



Editors' Note: Hodgson and Lukas and Freeman et al. discuss subspecialization in neurology and its potential impact on the future neurology workforces. Alfahad questions the diagnosis of amyotrophic lateral sclerosis (ALS) in the Teaching NeuroImage by Sharma et al. He argues that, because of the young age and lack of convincing upper motor neuron involvement in the patient reported, other potential ALS mimickers should be considered. The authors respond.

—*Chafic Karam, MD, and Robert C. Griggs, MD*

THE WORKFORCE TASK FORCE REPORT: CLINICAL IMPLICATIONS FOR NEUROLOGY

Trent S. Hodgson, Rimas V. Lukas, Chicago: Dall et al.¹ noted that the future supply of neurologists will be affected by the increasing subspecialization of younger neurologists. The decision to subspecialize is a highly personal decision that may not be driven by patient demand.² This, in addition to the geographic distribution of neurologists demonstrated by Dall et al., could lead to an oversupply in a few subspecialties and a shortfall in others. All of this will be in the context of upcoming economic challenges described in the recent report by the American Academy of Neurology Workforce Task Force.³ Given the potentially large impact of increasing subspecialization, it is important to monitor the effects of new accreditations that are being created for neurology fellowship programs by the American Board of Psychiatry and Neurology and United Council for Neurologic Subspecialties.⁴

Author Response: William D. Freeman, Jacksonville, FL; Kenneth A. Vatz, Chicago; Robert C. Griggs, Rochester, NY; Timothy Pedley, New York: We appreciate the comments by Drs. Hodgson and Lukas concerning factors that affect decisions trainees make about choice of subspecialties and the potential impact of those decisions on the composition of future neurology workforces. The unsettled economics of health care makes it unclear whether the number and types of neurology subspecialty fellowships, especially those associated with cuts in graduate medical education (GME) funding, will increase or decrease over time.

Significant reduction in GME funding could lead to decreased residency/fellowship slots due to the medical school/residency “mismatch.”⁵ Hodgson and Lukas correctly pointed out that the accreditation process itself may have resulted in increased subspecialization in neurology independent of the patient demand or medical need for these subspecialties.⁶ Much, however, will still depend on economic priorities as determined by policymakers, especially with regard to GME funding.

Ultimately, optimizing specialty and subspecialty allocations will likely require a restructuring of both the medical school experience as is already being done at many centers and GME, as well as the development of new models of financing both medical school and GME.

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TEACHING NEUROIMAGES: SNAKE EYES APPEARANCE IN MRI IN PATIENT WITH ALS

Tariq B. Alfahad, Washington, DC: Sharma et al.¹ presented an interesting radiologic finding—the “snake eyes” appearance—which has been reported in several neurologic disorders. However, I question the diagnosis of amyotrophic lateral sclerosis (ALS) since the patient was only 17 years old. Classic ALS does not occur in this age group. An ALS-like complication of HIV or human T-cell lymphotropic virus 1 has been reported.² In addition, we have seen

teenage girls who have diffuse symmetric brisk reflexes that are normal. The authors did not report whether the patient had any other signs of upper motor neuron (UMN) involvement, such as clonus, abnormal plantar response, or spasticity. Otherwise, the diagnosis of ALS cannot be established according to the El Escorial criteria.³ The differential diagnoses should include ALS-like mimickers such as Hirayama disease or O'Sullivan-McLeod syndrome.

Author Response: Pradeep Pankajakshan Nair, Puducherry, India: We thank Dr. Alfahad for his interest in our Teaching NeuroImage.¹ He questioned the diagnosis of ALS due to the unusual age at presentation and clinical features. Initially, we considered Hirayama disease because of upper limb wasting. However, the brachioradialis was wasted and the deep tendon reflexes in the atrophied limbs were exaggerated, including supinator jerk.

We agree that hyperreflexia is common in female patients of this age, but hyperreflexia in a limb and muscle that are wasted is considered pathologic. We also agree that classic ALS is usually seen in older adults. However, there are reports of juvenile sporadic ALS from Asian countries. Juvenile sporadic ALS is a rare form of motor neuron disease occurring in patients younger than 25 years of age. Zou et al.⁴ reported association of *FAS* mutation with juvenile ALS in Chinese patients.

We could not do genetic analysis in our patient. According to the El Escorial criteria, a diagnosis of probable ALS can be kept if 2 regions are showing UMN and lower motor neuron (LMN) signs with some UMN signs rostral to the LMN signs either clinically or by electrophysiology.⁵ We had EMG evidence of denervation in the lower limbs and UMN signs in the upper limb so this fulfills the criteria. Finally, we ruled out retrovirus before doing the EMG.

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Teaching Neuroimages: Snake eyes appearance in MRI in patient with ALS

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