Multiple issues in inherited connective tissues - more than ‘just’ hypermobility

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Background: Mechanisms of pain associated with joint hypermobility are poorly understood and include nociceptive pain from structural joint changes along with soft tissue injuries linked to impaired proprioception; central sensitisation associated with chronic pain and muscle weakness alongside deconditioning. Anxiety and depression are also thought to play a role in patients presenting with pain and hypermobility. We have observed an increase in the rate of orthopaedic surgical procedures undertaken in patients attending the hypermobility clinics compared to those attending the general rheumatology and chronic pain clinics. There is limited published data regarding orthopaedic interventions in patients with hypermobility related disorders especially those with confirmed genetic mutations.

Objectives: We aimed to evaluate the characteristics of patients in our hypermobility cohort focusing on those who had received prior surgical intervention in order to understand the underlying mechanism behind their presentations.

Methods: A retrospective review of medical records was conducted of patients attending a hypermobility clinic at our tertiary referral centre, University College London Hospital, between January 2018 and December 2018.

Results: There were 350 patients (300 females, 50 males) with a mean age of 36 years (range 18-71 years). 63% had a diagnosis of Hypermobility Spectrum Disorder or Hypermobility Syndrome and 37% had a type of Ehlers-Danlos Syndromes (EDS) (hypermobile, classical, vascular or other rare type). 46 patients (13%) had documented genetic mutations. 83 patients (24%) had undergone orthopaedic interventions including 9 who had EDS with confirmed genetic mutations. 54% of patients who had surgical intervention were under the age of 40. The total number of surgical procedures in the cohort was 227 (equating to 0.6485 interventions per patient). Of those requiring operative intervention, the average number of interventions per patient was 2.73. One third of patients had surgery on two or more joint groups, including 8 patients (2%) who had surgery in four or more joint groups. Knees (24%) and hips (23%) were the most common sites for operative intervention with 9% having surgery on their shoulders. 29% of its had significant hypermobility with a Beighton score of 7 and above but there was no correlation between Beighton score and number of surgical procedures. Only 2% of cases were referred from an orthopaedic team thereby excluding a referral bias.

Conclusion: Patients with hypermobility related disorders have a significant number of orthopaedic surgical procedures on multiple sites and at a young age, with indication of mechanical pathology playing an important role in their symptoms. The Beighton score does not appear to be a reliable predictor of surgical intervention. This is not surprising given that the score only covers 5 joint areas and excludes common surgical sites such as the hips and shoulders. Early diagnosis and a holistic non-operative approach combining physiotherapy and chronic pain management is essential to reduce the need for multiple surgical procedures.

References:

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