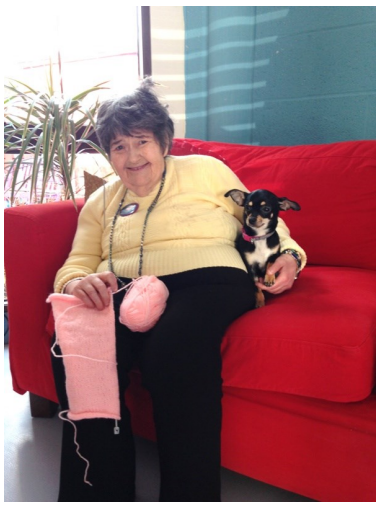
Ageing in PWS

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 194 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 68.

Anecdotally we have reports of a general slowing down in older people with PWS, but this is also evident in the general population.

We also have reports of dementia in a few people, but again these are anecdotal.

Improved behaviour has been noted in some, but not in others. In some cases, particularly in residential care situations, the interest in food and desire to eat has also diminished.

The two summaries on the following pages are from research carried out about older people with PWS.

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.

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

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

Comparison with younger people with PWS

- Functioning, behaviour and care dependency revealed worse functioning in the older than the younger groups.
- Scores in the older group were significantly lower on the following items:- personal hygiene, dressing, eating, being ambulant, mobility, grooming, memory, orientation, sleeping difficulties, physical complaints, hearing, vision, dependency on medical care and care dependency (note that deterioration in some of these areas would also be evident in the over 50 population without PWS)
- Scores on lack of self-confidence or poor self-esteem were statistically significantly lower than those for people under 50.
- No significant difference in eating behaviour between over and under 50s.
- No reduction in behaviour problems as people got older, as opposed to findings in some previous studies. More behaviour problems were noted in those with mUPD, which may overlap with the psychiatric disturbances that this sub-group displays.

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** included in this pack. This explains the importance of health checks.

Other points

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.

Psychiatric illness

- *No-one with deletion (4 people) had a psychiatric illness*
- *7 out of 8 mUPD had a history of psychiatric illness:*
- *Bipolar disorder with psychotic symptoms – 3*
- *Psychotic illness – 2*
- *Depressive illness without psychotic symptoms – 1*
- *Bipolar disorder – 1*
- *All with mUPD used psychotropic medication.*
- *One woman presented with symptoms highly suggestive of dementia*

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.*

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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