Prevalence of Arnold Chiari Malformation in Ehler’s Danlos Syndrome patients

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Poster #1989
Disclosure

I acknowledge my continuing obligation to disclose to AANS/NREF/NPA, promptly and in writing, any change in my circumstances.

I further acknowledge that if there is any case where my private interest conflict with the interests of AANS/NREF/NPA, I will indicate that I may have a conflict and abstain from any vote, speaking engagement, planning related to that issue.
Introduction

- Ehler’s Danlos Syndrome (EDS) comprises a group of genetic connective tissue disorders affecting the tissues supporting the skin, bones, and blood vessels.
- Characterized by joint hyper mobility, skin hyper extensibility, and fragility.
- Ehlers-Danlos syndrome (EDS) is a connective tissue disorder that may increase the incidence and severity of Arnold Chiari Malformation.
- Beighton score is a screening technique using a 9 number scale determining the severity of hyper mobility.
- Aim of study is to determine the prevalence of ACM in EDS patients.
Methods

- 756 patients diagnosed with EDS by genetic testing were randomly selected from 8/21/2015 to 8/20/2019 in electronic records and evaluated for ACM.

Source: mayfieldchiaricenter.com/chiari.php

Source: dizziness-and-balance.com/disorders/medical/EHD.html
Results

- Out of 756 patients, 702 (93%) were female and 54 (7%) were male. 23 (3%) were diagnosed with ACM of which 22 (96%) were female and 1 (4%) was male (mean age=32.7; SD=10.11)
- 21 had Beighton scores on record (mean= 7.2; SD=1.61)
Results

- A similar group of 21 patients with EDS without ACM with same age and gender were blindly selected from the same time period and their Beighton score was evaluated (mean=7.3; SD=1.9)
Results

- Of the 21 patients with EDS and CMI, 14 were diagnosed with Postural Orthostatic Tachycardia Syndrome by tilt table tests (HR>30 bpm) (mean=48; SD=15.1)
- The Wilcoxon rank–sum test was used to compare differences of Beighton scores between the groups; no statistical significance (p=0.8253)
Discussion

- Our results suggests a significantly higher prevalence of ACM in patients with EDS

- Our hypothesis concludes most neurologic symptoms in EDS patients could be due to ACM

- However, higher hyper mobility (that is high Beighton scores) does not necessarily mean that patients will also concurrently have ACM
Summary points

• A group of 21 patients with EDS without ACM was compared to another similar group of 21 patients with EDS and ACM

• The Wilcoxon rank – sum test shows no statistical significance between the two groups

• About 67% of patients with EDS and CMI also had POTS