

## Ehlers-danlos syndrome, proprioception, and pain: The effects of proprioceptive impairment on pain-related communication with pediatric patients with joint hypermobility syndrome/ehlers-danlos syndromes



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### Abstract

Proprioception is often referred to as the “sixth sense”, our conscious and unconscious ability to know the precise position and velocity of our bodies in space, even when we’re immersed in total darkness. While proprioception is necessary for motor control, its function is uniquely introspective, as motor and sensory systems must collaborate to continuously and accurately measure physical properties within the body, such as muscle length and tension, deep pressure, and limb velocity. Proprioception emerges from a complex system of mechanoreceptors, fibrous collagenous connective tissue, neural coordination, and the highly specialized vestibular system. Proprioception may become impaired for various reasons, including certain medical conditions (e.g. Parkinson’s disease, chronic and recurrent low back pain) and adolescents experiencing rapid growth or benign joint hypermobility. However, Ehlers-Danlos Syndrome, a connective tissue disease that results in tissue fragility and joint instability, demonstrates widespread proprioceptive impairment among patients. The severity of hypermobility is positively correlated with impaired proprioception, although the underlying mechanism is not well understood.

Ehlers-Danlos Syndrome is a collection of inheritable connective tissue diseases, consisting of thirteen subtypes caused by various mutations affecting the widespread collagen protein or a collagen-affecting enzyme. Each subtype presents diverse pathological variations, as well as a wide spectrum of severity and disability, although a recent cohort study of EDS patients of various subtypes revealed pain as a unifying experience, with 90% of patients reporting pain. Impaired proprioception is a dimension of the increased risk of injury among EDS patients; the laxity of tissues may result in proprioceptors sending incorrect sensory input to afferent nerves, which continuously inform and update the somatosensory maps produced by the somatosensory, motor, and parietal cortices of the brain.

A 2015 study revealed that EDS patients were less precise in estimating the precise position of their hands in relation to visible peripheral reference locations, and that this proprioception deficit was positively correlated with the severity of the patient’s hypermobility. The relationship between proprioceptive sensitivity and chronic pain in EDS patients was also explored. The study revealed that EDS patients were as accurate as controls in their ability to detect the magnitude of the pain, but consistently were unable to accurately identify the precise location of nociceptive (pain) signals.

Pain-related communication faces many barriers in pediatric populations, specifically for children and adolescents affected by hypermobility and proprioceptive impairment. Communicating with pediatric patients in a way that is developmentally appropriate and sensitive to cognitive and physiological communication barriers can reduce the risk of distress and medical trauma. Identifying and communicating one’s internal state requires the integration of sensory information, representational thought, and language, and this complex process is unrefined during the developmental stages of childhood. Adolescents may predictably experience difficulty in identifying the location, type, and intensity of pain, and the presence of hypermobility and impaired proprioception introduces an additional barrier to pain-related communication in pediatric populations.

### Biography

Randall Burks is completing her undergraduate degree at the Harvard University extension division. She has published articles on Ehlers-Danlos syndrome in the American Association for Anatomy (2020) and the American Journal of Rare Disorders (2020). Randall previously worked as a research intern in a developmental cardiology lab at the Medical University of South Carolina, focusing on mitral valve prolapse.



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