

## **Hidden Swallowing Problems in PWS: Subclinical Dysphagia in Persons with Prader-Willi Syndrome**

*Gross RD, Gisser R, Cherpes G, Hartman K, Maheshwary R. 2016. Subclinical dysphagia in persons with Prader-Willi syndrome. Am J Med Genet Part A 9999A:1–11.*

PWSA | USA fully funded this recently published research, which identified significant swallowing problems (dysphagia) in persons who have PWS. Our thanks to the authors for this important research, and we look forward to further studies to help us understand how to protect the health of persons with PWS.

The term “subclinical” means “without symptoms”. This means that problems can exist without anyone noticing unusual swallowing and without the individual being aware of any problems. While we have long been aware of the high risk of choking and the increased incidence of pneumonia in our population, the reasons have been unclear – and this has prevented us from knowing what actions to take for prevention.

Breathing and swallowing are complicated and connected actions, coordinated by numerous areas in the brain. Muscle strength and sensory skills are needed for effective swallowing. Most healthy people exhale (breathe out) after a swallow is completed, which helps protect the lungs. Try it yourself – it just happens naturally. But this may not be the case for everyone with PWS.

In this study, thirty individuals between ages 4-55 years underwent a radiology procedure called videofluoroscopy. They were tested while swallowing a thin watery liquid, and then a small cookie.

- 66% of subjects still had liquid in their throat after they were finished swallowing. Normally there should not be any left after a swallow.
- In eating the cookie, almost every subject had some bits of the cookie left in their throat after their first swallow. These people were unaware of this leftover food.
- All subjects had slow passage of the cookie into the stomach; it did not make its way down at a normal speed or in a normal pattern.
- 30-40% of the swallows recorded were followed by inhaling, not exhaling, which increases the risk of food or liquid getting into the lungs.

PWSA | USA recommends that parents mention this research to their healthcare provider, and that you observe the swallowing behavior of your loved one with PWS. If you are working with a feeding specialist, they need to read this research as well. While we have yet to prove that swallowing studies are needed in every individual with the syndrome, such studies are essential if there are symptoms such as choking and coughing episodes, reflux and regurgitation, and lung infections. If in doubt, ask your medical provider for a referral to see a swallowing specialist or gastroenterologist.

**Based on this initial research we recommend that persons with PWS:**

- Pace and chase – slow down speed of taking bites, and take a sip of liquid between bites
- Eat with other people rather than dining alone
- ALWAYS eat at a table, sitting in a chair
- Have smaller meal size with planned snacks rather than big meals
- Cut food into small pieces; select easy to chew foods, lower in fiber
- Try toothpaste like Biotene which may improve saliva flow
- Stay hydrated between meals
- At the end of the meal, stay upright at the table.
- Have a drink to “flush” the esophagus.
- Be active for a while after every meal. Do not eat and then lie down; have the last meal well before bedtime to let gravity do its job

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