

**CORTICAL-BASAL GANGLIONIC DEGENERATION**

*DE Riley, AE Lang, A Lewis, L Resch, P Ashby, O Hornykiewicz, S Black*

**Neurology 1990;40:1203-1212**

We report our experience with 15 patients believed to have cortical-basal ganglionic degeneration. The clinical picture is distinctive, comprising features referable to both cortical and basal ganglionic dysfunction. Characteristic manifestations include cortical sensory loss, focal reflex myoclonus, "alien limb" phenomena, apraxia, rigidity and akinesia, a postural-action tremor, limb dystonia, hyperreflexia, and postural instability. The asymmetry of symptoms and signs is often striking. Brain imaging may demonstrate greater abnormalities contralateral to the more affected side. Postmortem studies in 2 patients revealed the characteristic pathologic features of swollen, poorly staining (achromatic) neurons and degeneration of cerebral cortex and substantia nigra. Biochemical analysis of 1 brain showed a severe, diffuse loss of dopamine in the striatum. This condition is more frequent than previously believed, and the diagnosis can be predicted during life on the basis of clinical findings. However, as with other "degenerative" diseases of the nervous system, a definitive diagnosis of cortical-basal ganglionic degeneration requires confirmation by autopsy.

---

Free Access to this article at [www.neurology.org/content/40/8/1203](http://www.neurology.org/content/40/8/1203)

**Comment from Ryan J. Uitti, MD, FAAN, Associate Editor:** *This manuscript provided a major contribution to the description of a newly recognized clinical entity: cortical basal ganglionic degeneration.*

# Neurology<sup>®</sup>

## **Cortical-basal ganglionic degeneration**

DE Riley, AE Lang, A Lewis, et al.

*Neurology* 2011;76;1634

DOI 10.1212/01.wnl.0000398198.48275.f4

**This information is current as of May 9, 2011**

### **Updated Information & Services**

including high resolution figures, can be found at:  
<http://n.neurology.org/content/76/19/1634.citation.full>

### **Permissions & Licensing**

Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at:  
[http://www.neurology.org/about/about\\_the\\_journal#permissions](http://www.neurology.org/about/about_the_journal#permissions)

### **Reprints**

Information about ordering reprints can be found online:  
<http://n.neurology.org/subscribers/advertise>

*Neurology*® is the official journal of the American Academy of Neurology. Published continuously since 1951, it is now a weekly with 48 issues per year. Copyright Copyright © 2011 by AAN Enterprises, Inc.. All rights reserved. Print ISSN: 0028-3878. Online ISSN: 1526-632X.

