

require surgery) has also been shown to be an effective therapy against tremor in ET.

DYSTONIA

■ CLINICAL FEATURES

Dystonia is a movement disorder characterized by sustained or intermittent muscle contractions of agonist/antagonist muscles causing abnormal, often repetitive movements and postures. Dystonic movements are typically patterned and twisting and may be associated with a “dystonic” tremor in which the tremor is most pronounced when the body part is moved in the direction of the dystonia. Dystonia is often initiated or worsened by voluntary action and associated with overflow muscle activation. Dystonia can range from focal minor contractions affecting only an individual muscle group to severe and disabling involvement of multiple muscle groups. The frequency is estimated to be 16 per 100,000 (~50,000 cases in the United States) but is likely to be much higher because many cases are not recognized. Dystonia is often brought out by voluntary movements (action dystonia) and can extend to involve other muscle groups and body regions not required for the intended action (overflow). It can be aggravated by stress and fatigue and attenuated by relaxation and sensory tricks such as touching the affected body part (*geste antagoniste*).

Historically, dystonia has been described as primary or secondary. However, because of a confusing and not always congruent combination of phenotypic and etiologic features, the older terms are no longer recommended. A Movement Disorder Society Task Force charged with redefining dystonia recommends classifying dystonia along the same main axes as ET: clinical and etiologic. On clinical grounds, dystonia can be categorized by age of onset (infancy, childhood, adolescence, early and late adulthood), body distribution (focal, segmental, multifocal, and generalized), temporal pattern (static or progressive, action-specific [diurnal and paroxysmal]), and association with additional features. Clinical description along these lines enables formulating specific dystonia syndromes (e.g., early-onset generalized isolated dystonia). From an