FINAL REPORT

THE EHLERS-DANLOS SYNDROME (EDS) AND INTRACTABLE PAIN CONNECTION

By

Forest Tennant, DrPH, MPH, MD

March 2024

A report from the Arachnoiditis Study and Education Project of the Tennant Foundation

BACKGROUND

Beginning approximately five years ago the author came to the realization that persons with hypermobile Ehlers-Danlos Syndrome (hereafter hEDS) were the majority of patients who sought intractable pain (IP) care. They all had been started on a number of pain relief drugs including opioids, neuropathic agents, non-steroidal anti-inflammatory drugs (NSAIDS), and muscle relaxants. Despite having seen many doctors and taking multiple drugs, they were usually in great, constant pain, and desperate to find some relief. When asked about their anatomic location of pain, the usual answer was "all over." Further questioning usually identified the locations as back, neck, or chest wall. As to what caused the pain or when it started patients often stated "It just sort of happened." One thing was clear, I was encountering a group of suffering individuals who were a mystery.

RESEARCH BEGINS

After hEDS patients became the major attenders at our intractable pain clinic, I began to search the medical literature and consult with anyone I could find who might know something about hEDS and pain. I didn't come up with much. Medical literature clearly described the genetic deficiency in hEDS as a defect in collagen production, but little was mentioned about pain. Most of the writings described the pain of hEDS as mild to moderate and the recommended treatment was exercises, NSAIDS, and avoidance of opioids. Simply, the medical literature didn't fit with my observations. The hEDS patients in my clinic had severe intractable pain mainly from adhesive arachnoiditis and other spinal conditions.

DEEP DESPAIR

Besides severe, intractable pain, the emotions and mind were clearly impacted. Almost all had considered suicide. Families and friends had pretty much deserted them leaving them lonely and alone, because they either didn't understand their plight or rejected them if they did. The misunderstanding, humiliation and rejection by the medical establishment was pretty much universal, and it highlighted the fact that medical practitioners didn't know much or understood hEDS and pain. Attached to this report are some comments received from medical practitioners by study participants.

NEED FOR THIS STUDY

This study was done to learn more about hEDS and intractable pain so that better treatment and possibly some prevention measures can be developed.

STUDY PROCESS

To conduct this study, we put out a call in December 2023 to our email universe which consists of over 4400 persons who either have intractable pain or a medical professional with an interest in IP. We asked persons who had been medically diagnosed with hEDS and IP to be a study participant. We accepted within two weeks, 45 study persons who had both hEDS and IP. We could have taken many more study participants, but 45 was enough to meet our goals. The 45 participants were from twenty different states and three foreign countries. All completed a detailed study questionnaire on their medical history.

GOALS OF THE STUDY

A. To Develop a Clinical Profile of Persons with hEDS and IP
B. What Treatments are Being Used?
C. Is There an Opportunity for Prevention?

RESULTS

Among the 45 study participants, 36 (80.0%) had adhesive arachnoiditis (AA) and the other nine (9, 20%) had IP due to multiple painful conditions including those that affect the spine. Summary Tables are attached for the 36 participants who had AA. The results are striking in that the participants were almost all females. A high percentage were bed bound and all described some difficulty with extremity function.

It is interesting that participants were aware that their hypermobility began in childhood or teenage years. Almost all had their first episode of pain before age 20. Many made comments that their pain in childhood or as a teenager was simply thought to be "growing pains." A diagnosis of hEDS wasn't given to most study participants until they were middle aged and had developed intractable pain.

The number of painful, pathologic conditions and medical procedures that participants had is striking. This group of 36 participants reported a total of 124 surgeries and 278 epidural injections.

The pain treatments used were primarily opioids and naltrexone. Ketorolac, methylprednisolone, and ketamine were the drugs used for pain flares. The majority felt their treatment had either brought stability or slowed deterioration. About a third felt their treatments were not working. Several mentioned they were undertreated.

SUMMARY AND SUGGESTION

This study involved a group of seriously ill, female participants. Their medical history is captured in the Tables. A quick survey of the Tables emphasizes the extreme suffering and impairment of these participants. It is no wonder that suicide is common among persons with hEDS and IP.

Participants in the study had obviously had hypermobility in childhood but weren't diagnosed with EDS until mid-life. An obvious conclusion is that a diagnosis of EDS in early life may lead to some preventive measures which will avoid the tragic consequences in adult life.

This is a small study from one source. Consequently, it cannot be assumed that it represents universal applicability. It does, however, point out that hEDS can result in intractable pain and a tragic outcome.

TABLE ONE		
SOME CHARACTERISTICS OF PERSONS WITH EDS, IP, AND AA		
	No.	**%**
Females	34	94.4
Males	2	5.6
Bed bound most hours	13	36.1
Had infectious mononucleosis	9	20.0
Had autoimmune disease	10	27.8
Had complicated pregnancies	21	58.3
Worsened with traumatic accident	8	22.2

TABLE TWO		
KEY EVENTS BY AGE **N = 36**		
Age informed they were hypermobile	Range (yrs) Mean Age	3 to 56 18.6
Age diagnosed with hEDS	Range (yrs) Mean Age	28 to 84 48.7
Age of first pain episode	Range (yrs) Mean Age	3 to 52 12.9
Age of development of intractable pain	Range (yrs) Mean Age	10-79 42.6

TABLE THREE ANATOMIC PAIN SITES AT FIRST PAIN EPISODE AND IP N = 36		
FIRST PAIN EPISODE	**No.**	**%**
Headache	4	11.1
Shoulder/elbow	3	8.3
Hands/wrist	3	8.3
Foot/ankle	6	16.7
Back	6	16.7
Neck	5	13.9
Stomach	1	2.8
Hips	3	8.3
Knees	10	27.8
INTRACTABLE PAIN SITES		
Back/spine	28	77.8
Neck/spine	5	13.9
Non-spine site	3	8.3
Note: Some study participants reported that pain began in more than one anatomic site.		

TABLE FOUR PAINFUL CONDITIONS THAT PRECEDED OR ACCOMPANIED AA N = 36		
CONDITION	**No.**	**%**
Arthritis	36	100
Neuropathies	36	100
Migraine	27	75
Fibromyalgia	22	61.1
Irritable bowel	21	58.3
Temporal mandibular joint pain (TMJ)	20	55.6
Bursitis	18	50.0
Sciatica	17	47.2
Loss of teeth	17	47.2
Plantar fasciitis	15	41.7
Tendonitis	13	36.1
Chronic regional pain syndrome (CRPS)	9	25.0
Costochondritis	8	22.2
Pancreatitis	3	8.3
Every participant reported they had experienced multiple conditions.		

TABLE FIVE		
SPINAL CORD AND CANAL CONDITIONS REPORTED BY PARTICIPANTS WITH AA N = 36		
CONDITION	**No.**	**%**
Herniated discs	29	80.5
Tarlov cysts	18	50.0
Scoliosis	17	47.2
Spinal fluid leaks	15	41.7
Tethered cord	12	33.3
Cauda equina disorder	12	33.3
Chiari malformation	5	13.9
Persons in the study reported that these conditions occurred either before or simultaneously with adhesive arachnoiditis. All participant reported more than a single condition.		

TABLE SIX		
PAIN TREATMENTS USED N = 36		
	No.	**%**
Opioids	26	72.2
Naltrexone alone	4	11.2
Naltrexone with an opioid	3	8.3
Used neither opioids nor naltrexone	3	8.3
Used ketorolac for pain flares	14	38.9
Used methylprednisolone or other corticosteroid for pain flares	9	25.0
Used ketamine for pain flares	5	13.9
Used anti-inflammatory medicinal	36	100.0
Perceived treatment to either stabilize or slow deterioration	23	63.9
Perceived treatment to be inadequate	13	36.1
Total No. of surgeries reported by all participants	124	
Total No. of epidural injections reported by all participants	278	

ADDENDUM

COMMENTS SAID TO EDS-INTRACTABLE PAIN OUTCASTS

- Cardiologists – "If you really have EDS",,,
- If you lose weight
- I'm crazy and anorexic
- Nothing is wrong with you
- Don't come back here
- You should go home and kill yourself because you'll be impossible to treat
- You just need to jog and eat more fruit
- You're born with it, so it doesn't cause any issues
- You just need to stress less
- There's nothing we can do for you
- You don't look like you're in that much pain
- A pain doctor gave me a signed contract that I needed to do her exercises and by 8 weeks, if I was compliant, I would not need opioids.
- All my pain is caused from my opioids (hyperalgesia)
- I need to exercise and eat better to feel better
- Cognitive behavior and physical therapy is all I need
- Just have to live with pain and accept it
- I don't have EDS and I shouldn't be taking so many opioids
- Well, you look great. You can't be in that much pain.
- You don't have autoimmunity.
- EDS doesn't cause pain.
- EDS is not disabling.
- You have hypermobility, not EDS.
- EDS cannot cause Tarlov Cysts or AA
- You don't probably have EDS. Even f you did, it wouldn't affect all this in your body
- If you will hang by your arms 1-2 times a day, your back pain will go away
- You don't have all these things wrong, you need anxiety medication
- Buck up, no one can have pain every moment of every day
- Have you tried removing gluten from your diet
- That EDS was useful and to my advantage because wouldn't tighten u when I get older with arthritis and that I should work on stage fitting myself into a suitcase