Physical Health Issues and the Aging Adult with Prader-Willi Syndrome

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(This is the second in a series of 3 articles that shares information about aging in adults with PWS with a focus on health.)

Many of the physical aspects of Prader-Willi syndrome impact body systems that can result in chronic health problems with an increased risk of complications as a person ages. Obesity impacts most (if not all) aspects of health. Low muscle tone and spine deformities can contribute to respiratory compromise as well as mobility issues. Hormonal abnormalities, low activity levels and nutritional deficiencies can result in low bone density, and an increased risk of fractures. Sedentary life style may contribute to lower extremity swelling and poor circulation. Diabetes mellitus is common and contributes to problems with eye sight, wound healing and other vascular issues. Constipation and other bowel problems may result from diet and low fluid intake as well as poor bowel habits. The following are five of the most common health concerns being seen in adults with PWS as they age. Prevention and management suggestions are also included.

Respiratory Insufficiency is the major cause of death in adults with PWS triggered by acute or chronic lung infections – not cardiopulmonary disease as originally believed (Eiholzer and Lee 2006). Spine deformities including scoliosis and kyphosis (“hunchback” deformity) can impact lung expansion and put a person at greater risk for respiratory compromise. Sleep apnea is common and can increase morbidity and mortality concerns especially when an adult is undergoing a surgical procedure. In addition to preventing and treating obesity, aggressive measures should be taken to prevent and manage lung infections and chronic lung conditions. Discussions with health care professionals should take place on the administration of annual influenza immunization as well as the pneumonia vaccine. Prompt evaluation of upper respiratory infections may necessitate initiation of antibiotics; encouragement of fluids; assurance that the adult is kept moving and expanding their lungs. Evaluation and treatment of sleep apnea is important. Extra care and monitoring may be required if the adult is undergoing a surgical procedure. Follow up with medical specialists is important.

Low Bone Density, Degenerative Changes and Mobility Issues can affect the aging population in general. The normal aging process results in decreased hormone production, arthritic changes and a combination of other risk factors that contributes to mobility issues and increases the risk of falling. Aging adults with PWS have a higher risk because many of these issues are inherent to PWS. Today, the use of growth hormone and supplementation of estrogen and testosterone in adults with PWS is more common. Calcium and Vitamin D supplements along with a greater focus on proper nutrition are also more common in an effort to prevent osteoporosis. Closer monitoring of bone density and the use of appropriate treatment is now becoming more prevalent. Weight bearing activities as well as medications are being tried to treat osteoporosis. Documentation of osteoporosis should be made before initiating these medications. Many of these medications have significant side effects. An increasing number of adults with PWS are also undergoing orthopedic surgeries to treat degenerative joint problems. Keeping a person mobile is critical. When an adult with PWS becomes dependent upon a wheel chair, their risk of developing other medical emergencies (blood clots in legs and/or pulmonary emboli) is greatly increased.

Back Row: Kate (43 yrs) And Jeff (48 yrs)
Front Row: Cindy (53 yrs) Jim (44 yrs)
It is also important to create environments which prevent the risk of falls. Guard rails may need to be installed in bathrooms and other areas. Throw rugs should be removed. Canes and walkers may be needed to stabilize balance. Proper fitting shoes with a good tread can also help. All medications should be assessed for their impact on balance. Proper lighting to prevent falls (especially at night) should be used. Vision should be checked and bi-focal glasses may need to be replaced by two pairs of glasses. In many cases, bi-focal lenses can alter depth perception and increase the risk of falling. Residing in a 1-story home or home where ramps and other measures to assist with mobility should be considered.

Peripheral Vascular Problems including ankle and lower extremity swelling and problems with blood pressure are being reported in many aging adults with PWS. These types’ of circulation problems are often seen in diabetes. Vascular problems also impact wound healing and can result in leg ulcers and cellulitis. If the adult with PWS has a problem with skin picking, these problems can become very severe. Preventing circulation problems is optimal. Walking is one of the best activities to promote lower extremity circulation. Prolonged sitting should be avoided. When sitting, make sure feet are resting on the floor or stool – avoid “dangling legs”. In severe cases, special stockings may be prescribed. However caution must be used to make sure these stockings do not impair circulation further or cause problems with wound healing. In some cases, these stockings have been used as a barrier to skin picking. Prompt attention to any cuts or sores in lower extremities and feet should take place. Avoid open toe shoes – especially thong-type sandals to prevent injury to skin. These types of shoes may also cause balance and stability problems. Nail care is imperative and should be done by a health care provider if the adult has diabetes. Hydration is important to assist with maintenance of blood pressure. Weight loss, exercise, lower salt intake, frequent monitoring of BP measurements are also important. Medication to lower elevated BP may be needed.

Complications of Diabetes can often be severe and impact every body system. According to the American Diabetes Association, the aging population, especially those who are overweight, may develop Type 2 diabetes. High levels of glucose in the bloodstream damages blood vessels throughout the entire body. This results in vision problems, kidney disease, and heart and vascular problems including stroke, high blood pressure and heart disease. Diabetes also impacts circulation and impairs healing – especially in lower extremities. Adults with PWS can be at increased risk to develop diabetes especially if they are overweight. Many persons with PWS are diagnosed with diabetes at a young age. This can add years of damage to blood vessels. Screening, early diagnosis and treatment are all important. The American Diabetes Association recommends screening with a blood test called Hemoglobin (Hgb) A-1C. This should be done 1-2 times a year depending on risk factors. Annual blood lipid and cholesterol testing is suggested. Diet and weight management is imperative. An active life style including exercise is also important. Once diagnosed with diabetes, it is important for the adult with PWS to adhere to any diet and/or medication regime that is prescribed.

Changes in Bowel Habits – Constipation is another health issue that many adults face as they grow older. Persons with PWS of all ages seem to struggle with gastrointestinal issues; constipation is common. Many of the same interventions used to manage other health issues also help this problem. Exercise, an increased activity level, adequate fluid intake, high fiber foods are just a few suggestions. Probiotic foods and supplements have also been beneficial. In many cases, daily fiber and/or laxative supplementation may be needed. Staff may need to monitor the frequency and consistency of a resident’s stool. Higher incidences of intestinal obstruction and ileus (slowing or stoppage of the intestine) have also been reported. Caution should also be used when using medications that slow the intestine – especially medicines used to treat diarrhea. Diarrhea can be a symptom of intestinal blockage.

Summary This article summarizes just a few of the more common physical health issues that are being seen and reported in aging adults with PWS. Many of the issues are the same as all of us age. The goal is to minimize problems so aging adults can maintain a higher quality of life.