

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

Lay Summary 6 – Sleep-related breathing disorders in children with Prader-Willi Syndrome

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 Prader-Willi Syndrome (PWS).

How common is SRBD in children with PWS?

Mild abnormalities of breathing during sleep are common in PWS, with estimates ranging from 25% to 100% of patients assessed. There is a tendency to obstructive sleep apnoea (OSA) which may be associated with the obesity seen so often in PWS, but there is also a tendency to have abnormal breathing control reflexes during sleep in PWS.

What are the risks of SRBD in PWS?

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.

We know that death from heart and lung (cardiorespiratory) failure, and sudden death are both more commonly seen in PWS, and it is possible that the disorders of breathing seen during sleep are a contributory factor to these findings.

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

We recommend that a screening test of overnight oximetry with carbon dioxide recording (capnography) should be carried out on all children with PWS on an annual basis. Children with abnormalities on the screening test should have a more detailed sleep study (cardiopulmonary sleep study or polysomnography). For more details about the tests please see Lay Summary 1.

What can be done about SRBD?

If SRBD are detected then a more detailed sleep study (polysomnography) should be performed. This should be done in a centre which has adequate facilities and expertise for both the study and the interpretation.

If nocturnal respiratory failure is demonstrated then a trial of Non Invasive Ventilation (NIV) should be offered. 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.

Children with PWS in whom Growth Hormone treatment is planned should have a preliminary cardiopulmonary sleep study or polysomnography before starting treatment.