

# Procerus Sign: Mechanism, Clinical Usefulness, and Controversies

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## Abstract

This sign is observed in atypical parkinsonism mainly in Progressive Supranuclear Palsy. The exact mechanism of this sign is not clear but the contraction of the Corrugator Supercilii muscle is the most important factor for the generation of vertical forehead creases. Though the wrinkling of forehead is a complex phenomenon involving multiple muscles but the ‘Corrugator sign’ would be a better term to describe this phenomenon. Despite the controversies regarding the mechanism and nomenclature, the ‘Procerus sign’ remains an important clinical clue for early Progressive Supranuclear palsy.

**Keywords:** Corrugator supercilii, dystonia, glabella, procerus, progressive supranuclear palsy

The “Procerus sign” means vertical wrinkling on the forehead in atypical parkinsonism, especially progressive supranuclear palsy (PSP) [Figure 1]. This imparts a “staring,” reptile-like, or “astonished” look characterized by lid retraction and infrequent blinking [Video 1]. This astonished look of the PSP patients is different from the hypomimic facial appearance of the Parkinson’s disease patients.

The Procerus sign was first described by Romano and Colosimo in 2001 in two patients.<sup>[1]</sup> The authors suggested that the vertical wrinkling on glabella and bridge of the nose was secondary to the dystonic contraction of the procerus muscle and could be a clinical sign of early PSP. The sign persists regardless of the eye closure and is not associated with blepharospasm. Shibasaki Warabi *et al.* also reported this sign in patients with probable corticobasal diseases. These authors also thought that the sign was secondary to dystonia of the procerus muscle.<sup>[2]</sup>

However, the generation of the vertical wrinkles is a complex event and involves the contraction of corrugator supercilii and orbicularis oculi muscles in addition to the contraction of the procerus muscle [Figure 2].<sup>[3-6]</sup> Procerus muscle originates in the nasal bone and inserts in the skin at the center of the forehead between the eyebrows. The procerus draws the medial angle of the eyebrows down, resulting in transverse wrinkling over the bridge of the nose.<sup>[2-4]</sup> Corrugator supercilii pulls the eyebrows

downward and medially, leading to the vertical wrinkling of the supranasal skin of the forehead.<sup>[4,5]</sup> This anatomical principle is the basis of surgical resection and chemodenervation of corrugator supercilii to treat vertical glabellar creases. The orbicularis oculi muscle has three parts, i.e., palpebral, orbital, and lacrimal part. The whole orbicularis oculi muscle complex pulls the skin of the forehead, temple, and cheek toward the medial angle of the orbit.<sup>[5]</sup> However, corrugator seems to be the most important muscle to produce the frown or vertical creases and reflects the “idea of the mind.”<sup>[7]</sup> The term “corrugator supercilii sign” instead of the “Procerus sign” would have been anatomically more appropriate though not perfect. The original authors also agreed to the fact that the vertical creases on forehead is a complex phenomenon and their aim was not to delve into the anatomical complexities but to highlight this important clinical sign in early PSP.

Batla *et al.* used the term “vertical wrinkling of forehead” instead of the “Procerus sign” to describe the complex action of multiple forehead muscles generating this sign.<sup>[3]</sup> They showed that the prevalence of this sign was 75% in the PSP–Richardson subtype and 40% in the PSP–Parkinsonian subtype. “Procerus sign” is claimed to be an early and relatively specific clinical sign of PSP.<sup>[1,3,8]</sup> Morimatsu

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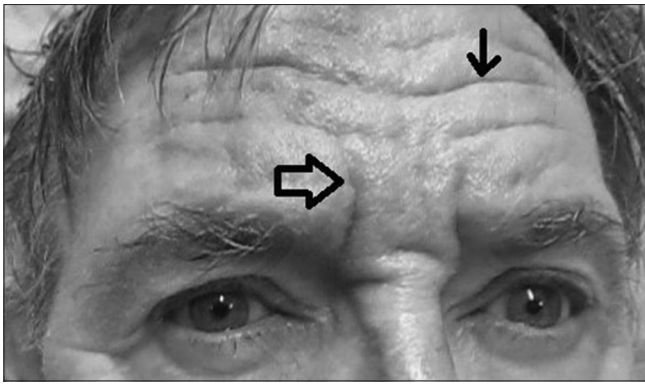
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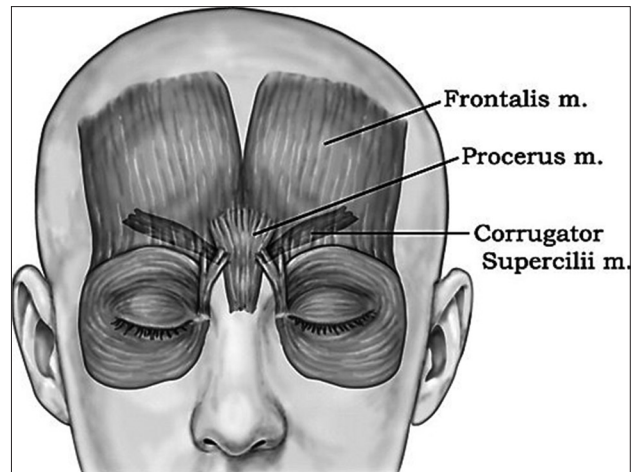
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**Figure 1:** Vertical wrinkling of the forehead (Procerus sign-large arrow) also shows prominent horizontal creases (small arrow)

opined that vertical wrinkling on forehead in the presence of blepharospasm is common with Meige's syndrome, so blepharospasm needs to be excluded to detect this sign, but Romano and Colosimo did not suggest the same.<sup>[9]</sup> However, vertical creases of forehead sometimes can be seen in normal people, so it is important to determine to what degree of vertical creases on forehead can be considered as normal.<sup>[9]</sup> Although this sign was also described in probable corticobasal diseases, misdiagnosis of PSP Corticobasal degeneration clinically is common as both are characterized by selective 4-repeat Tau deposition.<sup>[2,8,10]</sup> One patient with probable multiple system atrophy (MSA) was also found to demonstrate this sign.<sup>[2]</sup> Hence, it is possible that widespread degeneration outside substantia nigra (as occurs in Parkinson's disease) is required to produce this sign. However, recent literature suggests that PSP can present with predominantly cerebellar symptoms, making the distinction between probable MSA and atypical PSP clinically difficult.<sup>[11]</sup> Only histopathological studies can confirm whether this sign is a generic manifestation of tauopathy or specific to PSP.

What is the best way to describe this sign "Procerus sign" or "corrugator supercilii sign" or simply "vertical wrinkling of the forehead" needs to be settled. Another unresolved question is the exact mechanism of the sign. Is it dystonia and if yes which muscle is most important? There is no electromyographic study to support dystonia as the cause of this sign. The discovery of the underlying mechanism would probably help get the most appropriate name for this sign. There is a scarcity of literature on whether PSP patients with procerus sign have dystonia in other parts of body too such as neck/limbs/face. It is also unclear if PSP patients with Procerus sign and those without have differing scores on the PSP rating scale. Moreover, the exact prevalence of this sign in PSP is unknown. These are probably the reasons why this sign was not included by the International Movement Disorders Society in its most updated PSP clinical diagnostic criteria of 2017. However, despite these controversies, the "Procerus sign" remains a helpful clinical sign for early PSP. The presence of this vertical wrinkling on forehead should be considered as red flag sign for idiopathic Parkinson's disease and should make the clinician suspicious of PSP.



**Figure 2:** Anatomical diagram of the Procerus, Corrugator Supercilii, and the periorbital circular shaped Orbicularis Oculi muscle (reproduced with permission from Benjamin AB. *Cosmetic use of botulinum toxin in the upper face. Operative techniques in Otolaryngology-Head and Neck Surgery.* 2007;18;248-53)

### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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### Conflicts of interest

There are no conflicts of interest.

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