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### Chorea: whither comest it?

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# Commentary

## **Chorea: Whither Comest It?**

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*Chorea*—the term itself is a rather peculiar one. The word chorea is derived from the Greek word for dance and originally was applied to describe the dancelike gait of the dancing mania, a hysterical disorder of the late Middle Ages. The term is now used to describe an entire class of abnormal spontaneous movements. Each of these movements is a single, isolated muscle action: a short, rapid, uncoordinated jerk. These jerks can be proximal or distal and are of sufficient magnitude to move the involved limb. The simultaneous or successive occurrence of two or more such isolated movements can result in complex patterns of movement, and the superimposition of these movements on normal movements can cause a dancelike gait.

What little we know and understand about chorea is based on our knowledge of a single disease state: Huntington chorea. In Huntington chorea, the major pathologic features are limited to the corpus striatum and cerebral cortex. Striatal atrophy is the most striking change. Although particular involvement of the small neurons of the caudate nucleus is present, it is not unusual to see degeneration of the large caudate neurons as well. Associated with this marked neuronal loss is extensive proliferation of astrocytes. The other striking pathologic feature is diffuse cerebral atrophy. This takes the form of a general reduction in the population of cortical cells, with particularly heavy cell loss in layers 3, 5, and 6. It generally is accepted that the choreiform movements of Huntington disease are related to striatal pathologic changes and that the mental changes are related to cortical pathologic changes.

Thomas Sydenham first described acute chorea in 1685. Today the term Sydenham chorea is applied to instances of sporadic chorea that occur during childhood and adolescence. Over the years, it has come to be accepted that in most instances, this syndrome is of rheumatic origin. It is thought now that most cases of Sydenham chorea are a late complication of acute rheumatic fever and a late manifestation of immunoallergic tissue reaction between the streptococcal toxin and the patient that involves the nervous system [1]. Although rheumatic disease does account for the majority of these cases, a rheumatic cause cannot be shown in all.

Because of the favorable prognosis of Sydenham chorea, relatively few postmortem studies have been done. The CNS lesions are usually widespread and nonspecific. They include acute and chronic neuronal degenerative changes and vascular and inflammatory lesions [2]. In some instances, necrotizing arteritis is found. The brain often is involved diffusely in this process, but most early reports noted a predilection for the striatum [3]. More recent reports have recorded these same lesions of the caudate nucleus and putamen but have more emphasis on the involvement of the cerebral cortex.

The work reported by Kienzle et al. [4] in this issue of the *AJNR* is of far more than radiologic and diagnostic significance. It helps us understand the pathophysiology of the abnormal movement in Sydenham chorea and in other choreatic disorders. Their observations, that the chorea in Sydenham chorea is associated with observable changes within the caudate nucleus itself, strengthen the hypothesis that chorea, in general, is due to dysfunction of the striatum.

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This article is a commentary on the preceding article by Kienzle et al.

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