Book reviews

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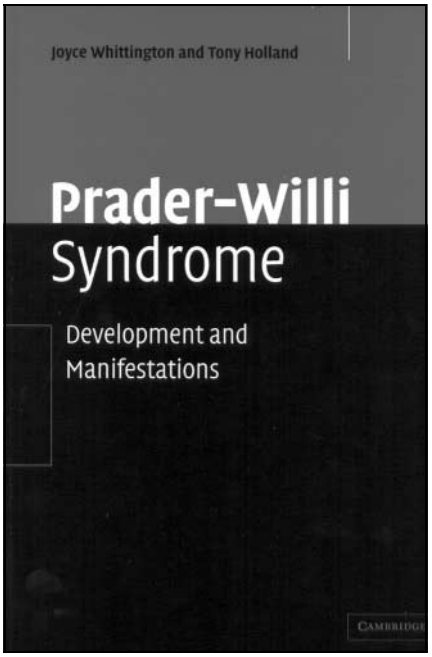
Prader–Willi Syndrome: Development and Manifestations

By Joyce Whittington & Tony Holland.
Cambridge: Cambridge University Press.
2004. 220 pp. £60 (hb). ISBN 0 52184 029 5



One finding from the Cambridge Prader–Willi syndrome (PWS) studies, summarised in this book, seems to be worthy of attention from psychiatrists in all specialties. It is that PWS caused by maternal uniparental disomy (inheritance of two chromosome 15s from the mother and none from the father) is associated with a prevalence of psychotic disorder that rises so steeply in early adult life that it approaches 100% by the age of 30 years. The finding has now been replicated. Like the association between Down's syndrome and Alzheimer-type dementia, there seems to be a clue here from research into an aspect of intellectual disability that has the potential to further knowledge in psychiatry more widely.

Prader–Willi syndrome was first described by Prader, Labhart and Willi in 1956. These Swiss paediatricians described a syndrome characterised by neonatal hypotonia, impaired sexual development, short stature, obesity and mental retardation. Further reports followed from around the world, and it became apparent that the obesity associated with PWS was a consequence of a severe eating disorder, and that strict dietary control from early childhood could prevent morbid obesity from developing.

Descriptions of behavioural aspects of the disorder ranged from early adjectival descriptions such as 'cheerful', 'somnolent', 'prone to temper-tantrums', through surveys of parent and carer organisations with questionnaires, to more rigorous studies using standardised behavioural assessments and comparison groups. It became apparent that PWS was associated with a behavioural phenotype that ranged from overeating (which is universal), to vulnerability to a variety of sleep problems, skin-picking, impulse-control disorders, compulsive and ritualistic behaviours and possibly vulnerability to severe psychiatric disorder.

Advances in genetics led to the recognition that PWS was associated with chromosome 15 abnormalities. Deletions, which were always of paternal origin, were found in association with about two-thirds of cases of PWS. The cause of PWS in the remaining third was eventually found to be maternal uniparental disomy, a finding that revolutionised thinking about human genetics and led to the concept of genomic imprinting. A small number of people with PWS are now known to have an imprinting error (in which the father's copy of chromosome 15 is 'marked' as though it were the mother's, leading to a situation akin to maternal uniparental disomy).

Research into behavioural aspects of genetic disorders (behavioural phenotypes) has advanced greatly over the past 20 years, and this book describes the culmination of several programmes of research into aspects of PWS centred on behavioural and psychiatric manifestations, but also including aspects such as prevalence and mortality rate (the latter found to be around 3% a year, far higher than for the general population).

The book is divided into three sections, giving an overview of the condition with some historical information, a review of genetics and biological aspects of the syndrome, an introduction to the Cambridge PWS project (the findings from which constitute the main part of the book) and concluding with a consideration of the future direction of research. It is written in a style that makes assimilation of the content easy for readers not versed in genetics or familiar with behavioural phenotype research. Its strength is also its weakness. By focusing almost exclusively on the Cambridge studies it provides an excellent overview of the findings from this research programme and sets them in the context of wider knowledge. There is relatively little emphasis on giving information about management strategies for psychiatric disorders or the practical applications of cognitive research for the education of people with PWS.

Declaration of interest

D.C. has collaborated with Joyce Whittington and Tony Holland on research into behavioural and psychiatric aspects of PWS.

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Somatoform Disorders: A Medicolegal Guide

By Michael Trimble.
Cambridge: Cambridge University Press.
2004. 254 pp. £50 (pb).
ISBN 0 521 81108 2

There have been many books published recently on somatoform disorders, but this one deals specifically with the medicolegal perspective. The book is timely because over the past decade a considerable amount of research has suggested that psychosocial factors are the key maintaining factors in disorders such as whiplash neck injury, upper limb pain (repetitive strain injury), chronic widespread pain and functional paralyses. Trimble starts well by taking a swipe at commentators such as Micale,

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