

Title: Ehlers Danlos Syndrome: A Retrospective Review of the Current Treatment Options in Pain Management.

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Abstract:

Purpose: The purpose of this study is to investigate the currently available treatment modalities for patients with Ehlers Danlos Syndrome (EDS) and their efficacies in pain and symptom relief.

Methods: Retrospective chart review of 98 patients seen in Physical Medicine and Rehabilitation clinic was performed. Treatment methods were extracted from individual patient charts and efficacy was grouped into 3 categories: improvement, no effect, or worsened symptoms.

Results: The most common treatments utilized were complimentary/alternative treatments (n=88). Occupational therapy and bracing were the most effective option with 70% of patients reporting improvement. Neuropathic modulators were the least well tolerated with 14% of patients reporting adverse effects.

Conclusions: While our study is able to identify positive and negative trends with certain modalities, it is vital to understand that EDS is not a uniform diagnosis among patients and that usually a combination of several different treatments is needed for optimal symptom control.

Introduction

Ehlers Danlos Syndrome (EDS) is a multifaceted disease, which can present with a variety of types of pain. Unfortunately, both the mechanisms and treatments for pain are poorly understood. At its nature, EDS is a disease of connective tissue causing joint hypermobility through laxity (1). Chronic pain is widespread in the EDS population with an incidence of 90% (3).

Methods:

A retrospective chart review was performed for all patients with EDS seen at the Physical Medicine and Rehabilitation Clinic between January 2015 and April 2019. Individual charts were reviewed and information pertaining to different treatment modalities was extracted. Treatment efficacy was grouped into 3 categories: improvement, no effect, or worsened symptoms. Individual modalities were grouped into several categories: physical therapy (PT), occupational therapy with bracing (OT), Non-Steroidal Anti-Inflammatory Drugs (NSAIDS), Acetaminophen (Tylenol), opiates and opiate-like medications, neuropathic pain modulators, muscle relaxants, and steroids. Statistical analysis was performed using GraphPad Prism utilizing the Mann Whitney U test.

Results:

The most commonly reported method was opiates and opiate-like pain medications (n=87), followed by NSAIDS (n=65), physical therapy (n=60), and occupational therapy (n=54). Occupational therapy with bracing was most effective with 70% of

patients reporting symptom improvement, followed by oral or injected steroids (48%) and muscle relaxers (42%). The least effective medications at reporting symptom improvement were neuropathic modulators (14%), acetaminophen (25%), NSAIDS (40%), and opiates and opiate-like pain medications(40%). Neuropathic modulators caused the most adverse effects (47%). The lowest percentage of adverse effects or worsening of symptoms was seen in acetaminophen (0%), muscle relaxers (4%), steroids (10%) and occupational therapy (11%).

Discussion:

Occupational therapy with splints and bracing should be a first line treatment as it has the highest percentage of patients reporting improvement and it was also relatively well tolerated, with only 11% of patients reporting negative effects. Its benefit can be attributed mostly to its role in improving proprioception and joint stability (17).

Despite being a commonly prescribed medication and a first line analgesic agent, our data suggests acetaminophen has minimal efficacy in the EDS population. There also appears to be a minimal role for neuropathic modulators as the adverse effects greatly outweigh symptom improvement. Historically neuropathic modulators have not been shown to be effective in EDS patients with the additional side effect of worsening dysautonomia (5), and perhaps should be avoided in this population.

The most commonly reported method was opiates and opiate-like pain medications, which was prescribed to 90% of patients, however only 40% of patients reported improvement in pain symptoms. Furthermore, in the EDS population, its side effects of decreased GI motility, and orthostatic need to be highly considered. Opiates may have a beneficial role in acute flares, however in the chronic pains of EDS, opiates should be avoided.

Muscle relaxants demonstrated moderate efficacy, with 42% of patients reporting improvement. Interestingly, these medications were one of the most well tolerated with only 4% of patients reporting adverse effects, despite its potential to worsen joint stability.

Conclusion:

Due to the complexity and variability of this condition, there is no one single treatment that works for every patient. Occupational therapy and bracing should be included in nearly all patients, while neuropathic modulators should be avoided. Opiates are the most commonly prescribed intervention in this population despite a relatively low benefit. Forming a close physician-patient relationship can facilitate trust and help patients understand that sometimes multiple treatments will be attempted before obtaining relief.

*References, Tables, and Figures to be provided upon selection for presentation