

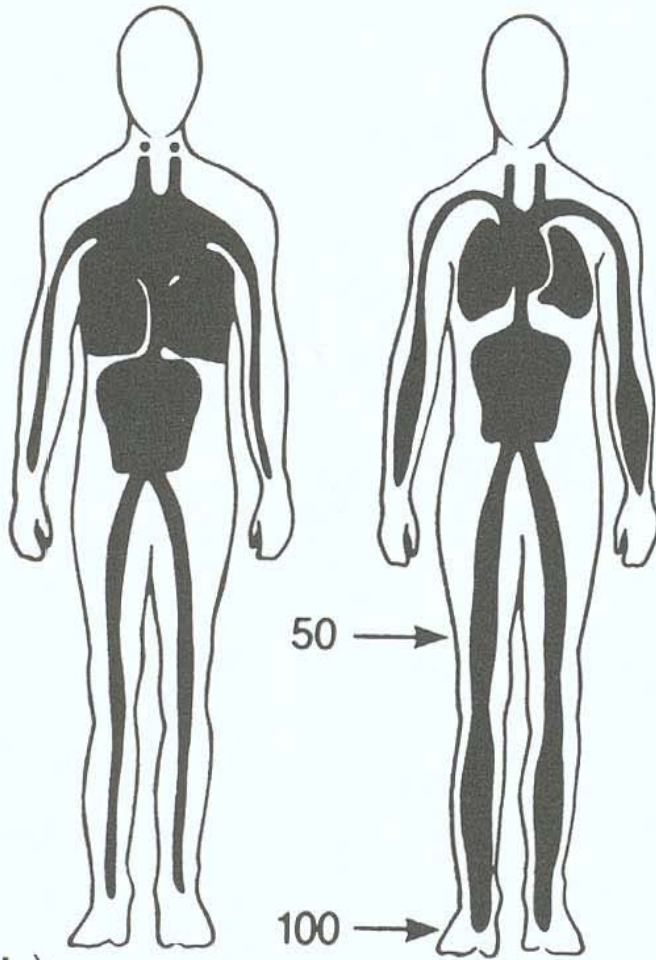
Postural Tachycardia Syndrome and Hypermobility Syndrome



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Supine

Standing



Central
venous
pressure

5 mmHg

0 mmHg

- ↓ CBV
- ↓ Arterial pulse pressure
- Mechanoreceptor unloading
- ↑ Sympathetic outflow
- ↓ Vagal tone
- Veno-arteriolar reflexes
- Activation of renin angiotensin system



Over the years it became evident that many of the the patients referred to the MCO Syncope/Autonomic clinic looked remarkably similar in appearance:

*Pale, fair skinned, caucasian women.
Usually blond haired, blue eyed, often tall and thin. Many complained of joint pain and easy bruising. Stretch marks were common.*



In the late 1990s investigators at the Johns Hopkins Hospital realized that many of these patients met the criteria for Type III Ehlers-Danlos Syndrome (now called the joint hypermobility syndrome).

J Pediatrics 1999;135:494-9

So just what is
Joint Hypermobility/Ehlers-Danlos
Syndrome?



Ehlers-Danlos Syndrome (Type III or joint hypermobility syndrome)

- Heterogeneous disorder of connective tissue
- Prevalence unknown, perhaps 1 per 5000
- Characterized by varying degrees of:
 - Skin hyperextensibility (not present in many)
 - Joint hypermobility
 - Cutaneous scarring
- Early varicose veins, easy bruising
- Easy fatigability and widespread pain common, of unclear etiology

Many EDS/JHS Pts also complain of

1. nausea and bloating (due to gastroparesis and GB disease)
2. orthostatic acrocyanosis
3. joint pain and dislocations
4. hernias
5. constipation
6. hemorrhoids
7. early arthritis
8. stretch marks



ORTHOSTATIC INTOLERANCE AND CFS ASSOCIATED WITH EDS

Among approximately 100 adolescents seen in the CFS/OI clinic at JHH over a 1 year period, they identified 12 subjects with EDS

11 females, 1 male

All had either POTS or NMH

6 classical-type, 6 hypermobile-type EDS

Rowe PC, Barron DF, Calkins H, Maumanee IH, Tong PY, Geraghty MT. J
Pediatr 1999;135:494-9

FEATURES ASSOCIATED WITH CFS IN 12 WITH EDS

Feature	%
Fatigue > 6 mo	100
Post-exertional malaise	100
Unrefreshing sleep	100
Impaired memory/concentration	92
Multi-joint pain	83
New headaches	83
Muscle pain	58
Sore throat	25
Tender glands	25

Revised Criteria for JHS (EDS III)

MAJOR CRITERIA:

1. A Beighton score 4/9 or more (current or historically).
2. Arthralgia for longer than 3 months in 4 or more joints

MINOR CRITERIA:

1. Beighton score of 1,2 or 3/9 (0,1,2 or 3 if aged 50+
2. Arthralgia (>3 months) in 1-3 joints or back pain (>3 M) spondylosis, spondylosis/spondyloisthesis
3. Dislocation/subluxation in more than one joint
4. Soft tissue rheumatism >3 lesions (epicondylitis etc.)
5. Marfanoid habitus
6. Abnormal skin: striae, hyperextensibility, thin, scarring
7. Eye signs: drooping eyelids or myopia
8. Varicose veins, hernia or utero/rectal prolapse

Diagnosis is made by the presence of:

1. two major criteria
2. one major and two minor criteria
3. four minor criteria
4. two minor criteria with an unequivocally affected first degree relative

Diagnosis excluded by presence of Marfans or the other EDS subtypes



Another picture from
a patient's childhood

Many of these patients
excelled at gymnastics
and dance









JOINT HYPERMOBILITY IS MORE COMMON IN CHILDREN WITH CFS

Study question: do children with CFS have a higher prevalence of joint hypermobility?

Beighton scores obtained in 58 new & 58 established CFS patients, and in 58 controls

Median Beighton scores higher in CFS (4 vs. 1)

Beighton score ≥ 4 higher in CFS (60% vs. 24%)

Barron DF, Cohen BA, Geraghty MT, Violand R, Rowe PC. J Pediatr 2002;141:421-5

Gazit Y. et al Dysautonomia in the joint hypermobility syndrome. Am J Med 2003; 115: 33-44

48 pts with Joint Hypermobility Syndrome(JHS) were compared to 30 healthy controls with a battery of Autonomic Tests : HUTT, Valsalva Ratio, HRV, catecholamine levels and baroreflex testing.

78% of JHS pts demonstrated Orthostatic intolerance and abnormal autonomic testing (on every one of the tests mentioned above), as compared to 10% of control subjects

They concluded that JHS/EDS III predisposed people to develop OI



Methods:

- **This retrospective study was approved by our local Institutional Review Board (IRB).**
- **Over a period of 10 years, 26 patients of POTS were identified for inclusion in this study.**
- **All these patients had features of Joint Hypermobility Syndrome (by Brighton criterion).**
- **A comparison group of 39 patients with other forms of POTS were also followed in the autonomic clinic during the same time.**
- **We present a descriptive report on the comparative clinical profile of the clinical features of Postural Orthostatic Tachycardia patients with and without Joint Hypermobility syndrome.**
- **The data is presented as a mean \pm SD and percentages wherever applicable.**



Results:

- Out of 65 patients, 26 patients (all females, 20 Caucasians) had POTS and JHS.
- The mean age at presentation of POTS in JHS patients was 24 ± 13 (range 10-53 years) vs 41 ± 12 (range 19-65 years), $P=0.0001$,
- Migraine was a common co morbidity 73 vs 29% $p=0,001$.
- In two patients POTS was precipitated by pregnancy, and in three by surgery, urinary tract infection and a viral syndrome respectively.
- The common clinical features were fatigue (58%), orthostatic palpitations (54%), presyncope (58%), and syncope (62%).



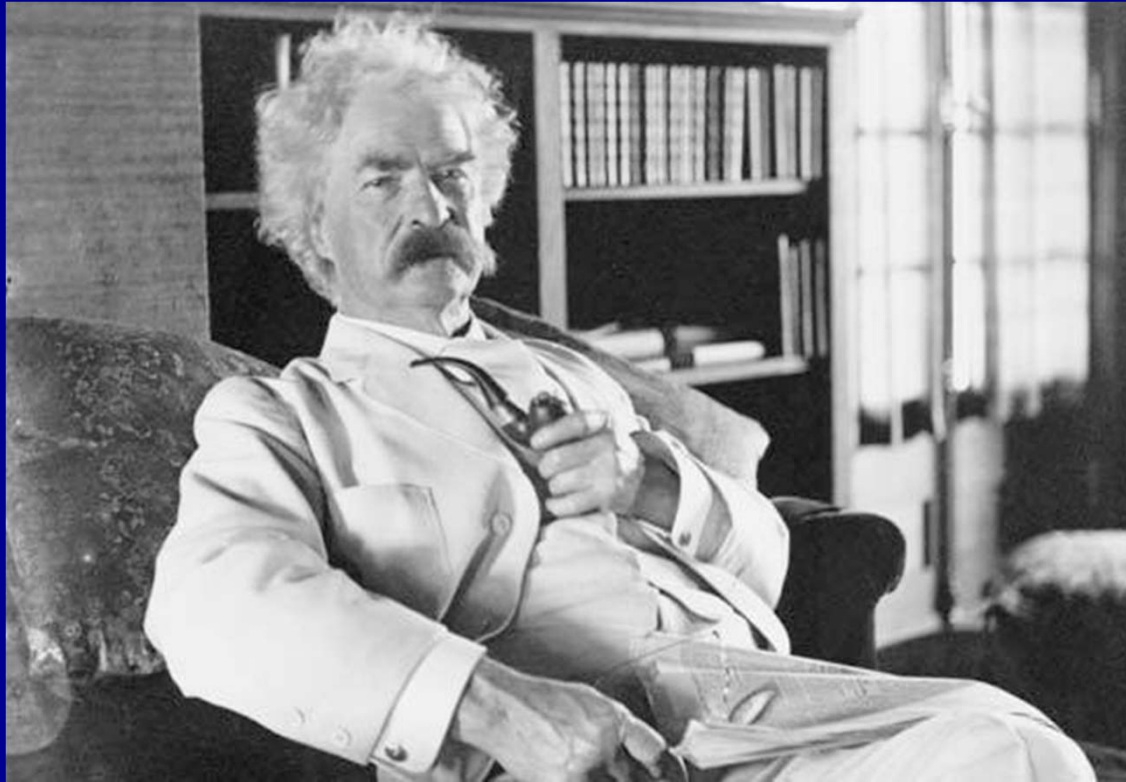
Conclusion:

- **Patients with POTS and JHS appear to become symptomatic at an earlier age compared to POTS patients without JHS.**
- **In addition patients with JHS had a greater incidence of migraine and syncope than their non JHS counterparts.**

*“We shall not cease from exploration,
and the end of all our exploring
will be to arrive where we started
and to know the place
for the first time... ”*



*T.S. Eliot
Four Quarte*



*“All you need in life is ignorance and confidence
And success is assured”*

Mark Twain