

Introduction: The prevalence of Joint Hypermobility Syndrome (JHS) worldwide is estimated to be 5-17%.¹ Hypermobility syndromes have been associated with a variety of comorbidities, and can be an important risk factor for Sleep Disordered Breathing (SDB). The usual risk factors for SDB, such as obesity, male gender, or post-menopausal status, are frequently absent in hypermobile populations, yet the impact of SDB on the quality of these patients' lives can be profound.

Methods: Charts of 49 consecutive patients with a Beighton hypermobility score ≥ 5 from a Neurology Institute were retrospectively reviewed. The review included 33 MRI's and 36 weight-bearing flexion and extension radiographs of the neck, obtained to evaluate cervical spine complaints. The presence of Fatigue or Sleep Complaints, SDB, Headache or Migraine, Disorders of the Cervical Spine, and Symptoms of Dysautonomia (DYS) was evaluated. All \pm values reflect the standard error of the mean.

Results: 41 patients (84%) had sleep or fatigue complaints. 30 underwent attended Polysomnography (PSG), and 5 completed Home Sleep Testing (HST), though PSG was requested for all. SDB was diagnosed in 31 patients (63%). 21 had Obstructive Sleep Apnea (OSA) (G47.33) and 10 had Sleep Apnea – Upper Airway Resistance Syndrome (OSA – UARS) (G47.30). For Patients with SDB the average age was 34.36 ± 2.3 years, and the average BMI was 27.14 ± 1.1 . See figures 1-3 for prevalence of SDB, Headache or Migraine, Disorders of the Cervical Spine, and Symptoms of Dysautonomia. On a T1-weighted MRI in mid-sagittal section, the following airway measurements were obtained and reflect the largest dimensions in centimeters: Retropalatal (0.47 ± 0.06), Retro-lingual (0.86 ± 0.05), and Retro-epiglottal (0.47 ± 0.04), and Epiglottal length (2.22 ± 0.06). MRI measurements for patients with SDB did not differ from those patients without sleep complaints. There was no difference in cervical spine mobility in patients with or without sleep complaints. However, there was a correlation in patients with Headache Disorders: see figure 4.

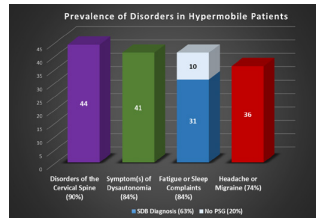


Figure 1: Bar Graph of the prevalence of commonly found disorders in hypermobile patients.

Number of Disorders Possessed by Hypermobility Patients

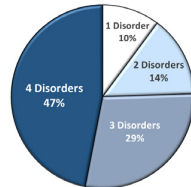


Figure 2: Pie chart of the number of complaints present in each hypermobile patient. Only SDB, Symptoms of Dysautonomia, Headache or Migraine, and Disorders of the Cervical spine were considered.

Figure 3: Table of the Prevalence of SDB, Headache or Migraine, Disorder of Cervical Spine, and Symptoms of Dysautonomia in Hypermobility patients.

	Female		Male		Total	
N	42	85.70%	7	14.30%	49	100.00%
Age	34 (± 0.09)		28.71 (± 6.46)		33 (± 2.00)	
BMI	27.11 (± 1.22)		22.93 (± 1.30)		27 (± 1.08)	
		% of F		% of M		% of Total
Fatigue or Sleep Complaints	36	85.70%	5	71.40%	41	83.70%
SDB:	28	66.70%	3	42.90%	31	63.30%
OSA	18	42.90%	3	42.90%	21	42.90%
OSA-UARS	10	23.80%	0	0%	10	20.40%
Headache or Migraine	34	81.00%	2	28.60%	36	73.50%
Disorder of Cervical Spine	40	95.20%	4	57.10%	44	89.80%
Symptom(s) of Dysautonomia	36	75.70%	5	71.40%	41	83.70%

Figure 4: Bar graph showing the prevalence of fatigue and sleep complaints in this series of 49 patients with JHS

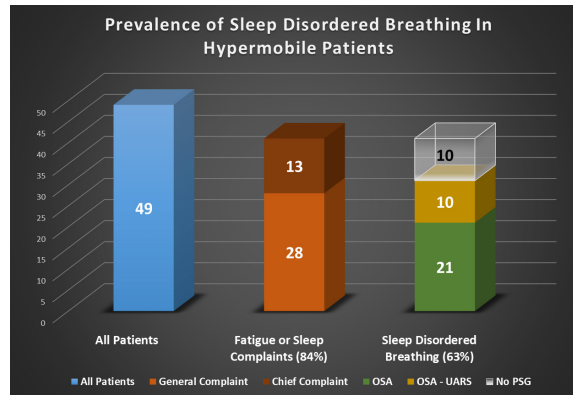
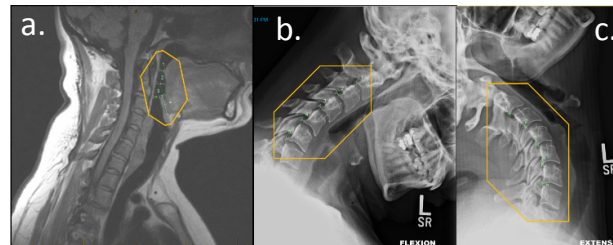


Figure 4: a. Example MRI airway measurement; b. Example flexion X-Ray; c. Example extension X-Ray; d. Table expressing headache and migraine prevalence in hypermobile patients with total cervical spine mobility less ($n=20$) or greater ($n=16$) than 1.25cm.



Total C-Spine Mobility	Total Headache and Migraine Patients	Headache	All Migraine Types	Chronic Migraine	Non- Chronic Migaine	Migraine History	Headache History
< 1.25cm (n=20)	13 (65%)	7 (35%)	5 (25%)	1 (5%)	4 (20%)	4 (20%)	0 (0%)
> 1.25cm (n=16)	15 (94%)	4 (25%)	9 (56%)	6 (38%)	3 (19%)	1 (6%)	1 (6%)

Discussion: The presence of SDB in patients with hypermobility disorders has been noted,² but the marked prevalence of fatigue and SDB in a population like the JHS patients in this series is a new finding. Fatigue by itself was not the most common complaint. In fact, many patients with JHS may suffer fatigue and SDB without seeking evaluation and treatment for those specific complaints. Fatigue in hypermobile populations has often been attributed to chronic pain, anxiety or depression, or nocturnal micturition.³

Unfortunately, in this series, HST (2/5) was considerably less effective in diagnosing SDB compared to an attended PSG (29/30), probably because HSTs do not accurately portray RERAs. Additionally, since 10 patients with fatigue or sleep complaints did not complete PSG, the actual prevalence of SDB in this cohort is likely to be greater than that measured (63%).

Most patients in this cohort of JHS patients had Fatigue or Sleep complaints, Headache or Migraine, Symptom(s) of Dysautonomia, and Disorder of the Cervical Spine. Whether or not these 4 major symptoms share a common cause must await further research.

Conclusion:

1. SDB is present in patients with hypermobility, and is the likely cause of fatigue and sleep complaints in these patients.
2. Hypermobility patients with SDB have an age well below menopause, and a BMI well below that generally associated with SDB.
3. In our series, airway measurements in patients with sleep or fatigue complaints did not differ from those without sleep or fatigue complaints.
4. HSTs are considerably less effective in detecting SDB in hypermobile patients compared to attended PSG.
5. Because of the frequent occurrence of milder, yet treatable, SDB in hypermobile patients, all patients with hypermobility and fatigue or sleep complaints should be evaluated with attended PSG.
6. Whether or not SDB, JHS, DYS, and Headache share a common cause must await further research.

References

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3. Castori M, Morlino S, Celletti C, Celli M, Morrone A, Colombi M, Camerota F, Grammatico. "Management of Pain and Fatigue in The Joint Hypermobility Syndrome (a.k.a. Ehlers-Danlos Syndrome, Hypermobility Type): Principles and Proposal for a Multidisciplinary Approach." *American Journal of Medical Genetics Part A*. Wiley Subscription Services, Inc. 11 July 2012.