Effects of Sleep on Adherence & Quality of Life in Children with Cystic Fibrosis  
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**Objectives**
- To develop a better understanding of sleep in children with CF and its potential impact on overall health, daytime function, treatment burden, and medication adherence
- To improve medication adherence in children with cystic fibrosis (CF) by identifying subjective quality of sleep factors that could be amenable to intervention

**Hypotheses**
- Children with CF demonstrate significantly worse sleep quality compared to age matched controls
- Children with CF and worse sleep quality are less adherent to their treatment regimens

**Specific Aims**
- To determine the effect of CF on the quality of sleep
- To determine the effect of poor sleep quality on medication adherence in children with CF
- To determine the effect of sleep hygiene and habits on medication adherence in children with CF

**Audience & Methods**
- Local participants followed in the Pediatric CF Center
- Pulmonary function and clinical data obtained from recent clinic visit
- Overnight admission to the clinical research unit sleep laboratory for sleep studies
- Standard polysomnography (PSG) with additional measurements
  - Nasal airflow tracings
  - Indirect calorimetry measurements for energy expenditure
- Quality of life assessment using CFQ-R

**Deliverables**
- Sleep parameters
  - Respiratory rate
  - Breathing patterns
  - Energy expenditure
- Quality of life assessment
- Correlations with clinical data (i.e., lung function)
- Adherence assessment
  - Medication refill data

**Impact**
- Children with CF at all levels of lung disease show changes in breathing during sleep compared to wakefulness
- Increased respiratory rate and inspiratory flow limitation during sleep
- Confirms our published findings in CF and matched controls
- Individuals with higher lung function had lower respiratory rate and higher flow limitation compared to those with lower lung function
- No correlation in this group between CFQ-R domains and lung function or breathing patterns during sleep

**Challenges**
- Delay with regulatory requirements
- Scheduling sleep studies/support
- Limitations with obtaining medication refill data
- Identifying validated and generalizable pediatric sleep quality instruments that can be used across a wide age spectrum
- Small sample size for statistical analysis and significance

**Demographic Data**

<table>
<thead>
<tr>
<th>Age (y)</th>
<th>Median ± SD (range)</th>
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<tr>
<td>16±2.6 (10-17)</td>
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<tr>
<th>% male</th>
<th>78</th>
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<tbody>
<tr>
<td>BMI (kg/m²)</td>
<td>20.6±4.2 (17.9-31.9)</td>
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<td>BMI z-score</td>
<td>0.14±1.11 (-1.5-2.44)</td>
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<td>FEV₁ (% pred)</td>
<td>89.0±30.8 (34-106)</td>
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**NREM Sleep Data**

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<tr>
<th>% TST</th>
<th>85.2±6.3 (73.1-94.2)</th>
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<tr>
<td>AHI (events/hr)</td>
<td>0.8±3.4 (0-9.8)</td>
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<td>SaO₂ %</td>
<td>97.6±2.5 (92.0-98.7)</td>
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<tr>
<td>tcCO₂ mmHg</td>
<td>41.7 ± 5.4 (38.4-51.2)</td>
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<td>EEmin, kcal/min</td>
<td>1.48±0.29 (0.92-1.93)</td>
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<tr>
<td>EE, kcal</td>
<td>483.4±145.4 (193.2-720.6)</td>
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<td>VE, L/min</td>
<td>8.7±6.1 (3.8-21.3)</td>
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<td>% IFL</td>
<td>70.9±29.6 (13.9-97.5)</td>
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<td>I, breaths/min</td>
<td>22.1±3.8 (15.4-26.5)</td>
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**Ideal Next Steps**
- Increasing recruitment & enrollment
- Focused effort on medication refill data toward adherence aim
- Identifying the most suitable sleep quality instrument
- Used in conjunction with CFQ-R