

## Intro:

Patients with hypermobility syndromes often have difficulty using Positive Airway Pressure (PAP) Therapy for treatment of their Sleep Disordered Breathing (SDB). To investigate this, the CPAP titrations of patients with hypermobility and SDB were studied in detail.

## Methods:

The attended PAP titrations of 19 consecutive patients with a Beighton hypermobility score of 5 or greater at a Neurology Institute were retrospectively reviewed. In this cohort of 19 patients, 6 had Sleep Apnea – Upper Airway Resistance Syndrome (OSA-UARS) (G47.30) and 13 had Obstructive Sleep Apnea (OSA) (G47.33). MRIs of the airway, and weight-bearing, flexion and extension X-Rays of the Cervical Spine were reviewed when available. All  $\pm$  values reflect the standard error of the mean.

## Results:

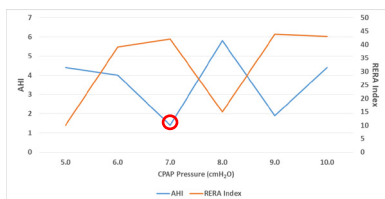
- At their hypopnea minimum, 15 of 19 patients' AHI was 0 and an average Respiratory Effort Related Arousal (RERA) Index of  $38.4 \pm 6.7$  remained. See figures 2b, 2d.
- During titrations, after an initial AHI minimum was achieved, 14 of 19 patients had increasing AHI with further increase in PAP. See figure 2a-d.
- Since the AHI and RERA indices measure fundamentally different properties of the PSG, the responses to PAP are not routinely predictable.
- The upper airway in patients with hypermobility can undergo significant changes with cervical flexion and extension, with a marked opening of the airway appearing during extension. See figure 3.

Figure 1: Cohort Data

### Hypermobile Patients With SDB and Attended PAP Titrations

	N	
Female	19	100.00%
Age	37.63 ( $\pm$ 2.6)	
BMI	28.24 ( $\pm$ 2.2)	
OSA	13	68.42%
OSA-UARS	6	31.58%

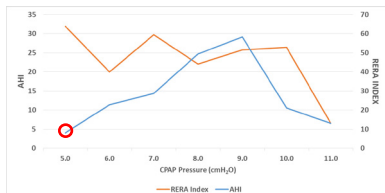
Figure 2a: Example Titration #1



○ = AHI minimum

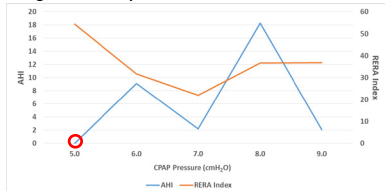
Example Titration #1		
CPAP Pressure (cmH <sub>2</sub> O)	AHI	RERA Index
5.0	4.4	15
6.0	4.0	44
7.0	0	44
8.0	6.0	15
9.0	4.0	43
10.0	6.0	43

Figure 2b: Example Titration #2



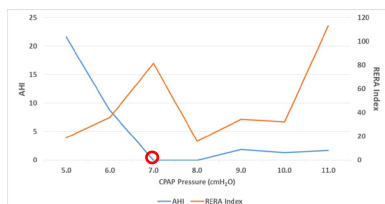
Example Titration #2		
CPAP Pressure (cmH <sub>2</sub> O)	AHI	RERA Index
5.0	5	30
6.0	10	40
7.0	0	64
8.0	20	59
9.0	25	44
10.0	10	51
11.0	30	52

Figure 2c: Example Titration #3



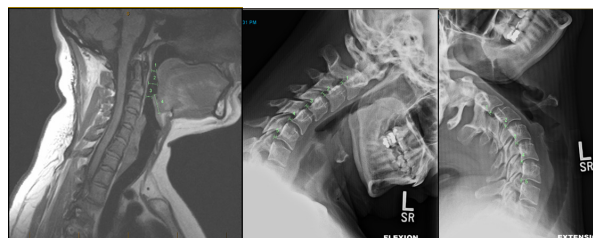
Example Titration #3		
CPAP Pressure (cmH <sub>2</sub> O)	AHI	RERA Index
5.0	18	18
6.0	10	31
7.0	0	54
8.0	20	22
9.0	20	37

Figure 2d: Example Titration #4



Example Titration #4		
CPAP Pressure (cmH <sub>2</sub> O)	AHI	RERA Index
5.0	22	22
6.0	10	36
7.0	0	81
8.0	2	16
9.0	2	34
10.0	2	32
11.0	2	113

Figure 3: Example MRI of Airway and Flexion + Extension X-Ray of Hypermobile patient.



## Discussion:

PAP therapy is the most common and effective treatment for SDB. In typical CPAP titrations, the RERA index and AHI should generally decrease and eventually plateau as the PAP is increased. The lowest possible pressure that reduces a patient's AHI and RERA index to a minimum is usually used for optimal SDB treatment.<sup>1</sup> However, the response to PAP is unpredictable in hypermobile patients, and many patients experience significant increases in AHI after an initial nadir is achieved. Also, many patients have a high RERA index at their AHI minimum.

The dissociation between the AHI and RERA index may be due to the fundamental difference between the way Apneas/Hypopneas and RERA's are measured. On the other hand, in hypermobile patients, this dissociation may be due to unequal compliance in different elements of the upper airway. For instance, even though portions of the airway may be expanded by PAP, increased PAP in hypermobile patients may lead to epiglottal closure.

Since many patients cannot complete an attended PAP titration for various reasons, sometimes they are treated with auto-titrating PAP devices, and ultimately fail PAP therapy. This may be due to the fact that auto-titrating machines cannot measure RERAs well because they cannot measure arousals and disruptions from sleep, though they do measure changes in airway resistance with formidable algorithms. Thus, in patients with hypermobility, the use of auto-titrating CPAPs may not resolve SDB and the resultant sleep fragmentation, and thus lead to non-compliance with therapy.

## Conclusions:

- PAP titrations in patients with hypermobility syndromes many times yield unpredictable results, and must be analyzed carefully if the SDB in these patients is to be resolved.
- This unpredictability could be explained by (1) variations in the compliance of different elements of the airway, (2) a fundamental difference in the way Apneas/Hypopneas and RERAs are scored.
- Given this unpredictability, auto-titrating PAP devices would not be expected to optimally resolve the sleep fragmentation so common in patients with hypermobility.
- Thus, patients with hypermobility syndromes should have attended PAP titrations so that optimal airway patency and resolution of their complaints related to SDB can be achieved.

## References

- Positive Airway Pressure Titration Task Force of the American Academy of Sleep Medicine. Clinical Guidelines for the Manual Titration of Positive Airway Pressure in Patients with Obstructive Sleep Apnea. Journal of Clinical Sleep Medicine : JCSM : official publication of the American Academy of Sleep Medicine. 2008;4(2):157-171.