Aging in the Adult with Prader-Willi Syndrome – A Focus on Physical and Cognitive Health

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(This is the first in a series of 3 articles that will begin sharing information about aging in adults with PWS.)

As adults with Prader-Willi syndrome (PWS) are aging, we are beginning to learn more about some of the common health issues many are facing. Researchers are starting to investigate the aging process in persons with PWS. It can be challenging to distinguish between health concerns that are inherent to PWS versus concerns that we all face as we age. Family history is also contributory to many health concerns and must also be considered.

The focus of this article is to highlight some of the more common physical and cognitive health issues being identified by researchers as well as professionals who are working with this aging population. According to Eiholzer and Lee (2006), some of the main medical concerns being reported in adults with PWS include physical disability (resulting in mobility issues), respiratory insufficiency, osteoporosis and scoliosis. The major cause of overall death in adults with PWS was respiratory insufficiency triggered by acute or chronic lung infections – not cardiopulmonary disease as originally hypothesized.

During the 2010 International Prader-Willi Syndrome Organization conference in Taiwan, researchers from The Netherlands shared findings from their study of 102 adults with PWS ages 18-66 years. Sinnema, Maaskant, et al. reported a majority (56%) were obese. In addition, the following health conditions were common - leg edema (56%), erysipelas - an acute streptococcal infection of the skin (38%), constipation (38%), diabetes mellitus (17%), osteoporosis (16%), pneumonia (14%) and hypertension (9%). These researchers also shared a high incidence of psychiatric episodes.

According to Rena Mills, RN Health Services Coordinator for Prader-Willi Homes of Oconomowoc (PWHO) who provides residential services to over 80 adults with PWS, premature aging seems to be a concern. By the time their residents are 40 years old, many have medical problems like those who are 60 years old. Mills also identified many of the same health concerns as mentioned above – osteoporosis/osteopenia, constipation, intestinal blockages, pneumonia, diabetes, cellulitis, low sodium levels and high/low blood pressures.

Along with medical challenges associated with the aging process, changes in cognitive function are also being examined to determine if the early onset of dementia may be an additional concern for adults with PWS.

Studies have shown that adults with Down syndrome (DS) are more likely to develop Alzheimer-type dementia than their peers without DS (Silverman, Zigman, Krinsky-Mchale & Schupf, 2008). Still other research suggests the prevalence of dementia in adults with intellectual disabilities (ID) who do not have DS is higher than the general population (Strydom, Chan, Fenton, et al., 2010). What does this mean for aging adults with PWS?

In March 2009 the State of Science on Dementia in older adults with Intellectual Disabilities reported, “aging and cognitive functioning has been studied in so few of the ID syndromes other than DS.” Sinnema and her research team from The Netherlands contributed to this critically needed knowledge base with their 2010 case study of a 58-year-old woman with PWS. The resulting assessment scores supported the presence of dementia.

PWHO currently supports 27 (33%) individuals with PWS in their 40s and 2 (2%) in their 50s; one woman diagnosed with dementia at age 54. An assessment procedure was developed to measure cognitive changes as they age, so the possible onset of dementia can be detected and early treatment implemented. This data will also be a future resource for understanding how aging influences the cognitive process in PWS.

Knowing what physical and mental health issues are common allows us to initiate preventive measures and plan appropriately for future care needs. There are two more articles in the series- one will focus on five of the most common health issues being seen in aging adults with PWS along with strategies to prevent and/or minimize their effects. Another will focus on dementia research, assessment, and treatment. This collaborative series will provide insight into how aging affects adults with PWS and the approaches to ensure they can remain healthy, productive members of their communities.