l stiff-man syndrome: abnormal brainstem reflexes suggest a physiological relationship

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Abstract:

Background and objectives: Hyperekplexia and the stiff-man syndrome (SMS) are both conditions with exaggerated startle suggesting abnormal brainstem function. Investigation of brainstem reflexes may provide insight into disturbed reflex excitation and inhibition underlying these movement disorders. Patients and methods: Using four-channel EMG, we examined four trigeminal brainstem reflexes (monosynaptic masseter, masseter inhibitory, glabella, and orbicularis oculi blink reflexes) and their spread into pericranial muscles in five patients with familial hyperekplexia (FH), two with acquired hyperekplexia (AH), 10 with SMS, and 15 healthy control subjects. Results: Both FH/AH and SMS patients had abnormal propagation of brainstem reflexes into pericranial muscles. All patients with hyperekplexia showed an abnormal short-latency (15ï 20 ms) reflex in the trapezius muscle with a characteristic clinical appearance (ñhead retraction jerkò) evoked by tactile or electrical stimulation of the trigeminal nerve, but normal monosynaptic masseter reflexes. Inhibitory brainstem reflexes were attenuated in some FH/AH patients. Four of 10 patients with SMS had similar short-latency reflexes in the neck muscles and frequently showed widespread enhancement of other excitatory reflexes, reflex spasms, and attenuation of inhibitory brainstem reflexes. Conclusion: Reflex excitation is exaggerated and inhibition is attenuated in both stiff-man syndrome and familial or acquired hyperekplexia, indicating a physiological relationship. Reflex transmission in the brainstem appears biased towards excitation which may imply dysfunction of inhibitory glycinergic or GABAergic interneurons, or both.