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LETTER TO THE EDITOR

Comment on "Quantitative measures of tissue mechanics to detect hypermobile Ehlers-Danlos syndrome and hypermobility syndrome disorders: a systematic review"

Rodney Grahame[1] · Irfan Malik[2] · Alan Hakim[3] · Myles Koby[4] · Fraser Henderson Sr.[4,5,6]

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Dear Sir,

We read with interest the article by Palmer [1] (Palmer, S., Denner, E., Riglar, M., Scannell, H., Webb, S. and Young, G., 2020. Quantitative measures of tissue mechanics to detect hypermobile Ehlers-Danlos syndrome and hypermobility syndrome disorders: a systematic review. *Clinical Rheumatology*, *39*(3), pp.715–725). Palmer et al. question the validity of the Beighton Score, which assesses hyper-extensibility in a limited number of joints, but does not include the most affected joints. Furthermore, the present criteria for hEDS (EDS—hypermobility type) and HSD (the hypermobility spectrum disorders) do not address the wider issues of connective tissue pathology. Palmer et al. reviewed 203 potentially relevant studies, of which 4 were chosen for systematic review; three described at least one measure of tissue mechanics that differentiated subjects with hEDS/HSD from healthy controls. These included assessment of skin, muscle and tendon.

We draw attention to another measure of tissue mechanics that of the measureable hyper-extensibility of craniocervical ligaments causing "floppy head syndromes" in this population. To address the differentiation of pathological hyper-extensibility from benign hypermobility, a consensus conference was sponsored jointly by the Office of the Medical Commissioner of the National Health Service, Bobby Jones Chiari Syringomyelia Foundation, the Ehlers-Danlos Society and Lady Trish Malloch-Brown. Lectures and arguments were put forward to address the presentation and diagnosis of craniocervical instability and ventral brainstem compression in the EDS population. A robust "round table" discussion enjoined by representatives of neurosurgery, neurology, neuro-radiology, genetics, rheumatology and physiatry reached the following consensus:

1. That hereditary connective tissue disorders—including but not limited to Marfan syndrome, the Ehlers-Danlos syndromes, Down syndrome, osteogenesis imperfecta—are characterized by ligamentous incompetence;
2. That in a small subpopulation of subjects with a hereditary connective disorder, ligamentous incompetence may result in radiological evidence of craniocervical or spinal instability, basilar invagination or ventral brainstem compression;
3. That dynamic imaging is necessary to demonstrate ligamentous incompetence at the craniocervical junction; dynamic imaging may include flexion extension MRI, flexion extension or rotational CT or in some cases fluoroscopic imaging;
4. That a diagnosis of radiological instability, basilar invagination or ventral brainstem compression should be characterized by radiological metrics which have been established in the literature;
5. That Subjects with hereditary connective tissue disorders who receive a radiological diagnosis of craniocervical instability, basilar invagination or ventral brainstem compression should undergo a neurosurgical evaluation for the presence of clinical findings of instability;
6. That Subjects with appropriate clinical findings and congruent radiological findings of craniocervical instability,

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✉ Fraser Henderson, Sr.
henderson@fraserhendersonmd.com

[1] Centre for Rheumatology Research, University College London, London, UK

[2] Neurosurgery, King's College Hospital, London, UK

[3] Rheumatology, Platinum Medical Centre, The Wellington Hospital, London, UK

[4] Doctors Community Hospital, Lanham, MD, USA

[5] Neurological Surgery, University of Maryland Prince George's Hospital Center, Cheverley, MD, USA

[6] Metropolitan Neurosurgery Group, 1010 Wayne Avenue, Suite 420, Silver Spring, MD 20910, USA

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basilar invagination or ventral brainstem compression who meet standard surgical indications should be considered for craniocervical, or atlanto-axial, reduction, stabilization and fusion;

7. That members of this colloquium elect a panel to work with the National Health Service to commission the use of appropriate imaging techniques in subjects suspected of harbouring craniocervical instability, basilar invagination or ventral brainstem compression, to confirm or rule out the diagnosis.

Respectfully,

Rodney Grahame
Irfan Malik
Alan Hakim
Myles Koby
Fraser Henderson Sr.

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Compliance with ethical standards

This manuscript does not contain clinical studies or patient data.

Conflict of interest/competing interests FH Sr. is a consultant for Life Spine Inc., and has developed patents pertaining to the craniocervical junction. All other authors report no conflicts.

Reference

1. Palmer S, Denner E, Riglar M, Scannell H, Webb S, Young G (2020) Quantitative measures of tissue mechanics to detect hypermobile Ehlers-Danlos syndrome and hypermobility syndrome disorders: a systematic review. Clin Rheumatol 39(3):715–725. https://doi.org/10.1007/s10067-020-04939-2