2. Rogaev EI, Sherrington R, Rogaeva EA, et al. Familial Alzheimer's disease in kindreds with missense mutations in a gene on chromosome 1 related to the Alzheimer's disease type 3 gene. *Nature* 1995;376:775-778.
3. Levy-Lahad E, Wasco W, Poorkak P, et al. Candidate gene for the chromosome 1 familial Alzheimer's disease locus. *Science* 1995;269:973-977.
4. Cruts M, van Duijn CM, Backhovens H, et al. Estimation of the genetic contribution of presenilin-1 and -2 mutations in a population-based study of presenile Alzheimer disease. *Hum Mol Genet* 1998;7:43-51.
5. Finckh U, Muller-Thomsen T, Mann U, et al. High prevalence of pathogenic mutations in patients with early-onset dementia detected by sequence analyses of four different genes. *Am J Hum Genet* 2000;66:110-117.
6. Lao JI, Beyer K, Fernández-Novoa L, Cacabelos R. A novel mutation in the predicted TM2 domain of the presenilin 2 gene

in a Spanish patient with late-onset Alzheimer's disease. *Neurogenetics* 1998;1:293-296.

7. Goate A, Chartier-Harlin MC, Mullan M, et al. Segregation of a missense mutation in the amyloid precursor protein gene with familial Alzheimer's disease. *Nature* 1991;349:704-709.
8. Shirotani K, Takahashi K, Araki W, Maruyama K, Tabira T. Mutational analysis of intrinsic regions of presenilin 2 that determine its endoproteolytic cleavage and pathological function. *J Biol Chem* 2000;275:3681-3686.
9. Thinakaran G, Harris CL, Ratovitski T, et al. Evidence that levels of presenilins (PS1 and PS2) are coordinately regulated by competition for limiting cellular factors. *J Biol Chem* 1997; 272:28415-28422.
10. Bird TD, Levy-Lahad E, Poorkaj P, et al. Wide range in age of onset for chromosome 1-related familial Alzheimer's disease. *Ann Neurol* 1996;40:932-936.

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Figure. (Top) Upper facial muscles anatomy: procerus muscle originates in the nasal bone and inserts in the skin in the center of the forehead between the eyebrows. (Bottom) Marked procerus contraction in two patients with clinically probable progressive supranuclear palsy.

Procerus sign in progressive supranuclear palsy

S. Romano, MD, C. Colosimo, MD, Rome, Italy

Progressive supranuclear palsy (PSP) is characterized by vertical supranuclear gaze palsy, axial rigidity, akinesia, dysarthria, fronto-limbic dementia, and postural instability.¹ A typical facial expression, described as “astonished,” “worried,” or “reptile-like,” has been described²; this peculiar expression, so common in PSP and different from the lack of facial expression (hypomimia) typically seen in patients with PD, has not been well characterized. The expression may be due to a focal dystonia of the procerus muscle as well as to a combination of very reduced blinking, spasticity, lid retraction, and gaze palsy. Procerus is a facial muscle that originates in the nasal bone and inserts in the skin

in the center of the forehead between the eyebrows; it acts forming vertical wrinkles in the glabella region and bridge of the nose (figure, top). The figure (bottom) shows two patients with clinically probable PSP according to the National Institute of Neurological Disorders and Stroke-SPSP criteria.¹ Both patients show this typical sign, occurring without concomitant blepharospasm. Indeed, the wrinkling is present with open and closed eyes. This sign could be called “the procerus sign.”

1. Litvan I, Agid Y, Calne D, et al. Clinical research criteria for the diagnosis of progressive supranuclear palsy (Steele-Richardson-Olszewski syndrome). Report of the NINDS-SPSP International Workshop. *Neurology* 1996;47:1-9.
2. Friedman DI, Jankovic J, McCrary JA III. Neuro-ophthalmic findings in progressive supranuclear palsy. *J Clin Neuro-ophthalmol* 1992;12:104-109.

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