

Polysomnography is Crucial to the Diagnosis of Sleep Disordered Breathing Syndromes in Patients with Hypermobility Syndromes

Introduction

Patients with hypermobile Ehlers-Danlos syndrome, as well as Hypermobility Spectrum Disorder (HSD) have narrow, flexible airways, predisposing them to sleep disordered breathing (SDB). They do not typically present with the usual risk factors for SDB including obesity, male gender, and hypertensive disorder. Hypermobile patients frequently have a milder form of SDB that is not detected on a Home Sleep Test (HST), thus requiring the sensitivity of a Polysomnogram (PSG). HST's provide a Respiratory Event Index (REI) which is used to diagnose Obstructive Sleep Apnea (OSA). In contrast, PSG's provide multiple sleep indices including Apnea Hypopnea Index (AHI), Respiratory Effort Related Arousal Index (RERA Index), and the Arousal Index. Due to the increased sensitivity of PSG's, milder forms of SDB, including Upper Airway Resistance Syndrome (UARS), can be detected. PSG's are essential to the diagnosis of SDB in hypermobile patients.

Methods

123 hypermobile patients completed an HST between March 15, 2016 and March 15th, 2018. The average REI captured through HST versus the follow-up PSG, when available, were calculated to compare the sensitivity in patients diagnosed with HSD. The outcomes were identified and compared between patients with a diagnostic HST versus an inconclusive HST.

Results

- 1. Seventy-one (57.72%) hypermobile patients had an inconclusive HST.
- 2. Of those who subsequently had a PSG (24 being lost to follow up), 10 were diagnosed with UARS, and 37 with OSA.
- 3. In those with an attended PSG, there was an approximately fourfold increase in sensitivity of the corresponding index in PSG versus the HST (8.1 AHI vs. 1.9 REI).

	Female	Male	Total
Ν	109	14	123
Average Age	38.3 (±10.8)	34.9(±9)	38 (±10.7)
BMI	289.5(±6.4)	29.5(±8.2)	29.5(±6.6)

Figure 1: Table expressing demographic data of patient population

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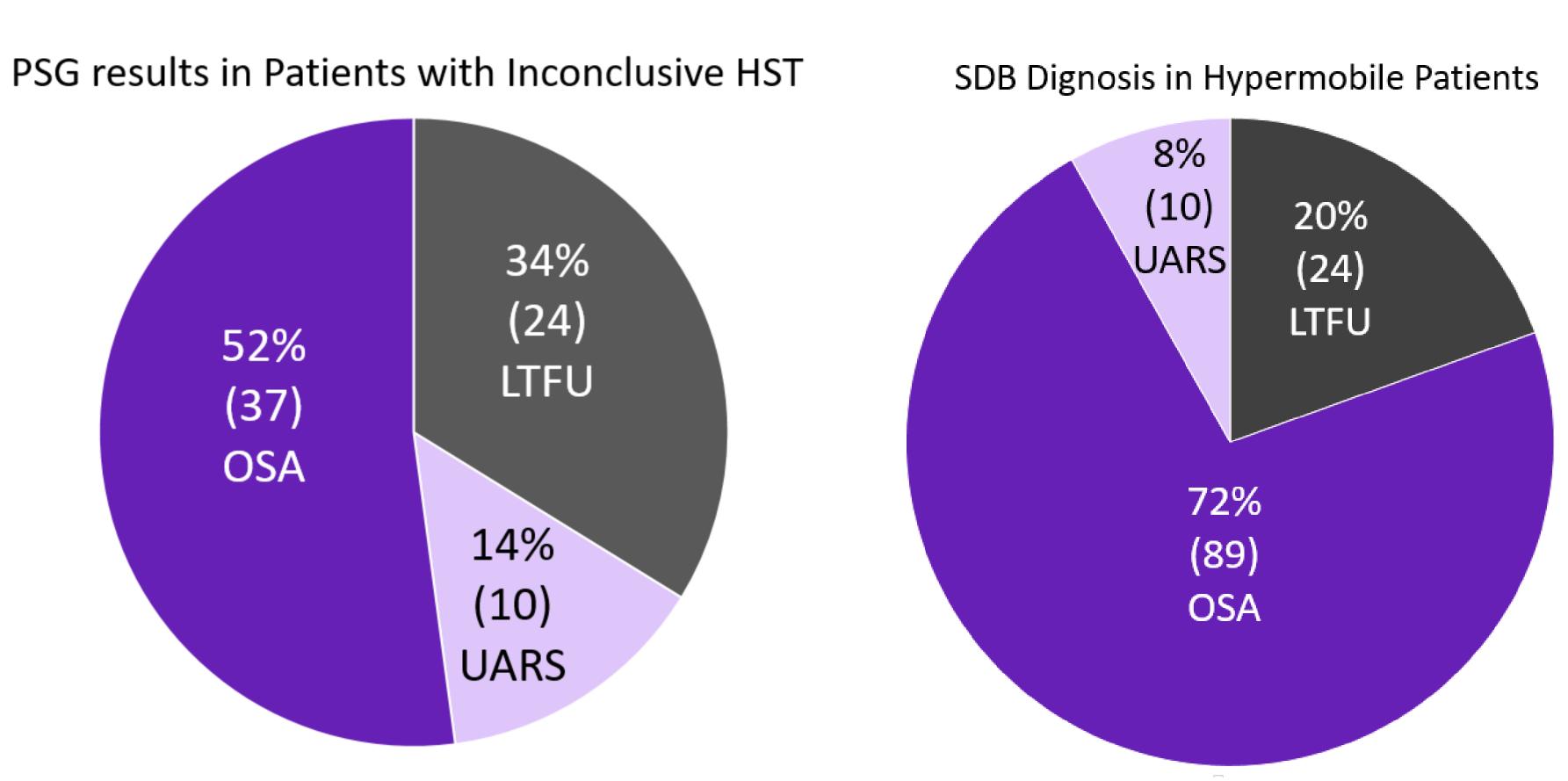


Figure 2: Diagnosis of SDB on those hypermobile patients that had a PSG (N=71)

Figure 3: Diagnosis of SDB in all hypermobile patients (N=123)

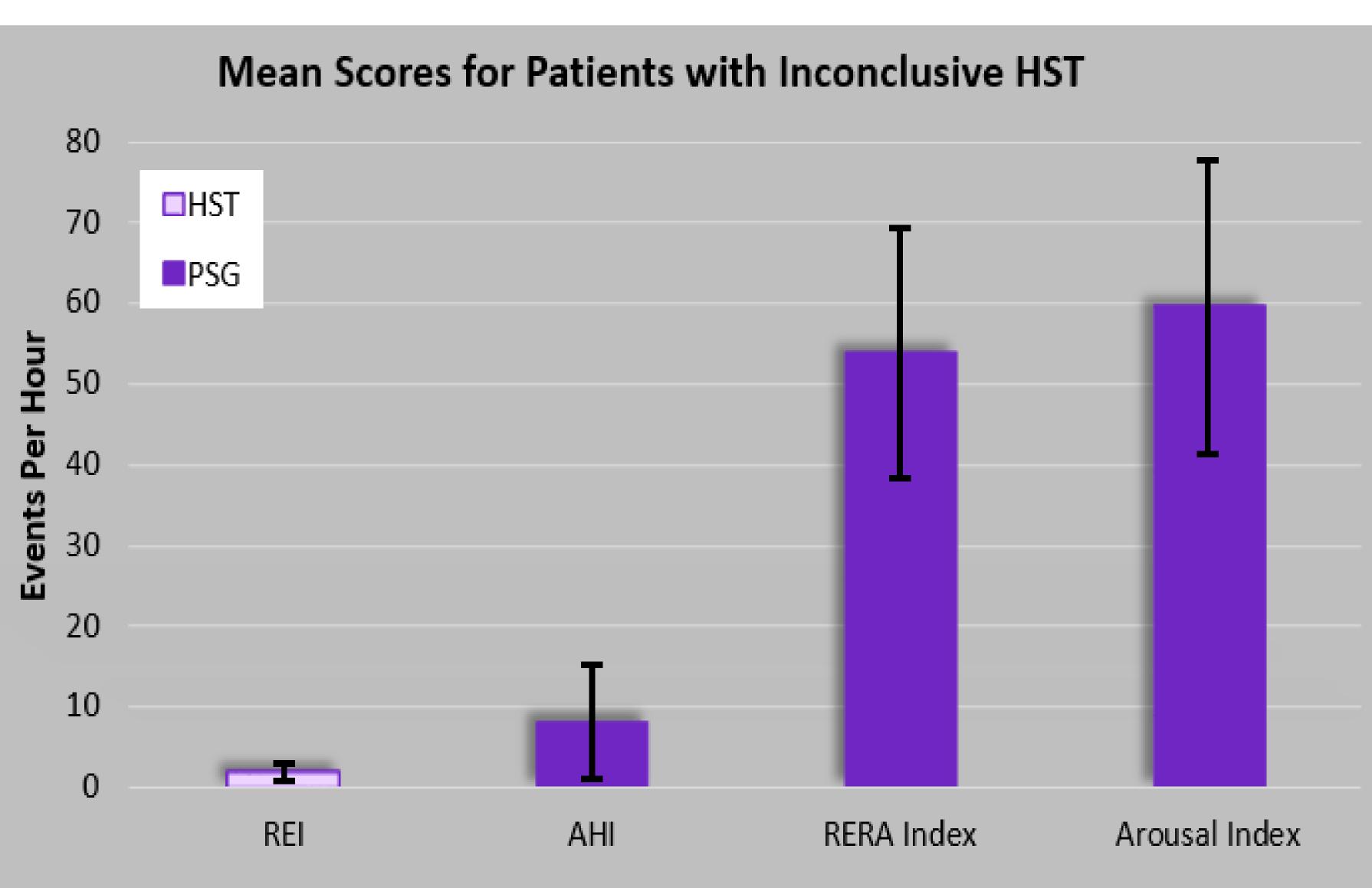


Figure 4: Bar graph indicating the average REI, AHI, RERA Index, and Arousal Index of hypermobile patients with an inconclusive HST (N=71)

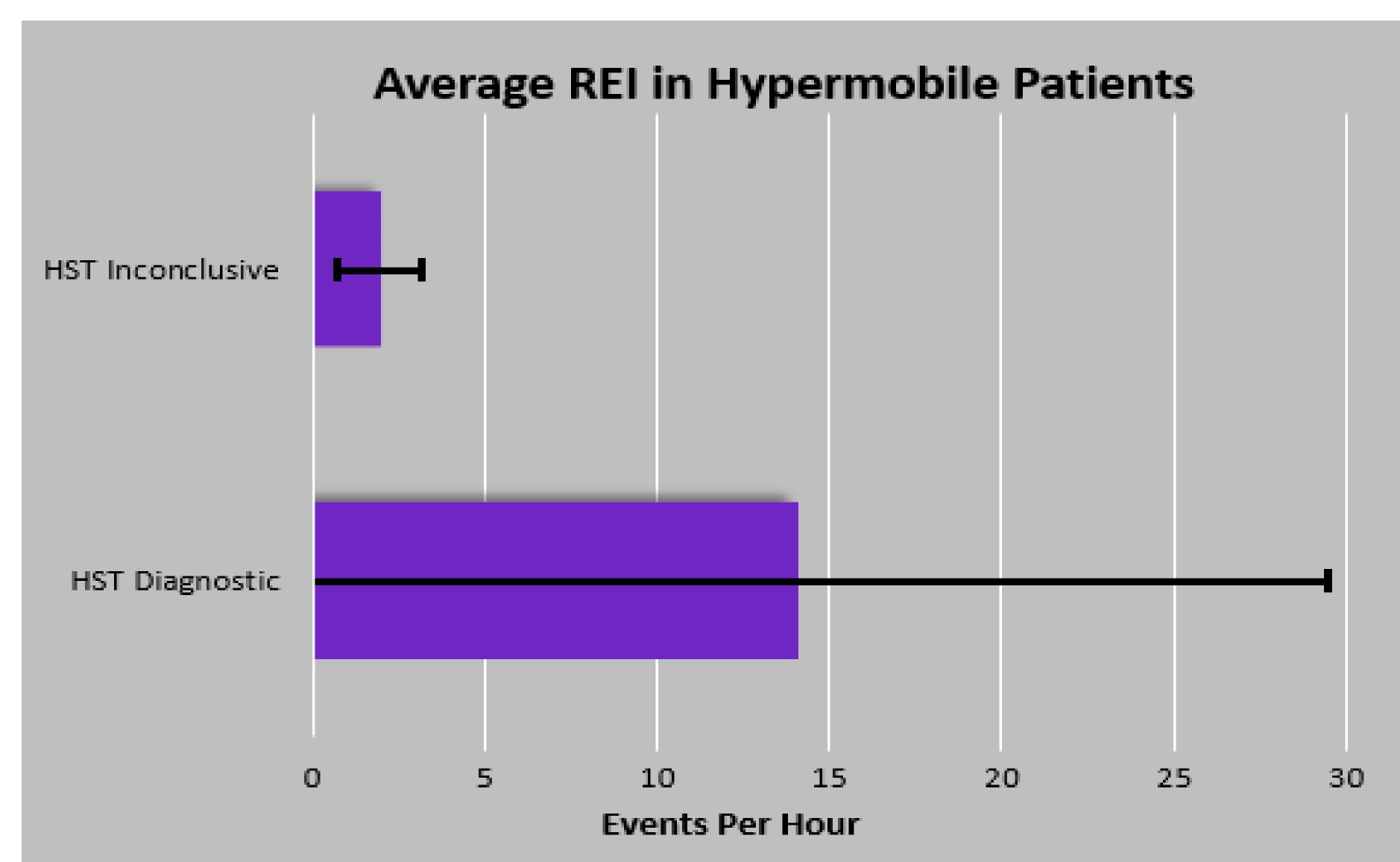


Figure 5: Bar graph indicating the average REI of hypermobile patients with an inconclusive and diagnostic HST (N=123)

Discussion

The presence of SDB in hypermobile patients has previously been reported. This population does not typically have the standard risk factors associated with SDB. They tend to present with milder forms of OSA or UARS due to their abnormally flexible oral airway.

HST's are capable of diagnosing moderate to severe SDB only, and for this reason, and because of other comorbidities in patients with hypermobility (BMI, neck circumference, etc.) are not effective at diagnosing milder forms of SDB. According to our data, only those patients with a BMI of greater than 30 was this tool found to be effective on a consistent basis. In those with a BMI of less than 30, there was no statistically significant correlation between BMI and HST effectiveness.

Further, 24 patients were lost to follow-up after an inconclusive REI, thus delaying care for what is otherwise a treatable cause of fatigue and other complications of SDB.

Hypermobile patients are difficult to treat with standard CPAP therapy, and because of differential airway flexibility characteristics, usually fail auto-titrating CPAP therapy. Combination therapy with a mandibular stabilization device and low CPAP therapy may be the most effective treatment in these difficult patients, though further study is required to ascertain the best non-surgical option for treatment of SDB in these challenging patients.

Conclusions

- evaluation.
- 2. Attended PSG's are necessary to diagnose SDB in hypermobile patients, with HST frequently failing to demonstrate SDB in these patients.
- 3. In hypermobile patients, HSTs are frequently a waste of time and money, and at best delay diagnosis of SDB. 4. For efficiency and improved patient care, in hypermobile patients, attended PSG is the diagnostic procedure of choice.

1. Hypermobile patients frequently have sleep complaints that need