Neurological Complications in EDS/HSD Sunday September 26, 2021

Management of Headache

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Primary refractory headaches, neurostimulation, headache education.

- Headache ICHD – III Criteria is what she uses for diagnosis
- Many types on the slide (wow)
- Migraine –
    - A polyphasic brain disorder with many aspects where the headache itself is only one part.
    - Intercal phase, premonitory, aura – numbness, tingling nausea, headache, postdrome, intercal phase
    - Pathophysiology – still largely unknown
        - Abnormal cortical activity, abnormal brain stem function, cortical spreading depression, activation of the trigeminal nerve – headache pain results.
        - Central sensitization and vasovagal responses
    - Management:
        - Education
        - Lifestyle mod and man
        - Pharma
        - Psychological
        - Surgical
    - Pharmacological treatment Acute/abortive or long term preventative
        - Acute/abortive – nsaids, paracetamol, triptans (can be contraindicated in patients with cardio issues), ditans, gepants, neuromodulators
        - Long term – usually takes 3 months to try to see if they work
            - Old school
            - CGRP receptor antagonists and monoclonal antibodies, nerve stimulation treatment
        - Opioids can make headaches much worse
    - 75% with EDS/HSD have migraine compared to control (43%)
    - Presentation is more florid, more frequent, more disabling
    - Medication overuse headache likely a contributor in patients suffering from recurrent musculoskeletal pain
    - Preventative: AVOID antihypertensives (beta blockers/candesartan)
    - Acute treatments: medication overuse needs to be addressed.

- Tension Type Headache (TTH
    - Bi lateral, not normally with the features of migraine like aura and light sensitivity
    - Acupuncture and cog behavioral therapy are non pharmacological treatments.
- New Daily Persistent Headache
    - One this day, of this month, at this time, I developed headache and it has never gone away.
    - Causes – primary (30%) secondary (70%), very rare comparatively
        - Life changes, CSF pressure disorders, Infections, Vascular disorders, stressful life event
    - Management: depends on the clinical phenotype (features present)
- Why are primary headache disorders over represented in HSDs?
    - Sleep disorders (67%), Anxiety disorders (32%), MCAS (24-66%), medication overuse,
    - Inflammation (cytokines), Abnormal blood vessel function, Autonomic Dysfunction, Altered central pathways (lowered threshold for pain with fibromyalgia)
    - "We know that Histamine is a potent trigger for headaches" direct quote.
- Headache is not only painful, it is disabling.
    - WHO study –
        - Migraine 6th highest cause of worldwide of years lost
        - Headache disorders third highest overall
    - In the US and UK only half of those identified with migraine have seen a doctor in the last year, only 2/3 are getting correctly diagnosed and of those they are solely relying on over the counter medications
- Barriers to effective care
    - LACK OF KNOWLEDGE AMONG HEALTHCARE PROVIDERS
        - Undergrad med education not trained
        - Worldwide only 40% of those with migraine or TTH are professionally diagnosed
    - POOR AWARENESS IN THE GENERAL PUBLIC
        - Not perceived by the public as serious because most are episodic, don't cause death and are not contagious
    - COST FOR TREATMENT
        - Not actually that high.
        - Better awareness would lead to better treatment.
        - More research is needed to better identify sub pathogenesis in those on the hypermobility spectrum with headache.

Dr. Manjit Matharu – Orthostatic Headaches

- Occurs when sitting down or standing up. Headache improves significantly when the person lies down. Also positional and postural. Not a diagnosis but a SYMPTOM
- Causes
    - **CSF Leak**
    - **POTS**
    - Cervicogenic headache – report that any neck position leads to pain in any position
    - Idiopathic
- CSF Leak
    - CSF cushions the brain and spinal cord with meninges
    - When meninges are damaged it can allow CSF to leak into the body lowering the volume and pressure
    - These changes cause the pressure in the brain to drop which stretches the pain in sensitive structures – the brain sinks within the skull
    - Orthostatic headaches develop when a person is upright because a lot of circulating CSF surrounds the spinal cord, laying down allows it to return to normal
    - Can be traumatic, spontaneous intercranial hypotension, iatrogenic: caused by medical procedure
- Spontaneous Intercranial Hypotension
    - Community prevalence of 1 in 50,000 but he suspects it is much higher
    - Highly disabling, severe headache as soon as they are upright
    - Various diagnostic criteria has ben proposed – contributes to uncertainty and reliability in diagnosing and treating patients.
    - Treatment pathways outlined in literature – validity largely uncertain
    - First Comprehensive systematic review in literature published recently
        - Results published earlier this year see: D'antona L, et al. Clinical presentation, investigation findings and treatment outcomes of spontaneous intercranial hypotension
- Clinical Presentation
    - Mean ache 42 (2 – 88 years)
    - More common in Female
    - Risk factors
        - Connective tissue disorders 37%
        - Spinal pathology (43%)
        - Bariatric surgery 3%
        - Mean Duration of symptoms at 31 days on the day of presentation
        - headache 97%, orthostatic in 92, non-ortho in 8
            - 33% pain in the back of the head
            - 30% diffuse

            - 21% frontal
            - 11% fronto- occipital
            - 8% temples
        - Many patients reported hearing disturbances! Muffled sounds, vertigo, tinnitus. Can be harmful to making diagnosis because it confuses doctors.
- Diagnosis
    - Rule out a CSF Leak –
        - MRI
        - Lumbar Puncture
            - Take a sample and measure pressure
        - Myelography
            - Lumbar puncture to inject dye and use CT or MRI or special x rays to SEE where the leak is
        - Tilt table test
            - Measures the HR and BP and observing symptoms to observe the onset of headache and diagnose POTS
- Investigations in the systematic review
    - Brain MRI
        - Sagging -
        - Pituitary gland larger than normal
        - Large blood vessels
        - Pachymeningeal enhancement
        - Subdural collection
        - Completely NORMAL – a normal MRI cannot definitely exclude CSF leak
    - Spinal neuroimaging – can diagnose in half of patients
- Treatments:
    - Conservative – make lifestyle changes
    - Medication -
    - Invasive
        - Epidural Blood patches
        - Surgery
- Headache in POTS
    - HR increment greater than 30 within 10 minutes of head up tilt or standing (greater than 40 if 12-19).
    - Presentation
        - Light headed
        - Muffled hearing
        - Tremulous
        - Nausea

        - Sweating
        - Anxiety
        - Syncope
        - Headache
        - Pallor
        - Tiredness or weakness
    - Global study suspected half of pots patients have headache. More specific questioning by physicians of POTS patients revealed that ALL of them had headaches.
- Most common headaches are migraine or orthostatic headache
- For Hypermobile patients:
    - POTS is usually treated before the headache which creates problems because the medication can exacerbate the migraine.
    - Diagnostic efforts for CSF leaks are more invasive and therefore avoided more frequently.
    - Must increase fluids more completely
    - Some patients still have orthostatic headaches even with treatment of POTS, we don't understand why (**I personally suspect mast cell activation and histamine control issues in the brain that is untreated, based on collective research published this year).**

Q and A segment

- What do you recommend is the best way to find a pattern for triggers
    - May be no trigger
    - Migraine – diet is at the TOP!!! Is that the trigger, or part of the attack? Before a person gets an attack they go through a premonitory phase with a craving for food. They eat very specific things like chocolate or cheese. You get the craving, you eat the food and then get pain and assume that the food triggered the attack. The pain and attack actually started much sooner.
    - Hunger, sleep deprivation, not enough exercise menstruation, but in the vast majority of patients the attacks are spontaneous.
- Are some opioids more likely to trigger mast cells and histamine release than others?
    - Not sure whether opioids trigger mast cells or why, we know that they exacerbate headaches. We know that long term use increases headaches. Shouldn't be used for management of headaches.
- How are hormones involved in migraine in EDS?
    - Not any specific relationship with the hypermobility and hormones, but hormones in general are triggers. Very individually based treatment approaches.
- How common is the link between idiopathic IH tension and ED syndromes?

    - My guess is that it's slightly more common I think. I see a lot of CSF leaks. There seems to be an association, yes, but not studied.
- Description of ear involvement and question about how that was a part of the migraine
    - We cannot comment on individual patients and experiences. See an ENT to check for ear problems, but pressure and inner ear issues are frequent in migraine
- What is the average duration? Can you have autonomic dysfunction and CSF leak
    - Don't put off evaluation for CSF leak even if bed rest seems to improve the issue. The longer they go
- My oxygen therapy question
    - 80% response rate for **cluster headache** with hyperbaric oxygen therapy
    - In migraine and other primary headaches the response has been negative, they tried it and had no response. Oxygen therapy in primary headaches has a largely negative response reported by the clinicians who have tried it, even though it has not been largely studied.
- Alternative medications and supplements
    - Lifestyle is very important
    - Mindfulness techniques are
    - Riboflavin, and other supplements can be affective
    - Then move on to preventatives if they wish
- How do you tell the difference between OH and Migraine in order to know how to treat it?
    - Generally not a problem. Migraine improves with laying down but improvement is not dramatic and pain is still significant. They go away and reduce light and movement.
    - Orthostatic Headache the change is very dramatic when lying down
- How do we as patients advocate away from functional neurological disorder diagnoses to better care and treatment
    - In spite of HUGE amounts of literature showing that headaches are NOT psychogenic, physicians can still assume this. The literature and CGRP responses have been significant.
    - If a doc says "why don't you see a psychiatrist" you know they haven't gotten the picture yet. Go see someone else.
    - Study has identified 4 genes for cluster headaches
    - Just starting to demonstrate that CSF leaks are more common and being studied now, very important for hypermobility.
    - As we continue, it's not going to be long for the unbelievers to not be able to hold up their views that the pain is made up.

(With the mast cell question, it's interesting how SURE they are about not over "evoking" of mast cells in headache whereas Dr.s Maitland and Kwan see this as very involved and needing consideration).