the wide variety of clinical pictures seen in case reports. Another hypothesis would be to suggest that each individual carries a particular set of susceptibilities to various psychopathological phenomena. The findings that sleep deprivation may induce mania in those with bipolar affective disorder (Wehr et al., 1982, 1987; Wehr, 1991) and may induce florid psychotic episodes in known schizophrenic patients (Koryani & Lehman, 1960; Bliss, 1967) would support such a hypothesis. When there is an absence of a psychosis after sleep deprivation, as has been seen in many case reports of prolonged total sleep deprivation in normal people, it would therefore represent a minimal susceptibility to psychopathology.

Any such hypothesis is important for clinical practice. It would point to a greater need in history taking for awareness of patients' sleep patterns and especially the relationship to illness. Further studies exploring this relationship would be helpful, particularly in areas such as puerperal psychosis where sleep deprivation before illness is common, and there is a similar variability in clinical presentation.

Acknowledgements

I would like to thank Dr S. Mahapatra for his help and for allowing me to report on one of his patients and for the kind assistance of Dr A. Zigmond.

References


Prader-Willi Syndrome and Psychoses

DAVID J. CLARKE

Prader-Willi syndrome (PWS) is associated with an insatiable appetite and (often) other maladaptive behaviours (self-injury, sleep disorders, insistence on routines, and temper tantrums). Psychoses are not a recognised feature. Most affected people have a chromosome 15 abnormality (deletion, disomy, structural rearrangement, etc.). Three people with PWS who developed psychotic disorders in early adult life are described. The nature of the psychoses and the significance of the association are discussed.


Prader-Willi syndrome (PWS) was first described in 1956 (Prader et al., 1956). Affected children are hypotonic and feed poorly as neonates. In early childhood the feeding difficulty resolves and an insatiable appetite develops, resulting in gross obesity unless a strict diet is maintained. Affected adults are of short stature, with hypogonadism, cognitive impairment, small hands and feet, and characteristic facial dysmorphology. Most affected people have a cognitive impairment, and their IQs are typically in the range of 60 to 90. Some people with PWS have low IQs (sometimes apparently as a result of perinatal complications), while others have IQs within the normal range, and a sizeable minority gain sheltered employment.

References


ACKNOWLEDGEMENTS

I would like to thank Dr S. Mahapatra for his help and for allowing me to report on one of his patients and for the kind assistance of Dr A. Zigmond.

REFERENCES

The incidence of PWS has been estimated at around 1 per 10,000 live births and it is associated in about 70% of cases with a cytogenetically detectable deletion of the long arm of chromosome 15 (del 15q11-13). Some affected people have deletions which are detectable by molecular, but not cytogenetic, studies (Robinson et al., 1991). Others inherit two chromosome 15s from their mother (maternal disomy), implying that genomic imprinting may be the genetic mechanism underlying some cases of PWS. Other chromosome 15 abnormalities have also been described in association with PWS. When a deletion is associated with PWS it is invariably of paternal origin. Apart from possible differences in pigmentation, no consistent differences have been found between people with PWS and a chromosome 15 deletion and those without.

Food-seeking behaviour is prominent among adults with PWS. Food stealing (or the stealing of money to obtain food) is not uncommon. Some people with PWS eat items which are usually considered unpalatable (such as discarded, rotten, frozen or pet food). People with PWS have been described as having 'cheerful' personalities, although temper tantrums and rage reactions are also frequently reported in association with the syndrome (Greenswag, 1987; Clarke et al., 1989). One report described the successful use of carbamazepine to treat intermittent explosive disorder associated with PWS (Gupta et al., 1987). Sleep abnormalities and self-injury (through picking at skin) also occur. One study found that over 90% of people with PWS self-injured by picking or scratching skin, sometimes resulting in persistent sores or cellulitis (Clarke et al., 1989). The oldest people with PWS in recent surveys have been in their mid-40s; a common cause of death is heart failure secondary to obesity.

Whitman & Accardo (1989) have suggested that the behavioural and emotional symptoms which emerge during adolescence and early adult life in PWS constitute a "distinct neurobehavioural stage", and that PWS should be viewed as a three-stage disorder: stage 1, dysmorphology, failure to thrive and hypotonia; stage 2, eating disorder and cognitive impairments; and stage 3 intensified eating/weight problems, behavioural and emotional disorders, and psychosocial problems.

Kollrack & Wolff (1966) described a 20-year-old man with PWS who had two brief psychotic episodes with a return to his usual level of functioning between episodes.

Three people with PWS are reported. They had the chromosome 15 deletion characteristic of PWS, and many of the behavioural abnormalities described above. In addition, they developed psychoses in early adult life.

Table 1

<table>
<thead>
<tr>
<th>Case</th>
<th>B</th>
<th>C</th>
<th>D</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sex</td>
<td>M</td>
<td>F</td>
<td>F</td>
</tr>
<tr>
<td>Height: cm</td>
<td>160</td>
<td>147</td>
<td>142</td>
</tr>
<tr>
<td>Weight: kg</td>
<td>71.8</td>
<td>82.5</td>
<td>126.1</td>
</tr>
<tr>
<td>Weight:height ratio (WHR): kg/cm</td>
<td>0.449</td>
<td>0.561</td>
<td>0.888</td>
</tr>
<tr>
<td>Hypogonadism</td>
<td>Yes</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>Dysmorphic face</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Small hands</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Neonatal hypotonia</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Neonatal feeding difficulty</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Marked urge to overeat as adult</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Frequent abnormal loss of temper</td>
<td>Yes</td>
<td>Yes</td>
<td>No</td>
</tr>
<tr>
<td>Self-injury through skin picking</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Chromosome 15 Del 15</td>
<td>Del 15</td>
<td>Del 15</td>
<td>Del 15</td>
</tr>
<tr>
<td>(q11-13)</td>
<td>(q11-13)</td>
<td>(q11-13)</td>
<td></td>
</tr>
</tbody>
</table>

WHR is a measure of obesity corrected for height. For example, a 175 cm (5'9") man with a weight of 70 kg (11 st 2 lb) has a WHR of 0.40.

1. Menstruates regularly. Receiving progestogen treatment for pre-menstrual tension.
2. Menstruated irregularly.
3. Had tempers which were abnormally severe, but not frequent.
and kept asking “¿have you fed the dogs?” (the family had no dogs). The agitation, disturbed pattern of sleep, and unusual remarks continued until the time of the psychiatric assessment.

At interview, B said “They won’t leave off. I know what his name is. Leave off. It’s him. Kevin. I told him to get off my back. He’s kicking me in my leg, my bad leg. It gives way, fluid. I said ‘¿leave off, I want to speak to Jane’. I said to Jane ‘¿forget about Kevin, it’s his fault’. My mind won’t work. I told the tutors”. (Kevin and Jane were students at the college B attended.) On questioning, B said Kevin was kicking his leg (without being present in the room), but could not explain how Kevin could do this. He said that Kevin was also making him lose his voice by “shouting bad language in my ear”. He added that “¿there’s something down there, against my throat, I can’t get fluid down properly”. He was distressed and cried during the interview, and also seemed suspicious and angry. He was orientated in place and person. When well he had little sense of time, but his parents felt he had become less well orientated since the onset of the disorder. He was, however, able to give an accurate account of recent events in college and at home. There was no history of epilepsy or other serious illness. He was not pyrexial, and no organic cause was found to account for his mental state. His parents wondered if more demands had been placed on him at college, but recent changes seemed to be minor and unlikely to account entirely for the sudden onset of psychiatric disorder.

B’s general practitioner (GP) had prescribed a small amount of chlorpromazine, but his parents had been reluctant to give it because they were afraid it would cause dependence. After reassurance, they agreed to give B 200 mg of chlorpromazine daily in divided doses.

He was followed up by telephone and seen again after three weeks. His abnormal mental state had resolved completely ten days after the psychiatric interview and introduction of chlorpromazine. Shortly afterwards the chlorpromazine was stopped. There has been no recurrence of illness during the three years since he was first assessed.

Case 2

C’s early development was typical of PWS (see Table 1), with the change from difficulty feeding to overeating occurring gradually during her fifth year. There is no family history of developmental disorder or delay, or of psychiatric illness.

She attended a mainstream school to the age of seven years, from which time she received education in a variety of special needs settings (moves often occurring because of difficulties with food stealing and consequent weight gain). From the age of 18 years she attended an adult training centre (ATC) and an industrial therapy unit (ITU). She had no other serious medical disorder other than PWS.

One day, aged 24, C returned home from the ITU, pointed to a small piece of metal swarf which had caught in one of her garments, and started to repeat “¿I have not stolen it”. Despite reassurance from her parents, she became agitated and anxious, and started to cry. She was unable to sleep, refused to eat, and spent the entire night sobbing and repeating short phrases including “¿I’m being burned”, “¿I’m being poisoned”, and “¿I’m being drowned” (sic). She gave no reason for these statements. Her GP prescribed diazepam, initially with little effect (C often refused the tablets, or spat them out and concealed them). The agitation, crying, and unusual statements continued for about three days. C then became withdrawn, and would only eat or drink if food or fluid was placed in her mouth. Although previously able to wash and dress herself, she refused (or was unable) to cooperate with washing or dressing while ill. She lost weight. She was suspicious of family members, and kept repeating “¿you’re my Mum, aren’t you?” when she saw her mother. She believed her brother to be her stepfather, and seemed disoriented in time. She occasionally hit her mother for no apparent reason.

After about three weeks, C was admitted to a psychiatric hospital. No organic cause was found for her condition, and she was treated with chlorpromazine in doses of up to 450 mg per day. Initially she spent long periods crawling on the floor, but then started to eat, her sleep pattern reverted to normal, and she made a good recovery. She was discharged after about four weeks. C had no further overt episodes of mental illness. She receives fluphenazine decanoate 50 mg, twice weekly, and dydrogesterone for episodic aggressive behaviour which was found to occur predominantly around the time of her menstrual periods. She remains slightly suspicious and is prone to misinterpret remarks (assuming them to be hostile or derogatory). She is otherwise mentally well, and attends an ATC and ITU regularly.

Case 3

D was abnormally ‘floppy’ and fed poorly in infancy. Other diagnostic features of her PWS are summarised in Table 1. Her older brothers and younger sister do not have PWS or any other developmental disorder. She started to overeat and gain weight around the age of two years. At the age of 15 years, she had an intestinal resection to help control her weight. This was initially successful, but in later years D gained weight again. She did not have epilepsy or any other serious medical disorder other than PWS.

When she was 16 years old her father died as a result of a ruptured cerebral aneurysm. After D left school she attended an ATC.

A few months before her first episode of mental illness she received help from a clinical psychologist for obsessional behaviour, with rituals involving cleaning and the folding of clothes, towels, and so on for many hours a day.

One day in 1985 she came home from the ATC, had her tea, and sat down to watch television. Suddenly, she burst into tears and shouted “¿my kittens have died”. Her mother was unable to convince her that the family did not have any kittens. A few minutes later, D said “¿Turn the heating off. It’ll blow up”. She became extremely anxious and restless, and tried to leave the house. As the evening wore on she became more distraught, and threw objects around. Her mother summoned help from relatives, and her GP visited and sent D to an accident and emergency unit. She was assessed by the medical team, and no physical cause for her sudden change in mental state was found. She was
transferred to a psychiatric hospital. On admission she refused to occupy a bed near an electrical socket because she was convinced it would catch fire. Her appetite and pattern of sleep (after the first day) were normal. She was discharged after two weeks, receiving no medication.

D was subsequently referred to a consultant psychiatrist, who found abnormal beliefs and temper tantrums to be the main problems. She was prescribed thioridazine. The abnormal beliefs persisted – D often believed that members of her family were not who they claimed to be. She developed a habit of checking her mother’s wedding ring to ‘prove’ her authenticity. She became preoccupied with witches and the occult and continued to mention dead kittens, which she believed were buried in a neighbour’s garden. On occasions she seemed to experience auditory hallucinations. Her mother remembers one occasion when she said “Diane next door keeps talking in my ear, she’s saying something to me” and then covered her left ear and suddenly lost her temper. D did not return to her premorbid level of functioning after her illness, and her personality changed. She became suspicious and liable to misinterpretations and abnormal beliefs. On one occasion she refused to take tablets because they were impressed with the letter ‘S’. D was convinced this stood for ‘snake’ and meant the tablets would turn into snakes inside her.

By 1990 her weight had exceeded 120 kg. Because of her limited mobility and other problems she was admitted to hospital. She developed several physical illnesses, including cellulitis and septicaemia as a result of incontinence and skin-picking, and she died some months after admission as a result of a chest infection.

Discussion

The psychoses experienced by the three people described above had many features in common, including a sudden onset (although in one case this was preceded by the development of obsessional behaviours), agitation and anxiety, abnormal beliefs and, in two cases, auditory hallucinations. They differed in outcome; in case 1 the disorder remitted with no residual symptoms; in case 2 the patient’s relatives noticed some persisting changes in personality; and in case 3 abnormal beliefs and auditory hallucinations persisted or recurred. B’s illness (case 1) could be viewed as an affective disorder; many of the symptoms, the outcome, and the family history were compatible with such a diagnosis. The presence of passivity phenomena would not necessarily be inconsistent with a diagnosis of affective disorder. It is difficult to evaluate passivity phenomena in people with a cognitive impairment – the experiences may have been ‘as if’ B’s body was being interfered with, rather than true passivity experiences. In cases 2 and 3 delusions and auditory hallucinations were prominent. Although no Schneiderian first-rank symptoms were unequivocally present, the course of the disorder (with some persisting deficit) suggests a diagnosis within the schizophrenic group of disorders.

An argument could also be made for other diagnoses, most notably the cycloid psychoses. All three cases manifested abnormal beliefs not syntonic with mood, ‘pananxiety’, and motility disturbances (Cutting et al, 1978). There are similarities to Leonard’s (1961) description of anxiety psychosis (and in case 2 features consistent with the description of motility psychosis). The symptoms also resemble descriptions of cycloid psychoses given by Perris (1988) with “distressed perplexity” and “a polymorphous and shifting symptomatology” suddenly appearing “like a bolt from the blue”. Perris emphasises the “very acute onset” of cycloid psychoses, in which first-rank symptoms are common. An acute onset and heterogeneous symptoms are striking features of the cases reported here. The possible chronicity of the disorder in case 3 argues against the diagnosis, although it was not entirely clear whether D continually had abnormal beliefs, or whether they occurred in discrete episodes; her behaviour related to auditory hallucinations occurred rarely.

The association of psychotic illnesses with Prader-Willi syndrome may be due to chance, or due to factors related to cognitive impairment rather than a more specific effect of Prader-Willi syndrome. The lifetime prevalence of affective and schizophrenic psychoses is about five times higher among the population of people with learning disability than among the general population, although there are problems with diagnosis (especially of schizophrenia and related disorders, where complex subjective experiences have to be conveyed in order to allow the diagnosis to be made). Cognitive impairment may predispose to psychiatric illness through a variety of mechanisms. Abnormalities in brain structure or function may lead to both cognitive impairment and psychiatric disorder (or to cognitive impairment which in turn leads to psychiatric disorder). Cognitive impairment may be associated with rejection, disadvantage, or other adverse consequences which may themselves predispose to psychiatric disorder, or psychiatric disorder may arise from the impact of impairments and disabilities associated with cognitive impairment. These factors are likely to interact. However, none of the people reported had epilepsy, an autistic-like condition or a sensory impairment (factors which may predispose the learning-disabled population as a whole to psychiatric disorder). The cognitive impairment in Prader-Willi syndrome is relatively mild, and the people reported had mild or moderate mental retardation.

No social or psychological precipitating factor (such as rejection because of obesity or other PWS-related
behaviour) could be found in any of the three cases, and no organic cause or precipitating event was identifiable. A further case resembling those reported has been seen (a woman with PWS who, while on holiday, suddenly became anxious, agitated and convinced that she was going to be kidnapped). This case has not been reported here, because there was some evidence that her skin-picking had worsened around the time of the change in her mental state (possibly as a result of anxiety) and she developed cellulitis. It was, therefore, possible that her psychiatric symptoms were due to an acute confusional state.

The three cases reported here had the same genetic abnormality (del 15 (q11-13)). They varied in degree to which they were overweight for height: B had been maintained on a strict diet from an early age, whereas D, in spite of an intestinal resection, was overweight for her height (see Table 1).

It is possible that PWS and psychotic disorders are associated other than by chance. Because PWS is associated with genetic abnormalities which are localised to a particular portion of chromosome 15, the recognition of psychotic episodes occurring in association with the disorder may be of relevance to research into the genetic basis of the psychoses. It may be of particular relevance to the aetiology or pathogenesis of cycloid and other atypical psychoses, if the tentative suggestion that psychoses in PWS are of acute onset, with polymorphous symptoms, is supported by other reports.

Acknowledgements

I am grateful to Professor J. Corbett and Dr A. Holland for their comments on earlier drafts of this paper, and to Dr T. Webb who performed the cytogenetic studies. Part of the genetic component of this work was supported by a grant from the Mental Health Foundation.

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David J. Clarke, MRCPsych, Senior Lecturer in Developmental Psychiatry, University of Birmingham Department of Psychiatry, Queen Elizabeth Psychiatric Hospital, Birmingham B15 2QZ

(A First received August 1992, final revision October 1992, accepted December 1992)

A Partial Form of Lycanthropy with Hair Delusion in a Manic–Depressive Patient

H. VERDOUX and M. BOURGEOIS

A 45-year-old man was admitted with a hair growth delusion and depressive symptoms. The delusion persisted for three years and disappeared after a manic episode. This odd delusion has some similarities with lycanthropy.


The delusional conviction that a part of the body is transformed or dysfunctioning can be observed in various psychiatric diseases. It may also be a solitary symptom as in monosymptomatic hypochondriasis (Munro & Chmara, 1982). Although this type of delusion can affect any part of the body, it is infrequent in some parts and, according to the literature, delusion of hair transformation seems to be unusual. We report the case of a patient with a persisting hair delusion.

Case report

D, a 45-year-old man, was abandoned by his biological mother and adopted by a childless couple when he was two
Prader-Willi syndrome and psychoses.
D J Clarke
Access the most recent version at DOI: 10.1192/bjp.163.5.680

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