

Ehlers-Danlos Syndrome

In March 2017 world experts on all the types of EDS came together to agree on diagnosis, current understanding of surveillance, management, and research priorities for EDS. This information was published in the American Journal of Medical Genetics (AJMG). All of the articles in the supplement can be found through the Ehlers-Danlos Society at <http://onlinelibrary.wiley.com/doi/10.1002/ajmg.c.v175.1/issuetoc>

The Ehlers-Danlos Society and AJMG hope that primary care providers and specialists will use this information for diagnosis and treatment. This is considered the new standard for diagnosis.

One of the changes that happened was to divide the diagnosis of Ehlers-Danlos hypermobility type into two diagnoses. One diagnosis is hypermobility EDS (hEDS). The other diagnosis is Hypermobility Spectrum Disorder (HSD). The reason for this change is primarily related to research. One person in a family might have an hEDS diagnosis while another has a diagnosis of HSD. One diagnosis is not better or worse than the other. Both family members can have the same problems. We will use hypermobility disorders (HD) to mean both hEDS and HSD because **the same information applies to both diagnoses**.

Education and genetic counseling was provided to the family. We reviewed the signs and symptoms of hypermobility Ehlers-Danlos syndrome (hEDS), and Hypermobility Spectrum Disorder (HSD) including surveillance, management, treatment and prognosis. It should be noted that there are several kinds of EDS. There is no laboratory genetic test for hEDS or HSD. Diagnosis of HDs is based upon clinical findings and upon the family history. Based on Kimberly's medical history and physical exam, she **has hypermobility type EDS**

Some people believe they have "some classical EDS and hEDS" or "some vascular EDS and hEDS". Unless their parents each have a different type of EDS, there is no overlap. However, different family members with HDs can have different types of problems.

The diagnosis of hEDS or HSD often explains why a person has so many apparently unrelated health issues. However, there are no different treatments just because the person has a diagnosis of a HD. Below we cover common problems. Just because a person has hEDS or HSD does not mean they will have all of the problems. It should also be noted that a "functional disorder" means that we cannot find an obvious medical cause. It does not mean "all in one's head."

MUSCLES, BONES AND JOINTS:

<http://onlinelibrary.wiley.com/doi/10.1002/ajmg.c.31545/full>

<http://onlinelibrary.wiley.com/doi/10.1002/ajmg.c.31551/full>

The hypermobility disorders are characterized by generalized joint laxity (double jointed), repeated joint dislocations and sprains, spontaneous joint subluxation and joint pain. There is a natural history of the joint problems associated with HDS. Children may be hypermobile and clumsy but have little joint pain or joint pain mostly in their legs often called "growing pains." Handwriting may be a problem. Females often become more flexible after puberty and males are more likely to get stiffer. Pain may start to develop and may be diagnosed as fibromyalgia. Recurrent subluxations/dislocations or pain often develop in the shoulder, knee, and Iliotibial band syndrome (sometimes called "snapping hip" syndrome) is also common, and is frequently perceived by the patient as hip instability although it is actually not dislocating.

Adults may not be as flexible as they were in childhood but do experience chronic joint pain. Males generally have fewer problems with joint instability than females, possibly due to greater muscle mass. In general, increased muscle mass seems to reduce joint instability.

Proprioception, or the ability to know where body parts are, is also affected in HDS.

Joint pain is often worse after exercise. However, improving muscle strength and tone may help with this.

Sometimes surgery can be helpful. However, sometimes a joint is so lax that surgery is not likely to help. Orthopedic surgery should be done by a surgeon familiar with HDS. Common problems such as anterior cruciate ligament and meniscal tears in the knee, labral and rotator cuff tears in the shoulder, wrist instability, thumb joint subluxation, labral tears in the hip, and lateral ankle ligament tears should be treated in the same way as any other patient. If surgery is done, discuss the possible use of a donor (cadaver) tendon for better stability. Yours may just stretch out again over time.

Thoracic outlet symptoms are common in HDs. This can cause arm and hand pain. PT is essential. Botox may help.

Treatment for joint and muscle pain:

- It may be wise to avoid contact sports.
- Physical therapy and daily exercises can help improve balance, muscle strength and tone. This decreases joint pain and improves function and self-esteem. Exercise should be with light weights, multiple repetitions and in the mid range. Exercise should include core.
- Hydrotherapy can be helpful.
- People with flat feet might benefit from orthotics or shoes with good arch supports.
- Splints or braces may help some people. Finger splints may help with hand pain
- Hip pain with pain “clunking” as the iliotibial band subluxes over the greater trochanter is not a hip dislocation although it may feel like it. PT helps.
- Bunions are common. Unless they are causing problems, let them be.
- In general a graduated or slowly increasing exercise plan is recommended
- Avoid inactivity. Deconditioning can worsen pain.
- Some children may need a modified gym class. They may benefit from working on their home exercise program if the gym activity is likely to cause problems.
- Weight training should be with light weights with multiple reps in the midrange.
- Exercise does help but it may take several weeks to notice a difference and you will likely have more pain as your body gets used to the new exercises. Tylenol, ibuprofen, ice, heat and topical creams can help.
- Sit/Stand work stations may help adults who develop pain with prolonged sitting or standing.
- Monitor for and treat osteoporosis. Remember that prolonged use of PPIs for reflux increases the risk of vitamin D and calcium deficiency
- While discectomy might help, often the next disc begins to have more issues.
- If an activity causes more pain or dislocation, it should be modified or avoided. It is important to not overdo the exercise or therapy to avoid exacerbating the problem. However, a consistent program is critical.

CARDIOVASCULAR AND AUTONOMIC DYSFUNCTION

<http://onlinelibrary.wiley.com/doi/10.1002/ajmg.c.31543/epdf>

<http://www.prohealth.com/library/showarticle.cfm?LIBID=15872>

Autonomic dysfunction seems to be fairly common in HDs. The autonomic nervous system is part of our “fight or flight” mechanism. People with HDs seem to have problems with this. It can cause:

- Fast heart rate (palpitations),
- light-headedness, sometimes with a sense of being about to blackout (pre-syncope),
- visual impairment including altered acuity, partial or complete visual loss, “seeing stars”, tunnel vision and light sensitivity
- cognitive complaints including word-finding difficulties, limited concentration and poor memory (often described in lay terms as “brain fog”),
- chest pain,
- tremulousness,/shakiness
- chronic fatigue,
- exercise intolerance and post exercise malaise,
- swelling and/or discoloration (dusky purple/red) in the legs after standing for only relatively short (e.g., 5 min) periods of time
- peripheral vasoconstriction (cold, dusky hands, and feet),
- fainting (syncope),
- temperature dysregulation, (heat or cold intolerance/hot flashes, feeling cold)
- sleep disturbance.

Sometimes autonomic dysfunction causes postural hypotension and sometimes POTS (postural orthostatic tachycardia syndrome). They can both be called “Orthostatic Intolerance” or OI. The treatment is the same for either one.

Treatment for cardiac/ autonomic dysfunction

- If echocardiograms are normal once in childhood and once as an adult, no further echoes are needed unless problems arise
- For autonomic dysfunction
 - Drink more fluids (64 oz daily)
 - Eat more salt (2 grams daily)

- Zero calorie sports drinks, Propel, pickle juice, etc are used by many people
- Exercise—recumbent bikes, rowing machines, and swimming may increase muscle tone without causing as much dizziness. Dynamic exercise may help more than isometric. Target 75% of estimated maximal heart rate for 30 minutes 2-3 times a week. Include a warm up and cool down
- Low dose propranolol, Flonase and Midodrine may be used. Other medications include Ivabradine, stimulants such as Ritalin, hormonal contraceptives, pyridostigmine, clonidine.
- Stand up slowly
- Small frequent meals
- Compression stockings
- Identify and avoid triggers such as heat, alcohol, caffeine, long periods of rest, travel

A few people with HDs have mitral valve prolapse. For most people this is not clinically significant.

Some people with HDs have aortic root dilatation, either borderline or mild. Aortic root dilatation is more common in the 6-12 year old age group. Usually the dilatation is not progressive and treatment is not recommended. Most children grow into their aortic size by high school. If we have a normal echocardiogram in childhood and in adulthood and no new problems arise, no further follow up is needed.

PSYCHOLOGICAL ASPECTS <http://onlinelibrary.wiley.com/doi/10.1002/ajmg.c.31544/full>
http://www.cci.health.wa.gov.au/resources/infopax.cfm?Info_ID=53

There is an association between HDs and anxiety, panic disorders, and mood disorders. Obviously not everyone will have these issues but the risk is increased. The rates of pain, GI issues, autonomic problems and sleep problems are greater as well and may explain the increased risk. It is also possible the issues with the connective tissues and body reactions also lead to increased ability to feel pain. It is important to consider the body itself may be playing a role in what looks like a psychiatric problem.

Management of Psychological/Psychiatric problems

- Cognitive therapies such as cognitive behavioral therapy and mindfulness (some may benefit from a therapist, others may do well with books, online options or apps.)
- Improve knowledge about HDs Remember other people in support groups may have entirely different problems and reactions to treatments or medications. Do not assume bad outcomes because others have had one.
- Physical activity can improve mood
- Psychiatrist if mood disorders are also present

DENTAL AND MANDIBULAR

<http://onlinelibrary.wiley.com/doi/10.1002/ajmg.c.31541/full>

Dental problems such as multiple caries, enamel problems and gingival bleeding may occur. Sealants should be considered for children with HDs.

Temporomandibular joint (TMJ) pain is also common. Not only can the TMJ come out of joint, it can cause headaches and jaw pain. Gum chewing, nail biting, and clenching can all make this worse. If the jaw is really flexible it can cause sleep disordered breathing (similar to sleep apnea). A sleep study may be indicated.

Delayed onset and/or resistance to local anesthesia is a frequent complaint. Make sure to discuss your diagnosis of HDs with your dentist prior to any oral procedures. Do not open the jaw extremely wide and rest frequently with the jaw closed during cleanings or procedures.

Many people have a high arched palate and need braces. Teeth often move more quickly than usual. This means that use of the retainer is essential because the teeth will rapidly move back to their previous positions.

Treatment for dental problems

- Be sure your dentist knows and understands HDs. The link to the HDs dental article is listed above.
- Avoid having your mouth open for long periods during dental work

- Consider sealants
- For TMJ pain
 - Dentists can make mouth guards to help with clenching
 - Some physical therapists help with exercise programs
 - Avoid chewing on gum, ice, fingernails and other tough foods
 - Topical gels may be helpful
 - Try deep heat 10 minutes of a heating pad, 3 minutes of cold and 10 minutes of warm
 - Cold laser may help
- Use a soft toothbrush—consider electric
- Have your dental hygienist demonstrate appropriate flossing

BLADDER AND GYNECOLOGY

Bladder, rectal or uterine prolapse are found in many women who have EDS. Heavy menstrual periods, menstrual cramping and dysfunctional bleeding are also common. Some women find it helps to take an oral contraceptive, and some stop periods completely by taking the pill daily and not taking the placebo pills. Many women with HDs have liked the IUDs such as Mirena.

Bladder problems are quite common in women with the hypermobility form of HDs. Women might have stress incontinence (accidents while laughing, coughing or sneezing). They can also have urge incontinence (having a sudden urge to go and not making it to the toilet.) Often the problem is a prolapsed bladder. Kegel exercises can help strengthen the perineal muscles. Scheduled toileting can be helpful.. Go every 1-2 hours even if you don't feel the need to go. Double voiding can also help. Sit down and urinate. Stand up, then sit down and try going again. Pelvic physical therapy can also be helpful. Sometimes a gynecologist can fit a pessary to help hold the bladder in place. Sometimes surgery is needed.

Women can develop varicose veins around the uterus and ovaries. This is called pelvic congestion syndrome and can also cause abdominal pain.

Pregnancy and childbirth are not typically problems for women with HDs About 1/3 of woman with hypermobility syndromes will have rapid (less than 4 hours) labors. There may be increased bleeding after childbirth but not usually enough to require a transfusion. A hip support may be helpful.

Treatment for bladder problems and gynecologic issues:

- Kegel exercises
- Scheduled toileting
- Avoid constipation
- Double voiding
- Pelvic PT
- Ibuprofen 800 mg three times a day can help decrease bleeding and pain in adults
- Gynecologists have a better understanding of the effects of contraceptives and may be the best option to manage problems with periods.

GASTROINTESTINAL

<http://onlinelibrary.wiley.com/doi/10.1002/ajmg.c.31546/epdf>

Constipation, irritable bowel syndrome, gastroparesis, delayed gastric emptying are frequently found in individuals with HDs. Eosinophilic esophagitis and SIBO (small bowel intestinal overgrowth) may also be more common. Lactose, fructose intolerance and gluten intolerance may be found as well. Gastroesophageal reflux is also very common. Reflux may be treated with over the counter proton-pump inhibitors (PPI). If you benefit from a PPI, you should try to switch to Pepcid or Zantac after a couple months. Long term use of PPIs can decrease vitamin D and bone density. A GI consult may be necessary for treatment of delayed gastric emptying, eosinophilic esophagitis, irritable bowel or diagnosing food intolerance. Miralax, Benefiber, Milk of Magnesia or fiber supplements may be needed on a daily basis to maintain soft stools and adequate bowel emptying. Many people have tried Milk of Magnesia (MOM or Miralax (PEG) and reported they did not work. Often the doses were not big enough. People with significant constipation might want to try 1 capful of MOM or PEG 3 times a day for 3 days then continue with that dose daily or twice a day to keep the stool soft and regular. Scheduled toileting is important as well. Take advantage of your body's increase in bowel activity after meals and sit and give at least 10 pushes after every meal. Children may need a footstool to keep their balance.

Treating GI symptoms:

- Consider GI consult for irritable bowel, concerns for eosinophilic esophagitis, SIBO, lactose, fructose intolerance or celiac. (There is some evidence that celiac may be more common with EDS)
- The book Reclaim Your Life from IBS can be helpful.
- Control constipation. Milk of magnesia is more effective than Miralax and much cheaper, however many people do not like the flavor.
- Try PPIs (above) for reflux but after 2 months try switching to Zantac or Pepcid. Weight loss and staying upright for a couple hours after meals can help as well. Try elevating the head of the bed on blocks.
- Exercise and adequate fluids help with constipation.
- A low FODMAP diet can be considered. Most hospitals will have a nutritionist, some grocery stores do, and there is a lot of information on low FODMAP diets on line.

Nutrition

- Avoid concentrated sugars and artificial sweeteners
- Consider avoiding high fructose corn syrup unless tested negative
- Try to add probiotics—pickles, sauerkraut, Kefir, active culture yogurt,
- Try to get 8 servings of a variety of fruits and vegetables daily
- Try to eat whole foods, not processed foods.
- Liberalize salt
- Consider adding magnesium (chelated, 400-800 daily), B6 (50 daily), D3 (cholecalciferol 1000-5000 unit daily, monitor twice yearly), C (500 twice daily), B12 (100% USDA) , zinc (100% USDA), iron (100% USDA, l-lysine
- Spices such as turmeric, crushed pepper, rosemary may be helpful

SLEEP and CHRONIC FATIGUE

<http://onlinelibrary.wiley.com/doi/10.1002/ajmg.c.31542/epdf>

Many people with HDs have problems with sleep. Memory foam mattresses or mattress toppers and sleep number beds are popular. Unfortunately sleeping pills are usually not very effective if used on a regular basis. Cognitive therapy such as mindfulness and CBT can help. There may also be an increased risk of sleep apnea or sleep disordered breathing. Good sleep hygiene is essential. Try to maintain a regular schedule. Try to go to bed at about the same time every day. It is even more important to try to get up at about the same time every day. This helps set your body clock. Don't take naps during the day. This can keep you from sleeping at night. Try to exercise daily. This is best late in the afternoon. Avoid vigorous exercise just before bedtime. Spend some time on relaxation daily. Do not lay in bed and think about problems. Don't try to go to bed after some stimulating or stressful activity. A light snack in the evening may be helpful, but do not overeat as this may cause heartburn. Try to make your bedroom dark and quiet. People generally sleep better if the temperature is slightly cool. Have a routine for right before bed. A warm bath or shower can help people relax and feel sleepy. Don't drink things with caffeine after about 4-6 pm. Don't drink alcohol right before bedtime. You might go to sleep, but you won't sleep well. Don't smoke in bed or right before bed. Don't do anything in bed but sleep. If you watch TV, read, play computer games, or talk on the phone, you don't think of your bed as just a place for sleeping. Start turning off electronics about an hour before bedtime. The light they make makes it hard for the brain to make melatonin and that can make it hard to get to sleep if you don't feel sleepy, don't go to bed. If you have been awake more than about 15 minutes, get up and do something boring or quiet. TV and video games are stimulating--avoid them. The book [Take Charge of Your Child's Sleep: The All-in-One Resource for Solving Sleep Problems in Kids and Teens](#) by Judith A. Owens and Jodi A. Mindell might be helpful even for adults because it does explain sleep. Melatonin is an over the counter sleep aid. It works best with lower doses (1.5-5 mg) and taken from 1-3 hours before bedtime. You may need to experiment because different biorhythms affect the timing.

Chronic fatigue or myalgic encephalitis can be associated with HDs and with autonomic dysfunction.

Managing sleep:

- Exercise (physical deconditioning can cause fatigue during the day.) Be aware that everyone can be sore after starting exercise.
- Use good sleep hygiene. If you can't get to sleep or get back to sleep get up for a while
- Avoid caffeine within 6 hours of bedtime
- Getting up at the same time is as important as getting to bed. Check your mobile phone for sleep tools
- Avoid daytime naps
- No TV in the bedroom. No electronics within an hour of bed or if you have to get up at night.
- Try to avoid consistent use of sleep medicine.

- Consider a sleep study, especially if there is significant snoring, morning headaches, waking gasping for air at night. Having to get up several times at night to urinate can be a sign of sleep apnea.
- Control autonomic dysfunction—exercise, fluids and salt, and medicine if needed.
- Rule out medical problems such as chronic infections such as hepatitis, Lyme disease, endocrine, autoimmune, cardiorespiratory or neurological problems
- Cognitive behavioral therapy, mindfulness, progressive muscle relaxation, yoga, visualization
- Can try co-enzyme Q10, carnitine, alpha-lipoic acid, magnesium, NADH and vitamins.
- Consider avoiding cow's milk protein
- Melatonin can help people get to sleep. Smaller doses (1.5-5mg) work better than larger doses. Take it 1-3 hours before bedtime—experiment.
- Pace yourself. AVOID PROLONGED BED REST—you will keep getting worse. Exercise. You may need to start low but keep increasing the amount that you are doing until you meet your therapist's goals.
- In general, avoid stimulants, although some people will do better on meds like Concerta or Vyvanse.

MAST CELL DISORDERS

<http://onlinelibrary.wiley.com/doi/10.1002/ajmg.c.31555/epdf>

Mast cells play a role in allergic disorders. Recently they have been found to play a part in headaches, irritable bowel, gluten intolerances, osteoporosis, autoimmune problems, neuropsychiatric problems and interstitial cystitis. There may be a link between HDs and problems with overactive mast cell response. At this time, there is no clear criteria for diagnosing mast cell disorders.

Treatment of Mast Cell disorders

- Cognitive therapy such as cognitive behavioral therapy as stress may be a trigger
- Allergy meds such as Claritin, Allegra, or Zyrtec,
 - May need to take more often: 2-3 times a day
 - Singulair.
 - Ranitidine (Zantac) and famotidine (Pepcid) may help GI symptoms
 - It may take a few weeks to see a difference.
- Try to identify triggers. Common triggers include alcohol, heat, some drugs, IV contrast, bee and wasp stings, fever or infections, exercise, and stress.
 - Narcotics can be a trigger
- Exercise but avoid overexertion. Regular exercise can help
- Can try co-enzyme Q10, carnitine, alpha-lipoic acid, magnesium, NADH and vitamins (similar to chronic fatigue), quercetin

Some people with multiple drug allergies may not be allergic to the drug itself but to the other inactive ingredients found in the drug.

- Allergists are typically making the diagnosis and treating it, but it should be noted that the symptoms overlap with other kinds of problems and criteria for diagnosis are still being debated.

NEUROLOGICAL

<http://onlinelibrary.wiley.com/doi/10.1002/ajmg.c.31549/epdf>

Chiari malformations may be more common with HDs. However, they are not all that common. A Chiari malformation occurs when a part of the back of the brain extends into the spinal cord. This can cause headaches, especially in the back of the head. Headaches may be worse with cough or strain. There can be dizziness and problems with coordination and balance. Sometimes surgery is needed, sometimes it is not. The cervical spine may be too flexible. This is called atlanto axial instability when the laxity occurs where the head meets the spine. Again, this is rare. Neck pain and headaches are the most common sign—but of course headaches are common anyway. Headaches in the neck and back of the head are *very* common among people spending a lot of time on electronic devices. Fainting, nausea, problems swallowing and breathing problems may be more common. Usually there will be neurological problems such as hyperreflexia or hypoesthesia. Sometimes surgery is needed, but usually a neck brace is adequate.

There is some evidence that constantly overstretching nerves may cause nerve damage. This may cause a variety of problems that are seen in HDs. At this point more research is needed to decide what is happening and how to treat this.

There might be higher rates of dyslexia and dyspraxia in those with HDs Likewise degenerative disc disease (arthritis, herniated discs and spinal stenosis) are more common. It should be noted that MRI and CT scan findings do not correlate well with clinical and

surgical outcomes. A bulging disc on MRI does not mean that surgery is needed. Bulging disks are common even in people who do not have HDs.

There is increasing awareness of small fiber neuropathy. This may explain numbness in hands and feet. In addition to carpal tunnel, cubital tunnel (the funny bone nerve) may occur. Stretching the nerve, such as bending the arm and wrist while sleeping at night or holding the arm bent to look at electronic devices can stretch the nerve and cause numbness. It may even contribute to chronic fatigue.

Tarlov cysts are perineurial cysts that can press on nerves. They can be found accidentally on x-rays. Sometimes but not all the time they can cause pain or neurological problems. Those with Tarlov cysts that are actually causing problems may need to have them removed.

Treatment of neurological problems:

- Physical therapy can help strengthen muscles and improve alignment
- Good posture
- Avoid overstretching
- Physical therapy, tai chi, graded exercise, yoga (avoiding over stretching)
- Referral to Neurosurgery if there are neurological symptoms
- Cognitive behavioral therapy and mindfulness

OTHER:

- While surgery to correct joint problems may be needed, often surgery to tighten ligaments fails because they simply stretch again
- Surgery in general tends not to be a problem although additional sutures may be needed and they may need to be in for a few extra days.
- Monitor for and treat vitamin D deficiency, vitamin B12 deficiency, folate deficiency, iron deficiency
- Easy bruising and poor wound healing are common
 - Can try vitamin C 500 mg three times daily

PAIN:

<http://onlinelibrary.wiley.com/doi/10.1002/ajmg.c.31554/full>

<http://www.tcapp.org/>

<http://www.chronicpainpartners.com/>

Pain is a warning to our bodies that there may be a problem. Pain in EDS is often out of proportion to anything found on physical examination. Amplified Pain Syndrome (AMPS) seems to be fairly common with hEDS. Although pain is intended to be a warning, with AMPS the pain is out of proportion to the problem. This is a problem with the central nervous system. Physical therapy and cognitive therapy can help rewire the brain and spinal cord and decrease the “volume” of pain. While it is important to remember that pain is what a person says it is, it is also important to understand that with AMPS, there is no damage happening to the body. There is no pill that takes away pain. Pain management requires a variety of approaches including physical therapy and cognitive therapy.

Pain in EDS may be related to overextending joints, nerve pain from traction or compression, decreased proprioception, and there may be other problems we have not yet identified. Temporal mandibular joint dysfunction, Chiari malformations, laxity in the neck may all contribute to headaches—as well as things such as autonomic dysfunction.

Headaches and migraines appear to be more common. Frequent use of Tylenol or ibuprofen can lead to chronic daily headaches. If you are having headaches more than once or twice a week, you may want to consider preventive medicine. Migraine meds should be taken as soon as you think you are getting a migraine.

Effective treatment of pain requires many approaches.

- With AMPS, remind yourself that this pain is not damaging your body.
- Decrease focus on pain
- Physical therapy as described above can help many types of pain. Some treatment centers for chronic or amplified pain may require up to 5 hours of physical therapy daily. Consistency is key. Increased pain says you need to move more, not less. Get up and move but start with things you can do. Pace yourself.

- Cognitive therapies such as cognitive behavioral therapy and mindfulness are also very effective. This does NOT mean that pain “is all in one’s head”, or that some is somaticizing or a hypochondriac. Think of this as physical therapy for the brain. Cognitive therapies have research showing they are effective in reducing pain, anxiety, depression, sleep problems, dealing with irritable bowel and helping control mast cell disorder. There are programs for cognitive therapy in a variety of books, on line and developed for portable mobile devices for those who cannot find a therapist nearby, or who have limited time to go to appointments. Moodkit, Calm, and Pacifica are all highly rated phone apps.
- Pain medication can be helpful. However, they do not help treat the underlying cause of pain. Non-steroidal anti-inflammatory agents (such as Ibuprofen) are the first medication to try for joint pain. These should be taken on a regular basis for several weeks at large enough doses to decrease inflammation. For adults, ibuprofen 800 mg three times a day can be helpful. Meloxicam is another effective medicine. Some people do not need them on a daily basis but notice pain when storm fronts come through or when they are doing something unusual, such as a trip to Adventureland. If there is stomach upset, it may help to take a proton-pump inhibitor (such as Prilosec, OTC), which is also available without prescription, and take the medicine with a large glass of water or other drink or with food. Some patients find that prescription medications are necessary to help control pain. These include amitriptyline, Cymbalta, Lyrica, and Neurontin. Note that many of these are antidepressants but they are used for chronic pain as well. Topical gels including meds such as lidocaine and diclofenac have also been helpful. AVOID NARCOTICS. Narcotics should only be used for a short time. Over time they actually make people more sensitive to pain. Tramadol is a possibility but watch for constipation. Naltrexone has helped some people.
- Consider DMSO, NAC (N-acetylcysteine)
- Joint injections can be helpful. It should be noted that the most common forms of local anesthesia, xylocaine, and bupivacaine, are now known to be specifically and highly cytotoxic to chondrocytes and ropivacaine should be used preferentially for intra-articular injections. EDS patients are often resistant to lidocaine and bupivacaine a fact underappreciated by most physicians. Anecdotally, carbocaine tends to work better in EDS patients
- Low dose naltrexone can be helpful but may take 3-6 months to fully kick in.
- Be cautious with surgery. The cause of pain may not be what the surgery is trying to fix.
- Consider a headache specialist for frequent headaches.
 - Exercise, salt and increased fluid intake helps with headaches
- Weight control is important to decrease pressure on joints.
- Distraction is another excellent way of controlling pain. Inpatient and outpatient treatment centers for chronic and amplified pain syndromes expect people with chronic pain to get out of bed and live through pain.
- hot or cold packs for 15-20 minutes at a time,
- massage
- PT may be able to provide ultrasound, electrical stimulation (TENS),
- acupuncture, acupressure
- balance boards, stork standing, exercise balls
- Limit time on electronics—flexing arms and bending head can increase pain
- Women should try a sports bra with a high racer back and front closure
- Reike, Pilates
- Consider kinesio taping
- Consider compression garments (such as you might find at a sporting goods store)
- Yoga and Tai Chi (avoid overstretching just because you can)
- Chiropractic can be helpful for many people. HOWEVER, the chiropractor needs to be aware of EDS and be careful and gentle with manipulations.
- Prolonged use of electronic devices can stretch the nerves in the arms and neck and cause numbness, nerve stretching, pain and complications from repetitive injury.
- If this all seems overwhelming, pick a couple small things to change to start
- Comfortability is a good website for pain in children, but the exercises may be helpful for adults too.
<https://www.thecomfortability.com/>