

Autonomic Dysfunction in Ehlers-Danlos Syndrome

Articulo-autonomic dysplasia is a unifying pathogenic mechanism in Ehlers-Danlos Syndrome (EDS) and related conditions in the clinical pattern termed arthritis-adrenalin disorder. Increased neuromuscular symptoms in females may be related to surrounding muscle support and joint connective tissue in males, leading to decreased male severity. The similar clinical profiles of joint, skeletal and dysautonomia, regardless of age our EDS diagnosis suggests the operation of an articulo-autonomic dysplasia (AAD) cycle, where lax vessels and lower body pooling elicit a sympathetic response and autonomic imbalance, which in turn affect small nerve fibers and enhance connective tissue laxity.

The findings of AAD are more frequent in females, but are paralleled by men. These include undergoing back surgery, slow healing, bladder issues, hernias, valve regurgitation, gallbladder issues, hives and reactive skin, food and medicine intolerance, and hypothyroidism. The greater flexibility and fragility of connective tissue in women is conveyed by measures ranging from historical performance of hyper mobility tricks to physical performance of Beighton maneuvers. Neuromuscular and dysautonomia symptoms more frequent in women include migraines, muscle aches, weakness and atrophy, physically highlighting greater muscular development and support in men as a key factor in their reduced severity. The role of surrounding muscle for joint connective tissue constraint and protection correlates with the benefits of physical therapy and exercise for the treatment of EDS. Problems can include popping joints that may manifest as subluxations, polyarticular and symmetrical joint pain of knees, shoulders and ankles with rare swelling and erythema, joint injuries in mostly ankles and knees, fractures most frequently in distal limbs and disk degeneration or herniation.

Clumsiness from joint laxity, cumulative joint pain injury and skeletal deformations like scoliosis, toeing in or out and flat or high arches make the typical patient uncomfortable with sports and prone to inactivity. Evidence of skin fragility is another hallmark of EDS with easy bruising, unusual scars and early striae. Most unnoticed unless questioned or documented is soft or elastic skin which can be pulled in 1 inch folds from their jaw liner mid-forearm on physical examination. The common findings of migraines and daily headaches which may arise from blood vessel abnormalities like Chiari formation of crania-cervical stability leading to numbness, tingling and muscle aches can prompt fibromyalgia diagnosis. Seizures may actually reflect syncope more than epilepsy and may be related to poor balance.

Bloating, stomach pain and nausea begin early in life and continue later with gall bladder dysfunction and accentuated by mast cell activation disease that presents as eosinophilic esophagitis with frequent food intolerances.

It is common to misdiagnose the anxiety and tachycardia associated with Postural tachycardia syndrome (POTS) as functional disease. The bowel disorders and overlapping joint and autonomic symptoms seen in EDS often are confused with Crohn's and Celiac disease with the various associated psychological aspects. Genomic and immunological studies can help determine if the overlapping joint and autonomic symptoms have separate causes. It is also important to make sure there is not a vitamin D deficiency or hypothyroidism that can present as Hashimoto thyroiditis.

In addition to the chronic fatigue, anxiety-tachycardia and POTS, we see brain fog - poor focus and sleep disturbances. This can be disabling and much more severe in females with the occasional extremely affected male and together with bowel issues, weight loss, hives and

reactive skin and reactive airway disease - shortness of breath. Mild valvular regurgitation, mostly mitral prolapse in both sexes.

Findings related to a Marfanoid habits include an angular build, arm span greater than height and long fingers with consequent maneuvers like making the prayer sign behind the back. Deformations like neck kyphosis, scoliosis and lordosis are much more frequent in females except for pectus and toeing-in mainly or out, a likely contributor to clumsiness.