

1 What is essential tremor?

The main characteristic of essential tremor (ET) is shaking of the hands, although shaking can also affect the head, voice, face and legs. This is most easily illustrated by the difficulty affected individuals have in writing or drawing a spiral, as illustrated in Fig. 1.1.

ET is one of many different causes of tremor and is not a simple disorder. In this chapter, we will review the particular characteristics of the ET tremor, namely *postural* and *kinetic* tremor, and consider some of the other criteria used to define ET.

There is also increasing evidence that psychological stress makes ET much worse. The technical words to describe this include *sociophobia*, *harm avoidance*, and *psychosocial dysfunction*. However, these terms describe a person who avoids interacting with society, and in ET this can be reduced to self-conscious embarrassment. Throughout this book, we will argue that this anxious personality trait is part and parcel of ET, and in many cases has a much greater effect on living than the tremor itself.

The first step is to understand the terms 'postural' and 'kinetic' tremor.

- Postural tremor occurs when part of the body is held against gravity such as holding the arms in front of the body (both arms usually shake, bilateral tremor) or holding the head still.
- Kinetic tremor is also called action tremor and is observed during voluntary movement such as writing, drawing, pouring, etc.

Most importantly, ET does not occur at rest. Even in severely affected people, if the muscles are completely relaxed, no tremor will be detected, and people with ET do not shake when they are fast asleep.

Other key characteristics of ET which have been well documented include the following

- Variable tremor severity.
- A tendency for the symptoms to worsen during emotional and physical stress.

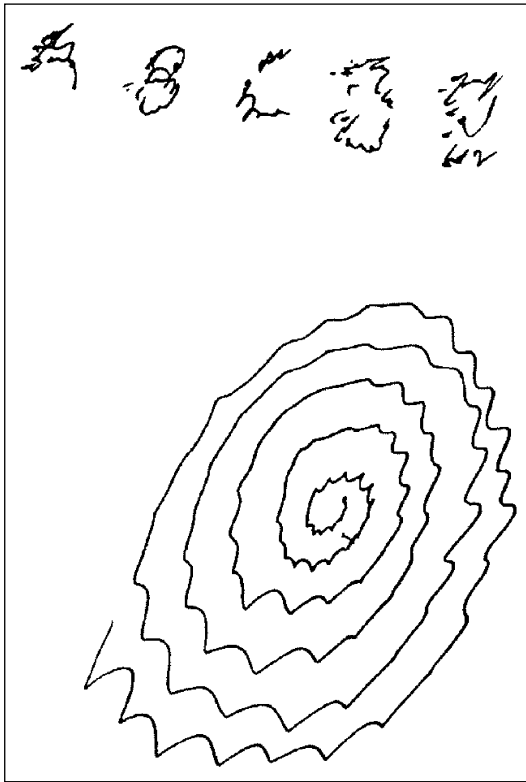


Figure 1.1 Spirograph/writing in ET. The postural and kinetic tremor in ET during voluntary movement in the arms, wrists and fingers is well illustrated in the difficulties with writing and drawing a spiral (spirography).

- A tendency for the symptoms to worsen with age.
- A variable age of onset, including childhood.
- ET often runs in families.
- ET can be *sporadic*—there are some cases of ET with no obvious family history. This suggests that environmental factors may play a causal role, although precisely which environmental factor(s) are involved is unknown.

ET is thus a very complex and *heterogenous* disorder. So that, except for the presence of the characteristic postural and kinetic tremor, everything else

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in ET is highly variable: severity; age of onset; family history (*genetics*); and other 'symptoms', all exhibit tremendous interindividual variation, both in ET populations as a whole and also within individual families. The use of the word 'complex' in this context simply means that the disorder is not fully understood.

In this and subsequent chapters, we will build up a picture of ET, and although the precise neurological defect or deficiency is not yet known, there are some tantalizing clues.

The history of essential tremor

ET was first reported in 1817 by James Parkinson, who realized it was a different condition from classical Parkinson's disease. The first detailed account of ET was published in 1887, although shaking hands as a movement disorder has probably been around since human records began. Since then, and as scientific approaches have evolved and been perfected, it has become clear that there are a number of quite different so-called 'movement disorders', each of which is manifest in slightly different ways and is caused by different genetic or environmental factor(s). The situation is further complicated because ET is not a simple disorder. The severity of the symptoms varies widely, as does the age of onset, further complicating the clinician's task of diagnosis. A correct and consistent diagnosis is crucial if optimal treatment is to be prescribed—medication that is effective in ET may not be effective for patients with Parkinson's disease, although there is some overlap in their treatments.

The name ET is also evolving. ET was once also called 'benign familial tremor', but the *benign* was dropped (in about 1991) following a study of 753 ET patients in Kansas, USA. A 'Sickness Impact Profile' (SIP) was used to measure the effects of the tremor on patients' ability to perform the functions of their day-to-day life. Using a questionnaire, patients with ET and those unaffected by ET were asked to assess the impact of tremor on their day-to-day activities including communication, work, emotional behaviour, home management, and recreation and pastimes—this gave the investigators a measure of 'sickness-related dysfunction'. A total of 145 patients with Parkinson's disease were also included in the study as a 'positive control'. As expected, Parkinson's patients had the highest dysfunction, but the study concluded that compared with the unaffected people, significant disabilities within all the categories of the SIP could occur in ET patients—ET is not '*benign*'. Similarly, more recent studies have suggested that a fair proportion of people with ET have no history of ET in their family. Although this does not prove that it was not inherited in these individuals, like *benign, familial* was also dropped, thus reducing the name of the condition to its bare essentials—ET.

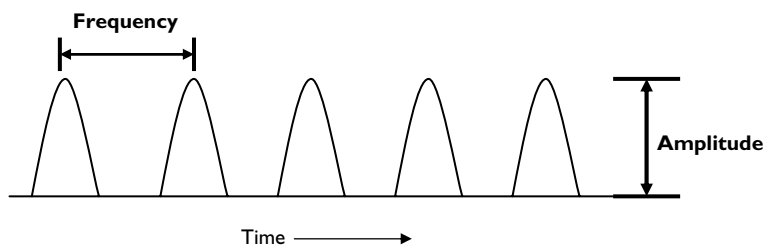


Figure 1.2 Wave frequency versus amplitude (see text). A schematic representation of a regular, oscillating electric pulse as one might find during tremor. The frequency is the time between pulses (1 per s = 1 Hz) and the amplitude defines the strength of the signal (i.e. severity of tremor).

The clinical definition of ‘classical essential tremor’

The bilateral postural and kinetic tremor that is so characteristic of ET implies that something is happening regularly—something is oscillating to cause muscles to contract periodically—with a given frequency during voluntary movement. One way of looking at this is to imagine regular pulses of electrical signals as waves generated over time (Fig. 1.2).

The time between the peaks of the waves defines the frequency (literally how frequent) with which the wave is repeating and this, like radio waves etc., is measured in Hertz (Hz). One Hz is one cycle per second. The height of the peaks is a measure of the amplitude (the strength) of the wave.

It is possible to measure electrical activity in muscles as electrical signals are used to initiate and control muscle contraction. The process is called *electromyography* (EMG), which can be roughly translated as the measurement (*graphy*) of electrical activity (*electro*) in muscles (*myo*). Pairs of tiny circular electrodes can be attached to the skin overlying a muscle or several muscles in the arm, and connected to an amplifier to record and amplify any electrical activity arising from the muscle. There is normally no movement-related electrical activity in resting muscle, and it is only when (electrical) signals are sent to the muscle from the brain via the spinal column and peripheral nerves to initiate a movement that electrical activity is detected from the muscle. EMG analyses of ET patients have indeed detected a regular, oscillatory electrical activity arising from the forearm muscles during postural or kinetic tremor (Fig. 1.3) which is not present in unaffected individuals. The frequency of this rhythmic electrical firing varies from person to person, but in ET it is usually somewhere between 4 and 12 Hz.

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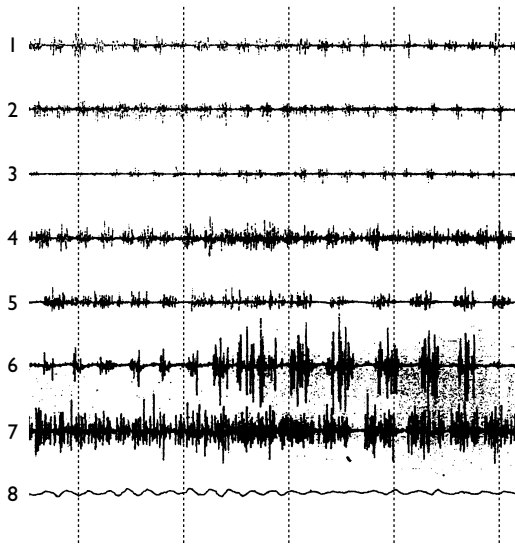


Figure 1.3 EMG recordings. A surface electromyogram (EMG) recording of tremor from several muscles in a patient's right arm held in front of the body to assess postural tremor. The vertical dotted lines are at 1 s intervals. The EMG traces show small electrical bursts occurring in the muscles interspersed by quiet periods. This is known as segregation of the EMG trace and is typical of tremor. The bursts are electrical signals given out by the muscles when they contract. These rhythmic muscle bursts cause tremor. The picture shows that the following muscles are affected by tremor in this patient: channel 1, deltoid; 2, biceps; 3, triceps; 4, forearm flexor muscles; 5, forearm extensor muscles; 6, first dorsal interosseous muscle of the hand; 7, abductor pollicis brevis muscle of the hand; 8, an accelerometer trace of the tremor occurring in the patient's hand. (An accelerometer is a small piezo-electric device that is attached to the back of the patient's hand with tape. It is commonly used to record tremor and works by measuring the acceleration and deceleration movements of the patient's hand caused by tremor in m/s/s). As the intervals between the dotted line are 1 s, so the number of oscillations within an interval can be directly converted into Hz. These traces are unusual as the tremor in some muscles is 5 Hz (traces 1, 2, 3 and 4), but 3 Hz in others (traces 5, 6 and 7). In a person who does not have tremor there would be no EMG bursts. Instead when the muscles were relaxed there would be no signal and, when holding a sustained posture, a continuous EMG signal would be apparent that is not broken up, i.e. not segregated into bursts. Thus the segregation of the EMG into bursts is the hallmark of abnormal tremor.

Continuing with the wave analogy, the amplitude of the oscillatory signal defines the strength of the signal, and therefore, in the case of ET, how badly one shakes. As will be described in greater detail in Chapter 4, drugs such as propranolol which are used to treat ET have little effect on the frequency of this signal but do reduce the amplitude—the shaking does not stop, but there is a marked effect on the severity of the tremor.

The 4–12 Hz tremor implies 4–12 shakes in an ET arm every second—as the EMG was recorded from the forearm muscles, which move (flex and extend) the wrist, this frequency is the number of oscillations the wrist makes per second. It is important to understand that any one muscle can either contract or relax, so a controlled movement of the wrist requires a pair of muscles to co-ordinate their activities—one muscle will move the wrist in one direction (agonist), whilst a different (opposed/antagonist) muscle is required to bring the wrist back to its original position. The 4–12 Hz bursts of electrical activity that can be detected in ET are only observed during voluntary movement, whereas in Parkinson's disease there is a 4–5 Hz tremor in the muscles even when they are at rest. This is known as a Parkinsonian *rest tremor*.

Essential tremor and emotional stress

The SIP study in 1991 mentioned above also found that although the physical symptoms in ET tend to be less severe than in Parkinson's disease, they cause relatively greater 'psychosocial dysfunction'. One literal translation is that ET patients have greater psychological (*psycho*) problems in public (*social*), and is consistent with the possibility that some of the problems in ET are due to self-consciousness/embarrassment—*anxiety* is another word that approximates to the feelings in ET. It also follows that although Parkinson's disease patients are far more severely physically disabled than those with ET, they are less embarrassed (anxious) by their condition. This raises two important questions. First, is psychosocial dysfunction part of ET or an indirect consequence of having the condition? Secondly, if ET patients could accept their condition and reduce their emotional stress in public, would this help them interact more comfortably with society?

Personality characteristics can be assessed in a number of ways. Most recently, individuals with ET were evaluated using a questionnaire to assess three personality traits, namely:

1. *Harm avoidance*: anxiety prone and loath to take risks.
2. *Novelty seeking*: bad tempered or docile.
3. *Reward dependence*: sentimental or aloof.

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ET patients were found to be normal for novelty seeking and reward dependence personality traits, but scored higher on harm avoidance, which describes a person who is pessimistic, fearful, shy, anxious, and easily fatigued.

With the exception of the *easily fatigued* part of that personality trait definition, it is a fair description of what it *feels* like to have ET. The harm avoidance personality trait was associated with the presence of the tremor but not its severity and so is unlikely to be simply associated with tremor-induced disability. What is perhaps highly significant is that three independent studies separated by 14 years (1991 and 2005) and using different questionnaires have come up with very similar results. The other study (2001) confirmed that ET patients tend to suffer from depression and 'psychosocial functioning', but also reported that this was noticed enough to cause disturbance in, or to provoke comments from, the patient's immediate family. These aspects of the ET personality are consistently noticed by the patients themselves and their immediate family, do not require ultrasensitive tests to detect, and thus have important influences on the daily lives of people with ET and their relatives.

Anxiety is a normal response to threatening or dangerous situations that are perceived by the senses. We are all familiar with the unpleasant physical effects brought on by a sudden fright. The response is rapid and uncontrollable (*autonomic*), and involves both neural pathways and the release of stress response hormones, including adrenaline (epinephrine), that are then carried throughout the body in the bloodstream to prepare the body for the emergency. The symptoms of fear and anxiety include sweating, panting, pupil dilation, increased blood pressure, increased heart beat, increased

Patient Perspective

Dry martinis with lots of ice were a favourite family tippie, and these were invariably served with an olive speared on a toothpick. As children, we grew accustomed to the musical sound of rattling ice in glasses, and secretly thought of James Bond who also liked his martinis 'shaken and not stirred'. The process of eating the olive was fascinating as there was a good chance that the olive would fly off before reaching the mouth, and a shaky toothpick anywhere near the eyes is very dangerous. However, the olive was always left to the last, and therefore tackled after the effects of the alcohol had kicked in, thus reducing the potential for accidents. We would never laugh openly (this would be unforgivable) but it was almost impossible not to see or imagine the lighter side. As is so often the case with children, this humour could be quite cruel.

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vigilance, increased reflexes, freezing, an open mouth (jaw drops), and unusual visceral feelings such as ‘butterflies in the stomach’ and a dry mouth.

It is ubiquitous finding that the symptoms of ET worsen during emotional stress, and several of the drugs that are effective in treating ET reduce anxiety and/or dampen the body’s response to stress. It is also well established that relaxation techniques such as meditation, yoga and hypnosis can be effective in times of stress. A sense of humour also helps relieve stress considerably. What is not well recognized is the possibility that for people with ET it is the self-conscious embarrassment of the physical tremor itself that causes, or contributes to, that stress in many (public) circumstances—this must be one major reason for the ‘sociophobia’ which is not specifically addressed by these relaxation techniques, and so they are not particularly effective.

Essential tremor and physiological stress

The tremor in ET also gets worse as a consequence of physiological stress. Physiology is the study of the functions, collectively, of the body. So physiological stress means anything which places a strain on the body’s function. For example, it is common knowledge that excessive alcohol consumption induces a physiological stress, as evidenced by the unpleasant hangover that occurs the following morning, when typically ET will be significantly worse. However, physiological stress has other implications. It may be transiently worse after strenuous exercise, but this usually does not last long. Much more difficult to cope with is the physiological stress of hormonal changes during puberty (Chapter 4) or women’s menstrual cycle—the signs and symptoms of ET do worsen before menstruation, and the thought of having ET, premenstrual tension and a hangover is difficult for a man to imagine. More

Patient Perspective

I first became aware of having a mild form of ET when I was about 8 years old, as physical exercise could cause some shaking. It was at puberty that shaking became quite noticeable, and this was partly due to being self-conscious but I also think it might be related to hormonal changes in the body. However, most of my life I have been convinced that I only have a mild form of ET and so have not let it interfere too much with my life and the things I do in public. I tend to shake when something stressful or unexpected happens, and then it is not only my hands that shake but my whole body and my voice. It is also getting worse with age.

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significant, however, is that physiological stress increases with age and is part and parcel of the process of growing old. Perhaps this partly explains why the severity of ET increases with age.

Ageing—essential tremor is a progressive disease, but is it a neurodegenerative disorder?

James Parkinson originally concluded that although there were similarities between Parkinson's disease and ET, they were in fact distinct conditions, separate movement disorders. What ET, Parkinson's, Alzheimer's and Huntington's diseases all have in common is that they all get worse with age—they are progressive. However, the symptoms of these disorders are all quite different. One big difference between the characteristic tremors seen in ET and Parkinson's disease is that the tremor in ET occurs during an action and disappears during rest, whilst that of Parkinson's disease is present at rest and may persist during a posture. Furthermore, in Parkinson's disease, tremor is accompanied by rigidity in the muscles and slowness of movement (*bradykinesia*), as well as unsteadiness. The combination of a rest tremor and rigidity in Parkinson's disease produces *cogwheel rigidity* in which an examiner can feel a cogwheel- (ratchet) like effect when passively flexing and extending a patient's wrist. The impacts of Parkinson's disease on a person's daily activities are more severe than those of ET. Nevertheless, the troublesome effects of ET are predominantly limited to a great extent on movement. In contrast, Alzheimer's disease affects the mind, causing memory loss as well as a progressive decline in personality and intellect. It is now appreciated that Parkinson's disease also produces dementia, but usually after a long period of physical disability. Similarly Huntington's disease usually manifests itself in mid-adult life as involuntary jerky movements of the arms, face, head, legs and torso, but ultimately significant cognitive and personality changes (*dementia*) occur. The progressive nature of these disorders implies that the neurological damage that causes the symptoms increases with ageing.

Jagger and King (1955) made the following remarks about the tremulous descendants of a Great Lakes ship captain who had essential tremor and migrated to Utah in 1860:

'In two men over 70 years of age, writing, shaving and buttoning clothes were not easy. Of the two, however, one had killed a deer with a rifle at 200 yards the previous autumn.'

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It is clear that different parts of the brain are being affected in these different disorders. ET involves parts of the brain involved in movement, Alzheimer's disease parts of the brain involved in thought, and Huntington's or Parkinson's diseases a combination of the two. What is most significant is that all these neurological conditions can be very difficult to diagnose, and absolute proof of the diagnoses of Parkinson's disease or Alzheimer's disease is only usually made following a post-mortem and a pathological examination of the brain. Different areas of the brain are damaged in Alzheimer's, Parkinson's and Huntington's diseases. This difference in the location of brain damage and differences in the mechanisms causing that damage, their *neuropathology*, accounts for the different symptoms observed. The progressive nature of Parkinson's, Alzheimer's and Huntington's diseases during ageing is therefore due to the gradual accumulation of nerve cell damage in various parts of the brain over time. Consequently, these diseases are classified as progressive neurodegenerative disorders.

There is no definitive evidence of progressive brain damage in ET. ET brains have been examined superficially over the last 100 years without revealing any consistent or obvious signs of brain damage. However, it is always possible to question the criteria used to diagnose ET in these studies (Chapter 2), and the pathological examinations were relatively superficial because the pathologists did not have modern neuropathological techniques and were not sure what areas of the brain to focus their investigations upon. Nevertheless, brain damage is easily detected at post-mortem in Parkinson's, Huntington's or Alzheimer's diseases, so the absence of any historical reference to clearly visible damage in the brains of people who had ET implies that the damage is very subtle. This absence of detailed neuropathological examinations of the ET brain has not gone unnoticed by the clinicians studying ET. The *Essential Tremor Brain Repository* has been established at Columbia University in New York, USA, so that patients with ET can, after death, allow their brains to be donated to a 'tissue bank'. This allows detailed studies to be carried out and the findings from ET brains compared with each other and with brains donated by people not affected by a neurological disorder. Preliminary results from this approach suggest that a defect in a small number of cells within an area of the brain called the *cerebellum*, which is involved in the control of movement, may be one common denominator in ET brains.

A further major complication in the argument that ET is a neurodegenerative condition is that, unlike Parkinson's, Huntington's or Alzheimer's diseases, ET can affect children and young infants. These very early-onset ET cases occur in infants whose brains are still developing, so it is difficult but not impossible to reconcile this finding with the view that ET is a progressive neurodegeneration.

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Even in the more severe and advanced cases of ET, the obvious symptoms are by and large restricted to postural and kinetic tremor affecting the arms, and perhaps also the head, voice, legs and face, without any obvious signs of the cognitive damage such as the memory loss or dementia that occurs in the latter stages of Huntington's, Alzheimer's or Parkinson's diseases. This suggests that either the brain damage in ET is very subtle and restricted to a few highly specialized brain cells (*neurons*) within functionally and anatomically defined region(s) of the brain, or the assumption that a progressive neurodegeneration is the main cause of ET progression is wrong.

Other symptoms in essential tremor

In patients with ET, abnormal electrical and/or biochemical activity has been detected in several key regions of the central nervous system and, in addition to movement control, many of these regions are involved in other neurological processes. Action tremor of the hands and other parts of the body are key features of ET, but an important issue is whether the neurological problems underlying ET cause any other, perhaps more subtle, effects on the body or brain. Furthermore, if this was found to be the case, then would these symptoms worsen during ageing? Many studies have tried to address this issue, but it is sometimes difficult to be absolutely convinced that the neurological problem is actually caused by ET rather than an indirect consequence of living with ET for several decades, or by the normal ageing process. Even if some associated abnormality is discovered, the fact that to date it does not feature in the current diagnostic criteria for ET, or traditional literature of ET, indicates that it is probably very subtle. On the whole, the patients and their families have not noticed it and, if they did, it is of relatively little importance in their day-to-day lives. Nevertheless, it is worth looking for evidence of other neurological problems in ET.

Potential neurological problems can be subdivided into: (1) those involved in mental functions such as cognitive ability, behaviour and personality; and (2) physical problems, for example, affecting walking or the senses.

Cognitive ability

Several studies have examined ET patients looking for evidence of 'cognitive' problems, and concluded that ET patients may have mild impairment of memory, attention, verbal fluency, cognitive flexibility and/or conceptual thinking tasks. This is alarming, but there is some consolation, as ET patients scored higher than unaffected people in several other tests such as 'verbal and non-verbal conceptualization and reasoning'. Furthermore, it is also worth

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remembering that the anxiety present in ET, the tendency to social reclusivity as well as the effects of treatments and co-existing illnesses, may have influenced the results. The cognitive evaluation studies were carried out within formal environments, with the examiner, a stranger, asking the affected ET individual a number of highly personal questions. If one accepts that ET individuals are inherently self-conscious, fearful and shy, the evaluation itself would have been stressful. This could possibly have affected at least some of the results obtained, for example verbal fluency and attention, so these data are difficult to interpret.

These studies report data for a large number of people, and the deficits tend to be mild in ET patients as a whole, but within that group they range from unnoticeable to severe. Furthermore, any cognitive impairment is always compared with the severe effects observed in other disorders such as Parkinson's disease and Alzheimer's disease, and is consistently found to be comparatively mild. Unlike Parkinson's disease, and Alzheimer's disease in particular, any cognitive deficit (such as memory impairment) in ET is only detectable using a battery of sophisticated and highly sensitive tests. The cognitive abnormalities are therefore of little importance to the patient, their families, the clinician or society, and are defined as *subclinical*—not bad enough to warrant clinical intervention.

Some cognitive decline is inevitable during ageing, so the decline observed in elderly ET patients is not out of the ordinary. At worst, on average, the age-related problems might be manifested slightly earlier in ET than in unaffected controls.

Walking

There is no clinical or anecdotal evidence to suggest that ET tremor has a noticeable effect on movement of the legs during walking. Nevertheless, investigators have turned to complex technology to try and detect abnormalities in the walking (gait) of people with ET. If it is not visible to the naked eye, then infrared cameras, video processors and infrared light-reflective markers attached to different regions of the legs can be exploited to compare all aspects of walking in ET with that of unaffected individuals in fine detail—measured in millimetres and degrees of rotation, etc. Patients walk on a treadmill while cameras follow every movement, and the patients can be asked to walk with their hands swinging freely or held behind their neck, to see whether balance is affected.

In one detailed study, a 'gait disorder' was detected in ET patients, but the defect was only detected in a subgroup of ET patients who had particularly severe tremors and also had '*intention tremor*' in addition to the usual-postural and (simple) kinetic tremors. Intention tremor is characterized by

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an exaggeration of tremor during a visually guided movement towards an object, so that the tremor increases as the hand approaches the target object. It is found in diseases of the cerebellum, an area of the brain situated in the back of the skull that is intimately involved in the control of movements. Intention tremor can be associated with other impairments of co-ordination, for example inaccuracy, so that there is a tendency for the hand to overshoot the target (*dysmetria*). Consequently, in these patients, the diagnosis of ET is debatable, or they represent a severely afflicted subgroup of the condition. The gaits of ET patients with classical postural and simple kinetic tremors were not different from those of unaffected individuals.

As the gait disorder is only noticeable in severely affected people, it may be a consequence of having severe tremor that includes *intention tremor*, rather than indicating the presence of a specific neurological defect in ET. Alternatively, one cannot help wondering whether having a shaking head will affect balance during walking.

Eyesight and eye movement

Poor or defective eyesight is not a symptom that has been associated with ET. If eyesight was a problem, severely affected ET patients would have mentioned it to their doctors, and this would undoubtedly have been reported if there was a significant association between ET and bad eyesight. Nevertheless, eye movement abnormalities have been detected, but again only in severely affected patients who had *intention tremor*, and not in typical ET patients who exhibited postural and simple kinetic tremors without *intention tremor*.

The initiation of eye movements in ET patients with *intention tremor* is significantly slower than normal (in milliseconds), although other eye movements (saccadic and gaze-holding functions) are unaffected.

Sense of smell

The sense of smell can be tested using a 'smell battery' in which patients try to recognize various different odours that are presented to them sequentially on scratch cards, each card coming with five potential answers, of which only one is correct. Using this type of test, a deterioration in the sense of smell is now appreciated to be a consistent early sign of Parkinson's disease. Initial studies also appeared to indicate that patients with ET may have a mildly defective sense of smell. However, recent reports have shown that this is not true and that the sense of smell in people with ET is normal. In fact, patients with inherited ET may actually have a better sense of smell than unaffected individuals. This finding may have important clinical implications because in

future it may be possible to use detailed tests of the sense of smell to help distinguish between Parkinson's disease and ET.

Hearing

Deafness in the general population does increase with age, as evidenced by the number of elderly people who rely on hearing aids. If one therefore wants to investigate whether hearing is impaired in ET patients, the majority of whom are elderly, then a number of potential experimental problems arise. One can only determine whether deafness is more or less frequent than expected compared with age-matched controls. One study measured hearing in ET patients, Parkinson's disease patients and healthy individuals using a standard hearing test (*Nursing Home Hearing Handicap Index*). ET patients were found to have an increased hearing disability compared with people with Parkinson's disease and healthy people, and this correlated with tremor severity, age and sex (male). However, this finding needs to be corroborated before too much credence is given to it.

Weight loss

Being underweight has as many health implications as being overweight, and patients with neurodegenerative disorders tend to be thinner than average. For example, in patients with Parkinson's disease, significant weight loss occurs even before the diagnosis is made. The scientific measure of whether an individual is overweight or underweight clearly requires that weight is related to the height (a measure of size) of that individual. The standard measure devised for the purpose of classifying people (normal, fat or thin) is called the 'body mass index' (BMI):

$$\text{BMI} = \text{Weight in kilograms}/(\text{Height in metres})^2$$

Compared with unaffected controls, ET patients are indeed lighter. The average ET patient BMI score of 26 was 5.5 per cent lower than the 27.5 BMI score of unaffected individuals. Although the average BMI score is clearly different, there is a range of BMI scores in each group, and this is expressed as the range above and below (\pm) the average which would include 95 per cent of all BMI scores in each group. Thus the range of ET BMI scores is actually 26.0 ± 4.3 (as high as 30.3 and as low as 21.7), and the control BMI is 27.5 ± 5.0 (as high as 32.5 and as low as 22.5). There is clearly enormous overlap between the ET and control samples, and a statistical test of the difference between the two showed that it was only of borderline significance.

It is probably not worth overinterpreting these marginal differences, which require confirmation. Even if they are accepted at face value, several

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obvious explanations for these findings come to mind:

1. ET patients use up energy shaking and trying to control their shakes—energy that would normally be stored in the body as fat. This extra energy expenditure in ET on average represents 5.5 per cent of food intake that is not be stored in the body, resulting in slightly lower weight.
2. ET patients on average drop 5.5 per cent of the food in each meal due to the shaky trip from the plate to the mouth. The average nutritional value of an ET meal is therefore in effect 5.5 per cent smaller than intended.
3. The ‘anxiety’ present in ET induces an increase in the metabolic rate of the body, which thus consumes more energy, so that there is less spare energy to be stored in the body as fat, etc.

Conclusion

ET causes a tremor that affects the arms and various other parts of the body. This tremor occurs during actions and not whilst resting. It typically occurs when the arms maintain a given posture (postural tremor) and when the arms make a movement (kinetic tremor). In some patients with ET, an intention tremor is also present. This is apparent when the hand makes a purposeful movement towards an object under visual guidance and manifests as an increase in tremor as the hand approaches the object. The self-conscious embarrassment and reluctance to expose the shaking in public (sociophobia and harm avoidance) is increasingly being recognized by patients, families and clinicians as one symptom of ET that can have a significant effect on living and coping with ET.

In the subgroup of ET patients with *intention tremor*, mild abnormalities of gait and eye movements have been detected with sophisticated apparatus. In addition, there is some weak evidence to suggest that there are subtle cognitive changes present in patients with ET. These other symptoms are by and large unnoticed by the affected individuals and their families, and are of no clinical importance.

