

Respiratory Complications in Children with PWS

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Tan H-L, Urquhart DS. Respiratory Complications in Children with Prader-Willi Syndrome. Paediatr. Respir. Rev. (2016).

This article is a summary of 52 publications from around the world about the breathing problems seen in children with PWS. Included were many of our American publications. By combining the findings in multiple studies, the authors could describe the challenges and the effectiveness of various treatments with greater confidence; a single study may be exciting news, but the best information occurs when more than one researcher finds the same results.

Infants and Oxygen

Why do so many babies need oxygen at night, and yet most do not need this when they get older? Parents struggle with the challenges of oxygen therapy, and may see no change in their baby with this therapy. However, breathing in infants with PWS is unique – along with their obvious muscle weakness, they have poor regulation of breathing from the hypothalamus.

In the hypothalamus, breathing is finely tuned to the body's need for oxygen through chemical receptors. A healthy hypothalamus prompts a deep breath and raised heart rate when there is too much carbon dioxide (CO₂) in the blood. If CO₂ is high, there is not enough oxygen for the body – and this balance is not well managed in babies with PWS. They also may be too weak to “blow off” the CO₂ with a deep breath, and their chemical receptors may not do a good job of detecting this need. Adding oxygen stabilizes infants' sleep apnea; this is not a permanent problem.

Obstructive Sleep Apnea

Obstructive Sleep Apnea (OSA) is common in both adults (41%) and children (57%) with PWS. In the general population, people with this condition are often obese, and the obstruction is due to a heavy neck pressing on the windpipe. The authors explain that obstructive sleep apnea in PWS can be caused by the facial features that are so typical of the syndrome – small chin, small nose, and small mouth. Add weak muscles in the neck and mouth, and thick saliva, and there can be OSA without obesity, and without any symptoms, such as snoring. If the tonsils and adenoids are large, removing them often improves sleep quality, but these other PWS features may mean breathing during sleep is not completely normalized even after removing the tonsils. Another sleep study may be needed.

Respiratory Infections

We know this is a challenge at all ages. These authors remind us of the high risk of aspiration (fluids going into the lungs instead of down the throat) and recommend obtaining a videofluoroscopy (also recommended by research funded by PWSA (USA)) if a child has recurrent chest infections **or** these signs while feeding: noisy or “wet-sounding” breathing during or after feeds, eye reddening/watering, color changes, apnea or changes in heart rate related to feeding.

In the summary, the authors support the early use of growth hormone, as they noted the improvements in many areas of respiratory health for children who are on GH. This article may be helpful to share with your health care providers.

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