Vestibulogenic imbalance

JOHN FOLEY

British Medical Journal, 1978, 1, 1244-1246

Summary and conclusions

Vestibular lesions may cause imbalance that long outlasts vertigo. Photographic analysis in five cases showed defects not only of the tilting reactions, which are of labyrinthine origin, but also of certain other postural reactions, notably the counterpoising and protective stepping reactions. All of these reflexes are unconscious and consequently such patients are rendered unstable in several ways simultaneously and may have difficulty describing their symptoms.

Introduction

It is well known that an acute vestibular lesion results in falling to the affected side, deviation of the eyes to that side in the slow phase of nystagmus, and vertigo. It is less well known that postural instability may outlast vertigo and nystagmus by years and that symptoms due to the mismatching of vestibular, proprioceptive, and visual information, such as instability or unreality, may be the patient's only complaint. In a case described by Levin,¹ a patient with progressive vestibular degeneration complained only of imbalance and severe headaches after exertion. Bonnier² redescribed Romberg's test as one also disclosing labyrinthine dysfunction, and Rademaker and Garcin³⁻⁴ showed that the tilting reactions in animals and man (tested on all-fours) are absent on the side of the lesion. Martin⁵ used the sitting position and found that it is the reflexes in the trunk and neck that preserve balance, while movements of the limbs are secondary, of proprioceptive origin, and by themselves incapable of preserving equilibrium. The tilting reactions are some of the large group of postural reactions mediated by the basal ganglia and operating on the antigravity and automatic stepping mechanism and are the only ones of purely vestibular origin; the rest are proprioceptive (or visual)—an example is the counterpoising reaction of the trunk against a limb movement. The protective stepping reaction can also be disturbed by a labyrinthine lesion.

Five cases are cited below, showing that several postural reactions may be disturbed by a labyrinthine lesion even though they (with the exception of the tilting reaction) have no direct link with the vestibular system. None of the patients had any neurological signs in the accepted sense (except deafness and a facial palsy) but all were found to have postural abnormalities.

Present study

METHODS

In addition to routine neurological examination the patients were subjected to differential caloric tests; Romberg's test; examination of counterpoising of the arm when sitting, standing, or on all-fours, the patient being viewed behind a plumb-line; and examination of the tilting reaction when seated on a simple tilting platform, a fall or deviation of more than 20 degrees being counted as abnormal.

(Normal people can resist a tilt of 30 degrees imposed at any speed the examiner is capable of, though it is usual to employ slow tilts of about 20 degrees/s; on the other hand, an abnormal subject begins to fall long before his centre of gravity falls outside his base.)

CASE 1

This patient was a 76-year-old woman with right herpes oticus. Two weeks after onset there was no nystagmus or incoordination of the limbs. She fell to the right without a protective step of the right leg, which was neither paralysed nor atactic; the protective step of the left leg was preserved. Her posture was maintained mainly by vision. Six weeks after onset the tilting reaction to the right was absent. In addition, after a tilt down to the normal side she was thrust across the midline into a secondary fall.

CASE 2

A 37-year-old housewife had had right facial palsy and deafness for many years due to a cholesteatoma. After an attack of sterile meningitis (with no focal signs) the ear was exenterated. She quickly mastered her imbalance except under test conditions. When tilted down-right with her eyes open her tilting reaction was normal, which demonstrated the power of visual fixation (fig 1(a)). When blindfolded, however, she fell in the direction of the tilt (fig 1(b)). She could sit on an indefinitely prolonged down-right tilt unless vision was obscured (fig 1(c), (d)). The fall that occurred when she closed her eyes could not have been due to any alteration in stimuli to her semicircular canals as there had been no movement of the head; this phenomenon could therefore be attributed to the asymmetrical drive of the utricle of the normal side, since the otoliths are sensitive to gravity as well as to linear acceleration and centrifugal force.⁶ When a patient with a defective tilting reaction falls the head is directed downhill; when a normal person is tilted into a fall, however, the head remains vertical (fig 2).

Two years after the operation the patient was re-examined. She had no difficulty walking or swimming, but when she was a passenger

FIG 1—Case 2: right labyrinthectomy. (a) Normal tilting reaction with eyes open. (b) Defective tilting reaction when blindfolded. (c) and (d) Sitting on slope with eyes open, but falling when blindfolded.

FIG 2—Tilting reaction of normal person when blindfolded. Head remains vertical and tilting reaction in neck and trunk persists even after protective reaction of arm has begun. (Tracings from film; interval approximately 0.062 s.)

in a car she tended to slew over at corners, though when driving herself she was stable, presumably because of proprioceptive information derived from her grip on the wheel. When kneeling she fell forwards on closing her eyes (fig 3(a), (b)). Standing blindfold, she would begin to fall to the right (fig 3). Counterpoising of the left arm was normal but counterpoising of the right arm was defective, with no shift of the trunk across the midline (fig 3(d), (e)). When on all-fours she was stable even with her eyes closed, but whereas counterpoising of the left arm was normal, counterpoising of the right arm was defective, though not sufficiently so to cause a fall (fig 3(f)-(k)).

FIG 3—Case 2 two years after operation. (a), (b) Kneeling with eyes open and closed: unilateral lesion results in falling forward. (c)-(e) Standing blindfold: patient falls to right (operated side); counterpoising of right arm defective. (f)-(k) Defective counterpoising of right arm in sitting and all-fours positions.

Almost three years after the operation there was a short-lived phase in which she fell to the left when ironing; the counterpoising reaction of the left arm was found to be defective (the tilting reactions were unfortunately not tested). This phase was brief, and then her arm-counterpoising reactions were normal, while her tilting reaction down-right was as defective as before. This brief interlude, in which she fell to the "wrong" side, suggested that the vestibular tonus generated by her right vestibular nuclei had temporarily exceeded that of the left—that is, she had "over-compensated" by the postural equivalent of von Bechterew's phenomenon, which was originally described in relation to nystagmus.⁷⁻⁸

CASE 3

This patient, a woman aged 21, suffered tachycardia in crowds, progressive imbalance to the right with eventual vestibulogenic seizures,⁹ and, later, hysterical attacks. She also complained of right otalgia. Right canal paresis was noted but there were no neurological signs, and an electroencephalogram and EMI scans were normal. In this case the disorders of limb and head counterpoising caused imbalance beyond the patient's comprehension; they included even forward movements of the trunk. She was diagnosed as having right vestibular neuronitis with imbalance and vestibulogenic seizures.

CASE 4

A 59-year-old woman underwent transtemporal removal of a small (1.5 cm) right acoustic neuroma. Five months later her tilting and counterpoising reactions were similar to those in case 3, though not so pronounced.

CASE 5

This patient, a woman aged 60, had a six-year history of occipital-pressure sensations, occasional vertigo, and progressive imbalance to the left. She was diagnosed as a case of left vestibular neuronitis. There were no neurological signs. Other findings were: left canal paresis; normal brain stem-evoked responses; normal skull radiographs and computerised axial tomogram; and nystagmus to the right in darkness on electronystagmography.

She had an obvious imbalance to the left when standing and sitting with her eyes closed, and counterpoising of her left arm was defective. The down-left and forward tilting reactions were also abnormal. The disorder of stance was of greater localising value than the tilting reaction. The infrequency or absence of vertigo in this and case 3 and the prominence of disequilibrium suggest that the utricular rather than the canal mechanism was mainly affected.

Discussion

The reflexes preserving balance and promoting locomotion were beautifully summarised by Martin.⁵ They are mediated by the basal ganglia and operate on the highly co-ordinated anti-gravity and reflex stepping mechanism of the spinal cord and brain stem. That these mechanisms entail complex sequences or patterns of movement was shown by André-Thomas *et al*.¹⁰ and Lundberg and Phillips.¹¹ They may be summarised very briefly as (1) the axis-erecting mechanism; (2) the counterpoising reactions protecting balance when a limb or the head is moved; (3) the tilting reactions acting against instability of the base; (4) the protective stepping reaction occurring once balance has been lost; (5) the righting or rising reactions, proprioceptive and visual; and (6) the locomotive reactions. Only the tilting reactions are of primary labyrinthine origin.

The five cases cited here indicate that after a labyrinthine lesion there may be a disturbance of postural reactions that seemingly have no direct connection with the vestibular system, and they demonstrate the widespread effect of "vestibular-input failure" on the general postural mechanism, which is normally dependent on matched proprioceptive, vestibular, and visual afferents. The deaf mute can learn to walk without tilting reactions,⁴ but these reactions are essential for riding, walking on rough ground, and walking or sitting on a slope when vision is obscured either by darkness or, for example, when reading or carrying a tray. If a tilting reaction is absent it is, of course, necessary to determine whether the lesion is in the vestibular complex itself or in the pathway to and through the basal ganglia. One, some, or all of the tilting reactions may be affected by a vestibular lesion. In the early stages all may be absent, the patient being at the mercy of every movement, though later it is usual to find a defect of the tilting reaction down to the side of the lesion only. The reactions to quick and slow tilts may be tested separately, for the former are a function of the vertical canals, and the latter a function of the utricle.⁶ Thus, provided there is no evidence of brain-stem disease, the slow tilting reaction or the reaction to sitting on a slope blindfold may be a simple test of utricular function. The tilting or "tipping" reactions¹² have not been employed clinically because of the misconception that "if one labyrinth is functioning normally the patient will react normally,"¹³ which is obviously untrue; Rademaker and Garcin¹ were explicit on this point.

Just as each vestibular mechanism may be regarded as providing a tonic input driving the ocular axis to the opposite side (lack of such tonus causing the slow phase of nystagmus), so the intact labyrinth may be regarded as providing a postural tonus in the trunk and neck musculature, and in a sense propping up the trunk. Lack of this tonus results in the fall to the side of the

injured labyrinth; alternatively the body may be regarded as being "driven" towards the abnormal side by the tonus generated by the intact labyrinth, which is in keeping with the violence of the fall in some cases. The secondary causes of instability result from the lack of a contribution that the vestibular system presumably makes to certain other postural reflexes even though some of these appear to have no direct connection with the vestibular nuclei. These susceptible reactions are the protective stepping reaction and some of the counterpoising reactions.

In the early stages after a unilateral labyrinthine lesion vision is needed to prevent falling, just as it is needed to halt nystagmus, and many patients adapt to their disability in the course of a few weeks; later such people, and even patients with bilateral labyrinthine deficit, can maintain equilibrium with little visual guidance except in water, on rough ground, or on horseback. There is experimental evidence that after unilateral labyrinthectomy the activity of the vestibular neurones on the contralateral side is at first enhanced but later diminished by cerebellar inhibition, and it is this inhibition that is responsible for "compensation."⁸ The vestibular neurones on the affected side might even gain a temporary relative ascendancy.

Most patients seem to achieve compensation for the falling tendency quite quickly, and also for the more subtle deficiencies in their postural reactions illustrated in this paper; but some, even though formerly athletic, seem incapable of coming to terms with a state of "vestibular-input deficiency" and continue to complain of inexplicable instability even if they do not have actual vertigo. Their complaints are diverse—of feeling unsafe, of drop attacks or blackouts, of unreality, of anxiety and nausea on exertion or when in crowds, or of headaches and neck pain. Many of these symptoms may be misinterpreted as evidence of

temporal lobe epilepsy, neuroticism, or even hysteria. The present cases, however, show that a vestibular lesion may result in instability without vertigo or anything more than transient nystagmus; but the persistent defects of postural mechanisms can be demonstrated by simple clinical means, and these postural deficits may constitute the only physical signs of the disorder.

I acknowledge the helpful criticism and advice of Dr James Purdon Martin, and thank Mr T T King for permission to quote case 4.

References

- ¹ Levin, P M, *Journal of Nervous and Mental Disease*, 1939, **89**, 335.
- ² Bonnier, P, *Comptes Rendus des Séances de la Société de Biologie et de ses Filiales*, 1895, **74**, 707.
- ³ Rademaker, G G J, and Garcin, R, *Revue Neurologique*, 1932, **39**, 637.
- ⁴ Rademaker, G G J, and Garcin, R, *Revue Neurologique*, 1933, **40**, 566.
- ⁵ Martin, J Purdon, *The Basal Ganglia and Posture*. London, Pitman Medical, 1967.
- ⁶ Tait, J, and McNally, W J, *Philosophical Transactions of the Royal Society of London*, 1934, **244**, 241.
- ⁷ Roberts, T D M, *Neurophysiology of Postural Mechanisms*. London, Butterworths, 1967.
- ⁸ Precht, W, in *Handbook of Sensory Physiology*, ed H Kornhuber, vol 6, pt 2, p 451. Berlin, Springer, 1974.
- ⁹ Behrman, S, and Wyke, B D, *Brain*, 1958, **81**, 529.
- ¹⁰ André-Thomas, et al, *Clinics in Developmental Medicine*, No 24, p 90. London, Heinemann, 1966.
- ¹¹ Lundberg, A, and Phillips, G C, *Journal of Physiology*, 1973, **231**, 90.
- ¹² Rådmark, K, *Acta Oto-Laryngologica*, 1944, suppl 52, p 133.
- ¹³ Pettersen, E, in *Handbook of Sensory Physiology*, ed H Kornhuber, vol 6, pt 2, p 267. Berlin, Springer, 1974.

(Accepted 6 March 1978)

CONDENSED REPORTS

Radiotherapy in the treatment of Hodgkin's disease

A R TIMOTHY, S B J SUTCLIFFE, A G STANSFELD, P F M WRIGLEY, A E JONES

British Medical Journal, 1978, **1**, 1246-1249

Summary and conclusions

Eighty-seven untreated patients with localised Hodgkin's disease seen from 1969 to 1975 were treated by megavoltage radiotherapy. All were followed for at least 33 months. Thirty-three patients were staged clinically and 54 underwent more extensive investigation by laparotomy and splenectomy.

ICRF Department of Medical Oncology, St Bartholomew's Hospital, London EC1A 7BE

A R TIMOTHY, MRCP, FRCP, Gordon Hamilton-Fairley fellow and honorary senior registrar
S B J SUTCLIFFE, BSC, MRCP, senior registrar
P F M WRIGLEY, PHD, MRCP, consultant physician

Department of Histopathology, St Bartholomew's Hospital, London EC1A 7BE

A G STANSFELD, FRCPATH, reader

Department of Radiotherapy, St Bartholomew's Hospital, London EC1A 7BE

A E JONES, FRCP, FRCP, professor

The projected five-year disease-free survival figures for patients staged surgically were 100% for the 17 with stage IA disease, 70% for the 19 with stage IIA disease, and 73% for the 15 with stage IIIA disease. These results were consistently better than those obtained in clinically staged patients. Five patients died, one of them without evidence of Hodgkin's disease.

As irradiation seems to produce excellent disease-free survival in most patients who are staged accurately at diagnosis, caution should be exercised in the routine use of adjuvant chemotherapy until the full risks of such treatment are clear. Combined modality therapy may be appropriate for patients with unfavourable features at presentation.

Introduction

In 1913 Finzi first suggested that prophylactic extended-field irradiation might cure patients with localised Hodgkin's disease. Some 26 years later Gilbert² reported long-term survivors in a group of patients with localised disease who were treated with x-rays and again advocated that prophylactic radiation should be administered to adjacent, clinically unaffected lymph nodes. Subsequent studies by Peters³ and Kaplan⁴ confirmed that extended-field and, after the advent of the megavoltage era,