

DEFINITION

1. **Benign Essential Tremor** (synonyms: **essential tremor, ET, familial tremor**) is a postural disorder predominantly affecting the upper limbs, and which may also involve the head and jaw. The lower limbs are affected in fewer cases. It is a monosymptomatic disorder.

CLINICAL MANIFESTATIONS

2. The onset is typically in middle age, but may be at any time from childhood to old age. In the aged it is often called senile tremor, but there is no evidence that senile tremor is a distinct entity. Essential tremor develops insidiously and persists for life. Progression is usually slow and sometimes negligible. In many cases, the tremor causes little disablement and is accepted as a harmless idiosyncrasy, so that a relatively small proportion of those affected seek medical advice.
3. The tremor is rhythmical and appears only whilst maintaining a posture, eg. when holding a cup or writing. It may be unilateral and often remains asymmetrical. In 50% of cases there is titubation (tremor of the head, usually of the nodding or "yes-yes" type) and tremor of the jaw. The legs are usually spared or only mildly affected. The gait and tests of co-ordination are usually normal. There is no other neurological symptom or clinical sign.
4. In a small proportion of patients, the tremor becomes more rapidly progressive and severe. The speech may be tremulous and difficult to understand. Eventually, such patients may be unable to hold cutlery or crockery securely enough to eat or drink.
5. The rate of oscillation in essential tremor is usually between 5 and 8 Hz (Hertz or cycles per second), which is slower than the normal "physiological" tremor of the outstretched hands and more rapid than the tremor of parkinsonism.
6. Essential tremor is hardly accentuated by movement, but it is aggravated by emotional stress, exertion and fatigue, partly due to activation of beta-adrenergic receptors in muscle and possibly also in the central nervous system.
7. In mild cases, the tremor may be suppressed by beta-blocking drugs. It is also suppressed by moderate doses (about two units) of alcohol but the mechanism is unknown. Anticonvulsant drugs may be effective in some cases, but about 30% do not respond to treatment. Antiparkinsonian drugs have no effect on essential tremor. Thalamotomy (surgery to part of the brain) is performed on a very small proportion of patients in whom the tremor is severe and refractory, but is not always effective.
8. Some cases of solitary head tremor and of writer's cramp may be variants of essential tremor. However, solitary head or jaw tremor was not found in a large series of familial case studies.

AETIOLOGY

9. The physiological mechanism causing essential tremor is unknown. It may involve instability in neuronal loops controlling the posture of affected parts of the body. Esoteric tests have shown increased activity in the cerebellum.
10. Essential tremor is not associated with any biochemical disorder and no abnormality has been found at post-mortem examination of the nervous systems of patients.
11. The condition is inherited and a positive family history is reported in more than 50% of cases. The responsible gene has not been identified, but the pattern of inheritance is that of an autosomal dominant with very high penetrance (a gene carried by either gender which, where present in an individual, nearly always causes the relevant condition).

CONCLUSION

12. **Benign essential tremor** is a coarse tremor, predominantly affecting the upper limbs and head, which most commonly appears in middle age and persists for life. It is not associated with pathological or biochemical abnormality, nor any other neurological disturbance. The tremor is inherited. There is no evidence that trauma (physical or psychological), climate or other environmental factors have a causative role.

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