

Pilot Study Examining Swallowing Function in Persons with Prader-Willi Syndrome

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(More details on the results of this important grant sponsored by PWSA (USA) cannot be published until after it is presented at a major conference in March which will follow up with a formal publication, but we have permission to share the following with our members.)

Conclusions: Persons with PWS are highly likely to have an undetected swallowing problem that places them at risk for asphyxiation of a food bolus (choking), and they require a specific type of swallowing evaluation. A clinical or bedside evaluation is not sufficient to detect dysphagia in this population. Videofluoroscopic assessment of swallowing function that includes the esophagus is necessary. Although it is important to rule out any disease process or anatomic problem that may exist, a traditional barium esophagram alone will not suffice because the procedure does not use solid food or natural eating positioning, etc. The traditional videofluoroscopic swallowing study (VFSS) which is also known as the modified barium swallow (MBS) or "cookie swallow" will not suffice because it does not always include the esophagus. Therefore, whoever conducts the study must also examine for esophageal clearance using food items. The other instrumental method that is commonly used to evaluate swallowing function is called FEES for fiberoptic endoscopic evaluation of swallowing. This type of exam cannot examine esophageal clearance and, therefore, will be inadequate for persons with PWS.

The recommendation for PWS-specific videofluoroscopic exams may not be widely accepted without offering compensatory strategies that have been shown to be effective. Clinicians may alter diets and eliminate solid food unnecessarily when significant amounts of residue are observed. Most speech pathologists and radiologists may not know how to compensate for esophageal dysphagia and may make reflux-related and unnecessary recommendations such as 6 small meals per day.

Current recommendations: Drinking SIPS of liquids during a meal or snack (i.e. liquid washes) MIGHT clear the mouth and throat of any remaining, undetected residue. Taking additional saliva swallows may also be beneficial in clearing food residue. Any person with PWS who complains of food sticking in their throat or chest, is diagnosed with rumination, reports ongoing episodes of regurgitation, has a choking history, needed the Heimlich maneuver, or shows other signs of dysphagia should give the above information to their physician or speech pathologist. Doctors and therapists can also contact Roxann Diez Gross, PhD, CCC/SLP for management recommendations. She will work with physicians and speech pathologists to ensure that proper evaluation and management occurs.

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