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Cognitive Changes in the Aging Adult with Prader-Willi Syndrome

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

With the increased awareness of Prader-Willi syndrome (PWS) leading to early diagnosis and treatment, life expectancy has grown dramatically over the years. When I first began working with adults with PWS in 1989, I remember one of the first things I learned was that many did not live past their teenaged years due to obesity-related medical complications. Back then, I worked with 15 people diagnosed with PWS with most being in their early twenties. The PWSA (USA) Web site on the *Basic Facts about PWS* reports, "If weight is controlled, life expectancy may be normal, and the individual's health and functioning can be maximized."

Twenty-plus years later, I am thankful to still work with many of those individuals, as we step into our forties together. We are entering the uncharted years of aging in PWS with the aim to achieve the same outcome as the frontier movements of a*wareness*- early detection and treatment- to ensure continued quality of life well into the golden years.

This article will explore what we have learned so far about how aging may influence cognitive processes, specifically- reviewing current research, assessing cognitive changes, and predicting effective treatment strategies.

What is Dementia?

According to Wikipedia, "dementia is a serious loss of cognitive ability in a previously unimpaired person, beyond what might be expected from normal aging." There are several types of dementia, but the most common is Alzheimer's (AD), which occurs in "50-80 percent of dementia cases." It is a progressive disease with symptoms getting worse over time. (*What Is Alzheimer's?* Retrieved from http://www.alz.org). For the purpose of this article, the reference of "dementia" pertains to the Alzheimer's type.

Dementia Prevalence:

In the general population, Alzheimer-type dementia usually occurs after the age of 65 (alz.org). Dementia studies are not as abundant in the area of intellectual disability (ID) syndromes compared to the general population studies, but in March 2009 the State of Science on Dementia released a comprehensive review of studies from 1997-2008 relating to aging and intellectual disabilities. The review was compiled by the International Association for the Scientific Study of Intellectual Disabilities (ASSID) Special Interest Research Group on Ageing and Intellectual Disabilities.

Some of this research is summarized below:

Earlier research suggested that "the prevalence of dementia (particularly AD) among the ID population may differ from the general population, at least in specific subgroups such as Down Syndrome (DS)" (Zigman, Schupf, Haveman, & Silverman, 1997). Later studies showed that the prevalence of dementia in the ID population was 6.1% in those aged 60 and over, which is comparable to the percentage in the general population (Janicki and Dalton 2000). The same study showed that adults with DS had much higher rates of dementia- 56% for those 60 and older.

The mean age of dementia onset:

- General population: 67 years
- Intellectual Disability population excluding DS: 67.2 years
- Intellectual Disability with DS: 52.8 years
- Intellectual Disability with PWS: Unknown

Where will the average age of dementia onset for adults with PWS fall on this statistical spectrum?

PWS Research Overview:

There was one PWS study cited in the State of Science on Dementia review completed by Sinnema, Maaskant, Van Schrojenstein, et. al. in 2008, which evaluated 74 individuals with PWS ages 18-63 years old and reported no cases of dementia.

At the IPWSO conference in Taiwan last year, several studies examining aging in PWS were presented. Whittington and Holland reported the results of their study *Recent Mortality Rates and Risk of Dementia in PWS.* They found that out of the 26 individuals who were 40 years and older, 22 showed no signs of dementia; one case of mild-moderate dementia; one potential case of mild dementia; and evidence of cognitive decline in one person.

Sinnema and her research team from The Netherlands presented a case study of a 58-year-old woman with PWS. The assessment scores supported "the presence of dementia in very late stages."

The researchers agree that more studies are needed to better understand the effects of aging in older persons with PWS. This knowledge will help determine the most beneficial ways to minimize and treat the symptoms.

Preparing for the Future:

Prader-Willi Homes of Oconomowoc (PWHO) currently supports 81 adults in residential care, in which 27% are in their 30s, 33% are in their 40s, and 2% in their 50s. There is one woman who was diagnosed with dementia almost three years ago at the age of 54.

According to the article *Stopping Alzheimer's Before it Starts* posted on www.medicinenet.com, "the prevention process should begin at approximately age 40. That is because on the average, Alzheimer's disease begins 30 years before the first symptoms appear." Because it is unknown at this time when onset may occur, PWHO has taken the approach of evaluating cognitive changes over time with the aim of early detection and treatment. The fundamentals of this procedure are as follows:

1. All clients at PWHO will have a mental status baseline score from "*The Short Portable Mental Status Questionnaire*" (SPMSQ).
   Date of assessment and cognitive range (normal mental functioning, mild, moderate or severe cognitive impairment) corresponding with score on the questionnaire will be documented.
2. All clients thirty-five years and older will complete the SPMSQ every year. If there is a change in range, further assessment will occur, as needed (i.e. retesting, additional evaluations, psychotherapist consult, psychiatrist consult, neurologist consult).
   The age of 35 for annual assessment was based on speculation- if aging in PWS would be comparable to aging in Down syndrome, with potential onset of dementia in the mid 30s.
3. Clients that present with any significant behavioral and/or mental health changes should complete the SPMSQ to assist in determining the possible causes of the decline in functioning.
4. If cognitive ranges decline and other medical or mental health causes have been ruled out, more in-depth assessments that measure cognitive and daily functioning will be administered.

To date, the assessment scores have shown no indication of early onset of dementia in the 30s or 40s. In diagnosing dementia and evaluating potential risk for early onset, an important contributing factor is family history, as was the case of the 56-year-old woman with dementia at PWHO.

Treatment:

The National Institute on Aging (NIA) is part of the National Institutes of Health, a Federal agency for AD research. The following statement is on their Web site, www.nia.nih.gov.

AD is a complex disease, and no single "magic bullet" is likely to prevent or cure it. That's why current treatments focus on several different issues, including helping people maintain mental function, managing behavioral symptoms, and slowing AD.

What are the recommendations for daily care of somebody diagnosed with AD? On WebMD, *Alzheimer's Disease: Daily Care of the Alzheimer's Patient,* the following suggestions are outlined:

- Provide physical exercise, proper nutrition, good general health, and socialization
- Plan daily activities to help provide structure, meaning, and a sense of accomplishment. It is always best to establish a routine with which the person can become familiar.
- Choose the best times to do activities according to the part of the day when the person is usually at his/her best.
- As functions are lost, adapt activities and routines. Keep activities familiar and satisfying, and keep instructions simple.
- Allow the person to complete as many things as possible by him/herself, even if you have to initiate the activity.
- Provide "cues" for desired behavior. For example, if you label a drawer according to what it should contain, the person is more likely to put things in the correct place.
- Keep the individual out of harm's way by removing things that could endanger them.
- As a caregiver, it is important to understand and act according to your own physical and emotional limitations. Be sure to take care of yourself, and allow yourself periods of rest and relaxation.

Exercise, nutrition, socialization, structure, routine, visual cues…does any of these look familiar? As a PWS community, we are already one step ahead of managing the symptoms of dementia, as they are the same approaches that are effective in supporting persons with PWS.

In addition to daily supports, medication has also been used to manage and potentially slow down AD. The State of Science on Dementia reported that "Donepezil is

the most commonly used anti-dementia drug used in intellectual disabilities to treat dementia, and there is some evidence of Rivastigmine." The woman diagnosed with dementia residing at PWHO has had a positive response to receiving Donepezil (Aricept).

As treatment strategies are planned, it is important to remember that dementia is a progressive disease in that "lost skills cannot be regained" (www.webmd.com). It is crucial to incorporate and continue with activities that are physically and mentally stimulating per individual's preference.

Summary:

What we know is that we need to know more in order to better understand how aging will progress in persons with PWS. There is not enough information to make any conclusive statements about how or when dementia will present.

Based on my research of the literature and what I have observed at PWHO, I see no evidence to suggest that dementia in PWS is comparable to Down syndrome with the potential age of mid-30s onset. More studies are needed to evaluate whether or not the onset could occur earlier than what is predicted in other ID syndromes and the general population.

Like PWS, dementia has no cure, but with early detection and the necessary supports in place, we can provide the best opportunity for the physical, mental and emotional well-being of the individual.

As we make this venture into unknown territory together, we will continue to do what the PWS community does best- *unite forces*- by sharing experiences, contributing to the knowledge-base, applying what is successful, and growing together- stronger and more informed than the day before.