Abstract

This study is part of a larger UK-wide study investigating psychiatric illness in people with Prader-Willi syndrome (PWS), and describes the longitudinal aspect of psychiatric illness, in particular psychotic illness, and examines the use and role of psychotropic medication. A total of 119 individuals with genetically confirmed PWS were included in the study. An informant-based questionnaire was administered for each participant to screen for a history of psychopathology. Those who screened positive were visited at their homes to obtain further information. This assessment included a full psychiatric history and mental state examination using the Psychiatric Assessment Schedule for Adults with Developmental Disability and the Operational Criteria Checklist for psychotic and affective illness to collect information regarding phenomenology and course of illness, and a modified life events questionnaire. At the end of the study period, informant-based telephone interviews were again carried out, up to 2.5 years after the initial screening. Information regarding medication usage was collected. The results confirm previous findings that psychiatric illness in people with PWS resembles an affective disorder. Individuals with the maternal uniparental disomy genetic subtype had a more severe course of illness than those with the deletion genetic subtype in terms of a greater risk of recurrence, more episodes, higher incidence and a possibly poorer response to medication with more side-effects. Individuals with a recurrent episode during the follow-up period had a poorer course of illness. Selective serotonin reuptake inhibitor medication is frequently used, and beneficial effects may reflect fundamental pathological processes in PWS. Mood-stabilizing medication was found to be of little benefit and reasons for this are examined. The longitudinal course of psychiatric illness and response to medication in people with PWS is fully described. Further research is needed regarding the effect of psychotropic medications, particularly mood-stabilizing medication. These data will enable informed decisions to be made regarding management options and provide information on the possible long-term outcome of illness.