oscillopsia with unsteadiness and vertigo and (c) brief attacks of spontaneous vertical oscillopsia and unsteadiness lasting 5–15 s. Patients with this latter presentation did not deteriorate over the 1–5 years of follow-up. There was evidence of some overlap in presentation between these groups, and this may explain the different clustering of presentations noted in earlier reports [2, 35], although brief paroxysms of oscillopsia as an isolated presentation have not been reported previously.

Of interest, many patients in this group were otherwise healthy and had only minor symptoms of imbalance. Normal eye movement examination, brain stem auditory responses and brain scans excluded the presence of neurological involvement, and the lesions were therefore presumed to be of labyrinthine origin.

The lack of family history or involvement of other organs did not suggest a familial aetiology, although a variety of pathological processes may be relevant in this group. As early as 1972, Graybiel and coworkers [17] postulated that inherited vestibular loss might occur as an isolated entity, and more recently Verhagen and coworkers [42] have reported three siblings with "familial congenital vestibular areflexia", two of whom did not volunteer symptoms. On the other hand, a predisposition to vestibular loss, secondary to an environmental trigger, may be inherited, as highlighted by the reports of familial susceptibility to aminoglycoside ototoxicity [19], and BVF can be a feature of the vestibular aqueduct syndrome [20].

Alternatively there could be a viral or vascular cause. Schuknecht and Kitamura [34], on the basis of temporal bone studies, have demonstrated that vestibular nerve and end organ damage, similar to that observed in viral disorders, is found in patients with so-called vestibular neuritis and that sequential bilateral vestibular neuritis could give rise to BVF [35]. Finally, arterial obstruction to the labyrinth has been shown to give rise to isolated vestibular changes in animals [23]; vascular factors may thus be relevant in some cases of BVF [2, 8].

Autoimmune disease

Auditory and vestibular disorders in autoimmune disease have been observed previously [15, 22, 26, 38, 39], and isolated bilateral auditory and vestibular failure has been reported as a presenting feature of Cogan's syndrome [27], polyarteritis nodosa [32] and the antiphospholid syndrome [43]. Moreover, sudden and sequential bilateral hearing loss has been reported in systemic lupus erythematosus, but vestibular function was not assessed [9]. Antiphospholipid antibodies were raised in three of four patients tested in this group and may cause recurrent arterial and venous thromboses [18]. The presence of an associated severe

SNHL, without neurological findings, would suggest ischaemia of the peripheral labyrinth or eighth nerve in our cases.

In this study, two clinical presentations of audiovestibular symptoms in autoimmune disorders emerged: (1) A rapid aggressive course of disease with acute, severe bilateral hearing loss and vestibular failure and (2) a relapsing, episodic form with progressive unsteadiness and asymmetrical hearing loss. Involvement of other organs (e.g. ocular, abdominal, orogenital ulcers, skin, joints) and positive anticardiolipin antibodies were present in both types. The audiovestibular symptoms either heralded the onset of the autoimmune disease or appeared later, once the diagnosis of autoimmune disease was established. It was of note that no patients in this group had signs of involvement of brain stem or cerebellar structures.

Other disorders

Our series included well-recognised ototoxic (aminoglycoside) and otologic (e.g. bilateral Ménière's disease, bilateral temporal bone fractures) causes of BVF. The remaining three patients had malignancies, one of which may have had a unique paraneoplastic syndrome.

Conclusion

BVF should be considered in patients who appear more unsteady than can be readily explained on the basis of their neurological signs and in patients with both episodic and continuous oscillopsia. Audiological evaluation is helpful in differentiating BVF associated with neuropathies, meningitis and autoimmune disorders (hearing loss common) from BVF associated with cerebellar pathology and of idiopathic type (no significant hearing loss). Eye movement abnormalities allowed differentiation of cerebellar disorders associated with BVF. Rare cases of pathology involving the labyrinth (e.g. bilateral Ménière's disease, fractures or metastases) and the CNS (e.g. infantile progressive multisystem degeneration) may be associated with BVF.

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