

RCPCH Working Party on Sleep Physiology and Respiratory Control Disorders in Childhood

Lay Summary 4 –

Sleep-related breathing disorders in children with neuromuscular disease

What are Sleep-Related Breathing Disorders?

Sleep-Related Breathing Disorders (SRBD) are problems with inadequate breathing at night, either due to inadequate muscle strength to take deep enough breaths, or to airway collapse during sleep causing obstruction to breathing, or a combination of the two.

Symptoms of inadequate breathing at night may be very difficult to identify in children with neuromuscular disease.

How common is SRBD in children with neuromuscular problems?

Some conditions are more prone to SRBD than others. In general, any condition which affects the diaphragm is more likely to affect breathing during sleep. In addition, boys with Duchenne Muscular Dystrophy have a high incidence of SRBD as they get older.

What are the risks of SRBD?

In normal children, SRBD can cause impaired growth and development, and is associated with poorer academic performance. It can also put extra strain on the heart, and occasionally causes pulmonary hypertension, a very dangerous condition with elevated blood pressure in the lungs. Children with neuromuscular disease who have SRBD have impaired quality of life and will die earlier if they are not treated.

What tests can be done to detect SRBD in children with neuromuscular disease?

We recommend that overnight oximetry recordings (measurement of blood oxygen levels using a soft probe wrapped around a finger or toe) should be carried out on all children with neuromuscular disease if there are symptoms of SRBD, impairment of diaphragmatic function, or a vital capacity below 50% predicted. In conditions such as myopathies, where the risk of early SRBD is particularly high, regular recordings should be carried out even in the absence of any of these indicators. If possible, carbon dioxide (CO₂) recordings should be carried out at the same time. Recordings should be carried out annually or more frequently if the situation is high risk or unstable.

What can be done about SRBD?

If SRBD are detected then a more detailed sleep study (cardiopulmonary sleep study or polysomnography) should be performed. For more details about these tests please see Lay Summary 1. In severe cases further tests should not delay the initiation of treatment.

Non Invasive Ventilation (NIV) is indicated for children with neuromuscular disease who have significantly low oxygen levels or high CO₂ levels at night. This involves the use of a soft plastic mask which fits over the nose or nose and mouth, or soft plastic devices which fit into the nostrils. A machine is attached to the mask or device and provides help with breathing while the child is asleep. Further assessment with polysomnography should be performed to establish the effectiveness of the NIV.

In Duchenne Muscular Dystrophy, the commonest of the neuromuscular conditions in children, there is a substantial body of evidence that shows significant gains in life expectancy if NIV is used to correct SRBD.