Treatment Of Sleep Disordered Breathing In Hypermobile Patients

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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.

Results:
1. At their hypopnea minimum, 15 of 19 patients’ AHI was 0 and an average Respiratory Effort Related Arousal (RERA) Index of 38.4±6.7 remained. See Figures 2b, 2c.
2. During titrations, after an initial AHI minimum was achieved, 14 of 19 patients had increasing AHI with further increase in CPAP Pressure. See Figures 2a-d.
3. Since the AHI and RERA index to a minimum is usually used for optimal airway patency and resolution of their complaints related to SDB can be achieved.

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. 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.


Conclusions:
1. 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.
2. 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.
3. Given this unpredictability, auto-titrating CPAP devices would not be expected to optimally resolve the sleep fragmentation so common in patients with hypermobility.
4. 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

Figure 1: Cohort Data

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<tr>
<th>Hypermobile Patients With SDB and Attended PAP Titrations</th>
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<tbody>
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Figure 2a: Example Titration #1

Figure 2b: Example Titration #2

Figure 2c: Example Titration #3

Figure 2d: Example Titration #4

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