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Hypermobile Ehlers Danlos Syndrome (hEDS)

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Case Presentation

hEDS presents in 2 forms

1. Asymptomatic form: this form exhibits hypermobility but doesn't have the severity of pain and dysfunction

2. Symptomatic form: this form begins in childhood or early adolescence and progresses in severity of symptoms

3. Phases of disease over lifetime

- Hypermobile phase: presents in childhood. Usually seen with multiple dislocations and generalized gross and fine motor delay
- Pain phase: 20-40 years of age. Headache, fatigue, pelvic pain often diagnosed as fibromyalgia
- Unfolds Phase – joint trauma leads to arthritis, decreased muscle mass, weakness and disability (Trinkle et al., 2017)

Pathological Significance

hEDS is a disease which impacts almost every body system and exhibits varying degrees of significance in each individual case. It is a complex disease with limited but growing understanding of multisystem relatedness and disease process connectivity. Much more work and research needs to be undertaken to understand the complex interworking of this disease origin and course (Tinkle et al., 2017).

Please see Table 1 for specific disease processes and the mechanisms of impact proposed for each one.

Signs & Symptoms

- Pain, muscle weakness, fatigue, limited mobility, spinal abnormalities and osteoarthritis (Trinkle et al., 2017)
- Acute and chronic, nociceptive muscle pain, spasms of tendon, generalized pain, jaw pain, lip numbness, myalgia, joint pain, loss of proprioception, GI, GU, GYN pain, dystonia (Chopra & Tinkle, 2017)

Pathophysiology

- Symptomatic pain hypermobility, joint connective tissue lead to trauma and inflammation to joint articulations (Trinkle et al., 2017)
- See body system subtypes on this table for specific system pathologies
- Limited studies of underlying cause, some evidence of esophageal dysmotility, visceral hypersensitivity (Beckers et al., 2016)
- Low resting blood pressure, limitations and absence of clinical trials, dysmotility, visceral hypersensitivity (Beckers et al., 2016)
- Ligamental structure and poor posture decrease spinal stability, instability and ligament dysfuntion at the atlantoaxial and atlantoaxial joints, peripheral nerve connective tissue degeneration, reduced intradental nerve fiber density, tenascia deficiency, excess CS production and reduced absorption (Henderson et al., 2017)
- Muscle fatigue, lactic acidosis, muscle weakness and increased myalgia (Henderson et al., 2017)
- Pathophysiology poorly understood (Trinkle et al., 2017)
- Hypermobility, lax ligamental structure and poor posture decrease spinal stability, instability and ligament dysfuntion at the atlantoaxial and atlantoaxial joints, peripheral nerve connective tissue degeneration, reduced intradental nerve fiber density, tenascia deficiency, excess CS production and reduced absorption (Henderson et al., 2017)

Nursing Care Implications

- Early recognition and diagnosis in children can have positive impact on interventions and disease impact over the lifetime (Scheper, Nicholoson, Adams, Tofol, & Pacey, 2017)
- Physiotherapy, low dose dyclonine, stress management, hormonal regulation, topical lidocaine, TENS, epidurals are discouraged (Chopra & Tinkle, 2017)
- Anesthesia and maintain optimal nutritional practices
- Neck brace, physical therapy, lumbar puncture, Craniosacral fusion, VP shunts (Henderson et al., 2017)

References

Beckers et al., 2016
Chopra & Tinkle, 2017
Seneviratne, Maitland, & Afrin, 2017

Conclusions

Physical deconditioning can contribute to cycles of weakness and pain exacerbations and disability—promote physical conditioning (Trinkle et al., 2017, pg 2)

Many more tools need to be done in the areas of research and scholarship

Primary care should focus on appropriate diagnosis using the updated diagnostic criteria and treatment should be directed at a multisystem, multidisciplinary approach (Castor et al., 2017)

Management of symptoms varies to differentiate between acute vs. emergent symptoms, prevention of complications, and management of pain (Trinkle et al., 2017).

Significance to Authorship

EDS is estimated to affect nearly 1 in 5,000 individuals with hEDS including 90-95% of those diagnosed. hEDS presentation is found in the primary care setting, but despite this, the disease is poorly understood and underdiagnosed. This author’s current family tree includes the genetic characteristics of hEDS. Personal experience with diagnosis and treatment proved difficult and prolonged as medical providers have a general lack of understanding of this disease and hesitate to treat the resulting complex patient needs. This scholarship is to raise awareness of hEDS to clinicians and improve delivery of care.