Aging in Prader–Willi syndrome: Twelve persons over the age of 50 years

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Abstract

The life expectancy of persons with Prader–Willi syndrome (PWS) has increased in recent years. Because of the paucity of reports on older persons with PWS, the natural history, the onset, and type of age-related problems are poorly understood. Twelve persons with a genetically confirmed diagnosis of PWS aged over 50 years are described (4 deletion; 8 mUPD). Data on physical, behavioral, psychiatric, and aging characteristics were collected through semi-structured interviews with the individuals with PWS and their main carers. Cardiovascular diseases, diabetes, dermatological, and orthopedic problems were common physical complaints in older people with PWS. Functioning in activities of daily living, psychological functioning, physical functions, and care dependence were substantially worse in the older age group (50+) compared to the control group (18–49 years). Seven out of eight persons with mUPD had a history of psychiatric illness. Behavioral problems were observed in the older age group. Given the combination of age-related physical morbidity, physical appearance, behavioral and psychiatric problems, and functional decline in our cohort, we hypothesize that premature aging occurs in PWS. The care for older people with PWS requires a lifespan approach that recognizes the presence, progression, and consequences of specific morbidity. Special medical surveillance of people with PWS from 40 years onwards would ensure that intervention and support is offered with respect to specific areas of decline at the earliest possible time.

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