

Disease

Pulmonary Function

Sleep Abnormalities

Ventilatory Control

Cerebral Palsy	No data available	OA, MA > CA, hypoventilation, Abn REM	No data available
Spinal muscular atrophy	↓ TLC, FRC ↓ MIP, MEP	CA, MA, hypoventilation	↑ occlusion pressure
Duchenne muscular dystrophy	↓ TLC, FRC ↓ MIP, MEP	OA, MA, CA, hypoventilation, Nonapneic hypoxemia	↓ HVR, Hypercapnic VR N or ↑ occlusion pressure
Myotonic dystrophy	N or ↓ FRC ↓ MIP, MEP	CA>OA, MA, REM-related hypoxemia, hypoventilation, EDS, sleep-onset REM	↓ HVR, Hypercapnic VR N or ↑ occlusion pressure

Clinical Manifestation (SDB)

- Asymptomatic
- Morning headaches, fatigue, exertional dyspnea, irritability, hyperactivity, impaired learning, vomiting, difficulty tolerating supine position, restless sleep [Heckmatt et al 1989, Labanowski et al 1996]
- Failure to thrive, nocturnal sweating, developmental delay, cor-pulmonale [Beckerman and Hunt 1992]
- These symptoms may be erroneously attributed to the NMD rather than SDB [Gozal 2000]
- Nonrestorative sleep and EDS may be the early sign of SDB

Clinical manifestation

- PE: Bell shaped chest, tachypnea, use of accessory muscles and paradoxical breathing
- Even patients with mild symptoms may have significant unrecognized SDB [Labanowski et al 1996]
- Investigations
 - PFT
 - Ventilatory control
 - Polysomnography
 - Others: Fluoroscopy, CXR, CBC, HCO₃

Clinical assessment

■ PFT

- Spirometry, lung volumes, muscle strength assessment
- Restrictive physiology
- Periodic assessment of PFT especially when lung volumes < 60% [Gozal 2000]
- PFT may not be predictive of SDB [Smith et al 1988, Heckmatt et al 1989, Manni et al 1989, White et al 1995]
- Other factors contributing to SDB : upper airway, intercostal and diaphragmatic hypotonia, poor airway clearance, pulmonary atelectasis, abdominal distention, GER, pulmonary aspiration, progressive malnutrition or obesity [Givan 2000]