

Dystonia Pathophysiology

The [pathophysiology](#) of [dystonia](#) is poorly understood. As opposed to secondary forms of dystonia, [primary dystonia](#) has long been believed to lack any neuroanatomical substrate. During trajectory planning for [DBS](#), however, conspicuous T2-[hyperintense](#) signal alterations (SA) were registered within the target region, even in young patients, where ischemia is rare.

Fifty MRIs of primary dystonia patients scheduled for DBS were analyzed. Total basal ganglia (BG) volumes, as well as proportionate SA volumes, were measured and compared to 50 age-matched control patients.

There was a 10-fold preponderance of percentaged SA within the globus pallidus (GP) in dystonia patients. The greatest disparity was in young patients <25 years. Also, total BG volume differences were observed with larger GP and markedly smaller putamen and caudate in the dystonia group.

BG morphology in primary dystonia differed from a control population. Volume reductions of the [putamen](#) and caudate may reflect functional degeneration, while volume increases of the GP may indicate overactivity. T2-hyperintensive SA in the GP of young primary dystonia patients, where microvascular lesions are highly unlikely, are striking. Their pathogenic role remains unclear ¹⁾.

Over the past years, research into the [neurophysiology](#) of the [basal ganglia](#) has provided new insights into the pathophysiology of [movement disorders](#). The presence of pathological oscillations at specific frequencies has been linked to different signs and symptoms in PD and [dystonia](#), suggesting a new model to explain basal ganglia dysfunction. These advances occurred in parallel with improvements in imaging and neurosurgical techniques, both of which having facilitated the more widespread use of [DBS](#) to modulate dysfunctional circuits. High-frequency stimulation is thought to disrupt pathological activity in the motor cortex/basal ganglia network; however, it is not easy to explain all of its effects based only on changes in network oscillations ²⁾.

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Bai X, Vajkoczy P, Faust K. Morphological Abnormalities in the Basal Ganglia of Dystonia Patients. Stereotact Funct Neurosurg. 2021 Jan 20:1-12. doi: 10.1159/000512599. Epub ahead of print. PMID: 33472209.

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Guridi J, Alegre M. Oscillatory activity in the basal ganglia and deep brain stimulation. Mov Disord. 2016 Aug 22. doi: 10.1002/mds.26714. [Epub ahead of print] Review. PubMed PMID: 27548437.

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