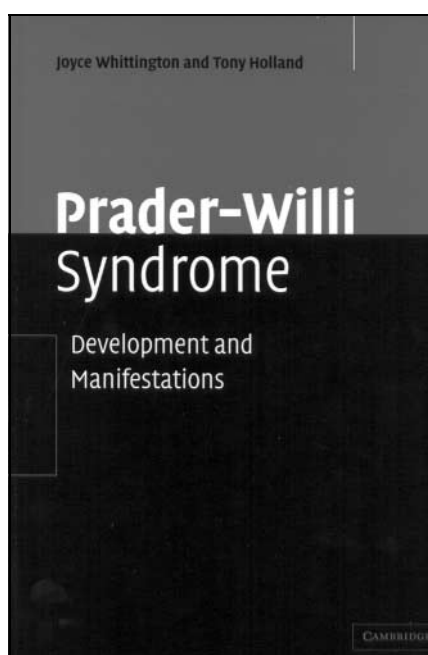


Book reviews

EDITED BY SIDNEY CROWN, FEMI OYEBODE and ROSALIND RAMSAY

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

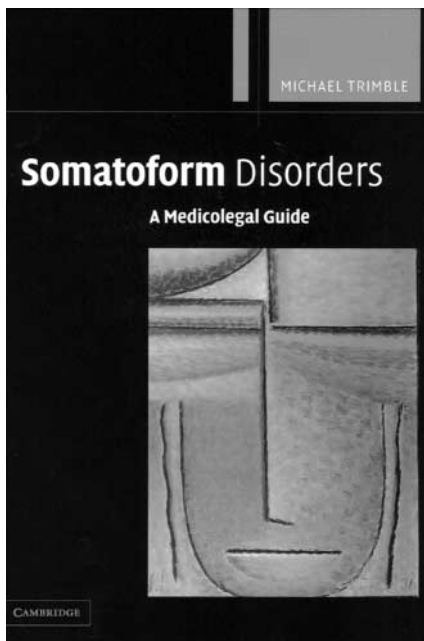
D.C. has collaborated with Joyce Whittington and Tony Holland on research into behavioural and psychiatric aspects of PWS.

David Clarke Consultant Psychiatrist in Learning Disability, Lea Castle Centre, Wolverley, Kidderminster DY10 3PP, UK

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 Micalé,



Showalter and Shorter, who suggest that 'Victorian faints, swoons and convulsions' are now rare and have largely disappeared from clinical practice. Any clinician currently working in the borderland between neurology and psychiatry would dispute this observation. Indeed, in October 2003 the first international conference on psychogenic movement disorders was held in the USA (Trimble and I attended), and it is clear that these disorders are becoming so common (1 in 30 of consecutive referrals to a movement disorder clinic) that even neurologists are taking note and wanting help. Trimble's elegant riposte to the social historians is that 'hysteria has never risen from the ashes: the fires have been burning brightly all along'. He goes on to criticise the diagnostic categories of both the ICD and DSM as being arbitrary (we all agree with that) and argues that, in a medicolegal setting, their use can be not only misleading but also mischievous.

Chapters on the clinical presentations and assessment are sound and well written, but I found most useful the chapters dealing with the legal context and issues of causation and compensation. Trimble outlines the shortcomings of Lord Woolf's reforms and the role of the single joint expert (SJE), who is unlikely to satisfy the needs of both parties in complex psychiatric cases. There are up-to-date accounts of compensation as well as the problems of legally aided claims. Issues of foreseeability and rules for secondary claimants/victims are described, with

helpful case vignettes, and Trimble introduces the term 'lexigenic' to emphasise the legal equivalent of iatrogenic illness. Chapters about mechanisms and causation include descriptions of recent imaging studies on hysteria and malingering.

This is a valuable text for psychiatrists involved in medicolegal work, especially those asked to provide reports on patients with unexplained gait disturbances, whiplash neck injuries and symptoms that are not matched by organic findings. The Department for Work and Pensions informs us that 70% of patients on long-term disability benefits have symptoms that are essentially medically unexplained: Trimble's book explains how to assess these patients when they are involved in a legal claim and how to prepare a coherent report for the court.

Christopher Bass Consultant in Liaison Psychiatry, Department of Psychological Medicine, John Radcliffe Hospital, Headley Way, Headington, Oxford OX3 9DU, UK

Self-Made Madness: Rethinking Illness and Criminal Responsibility

By Edward W. Mitchell. Aldershot: Ashgate Publishers. 2003. 272 pp. £50 (hb). ISBN 0 75462 332 7

To what extent could we hold people with mental illness responsible for their own disturbed states of mind? And what would this mean for those individuals with mental

illness who are charged with criminal offences? These questions are explored by Edward W. Mitchell in an erudite and complex analysis of what he terms 'meta-responsibility'. He offers both a conceptual analysis, and an empirical investigation of how mock jurors would judge a defendant who had committed a violent offence while non-compliant with prescribed psychotropic medication.

Mitchell found that his mock jurors gave intriguing answers to questions of meta-responsibility. Although some took the view that the defendant was criminally responsible because of the causal link between non-compliance and relapse, they also took the view that he could not be responsible *because* he had relapsed. There was also a suggestion that it was the decision to come off medication that should be judged for culpability, and not the offence itself.

The issues Mitchell raises are of profound importance for both psychiatrists and mental health service users. Current Department of Health policy emphasises the importance of involving patients in the management of their condition; of treating the patient as their own 'expert'. However, when it comes to mental health, government policy seems to assume that service users cannot be trusted to manage their own condition, and are uniformly incapacitous in terms of responsibility for criminal offences. Numerous homicide inquiries have criticised professionals who have tried to share clinical decision with their patients and allowed the patients some control over their medication.

As clinicians, we know that mental illness does not necessarily abolish the capacity to form meaningful and competent intentions. There must therefore be some pressure on those with mental illness who have acted violently to explain why their disorder should be both an explanation, and exculpation, for their behaviour. From my own clinical experience, these questions are as troubling for patients who have been violent when psychotic as they are for the rest of us.

The more expert and autonomous the patient seeks to be, the more responsibility they must take for their actions. In ethical terms, not to respect autonomy when present is both to act disrespectfully and to act unjustly. However, this approach is a radical challenge to the traditional belief that those with mental illnesses should not be held responsible for their behaviours.

