

# Positional Therapy in a Child with Down Syndrome and Obstructive Sleep Apnea: A Case Report

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## Abstract

Obstructive sleep apnea (OSA) is very common in children with Down Syndrome. We report an 11-month-old female infant with Down syndrome who presented with OSA and Hypoventilation. As surgery was not indicated she was referred for institution of CPAP. As her family had concerns about CPAP, she was started on medications (nasal steroids and leukotriene antagonists) and advised Positional Therapy. Repeat sleep study in lateral position performed 7 months later showed resolution of hypoventilation and significant reduction in AHI.

To the Editor,

Pediatric Pulmonology

Sub: Clinical Correspondence.

We are submitting an article titled “**Positional Therapy in a Child with Down Syndrome and Obstructive Sleep Apnea: A Case Report**” to your esteemed Journal to be considered for publication.

Obstructive sleep apnea (OSA) is very common in children with Down Syndrome. We report an 11-month-old female infant with Down syndrome who presented with OSA and Hypoventilation. As surgery was not indicated she was referred for institution of CPAP. As her family had concerns about CPAP, she was started on medications (nasal steroids and leukotriene antagonists) and advised Positional Therapy. Repeat sleep study in lateral position performed 7 months later showed resolution of hypoventilation and significant reduction in AHI.

To our knowledge, this is the first report that highlights the use of Positional Therapy with management of OSA in Down syndrome. This report demonstrates that Positional Therapy can be selectively employed in patients who are too young or cannot tolerate PAP. Positional Therapy can play a significant role in treating OSA being a low-cost and non-invasive intervention.

All authors are responsible for reported case and have participated in patient care, the concept and design; drafting & revising of the manuscript and have approved the manuscript as submitted.

We look forward to a favorable reply from you.

Sincerely,

Abigail Hadley, BS

Harish Rao, MD (corresponding author)

**Positional Therapy in a Child with Down Syndrome and Obstructive Sleep Apnea:  
A Case Report**

## Introduction:

Obstructive sleep apnea (OSA) is a disorder caused by complete or partial upper airway obstruction during sleep. In the pediatric population, OSA prevalent in ages 2- to 8-years-old commonly manifests as snoring, apneas, behavioral problems, and inattention/hyperactivity. A subset of these children can have significant hypercarbia and hypoxemia which, left untreated, can lead to pulmonary hypertension and cor pulmonale.<sup>1</sup> OSA is a frequent comorbidity in children with Down Syndrome (DS).<sup>1</sup> Treatment of OSA in children generally consists of upper airway surgery (adenotonsillectomy) and/or continuous positive airway pressure (CPAP).

Positional therapy for OSA is often used as standalone or as an adjunct to traditional therapies. It is the avoidance of the position that appears on polysomnography (PSG) to exacerbate sleep apnea. Patients likely to benefit from Positional Therapy (PT) often show an apnea-hypoxia-index (AHI) substantially higher in one position (usually supine) than in other positions<sup>2</sup>. Unfortunately, PT has not been well studied in children with the limited available literature mostly focused on children with obesity.<sup>2</sup>

## Report of Case:

An 11-month-old female infant with DS, plagiocephaly, and repair of Tetralogy of Fallot presented to sleep clinic following an abnormal PSG at 9 months. PSG was ordered due to history of snoring with high-pitched gasps with restless sleep to evaluate for OSA. PSG showed severe OSA with sleep related hypoventilation consistent with obstructive hypoventilation syndrome. During this study, she slept mostly on in supine position (Fig 1). Oxygenation was adequate with intermittent oxygen desaturation associated with obstructive events. Following this abnormal PSG, a pediatric otolaryngologist performed bronchoscopy to assess upper airway. However, upper airway surgery was not pursued due to lack of clear site or cause of obstruction.

Shortly after her bronchoscopy, she presented to pediatric sleep clinic for further management of her OSA to discuss institution of CPAP. Patient's expressed concerns that CPAP would not be feasible because of potential intolerance and due to the fact CPAP is not FDA approved in children weighing under 66 lbs. After discussing other treatment options, decision was made to trial a combination of medication (nasal steroid two sprays in each nostril daily and montelukast 4 mg qhs) plus PT (lateral position achieved with firm pillow placed against the patient's back) and to repeat PSG in three months. Risk of SIDS with lateral sleeping position was discussed but at 11 months, risk would be much reduced. CPAP option was also pursued in case this regimen failed to improve her OSA. Echocardiogram done at 10 months of age showed normal cardiac anatomy with no evidence of pulmonary hypertension (predicted pulmonary pressure gradient of 25 mmHg).

Patient was very averse to CPAP when seen in PAP clinic. CPAP education and desensitization initiated in case the patient would likely need CPAP in the future. She had her repeat PSG about five months from initial visit and treatment initiation. Results showed significant improvement from prior PSG. Her repeat PSG improved from severe to mild OSA with resolution of hypoventilation (Table 1). Transcutaneous CO<sub>2</sub> (TcCO<sub>2</sub>) was used during repeat study due to non-availability of end-tidal CO<sub>2</sub> monitors. During repeat PSG, the patient slept mostly in non-supine position (Fig 2). Considering her dramatic improvement on her current regimen, the decision was made to continue her current regimen at follow-up. Patient's mother also reported that the patient's snoring and sleep quality had also significantly improved on current regimen.

## Discussion:

First-line treatment for OSA in children and adolescents is typically adenotonsillectomy if indicated or CPAP treatment. However, this is challenging in younger children where adenotonsillectomy may not be indicated and CPAP may not be well tolerated. Additionally, this CPAP may be difficult to pursue, not desired by the family, and/or not sufficient to improve OSA.

Positional OSA is a phenotype of OSA where obstructive events occur predominantly in the supine position. Positional OSA has been well described in adults, affecting about 55% of adults with OSA<sup>3</sup>. This is because airway collapse is maximal in the supine position compared to lateral or prone positions in patients with

significant reduction in airway dimension occurring antero-posteriorly compared to circumferential reduction in airway dimension.

Positional OSA has not been studied well in children. In a cross-sectional study by Verhelst et al, overall prevalence of Positional OSA was 18.7%. Children with Positional OSA were significantly older, had a higher prevalence of obesity, smaller tonsils and lower AHI. <sup>3</sup>

Improved airway patency, particularly at the base of the tongue/larynx in left lateral position compared to standard supine position has been demonstrated in children during DISE<sup>4</sup>. In another study by Kirkham et al, children with positional OSA had higher odds of obstruction at the tongue base but not at other levels<sup>5</sup>. The airway collapse in the supine position is more significant in patients with DS secondary to hypotonia, mid-facial hypoplasia, and glossoptosis<sup>1</sup>.

Positional therapy (PT) is a reasonable approach to certain patients who demonstrate differences in AHI, oxygenation and ventilation depending on position. In fact, Xiao et al in their novel pilot study showed reduction in Obstructive AHI and oxygen desaturation in non-supine position compared to supine position using positional device therapy<sup>6</sup>. It is reasonable to trial patients not fully controlled on CPAP and/or with adenotonsillectomy on PT and monitor for improvements in symptoms or PSG. Identifying the OSA phenotype and the subset of OSA patients likely to benefit from non-supine position increases the success of PT. PT has evolved over time with the development of efficient and comfortable devices which can monitor sleep position changes as well as compliance.<sup>6</sup>

In this case, our patient showed excellent response to PT, along with an intranasal steroid and a leukotriene inhibitor with reduction in AHI and resolution of hypoventilation. Spontaneous resolution of OSA would have unlikely to have played a role considering her age and underlying diagnosis, though it has been described in older children (CHAT study). This case demonstrates that PT can be selectively employed and can play significant role in treating OSA in select patients with the benefits of being a low-cost and non-invasive intervention. To the best of our knowledge, this is the first report of PT to be successfully applied in a young child with DS.

**Tables/figures:**

Table 1 – Sleep study parameter comparison between the initial and repeat studies.

|  | PSG at 9-months-old   | PSG at 17-months-old   |
|--|---|--|
| <b>Height (cm)</b>                         | 67.0  | 77.6   |
| <b>Weight (kg)</b>                         | 9.6   | 10.8   |
| <b>Severity of OSA</b>                     | Severe  | Mild   |
| <b>AHI</b>                                 | 11.3 per hour   | 2.9 per hour   |
| <b>REM AHI</b>                             | 25.4 per hour   | 9.8 per hour   |
| <b>Mean oxygen saturation while asleep</b> | 98.0%   | 97.0%  |
| <b>SpO2 nadir in sleep</b>                 | 87%   | 92.1%  |
| <b>CO2</b>                                 | (End tidal) peak 54 mm Hg CO2 of 45-49 mm Hg; 56% CO2 of >50 mm Hg: 26% | (TcCO2) peak 48 mm Hg CO2 of 45-49 mm Hg: 25% CO2 of >50 mm Hg: 0.0% |
| <b>Sleep efficiency</b>                    | 90.1%   | 90.2%  |
| <b>Arousal index</b>                       | 14.4 per hour   | 20.7 per hour  |
| <b>Respiratory arousal index</b>           | 1.3 per hour  | 1.3 per hour   |
| <b>Sleep fragmentation</b>                 | Mild  | Mild   |
| <b>Periodic limb movement index</b>        | 0 per hour  | 3.2 per hour   |

OSA = obstructive sleep apnea; AHI = apnea-hypoxia index; REM = rapid eye movement

Figure 1 – Initial PSG at 9-months-old

(Hypnogram, SpO2, End tidal CO2, respiratory events, body position)

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Figure 2 – Repeat PSG at 17-months-old after positional therapy

(Hypnogram, SpO2, Tc CO2, respiratory events, body position)

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## Short title: Positional Therapy in Down syndrome and OSA

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*Abbreviations :*

OSA Obstructive sleep apnea

DS Down syndrome

PSG Polysomnography

PT Positional Therapy

AHI Apnea Hypopnea Index

CPAP Continuous positive airway pressure

#### Contributors Statement Page:

Dr Rao and Ms Hadley conceptualized and designed the study, collected data and drafted, reviewed and revised the manuscript; Dr Rao critically reviewed the manuscript for important intellectual content; and all authors approved the final manuscript as submitted and agree to be accountable for all aspects of the work

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