PWSA UK
PRADER-WILLI SYNDROME ASSOCIATION UK

Ageing in PWS 40+ years

Introduction

We know very little about older people with PWS, partly because in the past they rarely reached much older than 40. Nowadays, with better management, they are living longer, but the number of people with PWS known to the Association who are over 50 is still relatively small.

We are currently aware of 164 individuals with PWS aged 40 or older. Of these, only 56 individuals are aged 50 or older and of these, five people are aged 60 or older, with the oldest being 63. In recent times, the oldest person we know of died at the age of 68, but there is a published report of a woman with PWS living to age 74.

Signs of ageing in PWS

The PWSA UK Residential Care and Supported Living Forum (which consists of senior managers from these establishments) have discussed the signs of ageing that they have noticed in their services users with PWS from late 30s onwards:

- Older facial features (rapid occurrence often beginning in the mid to late 30s) – looking far less youthful and skin ageing
- Stooping posture
- Decrease in stamina
- Decrease in mobility
- Improved adaptive skills
- Less intense and less frequent behaviours
- Activities may lessen or change – as they do in the general population
- Motivation – variable between individuals – may go up or down
- Decrease in ability to process verbal information
- Possible decrease in language and listening skills
- Weight becomes more difficult to lose (possibly just females?)
- Chest infections take more time to clear

Continued ...

Can you help?

Please let us know of any physical or mental problems in your son or daughter which have occurred after the age of 40, or changes in behaviour – for better or worse.

This will help increase our understanding of what happens to people with PWS in middle and old age.

Further observations about ageing in PWS from the PWSA UK Residential Care and Supported Living Forum

- Although some members of the forum felt there was little or no difference in the ageing process to the general learning disability population, others felt that the ageing process seems quicker in those with PWS re physical appearance and also problems of older age in general population appear earlier.

- PWS may mask other issues which go alongside old age generally, and there may be under-reporting by individuals due to high pain threshold or not wanting to go into hospital.

- Good dietary management could be a factor in those who show fewer signs of ageing.
- Hormone regulation could be more difficult. We have little information about what happens at male and female menopause. Are replacement hormones required, or at what point should existing sex hormone treatment be discontinued?
- Lifelong anxiety levels causing greater stress may cause earlier ageing symptoms.
- Long term structured environment (usually in residential care) may be a factor in decreased challenging behaviour, rather than old age itself.
- We should not assume that because a person with PWS has enjoyed an activity or interest when younger that they will still enjoy it as they age – this is true for all people, with and without PWS.

Two summaries follow from research carried out about older people with PWS.

Ageing in Prader-Willi Syndrome: Twelve persons over the age of 50 years

Sinnema M, Schrander-Stumpfel CTrm, Maaskant MA, Boer H, Curfs LMG. 2012. (Am J Med Genet Part A 158A:1326-1336)

This study looked at 12 individuals with PWS aged 50 and older in the Netherlands. Individuals with PWS aged 18 – 49 years were used as a control group.

- Age range 50 – 66 years
- 5 males, 7 females
- 4 with deletion, 8 with mUPD (maternal disomy)
- 11 in community or residential facilities, one living at home with elderly mother
- Mean age at moving to residential facility – 19.4 years
- Mean BMI in persons with deletion was significantly higher than persons with mUPD – three of the latter had a BMI under 25.
- Mean maximum BMI was 36.5, with a range of 23.6 – 44.4
- 3 people smoked (cigarettes, pipe, cigars)

Health issues

- One woman died, aged 65, shortly after data collection, due to lung problems
- Half had diabetes mellitus, mean age of diagnosis 41.6 years
- No one in this group had received sex or growth hormone therapy
- No one had epilepsy or cancer

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Physical health problems	Total prevalence (N)
Hypertension	3/12
Stroke	3/12
Diabetes	6/12
Pneumonia	3/12
Excessive daytime sleepiness	8/12
Constipation	5/12
Reflux	2/12
Aenemia of unknown origin	2/12
Kidney problems (congenital)	1/12
Osteoporosis	2/12
History of any fracture	6/12
Primary amenorrhea (no menstruation at any time in life)	1/7
Scoliosis	5/12
Foot problems	10/12
Hip problems	2/12
Oedema	9/12
Erysipelas (skin infection)	6/12
Varices (varicose veins)	3/12

Health checks

The Dutch study underlines the need for regular health checks for adults with PWS. In particular:

- Cardiovascular disease
- Diabetes
- Dermatological problems
- Orthopaedic problems
- Sleep problems
- Osteoporosis

All usual age-appropriate screenings should be carried out (eg hearing, eyes, cancer etc, with possible exception of cervical smear tests for women with no history of sexual activity. You can give your son or daughter's GP a copy of Information for GPs

. This explains the importance of health checks.

Under-reported or undiagnosed problems

- Sleep problems and osteoporosis are likely to be under-reported and deserve special attention.
- Diagnosis of pneumonia is frequently delayed in older adults with PWS because of absence of fever.

Other points from the Dutch study

There were relatively more individuals in this study in the moderate-severe learning disability range.

These individuals may have required earlier intervention by being placed in structured residential settings at an earlier age, which in turn could have contributed to their longevity because of better weight management and prevention of serious medical complications.

The researchers hypothesize that there may be premature aging in PWS, especially where no sex or growth hormone is given. They state that aging in PWS starts at 50 or younger.

Ageing in people with Prader-Willi syndrome: mortality in the UK population cohort and morbidity in an older sample of adults

JE Whittington, A J Holland and T Webb – Psychological Medicine, Cambridge University Press 2014

This later research was a follow-up of previous research into a group of people with PWS of all ages in the UK which was carried out from 1998 to 2000.

Researchers found a mortality rate of at least 7/62 over 9 years (1.25% per annum; 20 untraced). Age at death was between 13 and 59 years.

Out of 26 people in this research group aged 40 or over, 22 showed no evidence of dementia, while the remaining four all had possible symptoms. All four were female, of maternal uniparental disomy (mUPD) genetic subtype, and have a long history of psychotic illness.

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