

Dr. Fraser Henderson, MD USA (yaaayyy!!!) introduction

Dr. Alan Hakim

- Musculoskeletal concerns at the neck in hypermobility related disorders
 - Joint, soft tissue and neurological concerns related to the head and neck
 - Cannot see these as unconnected from these!
 - Ligament laxity
 - Strength, posture, proprioception impairment
 - Increased range of movement in extension, flexion and rotation
 - Unstable movement
 - TMJ's, shoulder girdle and thoraco-lumbar spine
 - Pain and symptoms can radiate from here
- HSD's have high propensity for chronic headache and neck pain!
- Some individuals present with CCI (CCJ)
- Headache and neck pain – common clinic symptoms
 - Malhotra A, Pace A, Ruiz Maya T, Et al. Study used in presentation
 - 140 patients with hEDS or HSD over 2 years (23% female avg 34, 13-72 year range)
 - 93 (2/3) have headache or neck pain
 - 4/5 have migraine and 1 in 5 have mix of tension or cluster
 - 3/5 of the 140 have cervical spondylosis (wear and tear)
 - 1 in 14 have evidence of instability CCI
 - Of the 140, nearly half had BOTH
 - Over 80% (40% of the total group) have a combination of migraine AND cervical spondylosis
- Global Registry of the EDS society (1334 people)
 - Universal muskulo skeletal pain
 - 90% have headache/migraine
 - 75% have autonomic dysfunction
- Chronic Migraine
 - Gen prev. half to 1.5, most in females, peak in 40's, 7-8% of all migraine
 - OVER represented in EDS and HSD attending clinics (matching study with Hakim and Grahame R. 2004 associated with arthritis and more).
- Cervical Spondylosis – is it over represented or unusual in eds/hsd?
 - Symptoms most commonly reported are:
 - Pain
 - Dizziness
 - Weakness and tingling in arms and legs
 - Headache
 - Vertigo and unsteady gait

- Nausea and vomiting
 - Neck stiffness
 - What is unusual is that CS is as common in EDS and HSD is as common as in the general population but seems to be rising in younger ages 32 to 34 years of age. **Five to ten years younger than the gen pop.**
 - Likely to see cord compression and trapped nerve roots, but this is rare (thankfully he says)
 - X ray demonstration showing a slip forward of the vertebra **listhesis or slip**
 - MRI demo – you can see that the disc is slightly protruded, **THIS IS ENOUGH** to pick up on symptoms from the patient’s history.
- Tension Headache
 - Overall, 10-20% gen pop
 - Frequent 10%
 - Chronic 1%
 - Studies currently suggest that it is NOT over represented in EDS/HSD (Malhotra et al. needs more study done. He sees it more in his clinic, but intuitively you’d think there would be more because of the increased tension and movement issues in Hypermobility)
- Hypermobility Neck
 - Minor instability
 - Moderate Instability
 - Management similar in both. Possibly masked by gen primary headache
 - Severe instability leads to
 - Cervical medullary and other co conditions
 - Cerebral and cerebellar cord signs, radiculopathy
 - No studies looking at instability in the first few groups. They tend to be asymptomatic yet make up the vast majority of patients.
 - **Moderate Instability of interest to me**
- **Cranio Cervical Junction**
 - **All in this region Atlas, Axis, brain stem, upper cord, encephalic vasculature, CSF cisterns**
 - Malformation and malalignment at the CCJ cause pressure or tension changes
 - C1/2 nerve roots
 - And more
- Henderson FC et al, 2016, 2017, 2021
 - The vast majority of patients with these disorders do not have signs of overt cord compression or nerve root compression. Yet!

- Wobble head, head too heavy, neck cracking clicking and unease/foreboding that moving the head causes symptoms
- CMS
 - **Trouble is in Clinic, They overlap with other things!**
 - Headache
 - Neck pain
 - Double vision
 - Vertigo
 - Tinnitus
 - Memory loss
 - Swallowing
 - Breathing
 - Auto dysfunction
 - Numbness in arms and legs
 - Arms and legs feel weak
 - Clumsy, unsteady gait
 - **Imaging very difficult, unreliable**
 - **We tend to treat based on symptoms and signs.**
- **Symptoms worsen or appear only when a person is upright or performs specific movements**
- **Upright and dynamic imaging allows assessment of changes in alignment/angulation on physiological loading and movement, not seen in static and recumbent imaging**
- Types of radiological test
 - Dynamic plain radiographs (x ray)
 - Ext/flexion supine MRI
 - Upright dynamic MRI
 - Clivo-axial angle, Grabb mapstone oaks, harris line
 - 3d rotation CT CCJ
 - Vascular imaging
 - CT angio, MRA/V, doppler US
- All of our knowledge of imaging is in people with symptoms. We don't have enough evidence in people who have hypermobility and EDS but currently with NO symptoms.
- Studies Coming!!! **See slide**
- **Pain management general principles (very few studies showing this in the EDS/HSD pop)**
 - Physical therapies
 - Psychological therapies
 - Drug therapies
 - Procedures – nerve blocks facet injections, multidisciplinary

- General physical therapy principles
 - In hEDS/hSD focusing on joint control
 - Strengthening
 - Posture education
 - Proprioception
 - Mobilization/immobilization
 - Maybe bracing
 - Can draw from whip lash injury evidence
 - Power/strength is impaired
 - Muscle composition is altered
 - Sensation is disturbed/altered
 - Muscles do not activate as quickly – delayed responding
 - Response to strengthening stability of c-spine muscular system
 - Emerging therapy
 - Prolotherapy – inflaming leading to remodeling tissue and might be helpful
 - Stem Cell Therapy – PICL injecting ones own bone marrow derived stem cells into the damaged ligaments **grey literature – client endorsements of this so far. Waiting for literature.**
- Surgery – **the ultimate final way of dealing with instability**

Dr. Henderson’s Presentation

- Case study, 26 year old woman, formerly a vocalist
 - Working on a masters in music, headache, urinary issues, swallowing issues, and much much more.
 - Finally got an MRI showing Chiari malformation Type 1
 - Basilar invagination, atlantoaxial instability, cervical segmental instability
 - Also had tethered cord syndrome
- Ligamentous laxity results in deformation of the nervous system in EDS (reported by “vanderepepe?” years ago.
- Helpful Measurements proposed flexion/extension CT or MRI
 - 1. Clival-axial angle (CXA)
 - Ventral brainstem compression – if Grabb-Oaks measurement is greater than 9mm (very sig. in low lying cerebellar tonsils)
 - Harris Measurement (very important) Abnormal greater than 12mm
 - Vertical Harris Measurement, 12mm or more is pathological

- Normally, the basion pivots over the mid odontoid, translational between cranium and odontoid is pathological, measurement greater than 1mm **there should be NO translation at this junction!**
- All cases of EDS there is chronic Craniocervical instability, translation on imaging of greater than 1mm is pathological
- When do we recommend fusion?
 - Non operative therapy and treatments are exhausted
 - Disabling headache or neck pain
 - Symptoms of the Cervical Medullary syndrome, neurological deficits and measurements are present
- Cervical Medullary syndrome
 - Also, dysarthria, dysphagia, choking, altered breathing, syncope
 - In addition to the usual issues
- Operative Procedure explained
 - Pre-post surgery symptom improvement in
 - Vertigo
 - Dystonias
 - Memory loss
 - Photosensitivity
 - Double vision
 - And more
 - Karnofsky performance score
 - Overall ability to perform daily activities
 - Surgical group had improvement
 - “Would you still choose surgery?”
 - Most strongly agreed.
 - Rarely disagreed.
- Atlantoaxial Instability
 - 86% of RA exhibit atlantoaxial subluxation (AAS) but it is a different ligament in EDS population!
 - We see LATERAL instability sliding of C1 on C2, greater than 3mm slide is abnormal
 - Can use CT and tilt the head right and left to measure the rotation, greater difference of more than 45 degrees is pathological
 - Greater than 80% facet subluxation = unstable
 - In EDS, excessive rotation, twisting of spinal cord and brain stem, kinking of the “ “ artery.
 - Indications for C1C2 fusion
 - Occipital neuralgia
 - Syncope

- Intermittent dysesthesias
 - Stick them with a pin they say “that’s sharp but it doesn’t hurt”
 - More on this slide is very good. Grrrr going too fast
 - *See study by Henderson et al. on atlantoaxial instability.....*
 - After surgery, patients showed no syncope after having it 1-3 times per week, daily living score significant improvement
- EDS almost always has degenerative disc disease and/or Osteoarthritis
 - Cervical instability/ stretch _____
 - Symptoms of high cervical instability may mimic CCI
 - Headache
 - Nausea
 - Poor memory
 - Convergence fatigue, and more
 - After surgery went back to school and got straight A/s
- Headaches due to tethered cord
 - 50% tethered cord pts “pulling down on the back of the head”
 - Headache worsens with bending, leg raising
 - Some tethering in the thoracic causing these
 - Lower spine:
 - Neurogenic bladder
 - Leg pain
 - Leg weakness and sensory loss or pain
 - May be radiologically occult (not visible)
- Vascular problems
 - Not only vascular EDS but classic, classic type, and kyphoscoliotic EDS
 - Loss of blood flow to the brain from vertebral artery kinking
 - Occlusion of veins draining the brain – removing the clot woke her up from coma
 - Sinus thromboses – used CT angiogram to diagnose
 - Vascular congestion – causing headaches and potential for increased bleeding

Whew. The end.

Q and A with Dr. Hakim (Henderson not online)

- Who do I speak to get diagnose and treatment, doctors I talk to think I’m making this up.
 - Ask questions about a specific symptom, why could this be happening here, very specific, BEFORE bringing up personal suspicions about the cause.
- Would CCI problems come from neck down to fingers, or reverse? And more questions
 - What could possibly happen to different types of nerves as they come out from different levels of the cervical-thoracic region basically.

- Nerves down the arm come from c5-8, wouldn't expect them to be directly involved in the CCJ. CAN be involved because of pinching and wear and tear in the neck that are felt with sensory changes all the way down the arm.
- Caveat with thoracic outlet syndrome which can be nerve or vascular –
 - Pain across the shoulder girdle, causing posture changes, causing pain and nerve issues down the arms.
- Lower cranial nerves – supplying face, tongue, swallowing, etc. absolutely effected at the CC junction instability. Check for something that is pressing or pulling on those nerves and look at the vascular supply in those regions.
- Henderson's Response –
 - Simple compression of the medulla and upper spinal cord can cause symptoms
 - Can be excessive rotation or translation at that level that leads to STRETCHING of the nerves causing problems (as opposed to compression!!!)
 - Restriction of blood flow to the brain stem
 - Vertebral artery can be compressed as well, that should be checked
 - You can have pressure from c4 to c5 that affect the face of the trigeminal nerve can affect the lower portion of that nerve nucleus and that causes sensory loss on the outside of your face. Very common, anatomically understood.
- Henderson – consider obstruction of CSF flow, and Endocrine issues
- Hakim – fascial release for management of flow and these issues
 - Whether that is doing anything decreasing intracranial pressure I don't know
 - But therapists who work on this talk about improving lymphatic flow.
- I've had symptoms of CSF and tonsillar hernia not on MRI. What do I do next?
 - MRI and CT Can be very misleading. Look like overcrowding, more space than really is because of the flare of spinal fluid.
 - 1, if you're looking at Chiari 1, look at T1 weighted imaging where CSF is dark. That shows the true level of the tonsils.
 - 2, an upright image will often show lower tonsil position than supine. Upright flex/ext is better.
 - He suspects that in EDS, fluid can leak through the nose. But it hasn't yet been proved.
- How do you, Gricelda, advocate for the care you need when people don't believe you.
 - Present symptoms first, not a diagnosis.
- Surgery is
 - Surgery is last option.
 - Avoid activities that trigger events.

- Wear a brace constantly for 1-2 weeks to let things calm down and realign, so you have to do isometric exercises for a few minutes several times a day to keep neck strength when doing this.
- Postural position is HUGE. Focusing on this for years can gradually rebuild and reposition the things causing issues.