Persons with Prader-Willi syndrome (PWS) have a higher than average death rate due to “respiratory events.” This can mean many things – choking on food, aspiration of liquids with swallowing, obstructive apnea, and pneumonia. Pneumonia is an infection of the lungs that can lead to lung failure and the need for mechanical ventilation (“breathing machine”), as the tiny air sacs in the lungs fill up with fluid. Pneumonia can be the result of a virus or bacteria. Inflammation of the lung (pneumonitis) is often confused with pneumonia. Pneumonitis occurs with aspiration of chemicals, typically acid, food or other secretions from the stomach. We don’t know how often pneumonia or pneumonitis occurs in persons with PWS, or what type of pneumonias are most common.

Healthy breathing requires brain control (knowing when to breathe deeply) as well as muscle strength – muscles of the mouth, throat, diaphragm and ribs. Muscle strength and brain control to synchronize the various muscles are very important in coughing, one of the most important protections against pneumonia and pneumonitis. Both brain control and muscle strength are weaker in persons with PWS. Because of the emerging evidence of aspiration (liquid going to the lungs, not to the stomach) with abnormal swallowing, we know that lung health can be easily compromised in individuals with PWS. If your loved one has experienced frequent “lung infections,” abnormalities of swallowing are important to consider as the cause – with liquids entering the lungs rather than being swallowed.

While most pneumonias are caused by viruses, there are immunizations available to decrease the risk of certain bacterial pneumonias. Prevnar13 is very effective in preventing pneumococcal pneumonias, one of the most common causes of bacterial pneumonia. Most children born after 2001 have already been immunized with Prevnar13 and Hemophilus influenzae type B (HiB conjugate vaccine), which works against another common cause of bacterial pneumonia, as a routine part of childhood care.

Prevnar13 is also recommended for people with certain underlying health conditions including chronic lung disease, diabetes and heart disease, as well as diseases that decrease immunity to infection. Whether the health challenges in PWS are sufficient to
warrant giving this vaccine to all individuals with PWS who may have missed these childhood vaccinations is unclear.

A second vaccine, Pneumovax, protects against additional strains of the pneumococcus. This second vaccine is currently recommended in the elderly and for children and adults with the same underlying health conditions listed above (lung disease, diabetes, heart disease, immune compromised diseases). Check with your doctor to see if these immunizations have been given, and if not, whether these would be recommended now.

Here is the CDC guideline to discuss with your provider:
https://www.cdc.gov/mmwr/preview/mmwrhtml/mm6140a4.htm

For more information about pneumonia, visit this link:
http://www.thoracic.org/patients/patient-resources/resources/what-is-pneumonia.pdf

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