Autism and Child Psychopathology Series
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Elias Tsakanikos
Jane McCarthy
Editors

Handbook of Psychopathology in Intellectual Disability

Research, Practice, and Policy

Springer

Autism and Child Psychopathology Series

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Handbook of Psychopathology in Intellectual Disability

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Foreword

The *Handbook of Psychopathology in Intellectual Disability* is on a topic that is both timely and important. These overlapping problems are also sometimes referred to as dual diagnosis. The focus of this specialty area is on mental health issues and how they are expressed among these people, relative to the general population. More recently, researchers have come to realize how interconnected developmental disabilities are to one another. This view of psychopathology and intellectual disabilities has fit nicely into the broader field of how mental health has evolved. Current thinking is that these phenomena occur on a spectrum and separate disorders as currently defined intersect.

As recently as the 1960s and 1970s, many experts in the field of intellectual disabilities were of the opinion that mental health issues could not occur in persons with intellectual disabilities. Various explanations for this position existed including a lack of insight on the part of the patient. Obviously, these attitudes were a major impediment to developing effective assessments and treatments. The nature of these problems, how they developed and what caused them, also went unresolved.

Professionals now recognize that psychopathology and intellectual disabilities coexist and are expressed somewhat differently than in the general population. Moreover, persons with intellectual disabilities are much more likely to suffer from mental health issues and at rates perhaps 4–5 times as often as the general population. This point underscores the importance of this topic.

Dr. Tsakanikos and McCarthy do a superb job of bringing together many of the leading experts in the area of psychopathology in persons with intellectual disability. They present a comprehensive review of the current state of the field. They present a comprehensive review of the current research and practice on this relatively new subdiscipline. Some of these topics have been well established in the field of intellectual disabilities. Psychological assessment in the form of IQ tests, for example, dates to the beginning of applied psychology. Binet and associates developed the methodology for modern testing. The IQ tests (Stanford-Binet) provide a method to identify children with intellectual disabilities. Using objective items, norms, reliability and validity approaches set the tone for future test. Measures of psychopathology specific to intellectual disabilities are of much more recent origin but have followed the same method of test construction and administration.

This book is timely because much of the available knowledge is so new. Two decades ago it would have been difficult to find enough information to write individual chapters on schizophrenia, substance misuse, dementia, ADHD or forensics. And these are just some of the examples. Now, large quantities of information are rapidly expanding what we know about psychopathology among persons with intellectual disabilities. These developments make for lively and informative reading across a host of topics. Other areas of study which are relatively new in the field of intellectual disabilities are rapidly becoming invaluable areas of clinical study. For example, epidemiology is a long researched and esteemed area of study. However, this field is a relatively new entrant in the subdiscipline of psychopathology among persons with intellectual disabilities. The general lack of recognitions of these co-occurring conditions and the lack of tools, until recently, to detect mental health problems in intellectual disabilities certainly have been major contributors to this situation.

Service provision is also a rapidly evolving area of study. Community services have expanded dramatically in recent decades. Entirely new areas of study have emerged such as competitive employment as society makes efforts to better and more effectively integrate persons with intellectual disabilities within everyday life. Mental health concerns play a significant role in community integration as well. Psychopathology is often a major impediment to healthy adjustment and living. Thus, advances in mental health care are major factors for enhancing quality of life.

Inpatient care has changed dramatically. Movement from long-term residential services is moving to short-term care and community support. Part of this difference is due to philosophical changes in society but also in advances in treatment. Pharmacological and psychological treatments have also been improving rapidly. These interventions are becoming both more specific to given disorders, but also these interventions have become more varied and more effective. One of the biggest issues is the gap between what we know and how well it gets implemented. That issue will continue to be a concern going forward. However, mental health care for persons with intellectual disabilities has a more positive outlook now than ever before. This handbook does an excellent job of reflecting the current state of the field.

Baton Rouge, LA Johnny L. Matson, Ph.D.

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Part I

General Issues and Assessment of Psychopathology

Introduction

1

Elias Tsakanikos and Jane McCarthy

Introduction

There is a growing recognition over the last 25 years that adults with intellectual disability (ID) are vulnerable to mental health problems due to particular biological, psychological and social factors. In this handbook we aimed to provide a comprehensive and up-to-date overview of research and critically evaluate the complexities in both aetiology and manifestation of psychopathology in those with ID. We also review assessment, interventions and service issues with an emphasis on translating research evidence into clinical practice and policy. The aim was to provide a reflection of current practice and research as well as implications for future clinical practice and service delivery. The handbook is written primarily for doctoral level professionals and doctoral level students in training across a range of disciplines such as psychology, psychiatry, special education, communication disorders, child development and social work.

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Content and Structure

The first section of this handbook covers general conceptual issues as well as assessment approaches of psychopathology providing a review of diagnostic trends and clinical dilemmas that mental health practitioners face when working with adults with ID and comorbid psychopathology. Key legislations, policies and scientific understanding of ID have evolved over time. Chapter 2 provides an overview of the history of ID as well as factors that determine epidemiology (Maulik, Harbour, & McCarthy, 2013). Chapter 3 outlines principles of clinical assessments with reference to specific mental health disorders, challenging behaviour and psychological treatments (Alim, Paschos, & Herne, 2013). Advantages and disadvantages of using structured assessments in clinical practice are being discussed in Chap. 4. A selection of assessments is compared in relation to their psychometric properties but also according to different models of assessments and staff skills (Moss & Hurley, 2013). Assessment issues are also discussed in Chap. 5 with respect to modern diagnostic criteria, leading operational definitions and models of understanding psychopathology (Hamelin, Maum, & Sturmey, 2013).

The second section of this book focuses on key aetiological issues. Neuroimaging has been successfully used to investigate in vivo brain anatomy and functioning. Chapter 6 provides an introduction to neuroimaging technologies

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investigating brain structure, functioning and brain chemistry with emphasis on mental health-related technological applications as well as neurological conditions commonly associated with ID (Ecker, 2013). Particular neurological conditions such as epilepsy are far from uncommon in adults with ID and can shed light on underlying mechanisms of atypical neurodevelopment. Patients with epilepsy and ID are at increased risk of developing additional cognitive dysfunction and psychiatric disorder. Chapter 7 reviews the complex interactions between mental health, neurocognition and epilepsy in people with ID and discusses clinical implications for the development of appropriate care management plans (Winterhalder & Ring, 2013).

Chapter 8 is comprehensive review of behavioural phenotypes and genetic syndromes. The idea that certain ID syndromes are associated with distinctive physical, behavioural and cognitive characteristics is not new. Nevertheless the genetic advances over the last decades have resulted not only in increased understanding of the aetiology but also phenotypic description of behaviour, and mental disorder has become more possible with technologically advanced tools. The great challenge for the years to come will be to translate understanding of the genetics of ID into understanding of the mechanisms underlying impaired cognition, behavioural and emotional disturbance, as well as to develop new targets for treatment (Paschos, Bass, & Strydom, 2013).

Any attempt to understand aetiology factors in psychopathology without a critical understanding of psychosocial issues surrounding the emotional and behavioural difficulties in adults with ID would be inevitably incomplete. Chapter 9 provides a critical review of psychological and social factors (including gender, level of functioning, ethnicity, life adversities, early family relationships etc.) that are likely to be implicated in the onset and maintenance of comorbid conditions. Bridging psychosocial and behavioural genetics research is the obvious future challenge. Psychosocial risk factors need to be better understood, as do protective factors such as secure attachments, supportive relationships achievements, social support and community participation (Magiati, Tsakanikos, & Howlin, 2013).

The third section of this handbook explores aspects of comorbid psychopathology in adults with ID along with related clinical and diagnostic issues. Chapter 10 discusses clinical presentation and management of schizophrenia in people with ID. Although Schizophrenia Spectrum Disorders are generally thought to be more difficult to detect and diagnose in adults with ID than in the general population; the estimated prevalence rate of these disorders is 3 times higher than the in general population (Hemmings, 2013). Chapter 10 presents common mental disorders (depressive and anxiety disorders, Obsessive Compulsive Disorder and Post Traumatic Stress Disorder) often seen in adults with ID. The prevalence of these disorders is similar if not higher than in the general population; however, diagnosis can be complicated by atypical presentation of symptoms. This chapter also describes the epidemiology, aetiology, classification and current guidance on management for common mental disorders in adults with ID (Hassiotis, Stueber, Thomas, & Charlot, 2013).

Chapter 12 explores the concept of personality disorder in relation to challenging behaviour and mental health and reflects on the usefulness of this diagnosis for people with ID. The boundaries between mental health, challenging behaviour and personality disorder are explored in an attempt to enhance understanding of the clinical value but also the limitations of this diagnostic category. The clinical outcomes for those with ID and personality disorder remain unclear especially in those with severe personality disorders (Flynn, 2013).

Significant improvements in health and social care has led to dramatic increase in the life expectancy of people with ID and a subsequent increase in age related conditions, including dementia. Chapter 13 discusses the causes and features of dementia in people with ID, as well as additional considerations that need to be taken in account when assessing and managing dementia in this particular patient group (Strydom & Sinai, 2013).

Chapter 14 deals with the issue of substance abuse in adults with ID, a particular under-researched area. When deinstitutionalisation started almost 40 years ago, policy makers and mental health practitioners were perhaps not ready to predict the issues that would be faced by people with ID given free access to and opportunity within society. This chapter examines the phenomenon of substance abuse in people with ID, considering legal, illicit and prescribed substances. Issues of prevalence and psychopathology are addressed along with biological, psychological and social factors frequently associated with substance use, and future implications for research and practice are critically evaluated (Taggart & Chaplin, 2013).

The forth section of this book explores developmentally and socially inappropriate behaviours associated with both neurodevelopmental conditions and challenging behaviour in adults with ID. Chapter 15 explores the relationship between ADHD (Attention Deficit Hyperactivity Disorder) and ID. Identification of neurodevelopmental comorbidities is important due to important implications for diagnosis as well as treatment and prognosis. The evidence for the validity of the diagnosis of ADHD in this population is critically discussed and evaluated (Xenitidis, Maltezos, & Asherson, 2013).

Despite the fact that rates of autism are generally higher among people, ID prevalence studies remain inconsistent as to whether the presence of autism spectrum disorders in ID results in greater risk for comorbid psychopathology. Chapter 16 provides a critical review of evidence showing that those with autism and ID are at greater risk for mental health problems than those without autism. The chapter also highlights differences in autism-specific coping strategies that may remain unrecognised in mental health research in autism and ID (Bradley, Caldwell, & Underwood, 2013).

Chapter 17 is devoted to the relationship between challenging behaviour and psychopathology in ID. Measures designed for the assessment of challenging behaviours in ID with or without ASD are critically evaluated as well as treatment interventions such functional assessment but also the controversial issue of psychotropic drug treatment (Rieske & Matson, 2013). Chapter 18 focuses on offenders with ID in prisons. Adults with ID that are likely to be overrepresented in parts of the Criminal Justice System (CJS) are mostly male, from deprived social backgrounds, have mild or moderate (non-severe) disability and often have mental health needs and comorbid neurodevelopmental disorders, including ASD and ADHD. Importantly, offenders with ID have well-recognised vulnerabilities such as not understanding the process or their rights, being suggestible during interview and making unwise, sometimes fatal, decisions at crucial points of the forensic pathway (Murphy & Mason, 2013).

The last section of the book covers treatment interventions and service development issues reflecting on ways of improving health provision and wellbeing among those with ID and comorbid mental health problems. Chapter 19 presents a review of the use of psychopharmacology in the management of problem behaviours and evidence for the effectiveness of psychotropic medication based on systematic reviews (Deb, 2013). Chapter 20 provides an up-to-date critical evaluation of the evidence base of psychological treatments people with ID. Effective psychological treatments may lead to improvement in the quality of life of patients with ID, and their carers, and can potentially result in economic benefits to society, such as avoidance of resources wasted on ineffective or harmful therapies (Sturmey & Hamelin, 2013).

Community services are reviewed in Chap. 21 where policy and different models of care are evaluated in the context of service delivery for adults with ID. This chapter examines these issues and looks at the development of evidence base for community mental health services and future models (O'Hara, Chaplin, Lockett, & Bouras, 2013). Chapter 22 provides a review of key issues concerning inpatient mental health services for people with ID. With the deinstitutionalisation movement, the purpose of hospitalisation has shifted to short-term treatment of acute psychiatric issues. The review focuses on research evidence on two main models of inpatient care: mainstream/general services vs. specialist units (Lake, Balogh, & Lunsky, 2013).

Implications for Research, Policy and Practice

One main objective of this book was to provide a reflection of current practice and research as well as the implications for future clinical practice and service delivery. Chapter 23 therefore concludes with a discussion of key themes for future research and clinical practice. In this final chapter, the importance of developing care pathways for people with ID and comorbid psychopathology is discussed with emphasis on care personalised to the needs of the individual. The editors and co-authors of this chapter propose that clinical research and practice would greatly benefit from the use of structured assessments and evidence-based interventions linked to measureable outcomes becoming part of daily clinical practice within these Care Pathways to support future research. Such a systematic, accurate and detailed routine collection of clinical, genetic and behavioural data has the potential to increase substantially our understanding of the complex interplay of genetic, biological and environmental factors in the presentation of comorbid psychopathology in people with ID and to inform policy and clinical practice (McCarthy & Tsakanikos, 2013).

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Epidemiology

2

Pallab K. Maulik, Catherine K. Harbour, and Jane McCarthy

Introduction

Both researchers and laypeople use a variety of terms to refer to intellectual disability (ID). The World Health Organisation identified mental retardation, mental handicap/disability, learning/developmental disability and mental deficiency/subnormality as some terms used to signify intellectual disability (WHO, 2007). Other chapters of this book deal with the diagnosis and classification of intellectual disability in detail. A shared definition of ID is "a disability characterised by significant limitations both in intellectual functioning and in adaptive behaviour, which covers many everyday social and practical skills" (American Association of Intellectual and Developmental Disabilities, 2011). ID starts before 18 years of age. A quantitative measurement is possible using standardised tests that measure the Intelligent Quotient (IQ). As discussed in more detail below, adaptive behaviour is also used to make a more comprehensive assessment of an individual's intellectual capabilities. Adaptive behaviour includes "conceptual skills [such as] language and literacy; money, time, and number concepts, and self-direction; social skills [like] interpersonal skills, social responsibility, self-esteem, gullibility, naïveté (i.e. wariness), social problem solving, and the ability to follow rules/obey laws and to avoid being victimised; and practical skills [which include] activities of daily living (personal care), occupational skills, health care, travel/transportation, schedules/routines, safety, use of money, use of the telephone" (American Association of Intellectual and Developmental Disabilities, 2011).

The estimated prevalence of ID is 1 % of the general population or about 70 million people with this condition in the world given the current population of seven billion (Maulik, Mascarenhas, Mathers, Dua, & Saxena, 2011). Even with modern medicine's understanding about the causes of ID and better knowledge about the epidemiology and history of the condition (Harbour & Maulik, 2010; Harris, 2006; King, Toth, Hodapp, & Dykens, 2009; Maulik & Harbour, 2010), around the world there are wide variations in the way people perceive intellectual disability. This variation in aetiologic understanding is reflected in the variety of legislation regarding the rights of people with ID in different countries of the world. The epidemiology of ID has many aspects that

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are determined by different socio-economic and socio-demographic characteristics. Often similar characteristics have played a role in the historical understanding of the condition and the way people with intellectual disability have been treated in their countries. Increasing awareness about people with intellectual disability over the years has led to some improvement in funding more research in the area, as well as development of better laws to protect those affected.

This chapter provides an overview of the history and epidemiology of ID. Other chapters of this book highlight different aspects of research related to ID and its public health implications, as well as details related to epidemiology of specific mental health problems in people with intellectual disability.

History of Intellectual Disability

Intellectual Disability in Ancient History

The first known reference to intellectual disability is in the Therapeutic Papyrus of Thebes (modern Luxor, Egypt), in 1552 BC. The papyrus suggests the understanding that intellectual disability could result from brain damage (Harris, 2006; Reynolds, Zupanick, & Dombeck, 2011). Ancient Greeks and Romans believed that children with intellectual disabilities were born as a result of having angered the Gods, and children with severe ID would be allowed to die of exposure as infants rather than permitted to grow up. However, children with intellectual disability who were born to wealthy Romans did have some protections; they had property rights and were permitted to have guardians (Harris, 2006). Before the eighteenth century, many people with mild intellectual disability who were socially competent received no special identification or treatment. People with more severe intellectual disability received protective care from their families or in monasteries. In some societies, people with more severe intellectual disability were thought to be capable of receiving divine revelation (Beirne-Smith, Patton, & Kim, 2006; Harris, 2006).

Rehabilitation and Reintegration

Jean-Marc Itard (1774–1838), a medical doctor in France, developed a systematic and documented programme of intervention for a mute and developmentally delayed child who was found in the forest of Aveyron, whom Itard named Victor. Modern scholars have suggested that Victor's was the first documented case of autism (Frith, 2003). Edouard Seguin (1812–1880), a student and colleague of Itard's, further developed Itard's skill-based programme and published *Idiocy*: *and its Treatment by the Physiological Method* in 1866. Some elements of Seguin's programme, like individualised instruction and behaviour management, are currently practiced. His work inspired Maria Montessori (1870–1952), a medical doctor like Itard and Seguin, and an innovator of early childhood education. Johann Guggenbühl established Abendberg, the first known residential facility for people with intellectual disability, in 1841 in Switzerland (Beirne-Smith et al., 2006).

Through the early and mid-1800s in the United States, the outlook towards possibilities to rehabilitate, train and reintegrate people with intellectual disability to "normal" life was optimistic. Optimism waned in the latter half of the 1800s, as people with severe intellectual disability were less able to adapt to the changes brought by industrialisation and urbanisation. The systematic programmes that had proven successful were diluted and more residential institutions were established. People with intellectual disabilities were termed "feebleminded" and were blamed for the ills that accompanied urbanisation, like poverty, illness and crime. A fearful, alarmist attitude towards those with intellectual disabilities developed. Criminal behaviour and intellectual disability were thought to be heritable, as were mental illness, tuberculosis, poverty, slums and prostitution. With the growth of the eugenics movement, which sought to improve the genetic composition of the human population, proponents focused on eliminating the possibilities for people with intellectual disability to reproduce (Beirne-Smith et al., 2006; Radford, 1991; Reilly, 1987).

Eugenics and Segregation

The eugenics movement was established with the 1869 publication of *Hereditary Genius* by Sir Frances Galton (1822–1911), a cousin of Charles Darwin's, which provided a theoretical basis for inherited intellectual disability (Beirne-Smith et al., 2006). Eugenicists held that intellect and personality are determined by nature, not by nurture, nor by environmental elements like nutrition, poverty or education. Therefore, reproduction should be managed to prevent the degeneration of the human species (Radford, 1991). Two books popular in the United States reinforced this myth of the heritability of intellectual disability. The first, published in 1877 by Richard Louis Dugdale (1841–1883), a sociologist who studied prisoners in upstate New York, was titled *The Jukes*: *A Study in Crime*, *Pauperism*, *Disease and Heredity* and suggested inherited criminality. The book presented detailed family trees and examined how environment and heredity had affected the families. The second book, *The Kallikak Family*: *A Study in the Heredity of Feeble-Mindedness*, was written by Henry Goddard (1866–1957) and published in 1912. Goddard's book presented five generations of family pedigree and suggested the heritability of "feeblemindedness" (Beirne-Smith et al., 2006). According to the eugenics movement, medical treatment interferes with Darwinian natural selection and allowed undesirable people to stay alive, which increased the burden on the society. Eugenicists felt that mentally retarded and mentally ill people were reproducing at greater rates than were the more valuable productive people and that these burgeoning populations of undesirable people were the cause of increasing costs for schools, prisons, hospitals and special homes (Bachrach, 2004).

Starting in the late 1800s and early 1900s, people with intellectual disabilities and others were confined to institutional settings to protect "normal" society and to control the institutionalised's reproductive ability. In the United Kingdom, Canada and the United States, the most common way of controlling the reproduction of people with intellectual disabilities was segregation through limits placed on marriage, immigration controls, sterilisation and, most of all, custodial institutionalisation (Joseph, 2005; Radford, 1991; Reilly, 1987). "Custodial institutions were most importantly the means by which the feebleminded were removed from a society in which they were perceived as a genetic threat and placed in isolated environments, completely segregated by gender" (Radford, 1991, pg. 454). In such institutions male and female residents were kept apart and many were sterilised. In Germany, the Nazi government's espousal of eugenics led to the 1933 compulsory sterilisation law, under which people with "congenital feeblemindedness" could be forcibly sterilised. The diagnosis of "congenital feeblemindedness" was very subjective (Bachrach, 2004; Sofair & Kaldijian, 2000).

Psychological Testing

The development of psychological tests starting in the early 1900s improved the identification of people with intellectual disabilities and also contributed to greater institutionalisation. In 1905 French psychologists Alfred Binet (1857–1911) and Théodore Simon (1872–1961) developed the Binet-Simon Intelligence Scale Test to identify schoolchildren who would need special services; the test they developed became the IQ (Intelligence Quotient) test. Greater use of intelligence tests made intellectual disability seem more prevalent, since it identified mildly disabled people who would otherwise go undiagnosed (Beirne-Smith et al., 2006).

In the twentieth century, research revealed non-genetic aetiologies and associations with intellectual disability, such as metabolic disturbances like PKU (phenylketonuria) and environmental factors, such as infection, trauma and endocrine disturbance. The heritability of intellectual disability was further discredited by studies of institutionalised individuals that found that more than half of them had parents without intellectual disability (Beirne-Smith et al., 2006). The Catholic Church opposed eugenic sterilisation in Germany and in the USA. The association of eugenics with Nazi "racial hygiene" further

discredited the movement (Bachrach, 2004; Reilly, 1987; Sofair & Kaldijian, 2000).

Deinstitutionalisation

Institutional settings can dehumanise residents with intellectual disabilities, and quality of life, adaptive behaviour and choice making can improve when people with intellectual disability move out of an institution into a community setting (Beadle-Brown, Mansell, & Kozma, 2007). The concept of "normalisation" was introduced in Scandinavia in the 1950s. Normalisation suggests that people with intellectual disability should have access to supports so as to be able to experience patterns and conditions of everyday life that are as similar as possible to those of mainstream society (Beirne-Smith et al., 2006). The proportion of people with intellectual disability living in institutional versus non-institutional settings has declined in many countries, including England, Scandinavia, Canada and the United States (Beadle-Brown et al., 2007). Transitioning people with intellectual disability from institutional settings to community settings requires attention to appropriate housing and coresidence selection, negotiation of staff needs with service users' needs, organising a culture of engagement in the home and in the community and focus on quality of life (Beadle-Brown et al., 2007).

International Agreements

In 1994, the United Nations passed the *Standard Rules on Equalisation of Opportunities for Persons with Disabilities*. This provided international standards for programmes, policies and laws for those with disabilities. More recently the United Nations passed the Convention for the Rights of Persons with Disability (United Nations, 2006). Around the world, in recent time there has been greater interest in early intervention, community-based rehabilitation, definition and diagnosis, human rights, and legislation and focus on deinstitutionalisation (Beadle-Brown et al., 2007).

Diagnosis and Classification of Intellectual Disability

Chapters 3 and 5 discuss the diagnosis and classification of intellectual disability, as do numerous studies (e.g. Harris, 2006; King et al., 2009; Maulik & Harbour, 2010). The previous section describes the historical evolution of the concept of intellectual disability and how early behaviour-based observational descriptions led to quantitative assessments based on IQ and then to a combination of both IQ assessments and behavioural observations to diagnose intellectual disability. Chapter 5 details how different diagnostic systems employ specific symptoms to reach the diagnosis of intellectual disability.

The International Classification of Diseases—Tenth Revision (ICD 10) (WHO, 1992), Diagnostic Statistical Manual of Mental Disorders—Fourth Edition, Text Revision (DSM-IVTR) (APA, 2000), International Classification of Functioning, Disability, and Health (ICF) (WHO, 2001), and the American Association of Intellectual and Developmental Disabilities (2011) all use a combination of IQ-based quantitative assessment and adaptive behaviour in their description of intellectual disability. "Adaptive behaviour" considers the individual's ability to cope and adapt to the demands of his or her physical and social environment. Intellectual disability is essentially a developmental disorder and its symptoms begin to manifest early in life. According to current diagnostic systems, symptoms must originate before 18 years of age. Based on IQ levels, ICD10 classifies the severity of ID as mild (IQ of 50–69), moderate (35–49), severe (20–34) and profound (<20) (World Health Organization, 1992). Researchers continue to debate the value of diagnostic systems based on IQ tests versus those based on adaptive behaviour. In our experience, both have value. While the IQ-based diagnosis is more strongly supported by theory, the adaptive skills diagnostic approach better accounts for an individual's adaptive capabilities, which is especially important for individuals with less severe forms of ID (King et al., 2009). In 1992, the American

Association of Intellectual and Developmental Disabilities (AAIDD) definition sparked an important debate when it classified ID according to the level of support the person needs: intermittent, limited, extensive or pervasive. The AAIDD also outlined ten different adaptive domains: communication, self-care, home living, social skills, community use, self-direction, health and safety, functional academics, leisure and work. It also increased the cut-off IQ level from 70 to 75, which effectively meant that at least twice as many people were classified as having an intellectual disability. As a result of this debate, AAIDD revised their IQ cut-off to two or more standard deviations below the mean, or an IQ of approximately 70 or less. The focus on support levels and adaptive domains remained part of case definition. Administering sophisticated assessments relies on sufficient availability of multidisciplinary personnel, of which there is a chronic shortage. From an epidemiological perspective too, the diagnostic criteria play a role in ascertaining the prevalence of the disorder in the community.

Epidemiology of Intellectual Disability

Prevalence

This section examines the epidemiology of ID using population-based data. King et al. (2009) found that the prevalence of intellectual disability varies between 1 and 3 % and identified some reasons for this variation. First, ID as determined using only IQ levels shows 3 % prevalence, but prevalence decreases when adaptive behaviours are also used for diagnosis. Second, a 3 % prevalence is plausible if the correlation between IQ and age is constant. However, for some medical conditions, IQ level changes with age. For example, the IQ of a person with Down's syndrome tends to be highest in the first year of life and then decreases through early and middle school years, whereas those with Fragile X syndrome start to show a decline at early adolescence—10–15 years (King et al., 2009). IQ level also varies as an individual grows and learns different adaptive skills. The third reason for variation in the prevalence of intellectual disability is that rates of ID depend on the criteria used to define ID, which are based on school and local administrative policies. Different schools send their students for evaluation based on their own policies, resulting in different prevalence levels across a varied age group. The most common age of identification is 10 years.

As discussed in more detail below, poverty and poorer socio-economic conditions are associated with greater likelihood of having intellectual disability, especially during antenatal and early childhood development, and variation in levels of socio-economic resources among different populations may explain many differences in prevalence of ID in those populations. Other factors related to the variance of ID prevalence are probably related to administrative policies. Administrative policies that seek to reduce stigmatisation may lead to over-identification of learning disorders in children with mild to moderate levels of IQ, who might otherwise be diagnosed with intellectual disability. Recent analysis (Boyle et al., 2011) of the National Health Interview Survey (NHIS) data from children aged 3–17 years in the United States showed that prevalence of learning disorders increased by 5.5 % while the diagnosis of intellectual disability reduced by 1.5 % between the 1997–1999 and the 2006–2008 waves of the NHIS. Finally, the mortality rate varies across different groups of individuals with ID. Those with more severe forms of ID have lower life expectancies, which leads to variation in prevalence rates.

A recent meta-analysis of 52 studies showed an overall prevalence of 10.37 ID cases per 1,000 population globally (Maulik et al., 2011). However, the rates varied according to a number of parameters (Table 2.1). Prevalence was highest in low and middle income countries, among children or adolescents, in socio-economically poorer regions like rural and urban slums and in studies that assessed ID based on psychological tests only. More males than females have intellectual disability. The female-to-male ratio in adults is 0.7–0.9, while in children/adolescents it is 0.4–1.0. The meta-analysis included population-based

Table 2.1 Proportion of studies and pooled estimates per 1,000 population by subgroups ($N = 52$) (Maulik et al. (2011))

	N	%[a]	Prevalence/ 1,000 population[b]	95 % CI of prevalence rate
Income group of country				
Low-income	6	11.5	16.41	11.14–21.68
Middle-income	17	32.7	15.94	13.56–18.32
High-income	29	55.8	9.21	8.46–9.96
Type of population targeted				
Rural	8	15.4	19.88	13.60–26.17
Urban	1	1.9	7.0	6.12–7.87
Urban slum/mixed rural–urban	17	32.7	21.23	16.34–26.11
Regional/provincial	23	44.2	7.85	6.98–8.71
National	3	5.8	6.23	5.48–6.98
Age-group of study population				
Adult	5	9.6	4.94	3.66–6.22
Child/adolescent	35	67.3	18.30	15.17–21.43
Both adult and child/adolescent	12	23.1	5.04	4.07–6.01
Type of study				
Cross-sectional	41	78.9	9.69	8.76–10.63
Cohort	11	21.1	13.21	10.70–15.72
Sampling strategy used to gather data				
Key informant report	1	1.9	2.61	−1.00–6.23
School based study	2	3.9	7.04	6.35–7.73
Hospital data or administrative registry	30	57.7	9.35	8.60–10.10
Random household survey	19	36.5	15.78	13.73–17.86
Measure used for diagnosis				
Psychological assessment	30	57.7	14.30	12.70–15.91
DSM/ICD	12	23.1	8.68	7.89–9.48
AAIDD/ICF/some disability criteria	10	19.2	6.41	4.89–7.93

[a]Values have been rounded so may not add up to 100 %
[b]Estimates based on meta-analysis using random effects model

data from studies that reported on ID in the community and excluded studies of subgroups such as people with specific genetic disorders and studies of people in institutional settings (e.g. prisons, long-term care facilities).

Factors That Affect Assessment of ID

To appropriately interpret the results of any epidemiological study, one must consider the study design, study population and assessment tools used in the research. The same goes for epidemiologic studies of intellectual disability.

Study Design

A cross-sectional study design, surveillance study or analysis of administrative data allows researchers to estimate prevalence or to characterise associations between ID and a specific condition like Down's syndrome. A cohort design is necessary to determine a causal relationship or to estimate incidence of ID. Randomised controlled trials have been used to study the efficacy of medications or other interventions in controlling psychological or medical problems associated with ID. For example, a systematic review of randomised controlled trials on the efficacy of medications to control epilepsy

in patients with ID found that anti-epileptic drugs were as effective in patients with epilepsy and ID as they were in patients with epilepsy without ID (Beavis, Kerr, & Marson, 2007).

Characteristics of the Study Population

As mentioned earlier, research has highlighted the importance of age in ID, and a meta-analysis found that prevalence of ID is highest during childhood and adolescence (Maulik et al., 2011). Age plays a role in both time of earliest diagnosis and occurrence of complications (King et al., 2009). Intellectual disabilities associated with different conditions, such as Down's syndrome or Fragile X syndrome, reach their peak at certain ages and are most likely to be diagnosed at those ages. While children with Down's syndrome show a declining IQ after the age of 1 year, those with Fragile X syndrome generally show a decline only after 10–15 years of age. Similarly, research conducted with a study population of people with a specific condition, like Down's syndrome, should be interpreted carefully. The specific condition may be associated with manifestations of comorbidities that differ from manifestations of the same comorbidities among people who do not have the specific condition. For example, people with Down's syndrome manifest symptoms of Alzheimer's disease at much earlier ages than do people without Down's syndrome (Patja, Iivanainen, Vesala, Oksanen, & Ruoppila, 2000; Zigman & Lott, 2007).

Gender is also associated with ID. Among children, boys have a higher prevalence of ID than do girls. This gender difference is more pronounced for mild mental retardation, which is 1.5 times more prevalent among boys than among girls. One reason suggested for this difference is that boys, especially those with mild ID, tend to be identified more because of their behavioural problems in school. Maulik et al. (2011) found that among both adolescents and adults, males were affected more.

Another aspect of the study population that affects the prevalence of ID is socio-economic conditions. Rural and urban slum populations have higher prevalence of ID than do wealthier populations. Furthermore, the environment in which a child develops affects his or her ability to develop adaptive skills and intellectual capacity, which in turn affects the results of any assessments done on children and adolescents with ID. Similarly, higher prevalence is also noted in low and middle income countries as compared to higher income countries (Maulik et al., 2011). A number of reasons can explain this. First, the opportunities for diagnosis of prenatal genetic conditions in pregnant women are lower among those from poorer socio-economic conditions, who are less able to access such services. Second, a number of nutritional deficiencies can lead to intrauterine growth retardation which can in turn lead to poor development of the foetal brain, which can then lead to cognitive impairment. Third, people with intellectual disabilities are also marginalised in the community and have fewer opportunities to earn a livelihood through employment. This leads to such groups being over-represented in the lower social classes (Hall et al., 2005). Finally, at a more macro-country level, low and middle income countries have fewer opportunities to screen for antenatal genetic conditions so are more likely to have more cases of ID (Dave, Shetty, & Mehta, 2005).

Instruments Being Used

The difference in prevalence based on type of instruments being used is also a key factor in research on ID. King et al. (2009) and Maulik et al. (2011) describe the reduction of rates of ID that would occur if assessment methods changed from assessment based only on psychological test (IQ) to assessment based on measurement of adaptive skills or based on both adaptive skills and psychological test. Any study or dataset that includes assessment of adaptive skills will show lower prevalence rates than those that are solely based on quantitative assessment of IQ. Using standardised diagnostic systems also increases the accuracy of diagnosis (Maulik et al., 2011).

The use of such diagnostic instruments also varies according to the type of study or data source being used—community-based studies, clinical studies, government or school-based administrative data. Case identification techniques vary according to the need and resources available.

Detailed assessment involves extra resources in time, personnel and cost and may not be uniformly administered.

Early Epidemiological Studies on ID

Some of the earliest epidemiological studies on ID are from Iceland and Denmark, where clergy identified 2.3 and 0.9 % of all individuals as "mental defectives" in each country, respectively (Hübertz, 1843). Later research found "intellectual subnormality" in 4 % of the individuals registered in Iceland's registries between 1895 and 1897 (Helgason, 1964). This group was comprised of individuals with an IQ below 90. The study found higher prevalence among males and those in lower socio-economic conditions. A study conducted in children born in Edinburgh, Scotland, between 1950 and 1956 found the rates of "mental handicap" (IQ < 70) to be 1/1,000 (Drillien, Jameson, & Wilkinson, 1966). Greater prevalence was seen among male children and among children from poorer households.

More Recent Epidemiological Studies

Prevalence Estimates

Figure 2.1 shows the forest plot of the 52 studies included in the meta-analysis (Maulik et al., 2011) subdivided according to the economic group of the country (World Bank, 2010). The studies were published since 1980. Overall prevalence was 10.37/1,000 (95 % CI 9.55–11.18/1,000 population). The rates are directly proportional to the income group of the country, with the highest rates in the low income countries. The meta-analysis highlighted some factors that determine the prevalence, such as income group of the country, diagnostic criteria, age of the study population, socio-economic strata of the study population, study design and sampling strategy and type of instruments used to diagnose ID (Table 2.1). Mild, moderate, severe and profound ID account for 85 %, 10 %, 4 % and 2 % of the population affected with ID, respectively (King et al., 2009).

Incidence Estimates

There are few longitudinal studies that report incidence rates of intellectual disability. Heikura et al. (2003) reported that two different cohorts in Finland, one from 1966 and one from 1985 to 1986, had similar cumulative incidences of 12.6/1,000 for mental retardation of any severity as measured over an 11.5-year period of follow-up for each study. The cumulative incidence for mild mental retardation was 7.5/1,000 compared to 5.0/1,000 in the younger cohort, whereas the rates for IQ < 50 were 5.1/1,000 in the younger cohort and 7.6/1,000 in the older cohort. The authors believe that earlier assessment of intellectual capacities by psychological tests had led to a higher number of children diagnosed with mild intellectual disability in the younger cohort. Compared to this study, 8-year cumulative incidence was found to be 9.1/1,000 in the USA (Katusic et al., 1996). While the study from Finland found higher rates for mild mental retardation compared to the US study, the rates for more severe mental retardation were similar. Heikura et al. (2003) opined that the US study may have missed some cases of mild mental retardation because of its shorter follow-up period, as the milder cases tend to be identified a little later in life. Differences in case finding and identification are also suggested as possible reasons for the differences in rates.

Mortality Estimates

Patja et al. (2000) followed almost 2,400 people with intellectual disability across Finland for 35 years and found their mortality rate to be 18/1,000 person-years. Those with more severe forms of mental retardation had lower life expectancies, but those with mild mental retardation had similar life expectancy to that in the general population in the first 3 decades of life. Among 2–9-year-old children low IQ and epilepsy were significantly associated with lower life expectancy. In those over 40 years of age, low IQ, ageing and visual impairment were significantly associated with lower life expectancies.

Another cohort study from Western Australia also established the same relationship between severity of mental retardation and life expectancy.

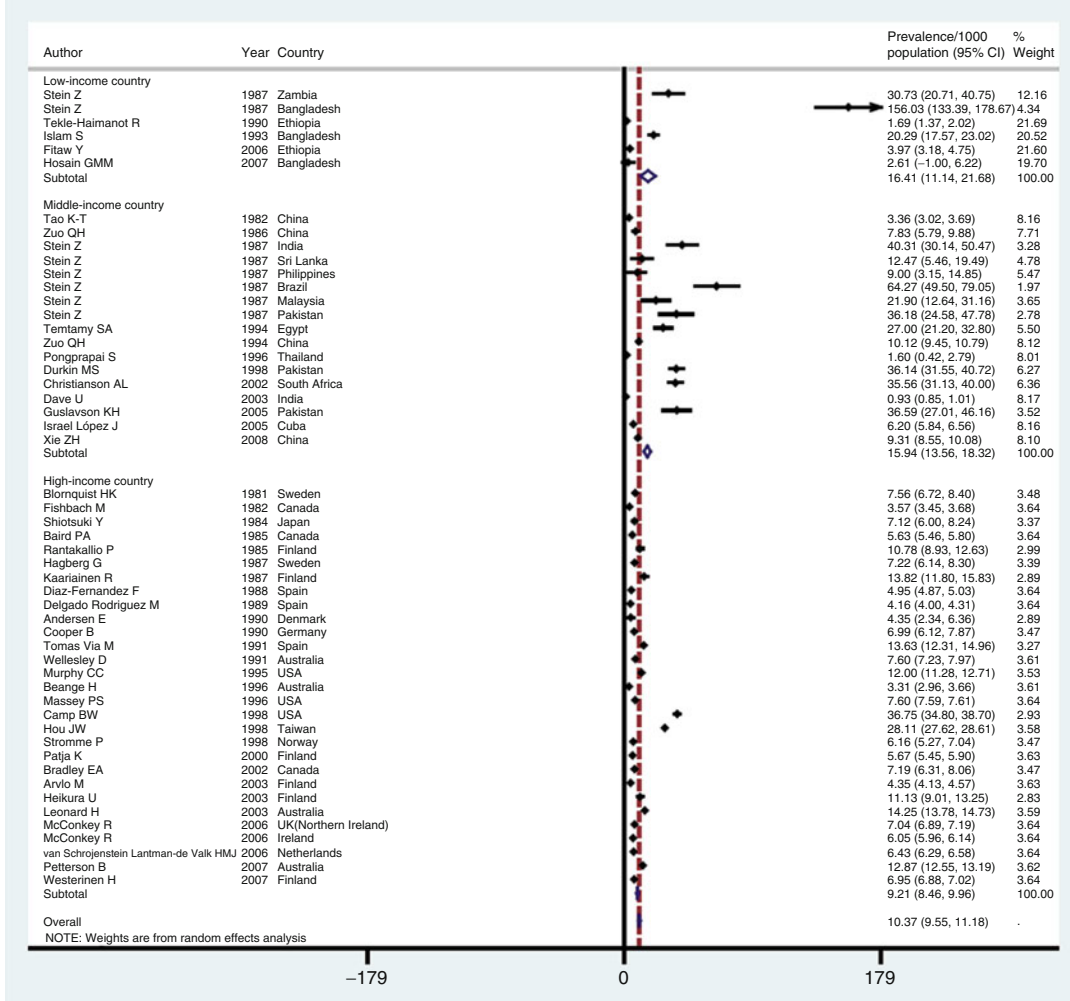

Fig. 2.1 Forest plot of studies on prevalence of intellectual disability by income group of country ($N=2$) (Maulik et al. (2011))

In this study, median life expectancy was 74 years, 67.6 years and 58.6 years, for people with mild, moderate and severe disability, respectively (Bittles et al., 2002). The median survival probability was 68.6 years in people with intellectual disability compared to 75.6 years and 81.2 years in males and females in the general population. Male survival rates were almost 5 years lower than the rates for females. Survival rates were also lower among those from the indigenous population. Whalley and Deary (2001) followed up data for more than 2,000 children with ID in Aberdeen, Scotland, and found that lower IQ at age 11 was significantly associated with lower survival rate at 76 years. Having an IQ level that was 1-standard deviation below the mean for the general population was associated with a reduced relative risk of survival of 0.79, and having an IQ of 2-standard deviations below the mean resulted in a reduced relative risk of 0.63. People with intellectual disability have an increased prevalence of comorbid physical illnesses like cardiovascular diseases, respiratory diseases, and Alzheimer's disease (Kilgour, Starr, & Whalley, 2010). However, these associations are confounded by other factors such as low socio-economic status, smoking, obesity

and other lifestyle factors. This does not include cases with more severe forms of ID who have genetic conditions that lead to premature death. Down's syndrome, which is one of the most common genetic causes for ID, has shown an increased association with celiac disease and Alzheimer's disease (Roizen & Patterson, 2003), which lead to lower survival rates.

Aetiological Factors and Physical and Psychological Conditions Associated with ID

Both aetiological factors associated with ID and comorbid physical or psychological conditions are discussed in detail in other sections of the book. In most cases the aetiology for ID is unknown. Genetic or physical disorders associated with the antenatal, perinatal and postnatal periods are equally responsible for almost 50 % of cases where a cause can be identified (Maulik et al., 2011). Down's syndrome is the most common genetic condition associated with ID, but other causes like brain trauma during delivery, birth asphyxia, intrauterine growth retardation, infections affecting the nervous system, hypothyroidism and iodine deficiency and lead poisoning are also commonly known to cause ID.

Harris (2006) reports that hearing impairment is present in 10 % of people with intellectual disability and that seizure disorder is present in less than 5 % to almost 30 % of people with intellectual disability, depending on the level of severity of mental retardation. Similarly, psychological problems are more than 4–5 times more prevalent in people with intellectual disability than in the general population. People with intellectual disability have psychological problems similar to those found in the general population, such as affective disorders, psychotic disorders, addiction disorders and other developmental disorders. Behavioural problems are also manifested more frequently in people with intellectual disability. A major problem of identifying physical and more importantly psychological problems in people with intellectual disability is correctly ascertaining the symptoms and diagnosing the conditions. Given the inability of some people with intellectual disability to express their distress—especially those with more severe forms of intellectual disability—proper diagnosis becomes critically important. The lack of trained personnel to facilitate that process adds to the problem.

Health Services Epidemiology

The World Health Organisation reports that only 39 % of countries have a specific national policy or programme of services related to intellectual disability (WHO, 2007). Mental health care for people with intellectual disability varies widely around the world with some countries having highly developed specialist mental health services (Bouras & Holt, 2010). In other parts of the world, there is a wide variation in services, such as in Asia (Kwok & Chui, 2008). This variation is in part explained by how healthcare systems are funded and how far deinstitutionalisation has progressed but is also due to the availability of expertise to develop services (Cain et al., 2010). The development of expertise is in part dependent on the infrastructures in place for the training of professionals providing mental health care. There are examples of effective training interventions using well-established training resources (Costello, Hardy, Tsakanikos, & McCarthy, 2010).

One of the key influences leading to the development of community-based mental health services for people with intellectual disability has been the extent in which countries have taken forward the closure of institutions in the form of the large hospitals. For example, in the UK, USA, Canada, Norway and Sweden, the closure of the institutions has led to more community-based community mental health and outpatient services, but in other countries the trend is towards more institutionalised provision (Chou & Schalock, 2007).

The evidence on the effectiveness of mental health services has been limited by few randomised controlled trials. The small evidence indicates that people with intellectual disability have more severe problems and receive more interventions than those without intellectual disability

(Chaplin, 2011). Also that those with severe ID tend to be lower users of inpatient mental health services than those with mild ID (Hemmings et al., 2009). In many parts of the world such as Asia, Latin America and Africa (Jeevanandam, 2009; Mercandante, Evans-Lacko, & Paula, 2009; Njgena, 2009), there is a dearth of research on the effectiveness of services to draw any conclusion on service developments in these parts of the world. For some parts of the world, poor resource allocation with the challenges of diseases such as HIV and war may make it a significant long-term challenge to develop mental health care for people with intellectual disability. We need to know much more about the prevalence and presentation of mental health problems in each country in order to plan services in the context of the health policy and resources available to an individual country.

Gaps in Knowledge and Future Research

While a number of areas related to the epidemiology of ID have lacunae that need to be plugged, a few broad gaps in extant knowledge are outlined below. Current knowledge about ID and facilities available for people with intellectual disability suggest that there are a number of gaps. While there are numerous epidemiological studies from the developed countries, there are fewer such studies from low and middle income countries. Studies from Africa, Latin America and most areas of Asia are lacking, and meta-analysis (Maulik et al., 2011) revels that there are variations across countries based on the economic prosperity; hence, there is a need for further research from low and middle income countries. Even within high income countries, there are large inequities and accessibility problems to available services across different economic strata of the society. The poor are often at a disadvantage in accessing services across all countries. Thus, there is a paucity of knowledge both about the magnitude of the problem and the specific issues relevant to marginalised groups of the society. Other determinants of ID are also poorly researched, currently, and future research should focus more on factors that have been associated with ID across different cultural settings.

Current studies suggest that knowledge about the aetiology of ID is also limited. Almost half of the cases have unknown aetiology and more research needs unravel the causes of ID. As genetic and environmental research develops further, it is hoped that more cases of ID will have identified aetiology and possibly also have the medical knowledge to prevent them or reduce the burden of such illnesses.

Information about health services specific for people with intellectual disability is limited. While some knowledge is available from high income countries, there is little literature suggestive of good evidence-based knowledge about service use from low and middle income countries. Studies that have included cost of services are also limited. One study reported that the cost of management of older people with ID is more than GBP 41,000/year (Strydom et al., 2010). Another on direct health cost for children and adolescents with Down's syndrome estimated costs to be more than USD 4,000/year (Geelhoed, Bebbington, Bower, Deshpande, & Leonard, 2011). However, more studies are needed to assess the cost of ID across different conditions and age groups and future research should focus on studies that cost good evidence-based practices.

Research is also lacking about the community-based services including educational and vocational services relevant to people with intellectual disability. What types of services exist, or what gaps in knowledge about the effectiveness of existing services are lacking. Research around national and international policies relevant to people with intellectual disability also needs to be conducted, and gaps in existing policies need to be identified.

Conclusion

The history and epidemiology of ID is an evolving field. Over the centuries as more awareness has developed about ID and the problems faced

by people with intellectual disability through scientific improvements and changing societal norms, both the definition and management of ID have changed. From being ostracised and viewed as deviants of the society, people with intellectual disability have developed their own voice and found strong support through various national and multilateral initiatives. However, a lot remains to be done both from a research and service delivery point of view for making a significant impact on the lives of people with intellectual disability across the world.

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Principles of Clinical Assessment

3

Nadja Alim, Dimitrios Paschos, and Michael Hearn

Assessment of Intellectual Ability and Functioning

Assessment of Intelligence

Deficits in intellectual ability and adaptive functioning are central to the diagnosis of intellectual disability (ID). The classification systems (DSM-IV TR, American Psychiatric Association (APA) (APA), 2000; ICD-10, World Health Organization (WHO), 1993; American Association for Mental Retardation, tenth edition (AAMR 10), Luckasson et al., 2002) are utilized to categorize ID. Explicit reference to onset of deficit prior to the age of 18 is made through the DSM-IV and AAMR 10, whilst this is only implicitly suggested through the ICD-10 classification. Models of disability (e.g. International Classification of Functioning, Disability and Health, WHO, 2001; AAMR 10 model of intellectual disability, Luckasson et al., 2002) conceptualize human functioning as determined by multiple factors including mental health status.

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Diagnosis of ID relies on classification of intellectual ability through IQ testing, the assessment of adaptive behaviour and confirmation of clinical aetiologies. People with ID exhibit heterogeneous cognitive profiles respective of IQ level, aetiology, associated disorders and psychopathological comorbidities. Common symptoms, however, constitute poor language (Fink & Cegelka, 1982), syntax and vocabulary (Borkowski & Büchel, 1983) and an inability to use language to support thinking and reasoning (Vakil & Lifshitz-Zehavi, 2012). Deficits in simultaneously dealing with multiple aspects of problem situations are common (Campione & Brown, 1984), whilst it is suggested that attention shifting may be impaired (Reed, 1996). Moreover, spontaneous appreciation of relations between pairs of objects is diminished (Paöur, 1992) and short-term memory is impaired (Belmont & Butterfield, 1974; Campione & Brown, 1984; Ellis, 1970). Difficulties in working memory and executive functioning may feature (Carretti, Belacchi, & Cornoldi, 2010; Numminen, Service, & Ruoppila, 2002; Schuchardt, Gebhardt, & Mäehler, 2010). Cognitive profiling of individuals with ID is hence vital. Factor-analytic studies of intelligence test scores indicate that general intellectual ability is a unitary construct referred to as 'g' which is normally distributed in the testing population (Dreary, 2000).

The dominant model of the structure of human intelligence in the psychometric tradition is the theory of fluid and crystallized (Gf–Gc) intelligence. The Gf–Gc theory was developed initially

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by Cattell (1943, 1963) but since has been elaborated by Horn (1976, 1985, 1998). Fluid intelligence reflects the capacity to solve problems for which prior experience and learned knowledge and skills are of little use. It is considered to be measured best by tests having little scholastic or cultural content such as perceptual and figural tasks or verbal tasks that rely on relationships among common/familiar words (Horn, 1998). Crystallized intelligence reflects consolidated knowledge gained through education, access to cultural information and experience. According to this theory, crystallized intelligence reflects an individual's fluid intelligence as well as access to and selection of learning experiences (Horn, 1998). Consequently, among people of similar educational and cultural background, individual differences in fluid intelligence should strongly influence individual differences in crystallized intelligence. Yet persons from different cultural backgrounds with the same level of fluid intelligence should differ in crystallized intelligence (Horn, 1998).

Evidence from factor-analytic studies further leads to the conceptualization of a hierarchical three-order model of intellectual abilities (Carroll, 1993). General intellectual ability (g) is positioned at the top, with second-order abilities, such as crystallized intelligence, situated below, and third-order abilities, such as vocabulary skills, occupying the lowest hierarchy level. Intelligence tests, however, do not offer pure measures of g, second- and third-order abilities but rather yield a global score with some organizing outcomes in terms of, for instance, verbal and performance abilities. The validity of IQ tests has been broadly accepted (Harrison, Kaufman, Hickman, & Kaufman, 1988), whilst it is argued that populations' performances improve over time (Flynn, 1984, 1985). In this sense, as long as floor and ceiling effects are avoided, compatible scaling and psychometric properties of these instruments are assumed (Baroff, 2003; Harrison et al., 1988). The most commonly used assessment instrument for adults is the Wechsler Adult Intelligence Scale (WAIS; e.g. Wechsler, 2008), due to earlier versions of tests like the Stanford–Binet not having been normed for older ages. Nevertheless, various studies have shown divergent results questioning the validity of test-based inferences.

It has been shown that the WAIS produces higher IQ scores than tests such as the Stanford–Binet (e.g. Roid, 2003). Here, Bensberg and Sloan (1950) found a discrepancy of 7–20 IQ points between the two measures, whilst Brengelmann and Kenny (1961) found WAIS scores heightened by an average of 8 points. Spitz (1988) as well as Nelson and Dacey (1999) found WAIS scores heightened by 12.4 and 14.6 points, respectively. Comparing the WAIS (e.g. Wechsler, 2008), Stanford–Binet (e.g. Roid, 2003), Leiter International Performance Scale (Levine, 1993), the Slosson Intelligence Test (Nicolson & Hibpshman, 1991) and the Vineland Adaptive Behavior Scales Vineland Adaptive Behaviour Scales (VABS; Sparrow, Balla, & Cicchetti, 1984), Silverman et al. (2010) found a systematic underestimation of severity of intellectual impairment through the WAIS. The WAIS was reported to significantly overestimate IQ in contrast to the Stanford–Binet (mean difference = 16.7 points; Silverman et al., 2010). Moreover, the Stanford–Binet results in comparison to Vineland, Leiter and Slosson assessments did not report significant IQ differences, whilst the WAIS IQ scores were again significantly higher in comparison to these tests (Silverman et al., 2010). Silverman et al. (2010) suggest it to be concerning that past evidence between the so-called 'gold standard' IQ assessment measures had been so easily dismissed and calls for further research into the validity of existing IQ measures.

The Ravens Progressive Matrices (RPM) test taps into analogical reasoning and metacognition (e.g. reasoning), fluid intelligence components required in the activation of working memory, which is impaired in people with ID (Swanson, Christie, & Rubadeau, 1993; Vakil & Lifshitz-Zehavi, 2012). Developmental changes in the prefrontal cortex determine working memory abilities, that is, to form and manipulate mental representations between objects and events (Baddeley, Emslie, Kolodny, & Duncan, 1998; Morrison, Holyoak, & Truong, 2001; Waltz, Lau, Grewal, & Holyoak, 2000). Facon, Magis, Nuchadee, and De Boeck (2011) in a comparative study of subjects with and without ID confirmed that the RPM measures the same constructs

in the groups under investigation, suggesting that a large majority of items have identical discriminative power and difficulty levels across the two groups. Vakil and Lifshitz-Zehavi (2012) explain lowered performances in people with ID through eye-movement particularities, suggesting that subjects without ID looked at RPM materials for a longer time before shifting to the options, whilst eye movement of subjects with ID showed higher rates of switching between stimuli. The authors conclude that people without ID spend less time processing analogy and adopt a more efficient monitoring process than those with ID. Vakil and Lifshitz-Zehavi (2012) hypothesize that people with ID are more likely to adopt a response elimination pattern to solving novel problems (i.e. eliminating incorrect alternatives in order to arrive at the correct answer by default) rather than a constructive matching procedure which appears widely applied by people without ID (i.e. participants analyse the components of the task prior to solving the problem and only then search for the alternative answers).

Assessments of Adaptive Skills

'Adaptive behaviour' refers to the functioning of an individual in his or her environment. As such, adaptive behaviour draws together a person's cognitive and personality characteristics. Assessments of adaptive behaviour typically focus on domains such as communication, self-care skills and interpersonal relationships. See Schalock (2002) for a summary of psychometrically robust adaptive behaviour assessment tools, and commonly used tools in clinical practice include the VABS: Vineland Adaptive Behaviour Scales (VABS; Sparrow et al. 1984).

The literature on adaptive behaviour has been extensively researched and discussed with regard to various presentations within the field of ID. Platt, Kamphaus, Cole, and Smith (1991) reported low correlations between intelligence and adaptive behaviour when measuring these constructs in children with intellectual disabilities. Bölte and Poustka (2002) conclude that these abilities embody distinct psychological entities which may vary for specific syndromes and specific forms of ID making up profiles of specific skills and disabilities. Levels and patterns of adaptive behaviour are known to vary between specific syndromes (Howlin, Charman, & Ghaziuddin, 2011). Balboni, Pegrabissi, Molteni, and Villa (2001) suggest that individuals with ID and a specific disorder of communication, social behaviour or motor abilities show significant deficits in the corresponding subdomains of the VABS (Sparrow et al., 1984) irrespective of their level of ID. Additionally, however, those with more severe levels of ID showed further deficits in secondarily affected disorders (e.g. daily living skills in cases of people with motor disorder) in contrast to those with mild ID (Balboni et al., 2001).

Autism Spectrum Disorder

Kraijer (2000) conducted a comprehensive review of adaptive behaviour studies of individuals with Autism/Pervasive Developmental Disorder Not Otherwise Specified (PDD-NOS). The author discusses these results and concludes that the presence of Autism/PDD-NOS in addition to ID does not appear to affect some domains of adaptive behaviour such as self-help and gross motor skills when comparing people with moderate vs. severe levels of ID, whilst other domains show differences between these subgroups (e.g. communication and behaviour). Kraijer (2000) reviewed 11 studies using the VABS to assess people with Autism/PDD-NOS and ID, and the author concludes that people with Autism/PDD-NOS obtain significantly lower levels on the communication and socialization subdomains, whilst no between-group differences could be found for the daily living and motor domain skills. Another study by Volkmar et al. (1987) found significantly higher levels of maladaptive behaviours in individuals with Autism/PDD-NOS than those with a single diagnosis of ID only.

Overall it may be suggested that individuals with Autism/PDD-NOS and ID show most severe impairments with regard to social skills and socialization, communication and maladaptive/problem behaviour domains when compared to other subjects with ID only. On the other side,

self-help/daily living skills and gross motor skills domains appear to be similar in individuals with ID with and without Autism/PDD-NOS.

IQ, Adaptive Behaviour and Mental Health

The assessment of mental health in conjunction with social and adaptive skill deficits and excesses may be regarded as important in light of the association between psychological symptoms and adaptive behaviour (Matson, Smiroldo, & Bamburg, 1998). A study by Manikam, Matson, Coe, and Hillman (1995) suggests that adaptive behaviour functions as a moderator variable, mediating the relationship between depression and intellectual functioning. In contrast, a study investigating the relationship between bipolar disorders and social and adaptive skills in people with ID found significantly more negative verbal social skills in people with ID and bipolar disorder than those with other psychopathologies and those without mental health problems; nevertheless, no differences in terms of adaptive behaviours between the three groups were noted (Matson, Terlonge, González, & Rivet, 2006). Psychopathology shows greater prevalence in people with ID, which may be explained through biopsychosocial and developmental factors such as personality styles, social stigmatization, genetic aetiology and neurological deficits (Dykens, 2000). Whilst to date, no evidence exists, it has nonetheless been hypothesized that psychopathology may be caused by cognitive deficits or that a common cause, such as nervous system damage, may result in both cognitive deficits and psychopathology (Goodman, 1993). Furthermore, social competence difficulties encountered by children can be related to the emergence of other psychiatric problems. According to Reiss and Benson (1985), social isolation, stigmatization and poor social skills increase the risk that individuals with ID will present affective problems. However, this relationship was not supported by the results of the Tremblay, Richer, Lachance, and Côté (2010) study since, according to interviews with informants, the profiles of participants do not significantly differ with regard to problem behaviour.

Aetiological Factors

A two-group approach with regard to aetiology factors supporting classification of ID highlights 'organic' causes vs. social disadvantage factors (Kaski, 2000; Volkmar & Dykens, 2002), whilst these may overlap constituting a range of aetiology factors in cases. The AAMR 9 introduced a multifactorial system for classifying aetiological factors such as biomedical, social, behavioural and educational (see Luckasson et al. (2002) for a summary of aetiological risk factors). A study of monozygotic twins without ID reared apart enabled the separation of environmental variables and genetic influences on subjects' IQ scores (Bouchard, Lykken, McGue, Segal, & Tellegen, 1990). The authors found high correlations of IQ scores, whilst no significant relationship was evident for environmental factors assessed. Wilson (1983) in a study comparing IQ developments between monozygotic and dizygotic twins as well as their non-twin siblings and investigating the pacing of spurts, plateaus and lags in cognitive development profiles suggests that cognitive development appears determined by an underlying genetic ground plan. This is further supported by an older adult monozygotic and dizygotic twin study (McClearn et al., 1997) revealing that even in older adulthood the influence of genes on general and specific cognitive functioning is noteworthy, whilst environmental factors are further suggested to be clearly evident. Environmental influences on intelligence developments in societies were further investigated by Flynn (1984, 1987, 1999) who suggested that, when assessing subjects using old versions of standardized IQ tests, significant rises in IQ scores could be reported between older and younger generations (Flynn effect; Flynn, 1984). Robustness of data collated was further enhanced by comparing and assessing adults rather than children (to avoid bias with regard to children nowadays reaching cognitive maturity sooner),

collecting data of large samples (to avoid sample bias), using culturally reduced tests (i.e. RPM so to avoid cultural bias) and using raw rather than converted scores (Flynn, 1987). Flynn (1987) again established significant IQ gains over time and further found that the most significant gains were those of fluid rather than crystallized intelligence skills. Overall, the extended research on the genetics of IQ development has widely suggested that variation in IQ in Western societies is highly heritable and that intellectual abilities increase from generation to generation (Plomin, DeFries, McClearn, & McGuffin, 2000). Increases in IQ scores have led to a criticism of using current IQ tests which do not take account of this rise and diagnostic criteria, which are still suggesting IQ cut-offs for people with ID to be at an IQ of 70 (Flynn, 2000). It has been argued that adaptive skills ought to account for a better predictor of profile associated with people with ID (Flynn, 2000).

Assessment at various stages of a person's life will aid differential diagnosis, highlight co-morbidities and may inform treatment as well as potential prevention of the disability. Early screening and a multidisciplinary approach to the detection of, for instance, metabolic disorders, specific clinical syndromes, language disorders/delay, seizure disorders and multiple disabilities are widely acknowledged (Simonoff, Pickles, Chadwick, Gringras, Wood, Higgins, Maney, Karia, Iqbal, & Moore, 2006; McKenzie & Megson, 2011).

Psychiatric Assessment of People with ID

General Principles

The assessment of mental health problems in people with ID follows the same principles of the psychiatric assessment for people without ID with appropriate modifications to adjust for lowered intellectual functioning, language limitations, sensory impairment and physical disabilities. Such modifications usually include flexibility in the setting and length of the assessment, use of pictures or symbols and relying more on observational data and informants' accounts. The interviewer would often need to use simple and unambiguous language and avoid the use of idioms or metaphors. Some people with ID may be suggestible and acquiescent, so leading questions are not helpful. Open questions, questions with multiple choices and contradictory questions may be employed to ensure understanding of what is being asked.

Because of the heterogeneity in cognitive abilities and language skills among persons with ID, it is difficult to use the same method for psychiatric assessment across the whole spectrum. Common psychiatric signs and symptoms, such as social withdrawal, lack of concentration, and stereotyped movement disorders, can be due to underlying brain damage rather than a superimposed psychiatric illness known as 'diagnostic overshadowing' (Reiss & Sysko, 1993). Having an accurate baseline of previous abilities, functioning and behaviours is therefore to detect changes that could signal the onset of a co-morbid psychiatric disorder.

Whilst reliance on informants' accounts would often be necessary, significant discrepancies in reported problems may emerge. Moss, Prosser, Ibbotson, and Goldberg (1996) conducted psychiatric assessment of 100 people with ID attending a specialist service and their key informants using the Psychiatric Assessment Schedule for Adults with a Developmental Disability (PAS-ADD). They found significant disagreements between patient- and informant-reported symptoms, with only 40.7 % of cases detected by both interviews.

ID is a common condition with a prevalence rate of around 1.37 % (Maulik, Mascarenhas, Mathers, Dua, & Saxena, 2011) and all psychiatrists, regardless of their specialization, are likely to be requested to assess a person with ID. People with ID are more likely to develop a psychiatric and behavioural disorder (Cooper, Smiley, Morrison, Williamson, & Allan, 2007; Smiley, 2005), and the point prevalence of psychiatric disorders in this population has been reported as high as 40.9 % (Cooper et al., 2007). Psychiatric disorders may be linked to ID in various ways

(Harris, 2006). The same neurobiological factors that cause ID may also predispose the person with ID to develop a mental illness. Poor problem-solving skills due to cognitive impairment may lead to inflexible patterns of behaviour. Mood dysregulation, aggression and low tolerance of frustration may be symptoms of an organic brain disorder such as frontal lobe damage.

Referrals of person with ID for psychiatric evaluations may originate from primary care, general psychiatric, medical and social care settings. Routes of referral and individual patient characteristics are sometimes related to certain diagnostic categories. Cowley et al. (2004) collected data on 752 persons with ID referred to a specialist mental health and ID service in South London. Older age, mild ID, admission to an inpatient unit and referral from generic mental health services tended to be associated with an increased presence of psychiatric symptoms, whilst severe ID, presence of epilepsy and residence with the family were associated with lower incidence of psychopathology.

An essential aspect of the initial assessment is to exclude any physical cause for psychiatric symptoms or behavioural problems as people with ID often have significant physical co-morbidities (Disability Rights Commission, 2006). A full physical examination, laboratory tests (blood tests, imaging) as required and the review of other medical conditions such as epilepsy are essential at the start of screening phase. Medication review may reveal side effects that sometimes mimic or cause psychiatric symptoms.

Epidemiological studies of psychiatric disorders in persons with ID report wide variation in prevalence rates, and this is most commonly attributed to sampling and case ascertainment bias. The use of standardized screening instruments and modified diagnostic criteria such as the DC-LD (Royal College of Psychiatrists, 2001) and DM-ID (Fletcher, Loschen, Stavrakaki, & First, 2007) can lead to better case ascertainment as traditional classification and diagnostic systems can undercount mental health problems in this population (Cooper et al., 2007).

Arriving to a diagnosis is an important part of the psychiatric assessment. In patients with ID this may be difficult after the first assessment and revisiting a working/provisional diagnosis is common. Many times, supports and intervention will be needed even when no definite or even provisional diagnosis can be made.

There are various standardized ways to organize a psychiatric assessment. A brief outline of useful areas to cover is shown below, but it is not a comprehensive list:

- Demographic details, source and reason for referral. Documentation of confidentiality and sharing of information agreements
- History of chief complaint with details of frequency and intensity of any challenging behaviours
- Review of psychiatric symptoms including psychotic, affective and anxiety symptoms
- Recent life events and precipitating factors
- Past psychiatric history (e.g. previous admissions, prescribing of psychotropic medication)
- Past medical history (e.g. review of co-morbid epilepsy, endocrine and metabolic disorders, sensory impairment and any other physical health problems)
- Family history of ID or mental health problems
- Personal history with emphasis on developmental and school history, special needs education
- Training and vocational record
- Relationships, sexual history
- Premorbid personality and functioning
- Substance misuse history
- Forensic or offending history
- Mental status examination
- Assessment of mental capacity to make decisions about treatment
- Clinical and structured risk assessment estimating the risk to self and/or others, as well as the risk of self-neglect, abuse and exploitation
- Physical examination
- Clinical investigations (e.g. neuroimaging, chromosomal analysis/array CGH, baseline physical investigations prior to pharmacotherapy such as glucose, lipid profile and ECG before starting antipsychotic medication)

The main categories of psychiatric disorders and their manifestation in people with ID are discussed below to inform clinicians of the issues that need to be explored in a clinical psychiatric assessment.

Schizophrenia

Schizophrenia-spectrum psychoses have been consistently reported to be over 3 times more prevalent in people with mild ID in comparison to the general population (Bouras et al., 2004; Morgan, Leonard, Bourke, & Jablensky, 2008). Schizophrenia is also associated with cognitive decline. Specific cognitive deficits have been linked with increased risk of schizophrenia, and a common neurobiological deficit, modified by genetic and environmental factors, is likely to be implicated.

People with ID due to specific genetic conditions are more likely to develop schizophrenia. This has attracted research attention as a possible way to understand better the link between disordered neurodevelopment and schizophrenia. Vogels et al. (2004) used the operational criteria checklist in a small sample of adults with Prader–Willi syndrome and psychosis to identify a subtype of psychotic disorder associated with early age of onset and polymorphous and changing symptoms. Chromosome 22q11.2 microdeletion (velo-cardio-facial syndrome) has been strongly linked to schizophrenia (Murphy & Owen, 2001; Philip & Bassett, 2011) with up to 20–25 % of affected individuals developing the condition. A shifting cognitive profile, anxiety disorders and predisposition to seizures have also been reported in persons with 22q11.2 and schizophrenia.

The diagnosis of schizophrenia relies mostly on accounts of characteristic symptoms that can be grouped together according to diagnostic classification. Schneider's first-rank symptoms are considered central to the diagnosis of schizophrenia, but they often refer to language-based, complex concepts that depend on self-reporting which makes it difficult to reliably elicit in people with ID. Furthermore, delusions of control could be hard to evaluate in people whose lives may often be entirely controlled by others. Loss of volition or poverty of speech may be of uncertain validity when applied to those with more severe ID. Moss, Prosser, and Goldberg (1996) reported schizophrenia symptom frequency, as detected by PAS-ADD, to be positively correlated with IQ and noted particular difficulties with identifying delusions and negative symptoms. Auditory hallucinations were the most likely psychotic symptoms to be reliably detected. However, caution is required when evaluating auditory hallucinations in persons with ID, especially in the presence of developmentally appropriate phenomena, such as speaking to oneself or having 'imaginary friends' (Pickard & Paschos, 2005). For those with mild ID, dimensional scales that have been developed for the general population such as the PANSS and PSYRATS can be used to assess some psychotic symptoms but less reliably for delusions or negative symptoms of schizophrenia (Hatton et al., 2005).

Mood Disorders

Depression is a common mental health problem in adults with ID. As it is also the case in the general population, depression in persons with ID is frequently undiagnosed. Increased prevalence rates have been reported (Richards et al., 2001) and vulnerability factors are likely to include higher rates of physical illness, socio-economic adversity and reduced problem-solving skills and support networks. Syndrome-specific vulnerability for depression has been previously advocated for people with Down syndrome; however, a recent literature review by Walker, Dosen, Buitelaar, and Janzing (2011) has casted doubt on this hypothesis.

The core features of depression can be reliably identified in those with mild ID, although atypical presentation has also been reported, e.g. hypersomnia and increased appetite (Deb, Matthews, Holt, & Bouras, 2001). In people with more severe cognitive impairment, challenging and aggressive behaviours are sometimes seen as 'behavioural equivalents' of depressed mood. Also, an excess of affective symptoms has been

identified in adults with ID and a history of self-injurious behaviours (Marston, Perry, & Roy, 1997). However, Tsiouris, Mann, Patti, and Sturmey (2003) reported on a sample of people with ID ($n=92$) with a third of participants having a diagnosis of depression that self-injury or aggression was not strongly associated with depression. Moreover, item and factor analysis of the scale indicated that the use of core DSM-IV symptoms was the best way to diagnose depression in their sample rather than reliance on behavioural equivalents or other proxy measures. As with other psychiatric disorders in people with ID, caseness is increased with use of specially developed screening tools for depression in people with ID. A systematic appraisal of such tools has been published by Hermans and Evenhuis (2010).

Bipolar affective disorder can be recognized and differentiated from other behavioural and psychiatric disorders in individuals with ID. For those with mild ID, ICD-10 and DSM-IV criteria can be applied (Cain et al., 2003). Rapid cycling bipolar disorder (more than four episodes of either mania or depression in a year) is thought to be more common (Vanstraelen & Tyrer, 1999). Manic episodes may present with the person's mood being predominantly irritable rather than euphoric.

The assessment of affective disorders should always include inquiries about suicidal thoughts or behaviours. Suicide in adults with ID has been reported, but very few detailed studies have been undertaken (Merrick, Merrick, Lunsky, & Kandel, 2006). Suicide rates in people with ID are lower than those seen in the general population, and suicide is extremely rare in those with more severe intellectual impairments. Hassiotis, Tanzarella, Bebbington, and Cooper (2011) looked at rates of suicidal ideation and acts in a general population sample. 16.1 % of participants ($n=1{,}053$) met criteria for borderline ID and were found more likely to report previous suicidal attempt and deliberate self-harm. Although these associations were not significant after controlling for income and age, their sample did not include people living in care homes, who are more likely to have higher rates of self-harm and psychiatric illnesses.

Anxiety and Stress-Related Disorders

Anxiety disorders have been described across the spectrum of ID. Behavioural equivalent (such as drinking excessively as a result of dry mouth) and autonomic arousal can be part of the symptoms in people with severe ID. Reid, Smiley, and Cooper (2011) estimated 3.8 % point prevalence of all anxiety disorders derived from a large-scale, population-based study. A recent history of life events was independently associated with having an anxiety disorder.

Traumatic experiences and life events have been described in people with ID (Martorell & Tsakanikos, 2008) who may be more vulnerable to PTSD, because of the increased incidence of traumatic experiences, such as sexual abuse. Such experiences often go unreported and people may not have the communication skills to describe their experiences, whilst post-traumatic stress disorder symptoms may be wrongly attributed to other psychiatric diagnoses (McCarthy, 2001).

Anxiety disorders can be more accurately diagnosed with use of structured questionnaires (Hermans, Van der Pas, & Evenhuis, 2011). Such an example is the psychometrically valid tool Glasgow Anxiety Scale for people with ID (Mindham & Espie, 2003).

Personality Disorders, Eating Disorders and Substance Misuse

Assessment for personality disorders (PD) is clinically important, but there are diagnostic challenges mainly because of difficulty in obtaining an accurate baseline of behaviours and a long-term account of functioning and symptoms and also due to significant symptom overlap with other psychiatric and behaviour disorders (Royal College of Psychiatrists, 2001; Alexander & Cooray, 2003).

Eating disorders, across the whole spectrum of abnormal eating behaviours, can be identified in this population. This may include, for example, anorexia nervosa in persons with mild and moderate ID and pica in those with severe and profound ID. More cases can be identified when modified diagnostic criteria are applied (Gravestock, 2003).

People with ID overall show a lower level of use of both alcohol and illicit drugs, compared with the general population (Barrett & Paschos, 2006); however, those with ID and substance misuse problems may suffer excess morbidity and mortality because of coexisting physical disorders.

Dementia

Dementia in people with ID is more common (Strydom, Livingston, King, & Hassiotis, 2007) and the higher incidence of early onset Alzheimer's dementia in people with Down syndrome is now firmly established and several possible risk factors have been suggested. An important component of the clinical assessment of suspected cases of dementia is to exclude any treatable condition and co-morbid physical (e.g. hypothyroidism) or mental illness (depression). The Mini Mental State Examination is unsuitable for this group. Some people with ID may have very limited cognitive and practical abilities, and a small decline may be missed at initial stages. Establishing a baseline, documenting the highest level of functioning that the person had and tracking any changes in the future can be more reliable with the use of specifically designed questionnaires such as the Dementia Screening Questionnaire for Individuals with Intellectual Disabilities (Deb, Hare, Prior, & Bhaumik, 2007).

Psychiatric Disorders in ASD

Symptom ascertainment when conducting clinical assessment of people with ASD may be hindered by limited engagement in the assessment process, language limitation, difficulties in identifying emotional states and concrete thinking. People with ASD comprise a heterogeneous group including many with varying degrees of ID (which is independently associated with increased rates of psychopathology). It is still not entirely clear if ASD is an additional risk factor for psychopathology. For example, whilst Bradley, Summers, Wood, and Bryson (2004) found that adolescents and adults with ASD and ID presented more psychiatric symptoms than those with only ID, Tsakanikos et al. (2006) found no differences in the number of psychiatric diagnoses in a large study comparing attendees of a specialist service with ASD and ID vs. those with only ID. Attention deficit hyperactivity disorder (ADHD) symptoms often coexist with ASD; however, DSM-IV excludes a mutual diagnosis of ASD and ADHD. This may cause uncertainties on whether to treat ADHD symptoms when they are excessive and burdensome in the absence of clinical diagnosis of ADHD.

McCarthy et al. (2010) investigated the association of psychiatric symptoms and behavioural disorder in adults with ID and ASD ($n=124$) with a group of adults with ID only ($n=562$). They reported a fourfold increase in challenging behaviour in adults with ASD as compared to non-ASD adults, but after controlling for level of ID, gender and age, there was no association between co-morbid psychiatric disorders and presence of challenging behaviour.

The Assessment of Challenging Behaviour

This section will review very briefly the strategies for assessing challenging behaviour. A more in-depth analysis can be found in Chap. 17. The dominant approach to understanding challenging behaviour is behaviour analytic. The historical antecedent to this approach draws directly on Skinner's (e.g. Skinner, 1953) description of operant learning processes. The assumption of the model is that behaviour is purposeful or has function, in the sense that it results in an 'outcome' for the person concerned. The literature has identified broad functions as follows: (a) contingent access to social interaction/attention or access to an item or activity (positive reinforcement), (b) escape from an interaction/event (negative reinforcement) and (c) automatic reinforcement for non-socially reinforced behaviours (Iwata, Dorsey, Slifer, Bauman, & Richman, 1994; Iwata, Pace, et al., 1994).

The strategies used in identifying these variables can be divided very broadly on two parameters: (1) descriptive and experimental approaches and (2) indirect and direct approaches. The first distinction concerns whether or not the researcher/clinician has any form of experimental control over antecedent or consequent variables (hypothesized to be in operation). The interested reader is referred to an influential early paper by Baer, Wolfe, and Risley (1968) that set the scene for the experimental analysis of behaviour; in particular these authors noted that in order to understand behaviour there needs to be a believable demonstration of experimental control. Direct approaches involve measurements of behaviour as it happens (e.g. recording on an ABC chart, carrying out systematic observations) as opposed to retrospective reporting on a scale or interview schedule. This brief summary will focus on the distinction between descriptive and experimental approaches to behavioural assessment.

The assessment of challenging behaviour invariably does not rely on any single method, rather a combination of approaches (e.g. records review, clinical interview and undertaking natural observations). What different methods should have in common though, according to Horner (1994), is (1) defining the challenging behaviour(s) of concern, (2) identifying antecedent events (triggers, setting events, discriminative stimuli (SD) and establishing operations (EO; see Michael, 1982, 1993)), (3) hypothesis development regarding maintaining variables and (4) direct observations (whether in natural setting or analogue conditions).

Descriptive Approaches

The defining feature of these approaches is that that the data derived are correlational in nature and inferences are made about functional relations, rather than deriving experimental evidence of cause and effect. In this brief review, the following approaches will be summarized: (1) rating scales, (2) informant-based interview, (3) scatterplots and (4) direct observations (including antecedent-behaviour-consequence, ABC, charts and more systematic time-based sequential analysis).

Rating Scales

Two commonly used rating scales are the Motivation Assessment Scale (MAS) (Durand & Crimmins, 1992) and the Questions About Behavioral Function (QABF) (Matson & Vollmer, 1995). The interested reader is also referred to the Motivation Analysis Rating Scale (MARS) (Wiesler, Hanzel, Chamberlain, & Thompson, 1985), although this scale is not being reviewed here.

The MAS is a 16-item scale, comprising Likert-scaled responses. The form assesses functional processes of access to attention or tangible items, escape and automatic reinforcement. Questions require the third-party respondent (who knows the service user well) to consider instances of challenging behaviour being more likely following particular antecedent events (e.g. following requests in the case of escape maintained behaviour) or being more likely to subside after certain events (e.g. access to interaction in the case of positive reinforcement). Item scores are totalled and hypotheses about function(s) are derived for one or more of the four reinforcement processes. Although the MAS has some intuitive appeal and is quick and easy to administer, there is some research that questions its psychometric properties in relation to factor structure (Bihm, Kienlen, Ness, & Poindexter, 1991) and inter-rater reliability (Sigafoos, Kerr, & Roberts, 1994). Conversely, Paclawskyj, Matson, Rush, Smalls, and Vollmer (2001) report reasonable correlation with outcomes for the QABF as described below.

The QABF is a 25-item scale, identifying the same reinforcement processes as the MAS, with the addition of questions that relate to challenging behaviour associated with pain. Respondents are required to answer questions ranging from never applies to behaviour occurring often in particular circumstances. Processes that are endorsed the most or attract the highest total scores are hypothesized to have some controlling function. Paclawskyj et al. (2001) compared the QABF, MAS and analogue approaches (see experimental approaches for analogues) and reported more agreement between the former and analogue

outcomes (i.e. 69.2 %) than with the MAS and analogue outcomes (i.e. 53.8 %) in those cases where the undifferentiated data from analogues were excluded. This suggests slightly improved concurrent validity of the QABF (i.e. concurrent with analogue assessment) over the MAS.

Informant-Based Interview: The Functional Analysis Interview

The functional analysis interview (FAI) (O'Neill et al., 1997) is probably the most widely used structured interview in clinical practice, undertaken with someone who knows the service user well. It does not derive a score but instead serves as a very useful tool for gathering information across eleven sections (e.g. descriptions of behaviour, ecology, frequency, possible functions of behaviour, communication strategies, health issues). However, despite its common usage, Floyd, Phaneuf, and Wilczynski (2005) report a scarcity of studies where the psychometric properties of the FAI are considered.

Scatterplots

The scatterplot was described in an early paper by Touchette, MacDonald, and Langer (1985) as a method of graphing the frequency and intensity of responses within given time windows (e.g. 1-h intervals). They describe this method as useful when it is difficult to identify precisely the controlling variables. In a series of three case studies, these authors show how this method is useful for identifying patterns of responding across time. Unfortunately, there is very little evidence in the literature regarding the psychometric property of this assessment approach.

Direct Observations

The use of ABC charts, for recording antecedent-behaviour-consequences, is a well-used approach in clinical practice. It does however attract criticism on several levels, chiefly focusing on issues around reliability and validity. In one early study, Sasso et al. (1992) reported a correspondence between the findings of ABC chart analysis and experimental functional analysis for a case report involving two participants. However, in their survey of functional assessment methodologies, Herzinger and Campbell (2007) reported on a systematic review of 58 articles (including descriptive and experimental approaches) and noted only eight articles where the use of an ABC chart had formed part of the assessment, and here, there were no cases involving the sole use of ABC charts, making comparisons with findings from other methods impossible. There are some indicators for use of ABC charts, in the absence of other approaches, for example, developing hypotheses around low-frequency behaviours, or for those occasions where direct observation by the clinician could be problematic (e.g. personal care of participant reactivity). Sturmey (2008) has made suggestions around improving ABC chart usage including staff training, the recording of absence of events, monitoring and feedback.

The development of computer software has been associated with improvements in the ability to carry out systematic observations. Software can allow the real-time recording (or coding of video recorded material) of onsets and offsets of key variables. As well as the recording of frequency, this allows the preservation of sequences of events and therefore, one can establish the temporal relations between occurrences of challenging behaviour and potentially controlling variables (assessed by conditional probability analyses), whether antecedent or consequent to challenging behaviour (Oliver, Hall, & Murphy, 2005). In an early comparison study, Lerman and Iwata (1993) reported that conditional probability analyses, based on observational data, were as effective as analogue assessments in identifying socially reinforced contingencies, but the descriptive approach was less effective at distinguishing between positive and negative reinforcement processes.

What all of the above approaches have in common is their descriptive nature, and as such, the argument runs that data derived from them is at best correlational (Sturmey, 2008) because there has been no experimental control.

Experimental/Analogue Approaches

Over the past 30 years, there has been a wealth of literature describing assessment and intervention case studies using experimental, or analogue, procedures.

The most well-known experimental variations are Iwata, Dorsey, et al. (1994) and Carr and Durand (1985). What these procedures have in common is the systematic control of variables, such as putative antecedent events. For example, the Iwata, Dorsey, et al. (1994) 'Alone' condition is a test for social positive reinforcement. Here, the experimenter is present in the session but only interacts with the participant contingent on occurrence of the target behaviour. The participant has unrestricted access to stimulating activities. Thus, this condition sets up the antecedent absence of social interaction (EO) and provides reinforcement for occurrence of the behaviour. The idea behind the approach is to compare rates of responding in this condition against rates in other test conditions (i.e. tests for social negative reinforcement, access to tangible items and automatic reinforcement). This approach is defined as the ABC model, where A and C are manipulated. The alternative by Carr and Durand (1985) is described as the AB model. Here, only the putative antecedent variables are manipulated and no reinforcing stimuli are programmed. The interested reader is also referred to variations where briefer sessions are used (Derby et al., 1992).

In their review of nearly 300 published papers using either of the above methods, Hanley, Iwata, and McCord (2003) noted that only 5 % of studies produced undifferentiated data outcomes that precluded an intervention. In a further systematic review, Herzinger and Campbell (2007) reported that analogue functional assessments, taken as a whole (whichever model used), were more effective at leading to interventions that suppressed challenging behaviour, when compared with descriptive approaches. Studies comparing conditional probability analyses of observational data with analogue approaches have suggested a further problem with the former approach, that there is an increased chance of inferring a (positive) reinforcement process because of increased likelihood of caregiver response, in natural settings, to occurrences of challenging behaviour even though the experimental analyses do not reveal this contingency as a controlling variable (Hall, 2005; Thompson & Iwata, 2007).

Whereas the analogue functional assessment method is largely held up as the gold standard for identifying function, it is not without its limitations. For example, there are training issues, the failure to identify more distil controlling variables (Horner, Day, & Day, 1997), poor ecological validity (Sturmey, 1995) and according to Paclawskyj et al. (2001) the difficulty of deriving clear functional relations for low-frequency behaviour. Hanley et al. (2003) addressed some of these criticisms (e.g. conducting assessments in natural settings and using people who know the service user within experimental sessions, i.e. preserving the reinforcement history that is most likely to apply in the natural setting). Despite the reported efficacy of analogue approaches over descriptive assessments, Sturmey (2008) notes such approaches are much less likely to be used in clinical practice (possibly because of some of the reasons shown above). However, Hanley et al. (2003) do note that on those occasions where analogue approaches produce undifferentiated data (i.e. no clear function), then there is a place for alternative strategies, such as descriptive observations in natural settings, for clarifying the idiosyncratic variables that might have a controlling function.

Assessment of Suitability for Cognitive Behaviour Therapy

It has been suggested that people with ID lack access to psychological and mental health services and that services available may not always meet the needs of their patients. Lack of evidence base of psychological practice at times of need for adequate services for people with ID and mental health problems has been criticized (Dodd & McGinnity, 2003; Sovner & Hurley, 1981). Public services generally promote the use of cognitive behaviour therapy (CBT) due to its increasing evidence base and short-term application at least with patients without ID and mental health problems (Roth & Fonagy, 2006). Whilst psychological services providing intervention to people with ID in the UK suggest the application of CBT over other therapeutic approaches

(Nagal & Leiper, 2005), empirically grounded behavioural approaches such as ABA may be more widely used at the expense of patient explorations of cognitions and emotions which could offer a path towards self-awareness and internalizing adaptive self-management.

Outcome research carried out in the general population often considered presence of ID as an exclusion criterion for trials (Willner, 2007). Certainly there is a lack of randomized control trials (Oliver et al., 2002); however, several case study series and an increasing number of controlled trials have shown improvements on standardized outcome measures relating to mental health problems such as offending behaviour (Willner, Jones, Tams, & Green, 2002), anxiety (Lindsay, Neilson, & Lawrenson, 1997) or psychosis (Hatton, 2002). At present, researchers from University College London are planning an RCT of individual CBT with people with ID vs. treatment as usual for presentations of depression and anxiety in adults with mild ID (Hassiotis, Serfaty, et al., 2011). Meta-analytic outcomes have shown that 13 % of all studies reviewed utilized a CBT treatment, whilst effect size was suggested to be 3.08 (Prout & Nowak-Drabik, 2003). Nevertheless, most CBT treatment studies appear to focus on behavioural rather than cognitive aspects to treatment (Willner, 2005).

Besides the paucity of studies, the variability of methodologies, intervention foci and therapy adaptation techniques challenge empirical research, true application of CBT and weaken the evidence base. A meta-analysis of this evidence (Prout & Nowak-Drabik, 2003) has been criticized for purely providing evidence for the behavioural aspect of CBT as a cognitive component was not included in most interventions (Sturmey, 2004). However, several recent studies using versions of Novaco and Taylor's (2005) modified anger management programme are emerging. This programme is devised specifically for adults with ID and includes cognitive components in addition to behavioural techniques. As yet, however, there does not seem to be consensus on the utility of this type of therapy and its success for people with ID. Isolating and investigating the cognitive processes within CBT (Nezu, Nezu, Rotherburg, DelliCarpini, & Groarg, 1995) and assessing these competencies individually may give more information on the effectiveness and aspects of CBT that might be helpful for those with intellectual disabilities.

Assessments of cognitive deficits and 'reality' of distortions may hence be suggested to encompass the psychosocial mapping of the client's distress (Alim, in press). Here, true motivation to engage in therapy as well as working collaboratively to change current levels of distress need to be gauged following a general paucity of self-referrals over carer referrals (Caine & Hatton, 1998). Moreover, research suggested that successful candidates for CBT require receptive language comprehension levels of 4 years 5 months (Reed & Clements, 1989). Dagnan and Chadwick (1997) as well as Joyce, Globe, and Moody (2006) suggested that the identification of emotions is vital for successful partaking in CBT. Whilst Dagnan and Chadwick (1997) focused on participants identifying 5 emotions (happy, sad, frightened/scared, anxious/worried and angry), Joyce et al. (2006) presented participants with 12 emotions (happy, sad, angry, afraid, bored, hurt, excited, disgusted, sneaky, surprised, thinking and interested). It was suggested that the original five-emotions presentation leads to higher success rates in participants. Reed and Clements (1989) further suggested an assessment of activating events as impacting on emotional responses to ascertain participants' ability to conceptualize that exposure to certain situations will lead to particular emotional responses. Quakley, Reynolds, and Coker (2004) further developed a task supporting an assessment of participant abilities to distinguish between thoughts, feelings and behaviours using hypothetical stories depicting all three elements. Whilst Quakley et al. (2004) used visual cues (in the form of three 'postboxes' in which the participant posts the 'thought', 'feeling' and 'behaviour' sentences of the scenarios), Sams, Collins, and Reynolds (2006) suggested that visual cues did not have any effect on task success. Finally Dagnan, Chadwick, and Proudlove (2000) created a task to assess participant's ability to mediate cognitions. Here scenarios are presented to the participant

including evaluative beliefs (core beliefs) and participants are then asked what emotion is likely to be the outcome (happy or sad). Half the evaluative beliefs are congruent with the situation and half incongruent. A second stage of the task gives the scenario and the emotion and asks the participant to choose which is the most likely evaluative belief (e.g. 'I am likeable'/'I'm not likeable'; 'I'm good at things'/'I'm bad at things'), again presenting a mixture of congruent and incongruent beliefs.

Use of a comprehensive assessment battery guided by the above principles and sound methodological research on the effectiveness of CBT for people with ID is yet required to ascertain whether this approach is useful for this client group. Suggestions about treatment eligibility for people with ID as depending on pre-therapy assessment success may predict the rise or fall of this therapeutic intervention for people with ID (Jahoda, Dagnan, Stenfert Kroese, Pert, & Trower, 2009; Oathamshaw & Haddock, 2007).

Conclusion

It is important to note that clinical presentations are heterogeneous and require individualized approaches both to information gathering, formulation and treatment. Once clinical diagnosis has been reached, the multidisciplinary assessment process clarifies the biopsychosocial underpinnings to the disorder, patients' functioning and interpretations of the world.

Organic conditions are various and multifold. Genetic conditions such as Williams syndrome or Prader–Willi syndrome mark brain development and impact heavily on clinical aetiologies. This will lead to marked discrepancies in adaptive and psychosocial functioning. Similarly, brain insults (infective or traumatic) at the developmental stage hinder human development and predispose certain mental disorders. Physical disabilities such as communication and sensory impairments further complicate the clinical picture and will need consideration at the assessment stage. Neuroplasticity (the shaping of cognitive structures through learning) will be impacted upon by internal and external factors. For example, parental facial reactions and social signals (Trevarthen & Aitken, 2001) as well as neurophysiological effects of stress (e.g. high cortisol) play a key role in infant brain maturation and it may be helpful to assess early life stressors and traumata. In adulthood, changes to brain functioning due to neurodegenerative conditions such as dementia, iatrogenic causes (unwanted medication side effects) as well as environmentally stimulated factors (e.g. substance misuse) will need to be considered in terms of significant impacts on cognitive and psychological functioning.

From a psychological perspective, relationships shape cognitive competencies such as metacognitive abilities and theory of mind (Heyes, 2003). Moreover, social isolation, quality of adult relationships and unhelpful environments hindering the development of adult relationships between people with ID (e.g. sexual relationships) disable psychological maturation, in turn leading to emotional and mental health problems obstructing psychological maturation. Unpicking these self–other schema and self-identities that will shape beliefs, attitudes and metacognitions can be vital in clinical assessment and formulation building. The impact of significant life events and acute psychological trauma will require thorough investigation irrespective of the person's level of intellectual functioning.

Finally, social ecologies further impact on health and may be related to socio-economic status, education and role identities (as relating to gender, disability, etc.). Issues of dependency and need for ongoing care and support are likely to impact on adult development. Organic factors as well as co-morbidly disabling social environments, paucity of self-development opportunities and breakdown of mental health may lead poor adaptive skill development. Lack of occupational opportunities for people with ID has long been noted to further exacerbate deterioration of mental health. The impact of social isolation and disdain on mental health and challenging behaviour marginalizes this patient population. Experiences of hate crimes and vulnerabilities will need to be considered during the assessment process. Social difficulties and challenging behaviour impact upon the expression of negative emotions among

staff in residential settings lead to high staff turnover, and it has been established that this exacerbates challenging behaviour (Langdon, Yágűez, & Kuipers, 2007). These social factors to formulation building offer further explanations to the shaping of identities and ways of social relating (Gillmore, 1990) which are impacted upon by cultural factors (Cohen, 2001).

The above picture is not an exhaustive amalgamation of biopsychosocial origins to mental health difficulties, and often clinicians face the dilemma of risk management at the expense of resolving underlying problems.

Following psychological assessments, clinicians will attempt to conceptualize a hypothetical formulation based on the person's core schemata as outlined earlier in this chapter. Its validity will be scrutinized throughout the therapeutic encounter.

This chapter has aimed to provide a comprehensive account of contemporary issues towards the assessment of mental health in individuals with ID. Nevertheless, the evidence base with regard to assessment and treatment of this patient population is only emerging so individualized assessment protocols based on existing evidence will best meet the needs of this heterogeneous and vulnerable patient population.

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Integrating Assessment Instruments within the Diagnostic Process

4

Steve Moss and Anne Desnoyers Hurley

Introduction

Traditionally, there has been a clear split in clinical practice between those who use structured or standardised instruments and those who do not. Nurses have a long tradition of completing a wide range of assessments and questionnaires covering a wide range of data such as mood, behaviour, symptoms of mental illness and aspects of physical health. Part of the psychologist's expertise is in using complex standardised assessments of overall intellectual functioning and specific cognitive areas. In terms of the core business of making assessments of mental health, whether they lead primarily to a diagnosis, or to a wider case formulation, most assessments made by clinicians are probably unstructured. That is to say, the clinician collects the information guided primarily by expertise and by the perceived requirements of the case.

The main aim of this chapter is to consider the potential benefits (and pitfalls) of using structured assessments in clinical practice and to give some guidance on how to choose them.

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The Significance of Instrumentation in Assessment

In the general population it has long been recognised that diagnosis can be an unreliable process. Bias can result from information about the reason for referral (Aboraya, Rankin, France, El-missiry, & John, 2006; Grove, Andreasen, McDonald-Scott, Keller, & Shapiro, 1981) or from the opinion of another clinician (Termelin, 1968), and there can be very significant differences between one country and another (e.g. Mackin et al., 2006).

Why is it that clinicians disagree to such a large extent? The answer may lie partly in the fact that there is no universal agreement about the collection of information or in the process by which diagnoses and case formulations are made. There can be huge variations in the type and quality of information obtained before making a diagnosis. Some teams or clinicians may focus mainly on current mental state, some are concerned more with the personality as a whole, while others concentrate on the situation in which the symptoms developed. There may be a predominant focus on a psychiatric approach to assessment, or the focus may be behavioural or psychodynamic. The relative weight given to history vs. present state can also alter the conclusions dramatically.

With all these variables to consider, it is clear that, even in the general population, it is desirable to have systematic approaches to the collection of clinical information and the drawing

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of conclusions from that information. The use of appropriate instrumentation offers immense benefits in this respect, but it is also clear there is a strong case for structuring the whole assessment process itself, to ensure all relevant avenues are adequately investigated.

Use of Instrumentation for Assessment of People with Intellectual Disability

People with Intellectual Disability (ID) present particular challenges to the assessment of mental health problems, these challenges having a fundamental role in determining where instruments should be used and what their characteristics need to be. First and foremost among these challenges is the problem of communication. Most people with ID, even those with a mild or borderline level, have problems with receptive and expressive language. Even if speech appears proficient, it is likely they will have problems with abstract concepts such as "feeling anxious" or "depressed", expressions that we would comprehend and use without a moment's hesitation. Time focus, and the ability to aggregate experience over a period of time to answer questions about the significance of symptoms, is also difficult. These limitations fundamentally change the nature of face-to-face interviewing. Assuming the person *can* be interviewed, any structured clinical interview will need to be carefully crafted to maximise the use of available language ability, and it will need to be coupled with appropriate training of users. Help with formulating ways of asking questions of people with limited language will clearly be of benefit.

These problems of communication lead to a second major issue, namely, that assessment of people with ID typically places a major emphasis on what *other* people say, rather than the major source of information being the patient's own report, as it is in people without ID. Since reports from informants are so important, there is a clear role for instruments that can ensure this information is of the best possible quality.

The Various Domains of Assessment

We only have to reflect on ourselves and our life courses to realise how precious and complex is our good mental health. Very often it is the confluence of many factors playing out over the person's life that have brought them to the point of showing significant mental health problems. There is usually a complex interaction between the physical organism (brain development, neurology, etc.), how that person has developed through interaction with parents and the wider world and the current environment in which they find themselves. Such complexity cannot be understood by a single model of assessment. For instance, the person may meet the criteria for a psychiatric disorder, but is the disorder a driving cause for other problems, or is it the *result* of other problems? Understanding the key elements of the case, and how they relate together, requires the use of a variety of perspectives to create diagnostic/formulation hypotheses of why the person is showing these problems at this particular time. Friedlander and Moss (2008) have illustrated this with a case example, showing a case formulation in relation to four fundamental assessment frameworks.

The Particular Issues Relating to Psychiatric Instruments

In terms of the population with ID, it is the psychiatric domain that causes the most significant problems. This is because the other domains of assessment are concerned primarily with information that does not rely on the person's own report, e.g. behaviours and their antecedents, factual details such as material circumstances and interviews to determine the dynamics of the person's family and wider world. The psychiatric perspective, however, is uniquely dependent on the person's own report of how they *feel*. Although we can to some extent observe and make inferences, nothing can replace the unique perspective of the person themselves. We can probably make reasonably accurate estimates about someone's low mood, but

does that mean they feel hopeless for the future? Nobody can really know except the person themselves. The more abstract the symptom, the more risky it is to make assumptions.

For the above reasons, the majority of this chapter is devoted to a consideration of instruments that, at least partly, relate to the psychiatric domain. We will shortly present some examples of assessments that we will subsequently compare in relation to a number of pivotal dimensions. Before doing so, however, it is appropriate to make a few initial comments about psychometric issues.

Psychometric Issues

Psychometrics is the mathematical analysis of psychological measurement. It is part of the wider field of statistical techniques concerned with measurements that *estimate* the truth, rather than providing a definitive answer.

Reliability is a measure of "dependability" or "consistency" of ratings for a symptom or behaviour. For example, reliability of ratings is calculated between raters, across raters and either at one time or multiple testing times. When instruments are not reliable, the ratings will differ significantly when the same individual is being assessed or when somebody else is doing the assessment. Thus, the reliability of any instrument is the first and most important measure to understand. Good reliability scores do not insure validity of what the test measures. However, if it is unreliable, it can never be valid.

Validity can be understood as the extent to which a test measures what it aims to evaluate. *Content validity* refers to the extent to which the test thoroughly evaluates the area of interest as determine by all studies on the instrument. *Construct validity* assesses how well the instrument measures the theoretical area it claims to measure and is studied in a number of ways. *Construct convergent validity* is often demonstrated by comparing results of a new instrument with another similar established instrument. For example, a new test measuring depression may be compared to the Beck Depression Inventory. *Criterion validity* refers to the relationship between the test and an independent external criterion. For example, does a total score on a test of psychopathology identify people receiving mental health services in a large population?

Although psychometric analysis is concerned with making estimates of what a gold-standard assessment would conclude, it is important to point out at this stage that instruments for mental health assessment are not only about estimating mental status in this sense. Many of the available instruments are designed to help clinicians raise the quality of the gold-standard assessment itself. These two axes, improving *estimates* of mental health and improving *full assessments* of individuals, are central to determining the choice of instruments.

Broad-Spectrum Psychiatric Assessments

We have not attempted comprehensively to review all the available assessments, but rather to give some of the notable examples. The instruments are presented first. Overall, our aim is to give some guidelines to help guide the potential choice of instruments in various situations. Instruments are presented in order of their publication date.

Reiss Screen for Maladaptive Behavior

The *Reiss Screen for Maladaptive Behavior* (RMB) (Reiss, 1988) or the "Reiss Screen" was one of the first instruments to assess psychopathology in people with ID. The screen is an "informant rating scale" so that a person directly involved with the individual completes the form independently and a professional scores and interprets it. The Reiss Screen contains 38 items rated ideally by two individuals who know the person well, and the scores are averaged. Items are rated on a 3-point scale for severity. The items result in eight subscales originally derived from factor analysis on a population, and they

are Aggressive, Autism, Psychosis, Paranoia, Depression/Behavioural Signs, Depression/Physical Signs, Dependent Personality Disorder and Avoidant Personality Disorder. The subscales roughly correspond to several DSM-IV categories. Critical or cut-off scores for the subscales are provided as well as a 26-item total score, and additional individual items of interest are noted but not included in scoring. These individual items of interest are drug abuse, overactive, self-injury, sexual problem, suicidal and stealing. The screen was designed to identify presence of a "mental health problem" in individuals with mild to profound ID. It has been translated into Dutch and Swedish. There are a number of reliability and validity studies finding moderate psychometric results with some disagreement on the factor structure (Gustafsson & Sonnander, 2002; Havercamp & Reiss, 1997; Sturmey & Bertman, 1994; Sturmey, Jamieson, Burcham, Shaw, & Bertman, 1996; Walsh & Shenouda, 1999). The Reiss Screen has been used in a number of clinical studies as well (Demark, Feldman, Holden, & MacLean, 2003; Kishore, Nizamie, Nizamie, & Jahan, 2004; Lunsky et al., 2010). This instrument is easy for an informant to complete independently. Scoring is straightforward. The Reiss Screen has most value for adults with mild to moderate ID.

Diagnostic Assessment for the Severely Handicapped-II

The *Diagnostic Assessment for the Severely Handicapped-II* (DASH-II) (Matson, 1995) was a pioneer in its aim to evaluate people with severe and profound ID. It is an "informant interview" scale (a trained professional interviews a person who knows the individual well). The DASH-II was developed on a normative population in a large facility. The instrument provides information on symptoms and behaviours that are related to DSM-III psychopathology, revised from an original version in 1991 (Matson, Coe, Gardner, & Sovner, 1991; Matson, Gardner, Coe, & Sovner, 1991). An informant rates 84 different items using a 3-point scale for frequency, duration and severity. There are 13 subscales and 7 that have direct relevance to psychiatric diagnoses: Anxiety, Depression, Mania, Schizophrenia, Stereotypies/Tics, Organic Syndromes and Impulse Control Disorder/Miscellaneous. Additional subscales are Self-Injurious Behaviour, Elimination Disorders, Eating Disorders, Sleep Disorders and Sexual Disorders. The DASH-II has been translated into Italian, Norwegian and Spanish. The DASH-II is considered to have good inter-rater and test-retest reliability with variability among subscales, and validity studies have been positive (Matson, Coe, et al., 1991; Matson, Gardner, et al., 1991; Paclawskyj, Matson, Bamburg, & Baglio, 1997; Sturmey, Matson, & Lott, 2004). The DASH-II has been useful in clinical areas such as schizophrenia, depression and mania (Bamburg, Cherry, Matson, & Penn, 2001; Matson, Rush, & Hamilton, 1999; Matson & Smiroldo, 1997). The variation in number of items for each area must be considered for interpretation of the scale (DSM-related subscales range from 6 to 17 items). Because it is used for people with severe and profound ID, any assessment of this population is challenging due to limitations in communication ability (Ross & Oliver, 2003a). The DASH-II, however, can provide important information for the clinician or researcher.

Developmental Behaviour Checklist

The *Developmental Behaviour Checklist* (DBC) (Einfeld & Tongue, 1995, 2002) is a widely used informant questionnaire for children and adolescents with ID. The DBC-P (parent or career version) has 96 items distributed on five subscales. The items were derived from case files of children with emotional and behavioural difficulties. Each behavioural description (e.g. "Distressed about being alone") is scored as follows: "not true as far as you know", "somewhat or sometimes true" and "very true or often true". The DBC has six subscales derived from factor analysis on a normative

population and they are Disruptive Behaviour, Self-Absorbed Behaviour, Communication Disturbance, Anxiety, Autistic-Relating Behaviour and Antisocial Behaviour. The DBC has strong psychometric properties (Bontempo et al., 2008; Hastings, Brown, Mount, & Cormack, 2001). Because it was developed on a population-based cohort and normative data is available, it can be used in epidemiological studies, research and clinical practice easily. The DBC-P (parent) is widely used and has been translated into 21 languages, and the DBC-T (teacher) has been translated into four languages. It is well designed and easy for informants to complete, as well as having clear scoring and interpretation guidelines. A bibliography is available from the website (Einfeld, 2007).

Nisonger Child Behavior Rating Form

The *Nisonger Child Behavior Rating Form* (NCBRF) (Aman, Tassé, Rojahn, & Hammer, 1996) is an instrument for assessing child and adolescent behaviour modelled on the *Child Behavior Checklist* (Achenbach & Edelbrock, 1983). It is an informant scale with a parent and teacher version. A unique feature of the NCBRF is a focus on positive social behaviours in a separate scale. It is therefore organised into two major sections with subscales: *Positive Social* (subscales are Compliant/Calm and Adaptive/Social); *Problem Behaviour* (subscales are Conduct Problem, Insecure-Anxious, Hyperactive, Self-Injury Stereotypic, Self-Isolated/Ritualistic and Overly Sensitive). The *Positive Social* is rated on a 4-point scale for frequency and *Problem Behaviour* is rated on a 4-point scale for frequency/severity. Psychometric properties are generally very good including internal consistency, inter-rater agreement and concurrent construct validity (Aman et al., 1996; Norris & Lecavalier, 2011; Lecavalier, Aman, Hammer, Stoica, & Matthews, 2004; Tassé, Aman, Hammer, & Rojahn, 1996). The NCBRF has proved valuable as a clinical tool (Croonenberghs et al., 2005; Tassé & Lecavalier, 2000). The NCBRF is well designed and easy for an informant to complete.

The PAS-ADD Assessments

The *Psychiatric Assessment Schedules for Adults with Developmental Disabilities* (*PAS-ADD*) have been in continuous development since the mid 1980s, a period that has seen the blossoming of the field of mental health of people with ID (ID). The work has had three major aims:

- Communicating better with people who have ID
- Identifying potential cases for assessment
- Involving more staff in the assessment process

The PAS-ADD 10 (Moss, Goldberg, et al., 1996; Moss et al., 1993) was designed to maximise the possibility of conducting an interview with the person themselves but was also to be conducted separately with a key informant. The *PAS-ADD* clinical interview (Moss & Friedlander, 2011) has now superceded the PAS-ADD 10. It is the most comprehensive of the assessments and is designed to produce full diagnoses under both ICD 10 and DSM-IV.

The validity of PAS-ADD 10 in relation to the clinical opinion of referring psychiatrists was reported in Moss, Prosser, and Goldberg (1996) and Moss et al. (1997). Inter-rater reliability of the ICD 10 version gave a mean Kappa of 0.65 for individual item codes and Kappa 0.7 for agreement on index of definition (clinical significance of the symptoms) (Costello, Moss, Prosser, & Hatton, 1997). The relationships between respondent (patient) and informant reports of symptoms, and the implications of deriving diagnoses solely from informant interviews, are discussed in Moss, Prosser, Ibbotson, and Goldberg (1996). The issues of using care staff as informants were discussed in Moss and Patel (1993).

The PAS-ADD Checklist

The PAS-ADD Checklist (Moss, 2002a) was designed specifically for improving case recognition. It is a 25-item questionnaire, couched in everyday language, designed for use primarily by care staff and families—the people who have the most immediate perception of changes in the behaviour of the people for whom they care.

Factor analysis of the checklist completed on a community sample of 201 individuals yielded eight factors, of which seven were readily interpretable in diagnostic terms. Internal consistency of the scales was generally acceptable. Inter-rater reliability in terms of case identification, the main purpose of the checklist, was quite good, 83 % of the decision being in agreement. Validity in relation to clinical opinion was also satisfactory, case detection rising appropriately with the clinically judged severity of disorder (Moss et al., 1998). Subsequent independent studies have further investigated the checklist's psychometric properties (Sturmey, Newton, Cowley, Bouras, & Holt, 2005) and established norms for an adult sample (Taylor, Hatton, Dixon, & Douglas, 2004).

Mini PAS-ADD

The general aim of the Mini PAS-ADD (Moss, 2002b) is to provide an instrument that can be used by a wide range of professionals, capitalising on their expertise and knowledge. The assessment is made accessible to this wide range of staff by providing a clear yet flexible structure and by the use of a detailed glossary of symptoms to guide coding. The Mini PAS-ADD provides in-depth information on Axis I psychiatric disorders, either by interviewing an informant or by collecting together knowledge already possessed. There is also a screen for autism spectrum disorders. This information can then be used in the subsequent formulation. Psychometric properties of the Mini PAS-ADD are reported in Prosser et al. (1998).

The ChA-PAS

The *Child and Adolescent Psychiatric Assessment Schedule* (*ChA-PAS*) (Moss, Friedlander, Lee, Holly, & Leech, 2007) is for the assessment of mental health problems in children and adolescents with learning disabilities. The ChA-PAS includes all the disorders covered by the Mini PAS-ADD but has also been extended to cover two major behavioural disorders, ADHD and Conduct Disorder. The ChA-PAS can be used to interview the children themselves if they are sufficiently verbally able.

Strengths of the PAS-ADD System

The PAS-ADD assessments have been used in a wide variety of research studies, but the primary aim of their development has been to aid the process of case detection and assessment in clinical settings. Aiding the process of collaboration between professionals has been a central theme. The Mini PAS-ADD and the ChA-PAS enable people who are not necessarily psychologists or psychiatrists to conduct clinical interviews to provide in-depth symptom information that can be used both in case formulation and for monitoring the impact of interventions. The PAS-ADD Clinical Interview provides a top-level assessment that is aimed primarily at clinicians, but does not preclude others being trained in its use. The whole system adheres closely to ICD 10 and DSM-IV(TR), so the information maps easily onto existing clinical practice. The Mini PAS-ADD has been translated into Dutch and German, with planned translations into Hungarian and Norwegian.

Instruments Focusing on Behaviour or Specific Symptom Areas

Aberrant Behavior Checklist

The *Aberrant Behavior Checklist* (*ABC*) (Aman & Singh, 1986, 1994) is a widely used instrument and may be considered to have set a standard for instrumentation concerning challenging behaviour and ID. It is an informant rating scale and assesses specific behavioural problems in people with all levels of ID from ages 6 to 54. The ABC contains 58 items distributed on five subscales and they are Irritability, Lethargy, Stereotypy, Hyperactivity–Noncompliance and Inappropriate Speech. The informant rates the description of a behaviour on a 4-point scale of severity. The original work was developed on a normative population in an institution (1986) and was updated for those living in community residences (1994); however, use of the normative data today is rare. The ABC is simple to complete and score. The outstanding reliability and validity of the ABC has been extensively demonstrated and it is often used to establish construct validity with other instruments (Aman, Burrow, &

Wolford, 1995; Aman, Singh, Stewart, & Field, 1985a, 1985b). The number of items in the scales is quite uneven, ranging from 4 to 16 and selective use of subscales is common. The ABC has been used in hundreds of studies and is translated into 25 languages. A bibliography is available from the website (Aman, 2010).

Behavior Problems Inventory (BPI-01) and Short Form (BPI-S)

The Behavior Problems Inventory (BPI-01)

The *Behavior Problems Inventory* (BPI-01) (Rojahn, Matson, Lott, Esbensen, & Smalls, 2001) is a 52-item informant interview rating scale for use with all levels of ID in children and adults. The BPI-1 was empirically developed and has four subscales and they are Challenging Behaviour, Self-Injurious Behaviour, Stereotypic Behaviour and Aggressive/ Destructive Behaviour. Each subscale has a general description of the area and a short description of each item, and this improves the information and judgments of informants. The items are rated on a 4-point scale for frequency and a 3-point scale for severity. Inter-rater reliability was low to very good, internal consistency was good for full scale and low to moderate for subscales, and validity studies are generally positive (González et al., 2009; Rojahn, Aman, Matson, & Mayville, 2003; Rojahn et al., 2010; Sturmey, Burcham, & Perkins, 1995; Sturmey, Fink, & Sevin, 1993; Sturmey, Sevin, & Williams, 1995). The BPI has been used primarily with people who have severe to profound ID with good results for clinical assessment and drug therapy evaluations (Hattier, Matson, Belva, & Horovitz, 2011; Snyder et al., 2002). The BPI-1 is a good choice when questions are related to the subscale areas.

The Behavior Problems Inventory-Short Form

The Behavior Problems Inventory-Short Form (*BPI-S*) has recently been developed to provide a more useful shorter version (Rojahn, Rowe, Hastings, et al., 2012; Rojahn, Rowe, Sharber, et al., 2012). This form has 19 fewer items. Sample verbal descriptions of severity were added to the Self-Injurious Behaviour and Aggressive/Destructive Behaviour subscales in an effort to enhance the objectivity of the ratings. Psychometric data showed very good results with the exception of the Self-Injurious Behaviour subscale, and the authors suggested this might be due to complexities in the construct of self-injury. This short form is easy to use and has a clear and concise presentation on one page.

Self-Report Depression Questionnaire

Reynolds and Baker (1988a, 1988b) developed the *Self-Report Depression Questionnaire* (*SRDQ*) based on *DSM-III-R* symptoms of major depressive disorder. The questionnaire is verbally administered to the individual. It is a 32-item instrument designed specifically for adolescents and adults with mild ID. The individual judges the frequency of each item as occurring: "almost never", "sometimes" or "most of the time". A 2-part pretest determines if the individual is capable of responding reliably to orally presented questions. The last item displays three faces asking the person to mark which face describes, "How you have been feeling?" The SRDQ has good psychometric properties (Benavidez & Matson, 1993; Esbensen, Seltzer, Greenberg, & Benson, 2005); however, Rojahn, Warren, and Ohringer (1994) found little agreement with the Reiss Screen and Diagnostic Interview of Children and Adolescents. Other studies have found the SRDQ to be useful in work on depression (Esbensen & Benson, 2005, 2006; Glenn, Bihm, & Lammers, 2003). The manual offers clear guidance for successful administration and interpretation.

Mood, Interest and Pleasure Questionnaire

Ross and Oliver (2002, 2003b) developed the *Mood, Interest and Pleasure Questionnaire* (*MIPQ*) for use with adults who have severe and

profound ID. The MIPQ was developed based on DSM-IV criteria for a major depressive episode and interpreted for ID. It is an informant rating scale with two subscales: Mood and Interest and Pleasure. Items are rated on a 5-point scale after being observed in the last 2 weeks. The questionnaire uses full sentences. For example, the first item is "In the last two weeks, did this client seem…. sad all of the time, sad most of the time, sad about half the time, sad some of the time, never sad". This approach is an excellent way to improve informant information. The scoring is easy and interpretation clear. Initial inter-rater reliability, test-retest reliability and internal consistency were good, and validity was demonstrated with the ABC (Ross & Oliver, 2003b). A recent Dutch translation was developed and inter-rater and test-retest reliability were good, and construct validity was consistent with the ABC; however, factor analysis did not confirm the 2-factor structure (Petry, Kuppens, Vos, & Maes, 2010). The MIPQ has been used in research on subjective well-being and challenging behaviour (Burbridge et al., 2010; Hayes, McGuire, O'Neill, Oliver, & Morrison, 2011; Oliver, Berg, Moss, Arron, & Burbridge, 2011; Vos, de Cock, Petry, Van Den Noortgate, & Maes, 2010).

Fear Survey for Adults with Mental Retardation

Ramirez and Lukenbill (2007) developed an adult fear survey schedule based on an earlier version for children (*Fear Survey for Children With and Without Mental Retardation* [*FSCMR*]) (Ramirez & Kratochwill, 1990, 1997). The Fear Survey for Adults with Mental Retardation (FSAMR) is a self-report instrument administered verbally to an individual with ID. It has 73 fear items, 6 items to test reliability of the person's response and 6 other items to assess acquiescent response set. The rating scale has the following subscales: Physical Assaults, Animals, Illness, Injury, Changes in Routine/Familiar Environment, Socio-emotional, Natural/Supernatural, People Nonspecific Reason and Idiosyncratic. The methodology is clear and addresses achieving rapport, eliciting self-report and establishing a preferred fear "word" (e.g. "afraid" or "scared"). The authors found good internal consistency reliability and moderate concurrent validity. Anxiety and fear are difficult to assess in ID (Hermans, Femke, van der Pas, & Evenhuis, 2011). A self-report instrument provides valuable information for those individuals who can respond to the verbal format.

Institute for Basic Research Overt Aggression Scale (IBS-OAS)

The *Overt Aggression Scale* (*OAS*) was developed as a way to assess aggression in psychiatric inpatients by improving on incident reports completed by informants (Silver & Yudofsky, 1991; Yudofsky, Silver, Jackson, Endicott, & Williams, 1986). The MOAS has four categories of aggression with items reflecting severity: *verbal aggression*, *physical aggression against objects*, *auto-aggression* (*physical aggression against self*) and *physical aggression against others*. It has been widely used in a modified form, the Modified Overt Aggression Scale or MOAS (Sorgi, Ratey, Knoedler, Markert, & Reichman, 1991). This revision used a 1-week retrospective evaluation of the patient's behaviour and, in addition, each item was scored for frequency. It is an easy instrument to use, and reliability and validity have always been considered to be quite good. A psychometric study of the MOAS used with ID found very good inter-rater reliability for verbal and physical aggression and good to moderate results for the other two subscales (Oliver, Crawford, Rao, Reece, & Tyrer, 2007).

Recently, a modified OAS for ID was developed at the Institute for Basic Research on Developmental Disabilities (IBR-MOAS) (Cohen et al., 2010). The scale contains demography and clinical data, a new category "Verbal aggression toward self" was added, and some wording was changed to be more consistent with ID assessment. In addition, a 4-point rating for frequency during the past year was developed. Good reliability and validity were achieved. A subsequent study used the IBR-MOAS and found valuable information regarding the relationship between

aggression and psychiatric illness (Tsiouris, Kim, Brown, & Cohen, 2011). This new instrument holds much promise for the assessment of aggression.

Psychiatric Instrumentation: Fundamental Considerations

Let us start by looking at the ways in which the various assessments differ. The following are some of the fundamental dimensions:
What is the instrument's theoretical approach?
What spectrum does it cover?
What depth of information does it offer?
Who makes the ratings, and how are they made?
What are the skill and training requirements?
How long does it take to administer?
What are its psychometric properties?

Theoretical Approaches to Assessment

Assessments derived from psychometric principles make no assumptions about relationships between the items, but use statistical analysis to derive factors. Their derivation typically starts from large number of items and use factor analysis to generate item groups. Examples of this are the ABC (Aman & Singh, 1986) and the DBC (Einfeld & Tongue, 1995). In comparison, clinical assessments relate to the fundamental pattern-recognition process of psychiatric assessment and basically seek to determine whether signs and symptoms match one of the diagnostic constellations, e.g. panic disorder and schizophrenia. Such assessments are usually based on DSM-IV(TR) (American Psychiatric Association, 2000) or ICD 10 (World Health Organisation, 1993) classification systems.

An important difference between these approaches is that observation-type instruments deal only with signs (the things that can be observed), while psychiatric assessment is normally concerned both with signs and symptoms (what a patient experiences). A further consideration is that the scoring categories of factor-based assessments sometimes bear little resemblance to the diagnoses made in clinical practice.

Include Challenging Behaviours as well as Psychiatric Symptoms?

The treatment and management of challenging behaviours is a fundamental concern for mental health service providers working with individuals who have ID and is a dimension that probably affects 10 % of all service users to a significant degree (Emerson et al., 1997). It has also been reported as one of the most common reason for referral to a psychiatrist (Hurley, 2008; Hurley, Folstein, & Lam, 2003; Jacobson, 1998). Assessments such as the Reiss Screen (Reiss, 1988) use a mixture of psychiatric and behavioural items, while the DC-LD Diagnostic Criteria for Learning Disability (Royal College of Psychiatrists, 2001) discusses the use of symptoms "equivalents", so that, for instance, an increase in challenging behaviour may be viewed as a symptom of depression.

The problem of using problem behaviours in this way is that their provenance is often very unclear. In the general population, someone who starts cutting him/herself *may* be depressed, but it would be unsafe to use this information if key diagnostic indicators were not present. Someone with severe ID whose head banging increases *may* be more anxious, or more depressed, or more stressed, but again it would be unsafe to use this diagnostically in the absence of clearer evidence.

Spectrum Covered by the Instrument

The main consideration to mention in this respect is that no assessment can be fully encompassing. Indeed, the danger of attempting to devise instruments to cover all eventualities is that they can become too cumbersome. A good approach is to use an assessment covering more common disorders and to invoke specific instruments to cover other problems that arise. For wide spectrum assessments, it is worth looking, in this respect,

at the provisions for dealing with disorders that fall outside the scope of the instrument. Is there some way in which other disorders can at least be flagged up?

The Depth and Detail of the Information

Human beings are capable of making very subtle distinctions in what they observe, but much of this ability is very difficult to quantify. Structured assessments seek to reduce this unreliability by defining specific pieces of behaviour to observe and record. However, reliability does not tell us about depth and detail. An assessment can be highly reliable, but not tell us what we want to know.

One way in which detail can be increased is to increase the number of items, breaking larger units of observation into smaller ones. If many nuances of behaviour are recorded, the possibility of making fine distinctions is theoretically increased. An example of this is the DBC, which raises the level of detail by defining a large number of individual behaviours. In this respect, instruments like the DBC are very different from assessments based on DSM or ICD. Diagnostic constellations such as depression or panic disorder are defined by the presence or absence of a relatively small number of signs and symptoms. A general principle of psychiatric assessment is that it takes a poly-diagnostic approach, that is, that different manifestations can lead to the same symptom or disorder being identified. Rather than breaking down behaviours into very small parts, the definitions are fleshed out by the glossary information in the diagnostic manuals. One way of looking at these different approaches is in terms of the point at which detail is transformed into clinical significance. Generally speaking, detail is good (provided it is reliable), but at some point the assessments must take all the details and interpret them into a formulation. Instruments like the DBC have a "framework-free" approach, using statistical methods to interpret the detailed information. Instruments based on psychiatric classification work within an already existing framework, which usually require the assessor to make fewer responses, but place more emphasis on interpretation *before* the response is recorded. Hence there is often greater need for skills and/or training when using them.

Who Makes the Ratings, and How Are They Made?

The choice of instruments often relates to the complex trade-off that has to be made between skill level, the availability of resources and access to relevant knowledge. Typically, clinicians are the ones with the greatest expertise in psychopathology, but the people with ID themselves, and their parents, are the ones with the greatest knowledge of the individual case. Health and social service staff such as community nurses, social workers and speech therapists often have close knowledge of the case as well. Self-report obviously requires the least staff time, but most people with ID do not have the necessary skills to complete a self-report assessment. Even for members of the general population, self-report methods suffer from the same problems of reliability of any assessment not guided by clinical knowledge and training.

The Psychometric Properties of Instruments

This dimension has been left until last because the issues are not at all straightforward. Psychometric data are often cited to give grounds for using the assessment in a subsequent study. On closer examination, however, there are some fundamental issues that can easily be glossed over if these data are taken at face value.

Reliability

Problems in reliability can result from two sources: problems in instrument development or problems in the rater. Many instruments have acceptable reliability but not for all items or subscales. Raters may have different levels of training, experience, knowledge, or motivation to complete the scale. It is imperative to choose motivated raters who know the individual well.

For self-report measures, there are unique threats to reliability (and validity) when used for people with ID. Question content cannot be abstract, socially reflexive, or require comparisons or quantitative judgments (Finlay & Lyons, 2001). Attempts to overcome these threats to reliability include strategies such as reading the questions, simplifying the content, visual aids to facilitate judgments and utilising a practice section to determine the person's ability to answer the questions.

Instruments can improve reliability in important ways. The first way is to insure the questions are understood. For example, the Reiss Screen (1988) has a definition and example of each item. The PAS-ADD assessments contain clinical interviews, so their reliability is susceptible to fluctuation in the way people ask questions and the way they code symptoms. Thus, clinical glossaries give tightly defined symptom definitions and severity codings, in conjunction with appropriately worded interview questions. Secondly, users are asked to undertake a course of training where they learn to use the semi-structured interview and use the clinical glossary.

Generally speaking, it must be bore in mind that research studies can usually achieve good reliability in a small, tightly trained team of assessors. The problems arise when the measure gets out into the real world, used by people who are distant from those who originally designed it. In clinical settings it is worth bearing this in mind and perhaps checking that users are achieving and maintaining reliability in a sustained way.

Validity

Validity must be evaluated within the context of the purpose of the test and the population for whom it was intended. We must ask the question, "How valid is this test for the interpretation and decisions I will make?" Validity is a more complex concept than it seems. A test for schizophrenia may measure criterion concurrent validity by identifying patients diagnosed with schizophrenia against other patient groups. Does everyone agree, however, on the independent measure of these individuals or that every person actually has schizophrenia? There are many conflicting opinions about how cases should be formulated from these various pieces of information. For example, some professionals eschew the value of psychiatric diagnosis in favour of a model based wholly on social factors. Others see psychiatric diagnosis as a primary aim.

Sensitivity and Specificity

In the field of mental health, screening tools need to be evaluated in terms of their ability to predict "caseness", that is, whether a gold-standard assessment would conclude that the person has problems above a defined threshold of severity. One wants to maximise sensitivity (reduce false negatives), but not include too many false positives.

It is important to recognise the difference between a scoring threshold for a screening instrument and a threshold for diagnosis. In the former case, (assuming the screen has good psychometric properties), someone scoring below the threshold may be assumed not to have significant problems in the areas covered by the instrument. A *diagnostic* threshold, on the other hand, is different. Under DSM-IV(TR), for example, a diagnosis of depression requires a minimum of five symptoms to be present, out of a list of nine. This does not mean that people scoring less than 5 have no problems. In some cases a clinician may well decide that the person's best interests are served by giving them a diagnosis, even if they do not strictly meet the criteria. Instruments that are designed for diagnosis on DSM-IV(TR) or ICD 10, rather than being screening tests, similarly need clinical interpretation.

Sensitivity and Level of ID

Clearly, the greater the level of ID, the fewer symptoms can be meaningfully identified. In the case of depression, this is not such a problem, because most of the diagnostic criteria can be observed. Low mood, loss of interest, tiredness and exhaustion, changes in sleep and psychomotor movement can all be reliably reported by informants. In contrast, the core symptoms of psychosis, hallucinations and delusions are highly abstract and cannot be inferred from external behaviour with any degree of certainty. Moss, Prosser, and Goldberg (1996) found the only first-rank symptom which could be detected with

any frequency in their verbally competent sample was auditory hallucinations. Almost none of the respondents or informants were able to give an account of thought disorder or passivity delusions which was sufficiently unequivocal to fulfil the diagnostic criteria.

Guidelines for Selecting Instrumentation

The vast majority of clinical services probably have time and manpower issues and would ideally benefit from more resources. As a result, the possibility of improving the speed and cost of assessment is usually of interest. It would of course be ideal if assessments could be made faster with no detriment to quality and that quality could be improved without making the process more time consuming. This ideal may to some extent be realised through the use of well-chosen instrumentation, but there will inevitably be a trade-off that has to be made. This final section of the chapter offers some logical and practical points to help guide the choice of assessments. In terms of this trade-off, points 1 through 5 focus primarily on the quality of assessment, while then remaining points have more of a focus on strategic planning.

Enhancing the Contribution of the People with ID Themselves

One of greatest differences between assessments in people with ID compared to the general population is the huge emphasis placed on what other people say, rather than on what the individuals themselves say. The reasons for this are obvious, but there is always a danger of underestimating the potential contribution the person can actually make. Nothing can replace the patient's own verbal report, so instruments that are specifically designed to interview people with ID can be of great benefit. Patel, Goldberg, and Moss (1993), using the PAS-ADD interview with older people with ID, found that 49 % of subjects could complete the whole interview, with a further 13 % being able to answer selected core items from the interview.

Introducing Instruments That Enhance the Skills of Staff

Instruments for experienced clinicians should have the flexibility to map onto existing clinical expertise. For all staff, quality will rise if the assessment is tailored to the skills of the staff using it, while at the same time enabling enhancement of skills to be achieved, perhaps in terms of reliability, validity, sensitivity, objectivity, level of detail or adequacy of coverage of relevant symptoms. Assessments requiring training in their use are usually the ones that require clinical judgement on the part of the user and hence the ones more likely to enhance skills. Training can also be beneficial in bringing together staff from different disciplines, working together on cases.

Raising the Motivation of Staff Through Increasing Their Involvement in the Assessment Process

Instruments that seek to enhance assessment skills, or fully capitalise on existing skills, are more likely to help staff feel more involved in the assessment process and recognise the importance of their own contribution. It is imperative that staff see the point of completing instruments; otherwise, focus will be lost and the quality of the information will suffer. "Refresher" training can be very helpful in this respect.

Providing Information That Was Hitherto Not Collected or Was Not Collected in a Systematic Way

Most case formulations require information relating to a variety of aspects of the person's life. These can include family factors, medical and genetic issues, behaviour problems and attachment issues. Appropriate instruments can help

ensure that these various areas are adequately and reliably covered.

Selecting Instruments as Part of Formal Assessment Protocol

Although most of this chapter has been about the characteristics of individual instruments, it is also important to stress that the overall assessment process can benefit greatly from the adoption of a structured approach. The relative contributions of psychiatric, behavioural, ecological, psychodynamic and historical factors are often very difficult to determine; indeed, it is often difficult to determine whether, for instance, a psychiatric disorder is a driving cause for other problems in the person's life or the *result* of other factors. This can dramatically affect the formulation, and often there are several logical formulations that appear to fit the data. Crucially, therefore, it is essential to collect relevant information over a period of time, rather than taking a snapshot of a single period. It is only when the information is seen in the longer perspective that it becomes possible to hypothesise which are the driving courses and which are the results. Overall, it is suggested that a protocol is desirable, by which all potential cases are investigated over a period of weeks, requests for appropriate information being made during that period. An example of such a protocol, developed by the Frazer Valley Health Team (Vancouver), can be found in Moss and Lee (2001). A discussion of case formulation within multiple frameworks can be found in Friedlander and Moss (2008).

Getting Non-staff Members to Provide Structured Information

The community of people with ID, their families and other immediate carers, is a large and essentially "free" resource on which to draw for information. Indeed, initial case identification relies heavily on the reports of carers (unlike the general population where people generally take their own problems to a GP who is then responsible for recognising the problem). Most people with ID are unable to recognise their own mental health problems, carers are often uncertain about the significance of the behaviours they have seen, and many GPs do not have in-depth experience of working with members of this population. As a result, appropriate case detection checklists can be of great benefit.

Following referral, carers can also complete checklists designed for non-trained raters, while the most highly functioning people with ID may be able to complete self-report measures.

Using Less Qualified People to Collect Clinical Information

Expert clinical time from psychiatrists and psychologists is usually at a premium, both in terms of availability and in terms of cost, so the possibility of collecting some of this information by less qualified staff is clearly of interest. All instruments provide data on psychometric properties, and there are many instruments that can aid the process of information gathering. However, it is an old adage, but one can rarely get something for nothing. Checklists can be easy and quick to complete, but checklists suffer from poor reliability. There may be a good statistical relationship between results on a test and the conclusions of a gold-standard assessment, but in clinical settings one is dealing with individual patients, not populations. Assessments like the Mini PAS-ADD that enable less qualified staff to complete sophisticated assessments can provide much better information but are of course more time consuming.

Overall, the balance between time to assess, skill level and quality of information needs to be carefully thought out. In this respect, the adoption of a "filtering" system (see below) can greatly improve efficiency while also maintaining the quality of information.

Adopting a Filtering Approach to Assessment

At the early stages of case recognition and assessment, the most important concern is to not miss problems. Sensitivity of the instrument is

paramount at this stage. Once the person has been identified as a potential case, some basic decisions can be made about the types of information that need to be collected (which may be in various domains including psychiatric, behavioural, ecological). In the earlier stages of assessment, basic information may be collected using simpler instruments such as checklists and questionnaires. As the assessment becomes more targeted, appropriate in-depth instruments can then be selected. In this way, the more time-consuming instruments can be reserved only for those cases that need them, and clinicians' time can be maximally focused on interpretation and case formulation. As mentioned earlier, a formal assessment protocol can help in the management of these various components of assessment.

Selecting Instruments to Measure Clinical Outcomes

This topic has been placed in a separate section because it is useful to contrast the issues of monitoring progress in an individual patient, compared with the evaluation of a service. Both of these have broadly similar aims, but there are also important differences that should influence the choice of instruments.

Measuring Change in Individuals

Monitoring outcomes for individual patients needs the use of measures that are highly reliable; otherwise, variability inherent in the measure itself could be interpreted as genuine clinical change. Simple checklists are not so suitable in this respect, unless the staff have trained in the use of the assessment and have been shown to have high reliability between raters. Also, it is important to remember that, in relation to psychiatric symptoms, two people can have very different levels of severity yet warrant the same diagnosis. This means that instruments to monitor the outcome of treatment should measure not just the present/absence of symptoms, but also their severity. Someone who comes to the service with a lot of severe symptoms may improve as a result of intervention yet still meet diagnostic criteria for the disorder.

Measuring Outcomes in Service Populations

Measuring change in a population is more tolerant of unreliability in the instrument than monitoring individual patients, so the full spectrum of instruments is available. Once again, there needs to be a careful consideration of the type of information required, and the trade-off between brevity and completeness. It is probably better to target the measures rather than to try and collect a lot of information in the belief that it may be helpful. This only serves to make the task more onerous. In this respect, one of the most important considerations is the motivation of the person completing the measure. If the task is seen as being "dumped on them" by a remote manager, it is highly likely that completion will be sloppy and unreliable, and the resulting conclusions will be suspect. It is vital that staff are made to feel involved in the assessments they are asked to undertake, that they are making a valuable contribution to the mental health of their clients.

Conclusions and Directions for Future Research

This chapter has sought to highlight the various ways in which the use of structured assessments can be of benefit in clinical practice. These benefits are not only about improving reliability and validity. Structured assessments can make for a more consistent approach between health and social service professionals, enhance staff skills, increase the sense of involvement and contribution in the assessment process and help ensure that all the relevant information is collected. In terms of assessing treatment response, structured instruments can provide a more unbiased report of any change than unstructured clinical observations.

Much of what has been said applies equally to the general population. However, the problems of communication with people who have ID, and the consequent emphasis on informant reporting, add greatly to the complexity of assessment. Helping people with ID talk about their problems, and balancing this with reports from other people, can be aided by the use of appropriate structured assessments, provided that thought and preparation is given to their selection and implementation.

We have reviewed selected instruments that are best examples of instrumentation relating to the psychiatric domain, each of which has its particular strengths. If the person with ID can speak for him or herself, instruments that utilise self-report or interview can be excellent choices. At the same time, instruments focusing on informant reports are valuable for organising information and for reliably reporting observations of the individual.

One thing that all mental health professionals are aware of is that mental health problems are not just confined to the presentations of signs and symptoms. There are many other interacting factors that need to be taken into account when interpreting the symptom information, many of which have been mentioned in this chapter. Although this is well recognised in both clinical setting and research, there is still a temptation to accept that a structured assessment somehow "encapsulates" mental health problems and that the designer has put in all the hard work so the results can be taken at face value. This may be an acceptable assumption in a research project where, for example, the prevalence of mental health problems is being compared between two groups. At an individual clinical level, however, structured assessments can be of great benefit, but cannot replace the process of weighing and judging the available evidence by an expert clinician.

In terms of future developments, there is no doubt that our concepts of mental health and mental illness will continue to shift and change over time and that structured methods will be a central part of research and development within the field. It seems likely, however, that there will always be a gap between what we can measure and categorise and what we can sense but cannot put into words or figures. There is perhaps a parallel with measuring intelligence. We can measure a person's IQ with a lot of accuracy, but why one person with high IQ succeeds in the world while another does poorly is something we may come to understand by knowing the people, but cannot really be reflected in terms of test scores. Similarly, interpreting the contribution of structured instruments must recognise that they are limited to measuring signs and symptoms, while it is the clinician who must put that information in context.

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Classification and Diagnostic Systems

5

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Overview

Defining criteria for intellectual disabilities (ID) and psychopathology have undergone continuous development since the first attempts to define such concepts. Over time, these diagnostic categories have evolved from relatively simple and rigid terms to complex, multidimensional terms that even now are growing and being redefined. Sections "Overview" and "Shifting Definitions of Intellectual Disabilities" of this chapter explore historical and modern definitions of ID and psychopathology. While the consideration of possible interactions between ID and psychopathology has an established history, researchers did not dedicate major efforts to understanding dual diagnosis until the 1980s. Current concepts and measurements of dual diagnosis, as well as diagnostic dilemmas, are explored in the "Definitions of Psychopathology" section of this chapter.

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Shifting Definitions of Intellectual Disabilities

Historical Definitions

The recorded recognition of ID can be identified as far back as 1500 BC, in the therapeutic papyri of Thebes, Egypt, which postulated that brain damage caused physical and mental disabilities (Scheerenberger, 1983). Since that time, the terminology and process of identification of ID have continuously evolved. British law distinguished idiocy from lunacy early on. An idiot was defined as "one that hath no understanding from his nativity" (Blackstone, 1765, p. 298), where as a person non compos mentis "hath glimmering of reason, so that he can tell his parents, his age, and the like common matters" (p. 293). Legal commentary opined that people who were deaf-blind were also functionally idiots, since "he is supposed incapable of understanding, as wanting those senses which furnish the human mind with understanding and ideas" (p. 293). If found to be an idiot by a jury, then the King could permanently seize that person's lands and possessions. Thus, the distinction between ID and mental health has long been distinguished legally. Over the last 200 years, accepted terms have

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included the following: idiocy, feeblemindedness, mental deficiency, mental disability, mental handicap, and mental subnormality (Schalock et al., 2007). Organized attempts to educate people with ID did not occur until the 1830s in France (Barr, 1904) when Edward Seguin, formally trained in medicine, was inspired by his mentor, Jean Marc Gaspard Itard's efforts in training a feral child. While Itard felt that his own attempts were fruitless, they impressed upon Seguin the possibility that people with ID could learn; Seguin thus opened a school for "educating the idiot" in 1837 (Barr, 1904). As Seguin's work gained recognition, American doctors and educators began establishing similar education centers (Trent, 1995). One such doctor, Hervey B. Wilbur, opened the "New York State Asylum for Idiots." In their first annual report, Wilbur identified four classifications of ID based on educative observations (Disability History Museum, 1852). "Simulative idiocy" referred to children who would eventually be able to enter typical schools and lead typical lives. "Higher grades of idiocy" referred to children who would eventually be able to attend typical schools and were likely to be civilly useful. "Lower-grade idiocy" referred to children who could be educated to do simple tasks under the supervision of others. "Incurables" were identified as children for whom the greatest goal would be any level of education whatsoever.

Wilbur was not unique in his attempts to classify symptoms of ID and use that classification system to determine supports; however, very few professionals had consistently defined guidelines available to them. Such discordance in nomenclature limited the educator's and physician's ability to make consistent diagnosis of and recommendations for children with disabilities; however, in 1904, the French Minister of Public Instruction commissioned Alfred Binet to create a standard measurement of intelligence to address this issue. The original Simon-Binet intelligence quotient (IQ) test consisted of 30 tasks ordered from easiest to most difficult and measured language skills, memory, reasoning, digit span, and psychophysical judgments. Later versions categorized scores further based on intellectual achievement level, later called "mental age" (Boake, 2002). The original Simon-Binet has served as a model of format and a source of intelligence testing exam content for subsequent intelligence tests, such as the Stanford-Binet and the Wechsler scales for children and adults (Boake, 2002). The normed IQ scoring system that is currently used emerged in 1916 in Lewis Terman's adaptation of the Simon-Binet, known as the Stanford-Binet (Horn & Fuchs, 1987). Current versions of such tests are commonly used to identify and classify ID based on standard deviations from a normative score. Historically, the cutoff criterion has been set at between 1 and 2 standard deviations from the mean, indicating an IQ score of 70 or lower as ID. Historically and currently, such IQ limitations must occur within the developmental period, typically from birth to ages 18–21, to render a diagnosis of ID (Schalock et al., 2010).

Despite their widespread use to identify below average intelligence, IQ tests have been criticized for having psychometric defects. As the definition of ID continues to evolve, professional groups, including the American Association on Intellectual and Developmental Disabilities (AAIDD) and the APA (American Psychiatric Association), have attempted to elaborate definitions of ID to include more than IQ score indicators by analyzing behavior development in ID.

American Association on Intellectual Disabilities

The AAIDD, formerly called the American Association on Mental Retardation (AAMR), began in 1876 as a group of interdisciplinary professionals concerned with research and treatment development in the field of ID. Since its inception, AAIDD has considered social, medical, intellectual, and behavioral research findings in their definition of ID. The current definition has three inclusive criteria: limitations in intellectual functioning, limitations in adaptive behavior, and age of onset prior to 18 (Schalock et al., 2010). AAIDD has proposed a dynamic framework of human functioning when considering the manifestation of ID, including the person's intellectual

abilities, adaptive behavior, health, participation in everyday activities, and the environmental and personal context of the individual (Luckasson et al., 2002). While AAIDD defines deficits in intellectual and adaptive functioning as approximately two standard deviations below the mean, they urge professionals to consider other factors when considering a diagnosis. Thus, the current definition is based on five assumptions: first, limitations in functioning must be considered in the typical community and age related context. Second, professionals must consider cultural and linguistic diversity, including differences in communication, sensory, motor, and behavior factors. Third, professionals must consider individual skills in relation to deficits. An assessment should not attempt to identify intellectual and behavioral limitations in isolation. Fourth, when identifying limitations, an assessment team should begin distinguishing appropriate supports. Finally, with the application of appropriate supports, there should be a correlated improvement in adaptive functioning of the person diagnosed with ID (Schalock et al., 2010).

The Diagnostic and Statistical Manual of Mental Disorders

The Diagnostic and Statistical Manual of Mental Disorders-IV (DSM-IV) defines ID analogous to AAIDD. The DSM, created by the APA, has served as a predominant tool for identifying and classifying mental disorders in the USA. The DSM-IV definition defines ID by the same three inclusive criteria as the AAIDD: deficits in intellectual functioning and adaptive behavior with onset during the developmental period. Like the AAIDD definition, such deficits are to be identified through standardized testing methods and are measured as two standard deviations or more below the mean. While the DSM-IV also includes codes to indicate level of functioning based on IQ (mild, moderate, severe, and profound), the proposed DSM-V definition may not (American Psychiatric Association, 2011). The authors of the DSM-V, which is scheduled for publication in May of 2013, give four arguments towards eliminating coding based on IQ score: first, the diagnostic criteria will have more functionality. Second, concerns over the inaccuracy of testing will be less of a factor, as there will no longer be a need to discern between two different codes. Third, they argue that using IQ score coding fails to consider adaptive behavior in setting thresholds. Finally, the DSM-V is attempting to be consistent with AAIDD practices (American Psychiatric Association, 2011). In addition to the three identified inclusion criteria, the DSM also uses a five-axis classification scale to identify overall levels of functioning. Intellectual disability is on Axis 2; Axes 1, 3, 4, and 5 include clinical disorders, medical and physical conditions, psychosocial and environmental factors, and a global assessment of functioning, respectively. Such axes may also be considered when diagnosing and assessing services for people with ID.

International Classification of Diseases

Like the DSM, the International Classification of Diseases (ICD) is also undergoing revisions and is to be presented to the World Health Organization (WHO) in 2015 for approval (World Health Organization, 2012a, 2012b). The ICD also uses coding systems and a multiaxial scale to diagnose and classify IDD; in fact, DSM codes have been developed in accordance with ICD codes in attempt to establish consistency between the two classification systems. One key difference between the DSM and the ICD is that the DSM is used primarily in the USA and is written by a team of American psychiatrists; the ICD is written by doctors from 66 different countries and is the largest world-recognized authority of diagnosis (Mezzich, 2002). The current ICD (ICD-10) refers to ID as mental retardation and bases diagnosis primarily on IQ scores as indicated by standardized intelligence testing; adaptive behavior measures are optional and are not a requirement for diagnosis. ICD-10 also provides functional expectations and descriptions for each scale as defined by intelligence testing (World Health Organization, 1996), though such functional

identifications have been identified as "quite minimal, especially for individuals in the moderate and severe categories" (Schalock et al., 2010, p. 75). Revisions for the ICD-11 have sparked considerable debate about the appropriateness of including ID in the ICD, specifically because if ID is to be considered a disability, then it possibly cannot be considered a medical disorder (Salvador-Carulla et al., 2011). The ICD Working Group has proposed to replace the term "mental retardation" with "intellectual developmental disorders," defined as "a group of developmental conditions characterized by significant impairment of cognitive functions, which are associated with limitations of learning, adaptive behaviour and skills" (p. 177). They also recommend that subcategories indicating severity continue and that problem behaviors be removed from the core classification. The team supports this decision in saying that severity codes are accessible around the world, and while behavioral scales may be relevant, they are in their infancy and are not available worldwide and thus have no current value to an international diagnostic measure.

Adaptive Behavior Definitions and Measures

While the ICD has not yet embraced adaptive behavior as a diagnostic measure, concepts of adaptive behavior have been used to define ID since the first AAMR definition in 1887 (Bruininks, Thurlow, & Gilman, 1987). Adaptive behaviors are "the behavioral skills that people typically exhibit when dealing with the environmental demands they confront" (Widaman & McGrew, 2000, p. 97). Adaptive behavior differs from intelligence in that measures examine everyday behavior, rather than maximum performance, and adaptive behaviors are considered to be amenable to treatment whereas intelligence should be relatively stable (Harrison, 1987). Adaptive behavior also differs from maladaptive behavior in that maladaptive behavior is not just behavior on the low end of an adaptive behavior scale; rather, problem behavior, such as aggression or disruptive behavior, is that which may serve as a barrier to acquiring adaptive behaviors through training opportunities (Bruininks et al., 1987). AAIDD classifies adaptive behavior into three main categories: *conceptual skills*, such as language, reading, and writing, and comprehension of money, time, and numbers; *social skills*, including interpersonal skills, self-esteem, gullibility, avoidance of victimization, law following, and social problem solving; and *practical skills*, such as activities of daily living, occupational abilities, travel skills, and use of a telephone (Schalock et al., 2010). Categories such as social skills and practical skills assume cultural relevance and appropriateness for the individual's age group. For example, a 2-year-old incapable of using the phone is not representative of a deficit but an 18-year-old from the same culture who was incapable of using a telephone would be identified as having a deficit in adaptive behavior.

While adaptive behavior measures are a key component of diagnosis, they also serve as a platform for identifying support needs (Bruininks et al., 1987). Indeed, it is implied in the AAIDD definition of ID that while diagnosing ID, professionals ought to be considering appropriate supports based on information garnered from adaptive behavior measures (Schalock et al., 2010). Numerous measures, formal and informal, have been developed to help service agency providers, including schools, residential providers, and vocational service providers, identify skills necessary for more independent functioning in specific environments that are subsequently used to identify training goals (Horn & Fuchs, 1987). In fact, some measures are designed specifically for identifying service needs rather than diagnoses. The Supports Intensity Scale (SIS) was developed in 2004 to "measure practical support requirements of adults with ID in 85 daily living and medical behavior areas" (AAIDD, 2012, Quick Facts section, para. 4).

When using an adaptive behavior scale for diagnostic measures, the AAIDD cautions professionals to use standardized measures that have been normed with demonstrated validity and reliability (Schalock et al., 2010). A diagnosis of ID using adaptive behavior measures indicates that

the person being tested scores around two or more standard deviations below the population average. Those administering the tests should attempt to measure the individual's typical, rather than optimal performance, and should use knowledgeable respondents who are familiar with the typical behavior of the individual being measured (Shalock et al., 2010). Adaptive behavior scales are susceptible to similar errors as those made on IQ tests, and thus, clinical judgment should be used especially if the scale is used as a factor in diagnosis.

Legal Implications of ID Diagnosis

The correct diagnosis of ID can have significant implications for the individual, services, and society. Diagnosis can (1) determine eligibility for lifelong funded support services; (2) restrict an individual's freedom or lifestyle, such as when someone is involuntary committed to a hospital; (3) make one exempt or nonexempt from certain laws, such as the death penalty; and (4) entitle or not entitle an individual to certain benefits, including Social Security (Schalock et al., 2007). Kanaya, Scullin, and Ceci (2003) discussed how the Flynn affect can be especially detrimental to the opportunities afforded to individuals who are diagnosed with borderline or mild ID. Depending on when the taken IQ test was normed, an individual scoring close to borderline levels may be determined ineligible for positions in armed services, disqualified from special education or typical education opportunities, or subjected to legal consequences such as the death penalty. Indeed, found that the execution of people with intellectual disabilities is a violation of the eighth amendment ban on cruel and unusual punishment; however, prior to the decision, only 13 of the 38 states with death penalty laws prohibited execution of criminals with ID (Snell & Voorhees, 2006). It is clear that appropriately identified diagnoses of ID can severely impact the life of the individual and family with the diagnosis. Examples such as the *Atkins v. Virginia* case provide further support for AAIDD's emphasis on using clinical judgment and multiple scales to identify and diagnosis ID, as a diagnosis or lack thereof may have a powerful impact on the trajectory of an individual's life.

Research Implications of ID Diagnosis

Diagnosis of ID also renders an individual eligible for participation in research oriented towards further understanding and developing appropriate supports for individuals with ID. Inclusion in research is often specified by specific diagnosis, IQ level, or level of adaptive behavior (Snell & Voorhees, 2006), though some have criticized that researchers use categories that are too broadly defined to apply research findings effectively to individuals with similar diagnoses or characteristics. Additionally, considering that the definition of ID has changed several times throughout the last 100 years, the applicability and reliability of previous research to current issues can be questionable (Reiss, 1994). Nonetheless, researchers have been able to successfully identify effective supports and interventions and further analyze factors that influence the presentation of ID. For example, some research on ID and adaptive behavior has identified that between 5 and 13 % of individuals with ID also have a mental health diagnosis (Bruininks et al., 1987). As research develops, consideration of psychopathology diagnoses for individuals with ID has demonstrated effects on how services are conceptualized and rendered. However, concepts of psychopathology have been as in flux as theories of ID. Thus, in order to understand issues of dual diagnosis, it is crucial that one examines the history of psychopathology itself.

Definitions of Psychopathology

Psychopathology: Concepts Versus Classifications

Psychopathology involves the empirical study of the causes and processes of mental disorders (Dorland, 2003). As with other branches of medicine, psychopathology involves the determination

of criteria that guide an accurate diagnosis for the purposes of treatment. In the case of psychopathology, that diagnosis is a mental disorder. Despite this seemingly straightforward aim, the study of mental disorders is largely dependent on the conceptualization of psychopathology that is being used (Maddux, Gosselin, & Winstead, 2005). Conceptions of psychopathology vary, and the history of the study of mental disorders is replete with evidence of the difficulty in effectively classifying mental disorders (Bergner, 1997; Maddux et al., 2005; Widiger & Sankis, 2000). The identification of psychopathology has been based on models of statistical deviance, maladaptive or dysfunctional behavior, distress and disability, social deviance, and harmful dysfunction (Bergner, 1997; Maddux et al., 2005; Widiger & Sankis, 2000). These approaches are briefly reviewed as well as the challenges involved with each of them.

Behaviors or experiences that are characterized as statistically deviant are those that are abnormal or infrequent within the population at large (Maddux et al., 2005; Wakefield, 1997). Statistical deviance has been used as a defining feature of criteria for mental disorders. Unfortunately, statistical deviance does not denote that a behavior is dysfunctional (Wakefield, 1997). If a behavior is abnormal but still functional for the individual, and adaptive within the environment in which he lives, the impetus for treatment could be seen to be lessened. A similar argument can be raised in the case of basing psychopathology on behavior that deviates from social or cultural norms. Thus, some conceptions of psychopathology have focused solely on determining whether the behaviors associated with a disorder are adaptive or maladaptive in some way (Maddux et al., 2005) or represent harmful dysfunction (Bergner, 1997; Wakefield, 1992; Widiger & Sankis, 2000). Similarly, some researchers and clinicians have defined psychopathology based on the subjective distress the individual with the disorder may be experiencing and whether the person is identifiably disabled in some way (Bergner, 1997; Maddux et al., 2005). The difficulty with these conceptualizations is determining how maladaptiveness, harmfulness, disability, or distress is to be empirically defined. Wakefield (1992) attempted to define harmful dysfunction in terms of the cultural standards for harm and deprivation, what he called the value criterion, and whether the mental mechanism(s) in question was functioning naturally, the explanatory criterion. Although such a definition does provide some direction for empirical standards for diagnosis, a more extensive definition may be needed to fully account for the various forms of mental disorders. Bergner (1997) stressed that an effective definition of psychopathology is one that:

> conveys other advantages such as (a) successfully addressing the problem of psychopathology's relativity to time, culture, and situation; (b) illustrating a straightforward entrée to an integration of existing theoretical approaches to psychopathology and treatment; and (c) providing a coherent principle of classification for mental disorders (p. 246).

As will be shown, Bergner's concerns have been met to different degrees throughout the development of classification systems for mental disorders both in the USA and abroad. However, conceptualizations of psychopathology and the methods and criteria used across classification systems continue to be criticized and revised as new research findings and practical complications arise. In fact, the model on which conceptualizations and classifications of mental disorders is based is still contentious.

Categorical Versus Continuum Models of Psychopathology

In the late nineteenth century, Emil Kraepelin proposed that mental disorders represented distinct biological illnesses (Greene, 2007). He was one of the first to suggest that the symptoms of these distinct disorders could be used as a firm basis for classification. Kraepelin's categorical, or taxonomic, conceptualization of mental disorders implied that there was no direct relationship between different mental disorders; an individual was either assessed as having one or not (Lilienfeld & Landfield, 2008; Maddux et al., 2005). Although Kraepelin's conception of psychopathology had a large influence on the development

of classification systems, such as the Diagnostic and Statistical Manual of Mental Disorders-III (DSM-III), the categorical model of psychopathology has been widely criticized in favor of a continuous or dimensional model. Many researchers have stressed the lack of scientific reliability and validity of these categories as well as the potential for overlap between one class of psychiatric disorder and another class (Greene, 2007; Krueger, Caspi, Moffitt, & Silva, 1998; Lenzenweger & Dworkin, 1996; Linscott & van Os, 2010). Research on the genetic foundations and expression of personality disorders, for instance, also bears out the continuum model of psychopathology (Livesley, 1998; Livesley, Jang, & Vernon, 1998) which suggests that "normality and abnormality, as well as effective and ineffective psychological functioning, lies along a continuum [where] so-called psychological disorders are simply extreme variants of normal psychological phenomena" (Maddux et al., 2005, p. 10).

This distinction between categorical and continuum models of psychopathology becomes more complicated once the implications of each to practical and theoretical concerns are considered. Although the continuum model may pose a better match to theories stemming from recent research findings regarding the cause of or relationship between different mental disorders, it is not necessarily the more practical of the two models. A categorical model of psychopathology may better meet "the pragmatic demands of administration, provision of care, communication, statistical analysis, and so on" (Linscott & van Os, 2010, p. 394). Distinct categories and diagnostic criteria promote the clinical utility of a classification system, allowing for clear-cut measurement, diagnoses, and treatment plans (Widiger & Sankis, 2000). The American Psychiatric Association (1994) is careful to point out in the DSM-IV that "there is no assumption that each category of mental disorder is a completely discrete entity with absolute boundaries dividing it from other mental disorders or no mental disorder" (p. xxxi), but the common use of the DSM-IV involves measurement that is strictly categorical (Lilienfeld & Landfield, 2008). Given this current disconnect between the conceptualization of psychopathology in theory and practice, it is unclear how these models will continue to be used within the field of psychopathology; however, the models do provide us with a sound background from which to understand the definitions used in the most common classification systems for mental disorders.

The Diagnostic and Statistical Manual of Mental Disorders

The Development of the DSM and Approach to Classification Over Time

Prior to the publication of the DSM-I in 1952, the classification of mental disorders was largely disorganized (Lilienfeld & Landfield, 2008). The DSM-I criteria were developed to establish a standardized way of diagnosing mental disorders. It was hoped that the operationalized diagnostic criteria would help promote greater inter-rater reliability (Lilienfeld & Landfield, 2008). The more detailed DSM-II was developed to better facilitate these goals. Although they were widely praised, both initial versions of the DSM suffered from relatively vague descriptive criteria and did not consider contextual factors that might be co-occurring and contributing to the level of dysfunction the individual was experiencing (Eysenck, Wakefield, & Friedman, 1983). Also, there were a number of references to Freudian theory in the first two versions which did not match with ongoing developments in the field of psychiatry (Lilienfeld & Landfield, 2008). In response to these criticisms, the DSM-III included more detailed criteria and guidelines and was reorganized to reflect a neo-Kraepelinian structure (i.e., mental disorders were grouped and distinguished based on the signs, symptoms, and natural progression of the conditions). Significantly, the DSM-III included standardized diagnostic and decision rules for each disorder that "delineated the signs and symptoms comprising each diagnosis and the method by which these signs and symptoms needed to be combined to establish each diagnosis [and] … outlined hierarchical exclusion rules for many diagnoses" (Lilienfeld

& Landfield, 2008, pp. 15–16). The DSM-III was also the first DSM to incorporate a multiaxial approach to diagnosis the use of which continues in more recent versions. Using the multiaxial approach, clinicians were able to consider other factors that have been found to contribute to psychological functioning (e.g., medical disorders, psychosocial stressors, and level of adaptive functioning).

In moving from the DSM-III to the DSM-III-R and DSM-IV, the APA incorporated a polythetic approach to diagnosis, a noted improvement (Widiger, Frances, Spitzer, & Williams, 1988). "In a monothetic approach, the signs and symptoms are singly necessary and jointly sufficient for a diagnosis. In contrast, in a polythetic approach the signs and symptoms are neither necessary nor sufficient for a diagnosis" (Lilienfeld & Landfield, 2008, p. 17). This modification had the advantage of significantly improving interrater reliability, a benefit that, in many minds, outweighed the increased number of symptom combinations possible for a given diagnosis (Lilienfeld & Landfield, 2008; Widiger et al., 1988). In the development of the DSM-IV, the APA also sought to relax a number of the exclusion rules introduced in the DSM-III in response to criticisms as to the validity of these rules and the difficulty many clinicians had applying them in practice. The DSM-IV also includes an appendix that references certain disorders that reflect differences across cultures (Lilienfeld & Landfield, 2008), a change that indicates an attempt on the part of the APA to meet some of the definitional criticisms by researchers such as Bergner (1997) discussed above.

The DSM-IV and Proposed DSM-V Definitions of Mental Disorder

The evolution of the DSM demonstrates the APA's commitment to modifying the definitions and classification methods in response to advances in research, theory, and practice (First et al., 2004; Stein et al., 2010). This is exhibited in the current definition of mental disorder used in the DSM-IV (see Table 5.1). As can be seen, the DSM-IV definition specifically addresses many of the conceptualizations discussed earlier in this chapter, with the exception of statistical deviance. Feature B specifically ties the behavioral pattern or psychological syndrome to a time, "present," "significantly increased risk" (Bergner, 1997), in relation to "distress" or "disability" or restriction of ability/freedom (Bergner, 1997; Maddux et al., 2005). Similarly, feature C addresses the cultural relevance of this behavior and D rules out the issues of maladaptive behavior (Maddux et al., 2005; Widiger & Sankis, 2000) or dysfunctional biological processes (Wakefield, 1992). Feature E also excludes interpretations of the behavior or syndrome as disordered if it is reflective of social deviance (Maddux et al., 2005). Thus, the DSM-IV definition clearly delineates how mental disorder is to be conceptualized; however, the operationalization of some of these conceptualizations is left open-ended.

Table 5.1 DSM-IV definition of mental disorder

Features
A. A clinically significant behavioral or psychological syndrome or pattern that occurs in an individual
B. Associated with present distress (e.g., a painful symptom) or disability (i.e., impairment in one or more important areas of functioning) or with a significantly increased risk of suffering death, pain, disability, or an important loss of freedom
C. Must not be merely an expectable and culturally sanctioned response to a particular event (e.g., the death of a loved one)
D. A manifestation of a behavioral, psychological, or biological dysfunction in the individual
E. Neither deviant behavior (e.g., political, religious, or sexual) nor conflicts that are primarily between the individual and society are mental disorders unless the deviance or conflict is a symptom of a dysfunction in the individual
Other considerations
F. No definition adequately specifies precise boundaries for the concept "mental disorder"
G. The concept of mental disorder (like many other concepts in medicine and science) lacks a consistent operational definition that covers all situations

Source: American Psychiatric Association (1994)

Researchers and clinicians have emphasized that this open-endedness is likely unavoidable with "precise boundaries" and "consistent operational definition[s]," a function of the ever-evolving properties of a concept like "mental disorder" (Stein et al., 2010; Widiger & Sankis, 2000).

Table 5.2 Proposed DSM-V definition of mental disorder

Features
A. A behavioral or psychological syndrome or pattern that occurs in an individual
B. That is based on a decrement or problem in one or more aspects of mental functioning, including but not limited to global functioning (e.g., consciousness, orientation, intellect, or temperament) or specific functioning (e.g., attention, memory, emotion, psychomotor, perception, thought)
C. That is not merely an expectable response to common stressors and losses (e.g., loss of a loved one) or a culturally sanctioned response to a particular event (e.g., trance states in religious rituals)
D. That is not primarily a consequence of social deviance or conflict with society
Other considerations
E. That has diagnostic validity on the basis of various diagnostic validators (e.g., prognostic significance, psychobiological disruption, response to treatment)
F. That is helpful in diagnostic conceptualization, assessment, and/or treatment-related decisions
G. No definition of "medical disorder" or "mental (psychiatric, psychological) disorder" perfectly specifies precise boundaries for the concepts or can provide consistent operationalizations that cover all situations

Source: American Psychiatric Association (2010)

This property of the definition leaves open opportunities for criticism and revision in light of new theoretical evidence, empirical findings, and practical concerns. For instance, some researchers have questioned the diagnostic validity and clinical utility of the "clinical significance" criterion that was added to the definition and the diagnostic criteria of many of the mental disorders covered in the DSM-IV (Spitzer & Wakefield, 1999; Stein et al., 2010). The term "clinical significance" is ambiguous from a measurement standpoint as it is unclear how one would classify "clinically significant distress or impairment in social, occupational or other important areas of functioning" (American Psychiatric Association, 1994, p. 7) (Table 5.2).

The difficulties in clarifying and defining psychological constructs and diagnostic criteria continue as the APA prepares for the publication of the aptly named DSM-V. For over a decade now, discussions have been underway in order to facilitate the development of the next version of the DSM (American Psychiatric Association, 2010). The preparations for and development of the DSM-V have involved multiple conferences to set research priorities, the ongoing planning and examination of criteria by different work groups, and the joint cooperation between researchers and clinicians from diverse fields and across multiple organizations (American Psychiatric Association, 2010). This process has led to the new "draft" criteria and definitions. The proposed revision of the definition of mental disorder, put forward by the Study Group on Impairment and Disability Assessment, can be seen in Table 5.1.

While the proposed definition retains most of the features that were highlighted in the DSM-IV definition, there are notable changes. The term "clinically significant" has been dropped from feature A, and references to "distress," "disability," and "impairment" in feature B have been excluded in favor of a criterion centered on assessed levels of functioning (i.e., global, specific). This revision to feature B may be the reason for the removal of feature D from the DSM-IV definition, as it specifically referenced dysfunction, but this remains unclear. References to culturally based behaviors, contextual events (e.g., stressors), and social deviance are still included in the proposed definition, although the wording of these features has been altered. A significant change from the DSM-IV definition is that the core features just discussed need to have recognized diagnostic validity and contribute to assessment and treatment planning. Such statements may be in response to criticisms over the previously ambiguous "clinically significant" terminology and to aid with the clinical utility of the DSM-V (see First et al., 2004; Spitzer & Wakefield, 1999; Stein et al., 2010).

As noted, this definition of mental disorder is only the most recent definition proposed for inclusion in the DSM-V. The current proposal may be further revised before the final publication of the DSM-V, forecasted for the spring of 2013 (American Psychiatric Association, 2010). The form of this definition is still relatively contentious, and an alternative definition has been proposed by select members from two of the

DSM-V work groups as well as other sources. The current status of the proposed definitions for the DSM-V again emphasizes the distinct challenges involved in defining a term as complex and far reaching as that of "mental disorder." As can be seen, many of the critiques and challenges to the definition of mental disorder mirror those involved in redefining intellectual disability. And, as with members in the ID community, the psychiatric community will no doubt eagerly await the finalization of the DSM-V in order to put to the test the new definitions and criteria that will serve as the foundation for much of the clinical and research work carried out in psychiatry for the next generation.

The International Classification of Diseases

The ICD is a classification system overseen by the WHO that has been developed for the purposes of identifying and researching diseases around the world (World Health Organization, 2012a, 2012b). Originally stemming from the recognition of the need for a method of classifying diseases, causes of death, and health records internationally, the ICD has been continually refined since its inception as the International List of Causes of Death. The ICD-10 is the current version and includes 22 classification categories or chapters (World Health Organization, 2012a, 2012b). Chapter V covers mental and behavioral disorders and includes classifications that parallel many of those found in the DSM-IV (e.g., Schizophrenia, Mood Disorders, Disorders of Adult Personality). Chapter V was originally developed in the 1960s and has been incorporated into the classification system since ICD-8 (World Health Organization, 1992).

As with the DSM, the process of defining terms reflective of mental and behavioral disorders in the ICD has been a complex one, having been driven by many of the same changes observed in research findings, theories, and practice. Although the definition of disorder used in the ICD-10 is more general than that specified by the DSM-IV, many of the elements are similar (see Table 5.3). As with the DSM-IV definition, the WHO has noted the imprecise nature of the term "disorder." Further, the definition includes references to "distress" and "interference with personal functions" (comparable to the terms "dysfunction" and "disability" used in the DSM-IV definition). Finally, as with the DSM-IV definition, the ICD-10 definition excludes social deviance as a sufficient condition for mental disorder.

Table 5.3 ICD-10 definition of disorder

A. The term "disorder" is used throughout the classification, so as to avoid even greater problems inherent in the use of terms such as "disease" or "illness"
B. "Disorder" is not an exact term, but it is used here to imply the existence of a clinically recognizable set of symptoms or behavior
C. Associated in most cases with distress and with interference with personal functions
D. Social deviance or conflict alone, without personal dysfunction, should not be included in "mental disorder" as defined here

Rather than being antagonistic to the DSM, the WHO has developed Chapter V of the ICD to reflect ongoing modifications to the DSM, often consulting with the Mental Health Administration in the USA, as its members work to revise new versions of the manual (World Health Organization, 1993). It is likely that cooperation such as this will persist as researchers and clinicians continue to grapple with the complex task of describing and defining mental disorders in the coming years, especially as the DSM-V is expected in 2013 and the ICD-11 in 2015.

Further Developments in Classification Systems and Intellectual Disabilities

Moving forward, it remains clear that classification systems for psychopathology will continue to evolve in response to new research findings. Most recently, numerous advances have been made in the fields of genetics and neuroscience that may foster improvements in diagnostic criteria and the understanding of the etiology of many mental disorders (Broyd et al., 2009;

Craddock & Forty, 2006; Insel et al., 2010; Uhlhaas & Mishara, 2007). There has also been increasing interest in the presentation and diagnosis of mental disorders in individuals with intellectual disabilities (ID) or dual diagnosis (Bhaumik, Tyrer, McGrother, & Ganghadaran, 2008; Fletcher et al., 2009; Griffiths, Stavrakaki, & Summers, 2002; Morgan, Leonard, Bourke, & Jablensky, 2008). Research on the similarities and differences in mental disorders, as expressed in individuals with ID, recently culminated in the publication of the Diagnostic Manual-Intellectual Disability (Fletcher, Loschen, Stavrakaki, & First, 2007). Individuals with ID already present with a number of distinct challenges that factor into diagnosis, assessment, and treatment. When comorbid psychiatric disorders are also present, a number of additional variables need to be considered, as will be described.

Application of Definitions of Psychopathology in Persons with Intellectual Disability

Development of Dual Diagnosis Concept

It is common to opine that prior to the 1980s, mental health issues in persons with ID were probably under-recognized, under-reported, and/or ignored. Yet this was not always so. For example, Penrose's (1938) Colchester survey found that 414 of 1,280 (32.0 %) people had some form of mental illness. The most common forms were epilepsy (16.4 %), psychoneurosis, and perversion (10.3 %), affective psychosis (1/8 %), and schizophrenia (3.8 %). In a similar manner to later studies, he observed that mental illness was negatively related to degree of ID. For example, the proportions of people with a mental illness excluding epilepsy were 37.8 %, 18.8 %, 9.0 %, and 7.2 % for people classified as "dull," "feeble minded," "imbecile," and "idiot," respectively (Penrose, 1961, Table 43, p. 253). Reviews in the 1960s, such as Tarjahn (1965), made no mention of the emotional or psychiatric aspects of ID. Some early studies even questioned if people with ID experienced common mental health problems such as depression. For example, Sovner and Hurley (1983) reviewed 25 studies of depression and mania in people with ID. They found that people with all degrees of ID could demonstrate signs and symptoms of affective illness, that researchers could apply DSM-III criteria for affective disorders, and that diagnoses could be made based on changes in behavior in people with severe and profound ID.

In the 1980s, the growing recognition of the emotional life of people with ID resulted in the development of the field of dual diagnosis. For example, psychometric assessments were developed, including general screens of psychopathology such as the *Psychopathology Instrument for Mentally Retarded Adults* (Matson, 1988), *Diagnostic Assessment for the Severely Handicapped-II* (Matson, 1995), the *Reiss Screen* (Reiss, 1986), the *Psychiatric Assessment Schedules for Adults with Developmental Disabilities* (Moss & Patel, 1995; Moss et al., 1993), and the *The Prout-Stromer Behavior Rating Scale* (Prout & Stromer, 1993). Following up on the development of general screens, instruments were developed to assess specific forms of psychopathology, such that today there are many instruments and observational measures of mood disorders in people with ID. In parallel with these developments, researchers constructed and evaluated psychometric measures of challenging behavior such as the *Aberrant Behavior Checklist* (Aman & Singh, 1985) and the *Behavior Problem Inventory* (Rojahn, Matson, Lott, Esbensen, & Smalls, 2001). The development of assessment protocols allowed researchers to conduct epidemiological studies which, in contrast to earlier research, often appeared to find surprisingly high rates of psychopathology (Rojahn et al., 2001). Reviews of specific disorders such as depression (Matson, 1983) set the agenda for future research. As interest grew, professional organizations responded. For example, the Royal Collage of Psychiatrists was instrumental in establishing chairs of psychiatry in Britain which formed the basis of specialized professional training for psychiatrists and for research. As the field developed, the question of applicability of standard

Table 5.4 Selected statistics on the number and prevalence of American children with various kinds of disabilities (adapted from US Department of Education, 2011)

	1980–1981			2008–2009		
Type of disability	Number of children (M)	Proportion of all children (%)	Proportion of children with disabilities (%)	Number of children (M)	Proportion of all children (%)	Proportion of children with disabilities (%)
All disabilities	4.14	101	–	6.48	13.2	–
Specific learning disabilities	1.46	3.6	35.3	2.47	5.0	38.2
Speech or language impairments	1.16	2.9	28.2	1.42	2.9	22.0
Intellectual disabilities	0.83	2.0	28.2	0.47	1.0	7.4
Emotional disturbance	0.37	0.8	8.4	0.42	0.9	6.5
Autism	0.02[a]	0.0[a]	0.2[a]	0.33	0.7	5.2
Traumatic brain injury	0.17[b]	0.4[b]	2.8[b]	0.35	0.7	5.5

[a]Data refers to 1995–1995
[b]Data refers to 2000–2001

psychiatric classification systems to people with ID was questioned. For example, most studies did not in fact use unmodified psychiatric diagnostic criteria with people with ID but modified them in numerous ad hoc ways (Sturmey, 1993). This has now become standardized with the development of several modified systems (Cooper, Melville, & Einfeld, 2003; Fletcher et al., 2007). Several journals now routinely publish research related to mental health issues in people with ID, and recently the *Journal of Mental Health Research in Intellectual Disabilities*, a journal specializing in this field, has been published. Finally, there has been some international dissemination of dual diagnosis concepts which has raised new challenges, such as how to identify simple resources for persons with dual diagnosis in developing countries. Thus, in just over 30 years, the field of dual diagnosis has gone from almost nothing to a fully developed field of research and practice.

The growth of the field of dual diagnosis resulted in the recognition of several practical and conceptual issues. To say that a person has both an ID and a mental health problem required reliable, valid, and simultaneous application of two sets of criteria. Unfortunately, the application of each criterion is problematic, and the application of the two sets of criteria may interact. The previous section of this chapter documented how both the conceptualization and measurement of ID has shifted radically over the last 50 years. These changes are not mere academic questions but are reflected in the administrative prevalence, patterns of diagnoses, and large-scale changes in the labeling of children. Table 5.4 summarizes selected statistics from the US Department of Education from 1980 to 1981 and 2008 to 2009. The number of children with all disabilities increased from 4.1 to 6.4 M which was an increase from 3.6 to 5.0 % of all children in education. Most children with disabilities were classified with Specific Learning Disabilities (SLD) or Speech and Language Impairments (SLI), consistently accounting for about 70 % of children with disabilities. The numbers of children with SLD, SLI, and Emotional Disabilities (ED) increased in line with increases in the population, as shown by relatively stable percentage of children served. In contrast, both the number and proportion of children with ID fell by nearly half, whereas the numbers and proportions of children with autism, TBI (Traumatic Brain Injury), and DD all increased dramatically. These disabilities

accounted for a small proportion of children with disabilities, except for autism which increased from close to 0 to nearly 5 % of children with disabilities.

These US national statistics and related studies provide good evidence that the criteria for ID are not applied reliably and accurately in US special education. Rather, service providers appear to use them pragmatically to obtain more resources for children who are difficult to serve in less restrictive, resource-poor settings. Evidence for this comes from MacMillan, Gresham, Siperstein, and Bocian (1996) who followed a cohort of 150 children whom they assessed at the beginning of the school year with a psychometric battery. There were 43 children with WISC-II IQs below 75 at the beginning of the study. Of 35 children with academic and behavioral problems, only six were subsequently classified as having ID and 18 were classified as having learning disabilities. MacMillan et al. suggested that the reluctance to classify children as having ID, despite meeting psychometric criteria for ID and the need for some form of additional supports, is reflected in the national statistics cited above and in evidence that the IQ of children with other disabilities, such as SLD has fallen (MacMillan, Gresham, & Bocian, 1998). The increased use of other disability categories, such as autism and TBI, may also reflect a more recent version of this trend observed some 15 years ago.

When adolescents with mild disabilities graduated from high school, approximately half disappeared from services (Landesman Ramey, Dosset, & Echols, 1996). This may reflect the removal of academic demands from their lives, failure to provide services for people with mild ID, or a lack of clear conceptualization of mild ID in adulthood by society and behavioral sciences (Landesman et al., 1996). There are several social ecological studies of adults with mild and borderline ID living independently in community settings. These studies suggest that many of these people do not receive services but often have significant problems in adaptive behavior and adaption to the demands of everyday life (Edgerton, 1993; Zetlin & Murtaugh, 1990). Some of these people periodically reenter services for people with disabilities, generic mental health services, or the justice system at periods of personal crisis, such as when key personal supports are lost or when mental health and behavioral issues arise.

Measurement Issues

Measuring Intellectual Disabilities

The adequacy of measurement systems also has large effects on whether someone is labeled as having ID and a mental health diagnosis. As mentioned earlier, the Flynn effect is the upward drift in the norms of IQ, educational, and other tests. These effects are large and affect both individuals and services: Examination of intelligence test results over a 46-year span indicated that American IQ scores increased an average of 13.8 points, nearly an entire standard deviation (Flynn, 1984). Additionally, individual scores have shown to vary based on which test was taken. For example, Lukens and Hurrell (1996) found an average eight-point difference between scores obtained from WISC-III and SBIS-4 for students with mild mental retardation. When considering typical standard deviation measurements and diagnostic cutoffs, eight points could determine whether or not an individual is eligible for supports appropriate for an ID diagnosis. Similarly, Kanaya et al. (2003) observed that when IQ tests were re-normed, students with both borderline and mild ID lost, on average, 5.6 IQ points, but if children were retested on the same instrument when the test had not been re-normed, no change in IQ was observed. Kanaya et al. argued that these changes have significant impacts on determining service eligibility, flipping some children in and out of eligibility depending upon the vagaries of testing practices.

These issues are also reflected in the epidemiology of ID at the state level. For example, Scullin (2006) observed large and steep reductions in the prevalence of ID in American states following the introduction of the WISC-III in 1991. The prevalence of ID fell by 12 % between 1981 and 1991 but in some states fell by fully 50 %. Subsequently, as the test norms drifted upwards,

the prevalence of mental retardation returned to its former level over a period of approximately 10 years. The reasons for the Flynn effect is unclear, but it could be due to improved test sophistication, sampling error, or cultural changes; however, the implication is that one's IQ score may be dependent upon when and which version of the test one has taken.

The reliability and validity of IQ tests and the conceptualization of intelligence as they apply to diagnosis of ID has received extensive attention, but similar concerns can also be raised regarding the measurement of adaptive behavior. For example, Thompson, Tasse, and McLaughlin (2008) evaluated the reliability of the SIS. Fifty-one interviewers interviewed 80 respondents concerning the supports for 40 people with disabilities for a total of 120 interviews. This was done as part of a workshop to train on the use of the SIS. There were three interviews for each of the 40 people with disabilities that permitted calculating inter-rater and test-retest reliability between two interviewers. Overall, reliability was quite high with 53 of 56 correlations being in the "excellent" range and three being in the "fair" range. Closer examination of the data in their Table 5.2 reveals that reliabilities for the Lifelong Learning Goals domain were lower than for other scales (range 0.35–0.88) and that mixed inter-rater correlations were generally somewhat lower than for other reliability measures, with overall reliabilities for the total index being only 0.54 and 0.59 for the total uncorrected and correct correlations, respectively.

Others have critiqued measures of adaptive behavior on the basis of lack of conceptual foundation. For example, Simeonsson and Short (1996) noted that "Instruments are largely developed on a pragmatic basis, defined by an aggregation of items that are often derived from clinical consensus of models that are validated empirically using factor analysis … there is a need for conceptual frameworks to guide the development and empirical validation of instruments … personal competence such as motivation and social inference are typically not included in measures of adaptive behavior …" (p. 139). Thus, even if adequately reliable measures of adaptive behavior were to be developed, the issue of construct validity, absent its conceptual clarification, would be wanting.

The third criterion—that delay should occur during the developmental period—is often easy enough to measure; however, legal definitions of the developmental period have ranged from 16 to 22 years. Under certain circumstances, practitioners may also have considerable problems in determining whether or not a deficit in adaptive behavior occurred during the developmental period. For example, when an adult with possible mild ID requests a particular service for the first time, determining whether or not a delay occurred during the developmental period may be difficult. School records may be incomplete or absent. Other challenges may include administering a test in the person's native language, tests that are not normed in the person's first language, and conditions that may onset during adolescence or early adulthood, such as TBI or degenerative psychiatric or other medical conditions.

Measuring Mental Health

Diagnosing mental illnesses has always been challenging. The continuing evolution of standardized diagnostic criteria and related instruments attest to this; yet, the application of diagnostic criteria to people with ID presents additional challenges, as shown by the use of modified diagnostic criteria (Cooper et al., 2003; Fletcher et al., 2007; Sturmey, 1993). One of the challenges in applying standardized diagnostic criteria to people with ID includes diagnostic overshadowing, the tendency to ascribe possible psychiatric symptoms as a feature of ID (Reiss, Levitan, & Szyszko, 1982).

There is a lack of systematic studies of reliability of psychiatric diagnosis in people with ID. At least one study, however, found relatively good reliability when mental health professionals were asked to diagnose depression or schizophrenia in adults with mild or moderate ID (Einfeld et al., 2007). In this study, these clinicians made a relatively easy discrimination of depression vs. psychosis. In more typical clinical situations,

however, clinicians would have to make more subtle discriminations such as depression vs. anxiety disorders. Thus, the external validity of such findings remains unknown at this time.

Whereas using standardized diagnostic criteria for people with mild and moderate disabilities with a reasonable language repertoire may be possible, applying standardized diagnostic criteria to people with severe or profound ID with minimal or no expressive language is far more difficult. Thus, various researchers have developed questionnaires (Ross & Oliver, 2003) and observational protocols to observe mood behavior. One common approach has been to use the notion of "behavioral equivalents." This may refer to the possibility that, under certain circumstances, challenging behavior represents an underlying mental illness that cannot be expressed with conventional symptoms or that the observable challenging behavior covaries with a mental illness and is a reliable index of a mental illness (Fletcher et al., 2007). For example, Sovner, Fox, Lowry, and Lowry (1993) treated self-injurious behavior (SIB) with fluoxetine based on the hypothesis that SIB was a symptom of an underlying depression. Similarly, Lindauer, DeLeon, and Fisher (1999) decreased the observable behavior indicative of negative mood and increased behavior indicative of positive mood in a woman with severe ID and a mood disorder by providing choices of preferred activities associated with positive mood.

The notion of behavioral equivalents has been popular for many years (Sovner et al., 1993; Sovner & Hurley, 1983), but empirical support for its use is varied and patchy. For example, a sample of 63 clinicians reported that practitioners found that modified DSM criteria were "easy" or "very easy" to use regardless of level of disability and helped them avoid using "not otherwise specified categories" (Fletcher et al., 2009). In contrast, empirical studies have found weak or negligible associations between hypothesized behavioral equivalents and typical symptoms of psychopathology (Sturmey, Laud, Cooper, Matson, & Fodstad, 2010a, 2010b; Tsiouris, Mann, Patti, & Sturmey, 2003).

Conclusions

The classification of intellectual disabilities and psychopathology each raise numerous difficult conceptual and measurement issues. When we ask the combined question concerning the co-occurrence of both conditions, these problems are magnified yet further. There is good evidence of very large changes in how society conceives of intellectual disabilities and how, in practice, there have been large shifts away from labeling people with mild intellectual disabilities to other apparently more palatable diagnoses that still enable the child to receive services. Over the last 30 years, there have been many new developments in measuring psychopathology in people with intellectual disabilities, and these new developments have usefully filled in a gap for both practitioners and researchers. This has been a useful beginning, but the measurement of psychopathology in people with intellectual disabilities has raised many questions that have yet to be resolved.

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Part II

Aetiological Approaches

Neuroimaging

6

Christine Ecker

Abbreviations

AD	Alzheimer's diseases
ADHD	Attention deficit hyperactivity disorder
ASD	Autism spectrum disorder
BOLD	Blood oxygenation level dependent
CBF	Cerebral blood flow
CSF	Cerebrospinal fluid
CT	Computerized tomography
DS	Down syndrome
DTI	Diffusion tensor imaging
FA	Fractional anisotropy
FMR1	Fragile X mental retardation 1 gene
fMRI	Functional magnetic resonance imaging
FXS	Fragile X syndrome
GLx	Glutamate+glutamine
ID	Intellectual disability
INS	Myoinositol
MCI	Mild cognitive impairment
MD	Mean diffusivity
MRI	Magnetic resonance imaging
MRS	Magnetic resonance spectroscopy
NAA	*N*-acetyl-aspartate
NMR	Nuclear magnetic resonance
SBM	Surface-based morphometry
sMRI	Structural magnetic resonance imaging
VBM	Voxel-based morphometry
VCFS	Velo-cardio-facial syndrome

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Introduction

Over the last 2 decades, neuroimaging has become a widely available, powerful and cost-effective tool for investigating the brain in vivo, both in the normal and pathological state. The appeal of neuroimaging ultimately lies in its power to produce very clear and detailed computerized images of the brain in vivo (i.e. in the living brain). These can then be used to investigate brain anatomy, functioning and connectivity in a wide range of disorders. Neuroimaging has therefore had a dramatic impact in psychiatry research and has significantly contributed to concepts and current thinking on a wide range of disorders, including intellectual disability (ID).

The aim of this chapter is (1) to introduce currently available neuroimaging technologies, (2) to summarize the insights neuroimaging has provided into disorders of psychopathology—and particularly those associated ID and (3) to outline potential applications of imaging techniques in the clinical setting.

Neuroimaging Methods

Magnetic Resonance Imaging

Magnetic resonance imaging (MRI) has by large substituted traditional computerized tomography (CT) scanning and is the investigation of choice for examining the brain in a noninvasive fashion.

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Unlike CT, MRI scanning is not based on the potentially harmful X-ray technology, but uses the *nuclear magnetic resonance* (NMR) or the *nuclear moment* of atomic nuclei to derive images of the brain. The nuclear moment can be best described using the analogy of a rotating spinning top. Just like the spinning top, atomic nuclei exhibit an *angular momentum*, which is also referred to as *spin*. The nuclear moments are usually randomly orientated and spin in all directions. However, when nuclear moments are brought into a strong magnetic field, the nuclei align along or against the magnetic field vector. The nuclei aligned along the magnetic field are in a state of low energy, and no signal can be detected (*equilibrium magnetization*). The nuclei orientated antiparallel to the magnetic field are in a state of high energy. When a burst of radio frequency waves is applied, nuclei transition between these two energy states forces them to emit energy. This energy is emitted as a radio wave and can be detected by the MRI scanner, a process known as *relaxation*. Depending on the specific radio frequency pulse applied by the scanner, the relaxation is of type T1 or T2, and the resulting images are said to be either T1 or T2 weighted. In T1-weighted images, less dense tissue appears darker (e.g. cerebrospinal fluid or CSF) while denser tissue types (e.g. bone, white matter) show increased signal intensity and hence appear lighter. This type of contrast is therefore particularly suited to investigate brain structure, as it makes it possible to separate brain tissue into its constituent components of interest (i.e. grey matter, white matter, bone/CSF). In T2-weighted images, the situation is reversed and lower signal intensity values are found in tissue types with enhanced fluid content. T2-weighted imaging is less common in MRI, but can be useful particularly in the early detection of oedema or stroke, which are difficult to see using a T1-weighted contrast. Only nuclei with an uneven number of protons and neutrons exhibit a net angular spin and are thus visible in NMR experiments. For imaging in biological systems, the nucleus that has attracted most attention is the atom hydrogen (^{1}H) or 'proton' NMR, as it is highly concentrated in the human body and highly sensitive (i.e. it gives rise to large NMR signals).

Structural Magnetic Resonance Imaging

Structural MRI or sMRI can be used to measure the physical structure or *anatomy* of the brain. Images are generally acquired as a series of 2D slices at a given orientation (sagittal, coronal, axial), which can then be rendered in three dimensions to cover the entire brain. Once images are acquired by the scanner, they are further processed computationally to make the data comparable across subjects or groups (e.g. patients vs. controls). Conventional sMRI preprocessing includes several important steps such as *normalization* (i.e. registration of individual brain to a template), *segmentation* (i.e. separation of white and grey matter), *modulation* (i.e. adjustment for volume changes during normalization), and data *smoothing* (i.e. blurring of the data). These steps are necessary to account for the large degree of natural variation in brain structure across individuals. The preprocessed images are then compared statistically, for instance using so-called *mass-univariate* techniques (e.g. voxel-based morphometry or VBM, Ashburner & Friston, 2001), which compare signal intensities between groups at each location in the brain. The resulting output shows regions of the brain that are larger or smaller in volume between patients and controls. These methods can be applied to both grey and white matter. Grey matter consists mainly of neuronal cell bodies, white matter predominantly contains myelinated axon tracts and is hence a measure of brain connectivity.

While traditional VBM approaches have been extremely successful at characterizing structural brain abnormalities in a variety of mental health conditions and psychiatric diseases, newer techniques such as surface-based morphometry (SBM) now allow us to describe brain anatomy at a much higher degree of specificity. Using SBM, it is now possible to compare groups on the basis of several parameters measuring brain anatomy, including *cortical thickness* (i.e. thickness of the cortical sheet), *surface area* or *cortical folding*. It has been suggested that these different parameters have distinct genetic determinants and differing developmental trajectories, and should hence be investigated in isolation in order to better

understand the neurobiological mechanisms of psychopathy and other conditions.

Functional Magnetic Resonance Imaging

Functional magnetic resonance imaging (fMRI) is one of the more recently developed forms of neuroimaging (early 1990s), which makes it possible to measure brain activation on the basis of relative changes in *cerebral blood flow* (CBF). Brain regions that are more active have an increased metabolic demand and hence consume more glucose and oxygen, which need to be delivered by the bloodstream. The fMRI signal, also known as the blood oxygenation level dependent (BOLD) response (Ogawa, Lee, Kay, & Tank, 1990), measures the small (<2 %) signal changes associated with the concentration of deoxyhaemoglobin in the blood as a result of the increased metabolic demand (i.e., increased consumption of oxygen). This change in metabolic demand alters the magnetic properties of the surrounding brain tissue and can be used as an endogenous MRI contrast agent. Thus, while the BOLD signal is closely related to neural activity, the origin of the fMRI response is undoubtedly vascular and hence provides an indirect marker of neural functioning.

fMRI can be used to examine neural activation while the brain is 'at rest' and in response to different *stimulation paradigms* (i.e. tasks that participants perform in the scanner). The choice of the paradigm is virtually unlimited and ranges from tasks involving emotional processing, perception, language, and/or memory. While participants perform the given task, the MRI system tracks the signal for the duration of the scanning session. In areas that are involved in processing the task, the fMRI response is expected to go up and down depending on the stimulation paradigm. The 'activity' of a brain region is therefore defined as how closely the time course of the signal matches the expected time course, which can be assessed using various model-fitting procedures (i.e. time-series analysis). The results of fMRI experiments are generally presented as so-called *activation maps*, which indicate the brain region(s) that are most active in response to the paradigm. These maps can be created within individuals or subject populations, but may also be used to compare different groups of subjects (e.g. patients vs. controls). The resulting difference maps indicate regions in the brain that differ in brain activation between groups and hence process information in a different way. These difference maps are of course related to various mental functions depending on the employed stimulation paradigm. fMRI thus provides an invaluable tool for investigating normal and abnormal brain functioning.

Magnetic Resonance Spectroscopy

Magnetic resonance spectroscopy (MRS) allows then noninvasive and in vivo exploration of the molecular composition of brain tissue and identifies certain molecular constituents (i.e. *metabolites*) involved in physiological or pathological processes. In 1950, Proctor and Yu proposed that the *resonance frequency* of a nucleus depends on its chemical environment, which produced as small change in resonance frequency of that nucleus (i.e. *chemical shift*) (Proctor & Yu, 1950). The value of the chemical shift thus provides information about particular molecular groups. For instance, different chemicals can be identified on the basis of their peak resonance frequency, height and weight at half-height (Lenkinski & Schnall, 1991). However, only a limited number of molecules are observable in MRS. Among others, these include (1) *N-acetyl-aspartate* (NAA), which is a molecular present in healthy neurons, (2) myoinositol (found in glial tissue), (3) glutamine-glutamate-GABA complex (neurotransmitters) and (4) choline compounds (marker in the synthesis and breakdown of cell membranes).

MRS is generally applied to regions of interest, which are chosen depending on a priori hypotheses. Adequate MR spectra can be obtained in periods of time as short as 10–15 min. There is a growing number of clinical applications; for example, spectroscopy can be used to detect and monitor brain tumours as well as certain neurodegenerative diseases (e.g. Alzheimer

and Parkinson Disease). One of the drawbacks of MRS is that information on specific chemical compounds can only be measured in a limited range of spatial locations in the brain and are currently not measures on a 'whole-brain' basis. Thus, it is essential to have a strong a priori hypothesis with regards to the specific brain region of interest. As described in further detail below, MRS has also been used to investigate a range of disorders associated with ID, such as certain developmental disorders (e.g. autism).

Diffusion Tensor Magnetic Resonance Imaging

Diffusion tensor imaging (DTI) can be used to examine the volume and integrity of white matter tracts and thus provides measures of large-scale structural connectivity in the brain. It is based on the principle of *diffusion*, which is the random motion of water molecules. Diffusion is either *isotropic*, if the motion is equal in all directions, or *anisotropic*, if the motion is restricted in a spatial direction. In the brain, water diffuses preferentially along the main direction of axons or fibre tracts (i.e. a collection of axons), while the diffusion perpendicular to pathways is hindered (Beaulieu, 2002). This directional dependence can be quantified for all spatial locations in the brain using diffusion tensor magnetic resonance imaging (DT-MRI), resulting in several outcome measures such as *fractional anisotropy* (FA; measures directionality of diffusion) or *mean diffusivity* (MD; measures he amplitude of diffusion).

Mathematically, the diffusion is characterized by a diffusion tensor, which contains information on the organization of orientated tissue in space and hence indicates the main diffusion direction. This information can then be used to reconstruct white matter fibre bundles using a technique called DTI *tractography*. Tractography is a non-invasive technique that reconstructs axonal trajectories by measuring the diffusivity of water along different directions and tracing a pathway of least hindrance to diffusion to form continuous pathways (Catani et al., 2007). These are virtually reconstructed in three dimensions and can then be compared across participants or patient groups. One drawback of DTI measures is that they are non-directional, and it is therefore not possible to determine whether a region A projects to a region B, or vice versa, or maybe there is a third region that acts that relays from A to B. Also, there can be a project with crossing fibres (i.e. different fibre bundles that cross at the same location), which can lead to reductions in signal.

DT-MRI has successfully been used to explore the microstructural organization of white matter in a wide range of conditions including schizophrenia (Phillips et al., 2009), epilepsy (Ahmadi et al., 2009; Concha, Gross, Wheatley, & Beaulieu, 2006) and autism (Pugliese et al., 2009).

Neural Correlates of Psychopathology: Insights from Neuroimaging Studies

Autism Spectrum Disorder

Autism spectrum disorder (ASD) is a life-long neurodevelopmental condition affecting approximately 1 % of the population. ASD is characterized by a triad of symptoms in impaired social communication, social reciprocity and repetitive-stereotypic behaviour (Wing, 1997). ASD is accompanied by several co-morbid conditions (e.g. anxiety disorders, depression, attention deficit hyperactivity disorder [ADHD]) and is also associated with ID (Simonoff et al., 2008). There is strong evidence coming from neuroimaging studies that ASD is accompanied by differences in brain anatomy, functioning and connectivity.

In terms of brain anatomy, it has been reported that individuals with ASD have increased brain volume and weight, which affects both grey and white matter. These gross anatomical differences are most prominent during early postnatal life and childhood (age 2–4) and are less apparent during adolescence and adulthood (Courchesne et al., 2001). There is also evidence to suggest that ASD is accompanied by neuroanatomical differences in specific brain regions, which are further associated with variation in clinical symptoms. For example, structural differences in

(1) language areas (i.e. Broca's and Wernicke's area) have been related to impaired social communication and language; (2) frontotemporal regions and the amygdala have been associated with abnormalities in socio-emotional processing and (3) orbitofrontal cortex and basal ganglia may be linked to repetitive and stereotyped behaviours (see Amaral, Schumann, & Nordahl, 2008 for review).

In addition, neuroimaging studies have contributed greatly to our understanding of *brain connectivity* in ASD. For example, it has been shown that there are extensive white matter deficits in ASD suggesting a general breakdown of communication between different areas of the brain (McAlonan et al., 2009). More specifically, it has also been suggested that ASD is a 'developmental disconnection syndrome' where higher-order association areas (e.g. in the frontal lobe) are atypically connected during development (Geschwind & Levitt, 2007). This has not only been demonstrated in sMRI studies, but also in investigations exploring *functional connectivity* (i.e. temporal coherence between brain areas) using fMRI. For instance, it has been reported that functional connectivity of medial temporal lobe structures is abnormal in people with Asperger's syndrome during emotional processing (e.g. processing of fearful faces) (Koshino et al., 2008). In addition, anatomical underconnectivity between frontal and parietal areas affects executive functioning and is accompanied by abnormalities in connecting fibres including the corpus callosum (Just, Cherkassky, Keller, Kana, & Minshew, 2007). Taken together, these studies provide a strong link between the brain functioning, anatomy and connectivity of the brain in ASD and general autistic symptoms and traits.

Lastly, MRS studies suggest that specific autistic symptoms may also be mediated by differences in brain chemistry. For example, Bernardi et al. (2001) recently demonstrated that individuals with ASD showed significantly lower concentrations of glutamate+glutamine (Glx) in the anterior cingulate gyrus and reduced myoinositol (INS) in the left temporoparietal junction (Bernardi et al., 2011). Abnormalities in neurotransmission in additional to atypical brain anatomy and connectivity may therefore underlie the pathogenesis of ASD although the exact aetiology of the condition remains elusive.

Research into the genetics of ASD suggests that ASD is among the most genetically determined psychiatric conditions of developmental and cognitive abnormalities with concordance between identical twins reported at nearly 90 % in some studies (Rosenberg et al., 2009). This high heritability has been linked to several common as well as rare genetic variants, which in turn have a differential impact on the development of the brain in ASD. For instance, many of the autism-linked rare genetic variants (e.g. copy number variations) play a crucial role in synaptogenesis and neuronal differentiation (reviewed in Abrahams & Geschwind, 2010) and hence influence the way the brain is shaped and wired in ASD. However, few of such rare genetic variants are 'causal' for ASD and have also been observed in unaffected individuals. For example, a recent study has also shown that a distinct autism-related CNTNAP2 'risk allele' is associated with reduced functional connectivity in frontal lobe networks, regardless of whether participants were autistic or neurotypicals (Scott-Van Zeeland et al., 2010). Thus, there is strong need for detailed phenotypic studies not only of patients with autism but also of unaffected individuals with more or less autistic traits who harbour such rare potentially causal mutations and their effect on brain anatomy and functioning. Future studies combining neuroimaging and genetic investigations are thus essential in order to elucidate the multifactorial aetiology of ASD.

Attention Deficit Hyperactivity Disorder

ADHD is a developmental disorder that persists into adulthood and is defined by age-inappropriate levels of hyperactivity/impulsivity and attention difficulties. Epidemiological studies of ADHD—and also hyperkinetic disorder—suggest that these disorders may be increased in

individuals with ID. For example, Simonoff et al. (2006) found that there is a negative linear relationship between ADHD symptoms and IQ and concluded that ADHD symptoms are increased in people with ID. Individuals with ADHD also generally show impaired higher-level executive functions of motor and cognitive inhibition (e.g. Rubia, 2011). Several neuroimaging studies have demonstrated that these impairments are mediated by structural and functional differences in the brain.

MRI investigations examining brain structure in ADHD have reported abnormal volume and cortical thickness in several brain regions of the *fronto-striatal* system, including differences in total brain volume, focal differences in prefrontal regions, the basal ganglia, the temporal and parietal cortices and the corpus callosum (reviewed in Nakao, Radua, Rubia, & Mataix-Cols, 2011). These changes most likely reflect a different neurodevelopmental trajectory of brain maturation. For example, children with ADHD show a delay in peak maturation of cortical thickness by 3–5 years, which is most prominent in frontal and temporal brain regions (Shaw et al., 2007). The differences in grey matter are accompanied by atypical white matter connectivity. Evidence coming from DTI studies suggests that, similarly to ASD, there are deficits in several white matter tracts in ADHD (reviewed in Konrad & Eickhoff, 2010) and that atypical connectivity might underlie some of the behavioural deficits observed in ADHD. In addition to structural differences, functional imaging studies have demonstrated reduced brain activation in the fronto-striatal system in ADHD. For example, reduced activation in ADHD has been reported in the inferior frontal cortex, the anterior cingulate, the caudate nucleus and in temporoparietal regions during tasks involving motor response inhibition, inference inhibition and various attention tasks (for review see Rubia et al., 2010). Also, atypical cerebellar activation has been reported by fMRI studies involving attention and timing tasks. Lastly, fMRI studies have shown that individuals with ADHD display a reduced degree of functional connectivity during attention and inhibition tasks, which may also contribute to the level of symptom severity observed in ADHD.

Overall, MRI studies have greatly contributed to our understanding of the neurobiological correlates of ADHD. Most importantly, these studies have demonstrated that neurocognitive deficits observed in ADHD do not only affect isolated brain regions but also the way these regions interact (i.e. are connected) both functionally and structurally.

Schizophrenia

Schizophrenia is characterized by severe perturbations in cognition, affect and behaviour and can also be accompanied by delusions and hallucinations. ID often co-occurs with schizophrenia. Although ID is not one of the core features of schizophrenia, it has recently been estimated that the risk of psychiatric disorder is increased among people with ID. More specifically, current estimates put the risk of schizophrenia in intellectually disabled populations at around 3 %, compared with a lifetime population risk of around 1 % (Hemmings, 2006). The prevalence of schizophrenia among people with ID is also higher than that of bipolar disorder (~1 %) and unipolar major depression (~1 %) (Morgan et al., 2010). There is also some evidence to suggest that individuals with schizophrenia show a progressive cognitive decline and that the cognitive changes with ageing in schizophrenia are also consistent with the hypothesis that schizophrenia is associated with accelerated ageing (reviewed in Kirkpatrick, Messias, Harvey, Fernandez-Egea, & Bowie, 2008). Neuroimaging studies have demonstrated that this cognitive decline is accompanied by excessive accelerated cortical thinning in widespread areas of the cortical mantle (particularly in temporal cortex and frontal areas) and that poor outcome in patients is associated with more pronounced cortical thinning (van Haren et al., 2011). This suggests that neurodegeneration may accompany the cognitive decline observed on the behavioural level.

Recent theorist have also conceptualized schizophrenia as a *neurodevelopmental disorder*, where interactions between genes and environmental factors interact over the during the time course of development and lead to abnormalities in the neural systems underlying the disorder. The concept of a neurodevelopmental disorder has also led to the notion of existing *prodromal stages* of psychosis (i.e. early symptom(s) that might indicate the start of a diseases before specific symptoms occur), which may offer the chance of early treatment and intervention. Here, structural and functional imaging biomarkers might play an important role in the future to identify those individuals that are at risk and/or to predict disease outcomes. Functional and structural MRI studies have identified several neuroimaging biomarkers that could be utilized to monitor the disease progress.

Studies investigating brain functioning in schizophrenia have highlighted two core findings associated with the disease. Firstly, individuals with psychosis consistently show a decrease of functional activation in the frontal lobe, which is also often referred to as *hypo-frontality*. Secondly, individuals with psychosis show an increase of activation in midline structures such as the anterior cingulate cortex. These core findings have been reported across a set of tasks including (1) executive functioning, (2) emotional processing tasks, (3) reward and condition tasks and (4) language tasks. *Executive functioning* includes a diversity of cognitive processes including attention, working memory, context processing and inhibition. Impaired executive functioning in schizophrenia has mainly been linked to reduced activation in the dorsolateral and ventrolateral prefrontal cortex, which have further been associated with inappropriate and bizarre behaviour, affect and emotional processing and formal thought disorders (reviewed in Goghari, Sponheim, & MacDonald, 2010). Schizophrenia is also accompanied by prominent *social and emotional dysfunction*, for instance interpreting the beliefs or intentions of others in order to predict and explain their behaviour. The neural substrate of social functioning in general comprises medial prefrontal regions and the amygdala-hippocampal complex. Impairments in the amygdala and hippocampus also lead to abnormal emotion recognition, a reduced ability of regulating affective states and misinterpreting neutral or ambiguous situations as threatening. Last, schizophrenia has been associated with the striatum (part of the basal ganglia), which may have a role in affective both negative and positive symptoms. Atypical functional activation and connectivity in these areas may therefore mediate several of the social and emotional impairments observed in schizophrenia.

Many of the brain regions displaying atypical functional activation in schizophrenia utilize dopamine as the main neurotransmitter, which has also led to the *dopaminergic hypothesis* of schizophrenia. For example, the frontal cortex, the amygdala/hippocampus and the striatum are part of the mesolimbic pathway, which modulates emotional processing (and reward in particular). Antipsychotic medications targeting the dopaminergic systems are therefore successfully used to ameliorate psychotic symptoms mediated by these regions.

Schizophrenia is also accompanied by several structural anomalies, which accompany the deficits observed on the functional level. Most importantly, schizophrenia is known to be characterized by mainly left-sided reductions in frontal grey matter volume and regional volumetric decreases in the limbic and para-limbic cortices, as well as the thalamus (reviewed in Mueller, Keeser, Reiser, Teipel, & Meindl, 2012). In addition, studies investigating structural connectivity in the brain of patients with schizophrenia show significant reductions in white matter connections of the fronto-striatal system, the corpus callosum and the parahippocampal gyrus. Taken together, the evidence provided by neuroimaging studies has led to significant advances in understanding the neurobiology of schizophrenia. The neural systems associated with the disease and their assessment using imaging technologies may therefore play an important role in diagnosis, predicting and treating the condition in the future.

Velo-Cardio-Facial Syndrome

Velo-cardio-facial syndrome (VCFS) is a disorder caused by a deletion at chromosome 22q11 and is therefore also known as 22q11 deletion syndrome. The manifestations of the disorder are both physically and mentally. For example, individuals with VCFS have a cleft palate, heat defects and a distinct facial appearance. VCFS is also associated with ID, psychosis and other mental illnesses. Interestingly, the learning disabilities commonly observed in children with VCFS are marked within syndrome variability and may vary depending on the mode of inheritance of the deletion.

There is evidence to suggest, that the deletion causes differences in brain structure, which in turn have been linked to specific VCFS symptoms. Traditional volumetric studies employing VBM have reported that individuals with VCFS show reduced amygdala volume, larger basal ganglia and structural alterations in the cerebellum and the fusiform gyrus (e.g. Campbell et al., 2006). These grey matter anomalies are accompanied by reductions in white matter predominantly in the basal ganglia and the cerebellum. More recently, there are also reports of cortical thinning in parieto-occipital regions and inferior frontal gyrus (Bearden et al., 2007) and decreases in cortical gyrification in the frontal, temporal and parietal lobes in individuals with VCFS (Schaer et al., 2006). Due to the high prevalence of psychosis, VCFS has also been suggested to be a useful neurodevelopmental model of schizophrenia. One region that distinguishes between VCFS patients with and without psychosis is the superior temporal gyrus, which is reduced in volume in the schizophrenia group relative to the unaffected VCFS individuals (Chow et al., 2011). Thus, neuroanatomical changes in this brain region might therefore be used as a valuable prognostic biomarker for the development of psychosis in VCFS in the future.

Down Syndrome

Down syndrome (DS) is genetic syndrome resulting from trisomy for human chromosome 21. Individuals with DS show a range of defects in many organ systems, including cardiac malformations, gastrointestinal anomalies and craniofacial and skeletal anomalies. DS is also associated with a high prevalence of ID and age-related cognitive incline of the Alzheimer's type. Overall, insights into the neural correlates of the syndrome come from a few neuroimaging studies and existing mouse models, which are particularly suited to investigate the specific neurobiological mechanisms of the condition. In general, the brain in DS shows a reduction in overall volume, which is proportional to their smaller stature (e.g. Kesslak, Nagata, Lott, & Nalcioglu, 1994). The neuroanatomical abnormalities in DS are most prominent in temporal and frontal lobes (Smigielska-Kuzia et al., 2011), but also include volumetric reductions in the cerebellum and amygdala-hippocampal complex (Pinter et al., 2001). The reductions in regional brain volume in DS are associated with impairments in specific cognitive processes. Grey matter density of the cerebellum and temporal gyrus has been reported to be associated with linguistic measures, while memory performance was predominantly associated with reductions of the parietal, temporal and occipital lobe (Menghini, Costanzo, & Vicari, 2011). There is also evidence for abnormalities in neural connectivity in DS. For example, Horwitz, Schapiro, Grady, and Rapoport (1990) found a reduced degree of functional connectivity between frontal and parietal lobes, which play a major role in visuospatial abilities that are affected in DS (Horwitz et al., 1990). Hence, trisomy for chromosome 21 affects a wide range of structural and functional differences in the brain and also leads to an accelerating ageing process in the brain.

Fragile X Syndrome

Fragile X syndrome (FXS) is one of the most common known causes of cognitive and behavioural disability with a prevalence of approximately 1 in 4,000 individuals. It is caused by mutations of the Fragile X Mental Retardation 1 (FMR1) gene on the X chromosome. Common behavioural problems observed in FXS are attentional dysfunction and hyperactivity, repetitive-stereotypic

behaviours, social anxiety and autistic symptoms (Reiss & Dant, 2003). Neuroimaging studies have demonstrated that the caudate nucleus and the thalamus are enlarged in individuals with FXS, while the cerebellum, amygdala and the superior temporal gyrus are reduced in regional volume (reviewed in Bray et al., 2011). These neuroanatomical differences are most likely the result of an atypical neurodevelopmental trajectory of brain maturation (Hoeft et al., 2010). It has also been shown in a recent longitudinal study that individuals with FXS display aberrant maturation in the prefrontal cortex, which followed a similar developmental trajectory as prefrontal cortex-related measures of cognitive functioning (Bray et al., 2011). In this study, cognitive measures included a verbal fluency task and a task measuring visuospatial abilities, which are impaired in FXS. Structural abnormalities in limbic structures such as the amygdala seem to mediate impaired emotional processing in FXS. For example, reduced amygdalar volume may play a role in abnormal fear responses (Paradee et al., 1999). Also, the volume of the insular cortex—a sensory integrative region that is critically involved in manifesting anxiety—is reduced in FXS and has been suggested to modulate an aversion of eye gaze, which is commonly observed in individuals with the condition (Cohen, Nichols, Brignone, Hall, & Reiss, 2011). Neuroimaging studies therefore suggest that mutations in the FMR1 causes a wide range of brain abnormalities including structural and functional differences, which mediate symptoms observed in FXS. Interestingly, the FXS phenotype varies between sexes as the disorder is X-linked (linked to the X chromosome), and both genders suffer from a varying degree of cognitive impairment. This varying degree of impairment has also been observed in the brain itself. For instance, individuals with higher levels of expression of the FMR1 gene also show more severe behavioural deficits and more atypical levels of functional brain activation (e.g. Rivera, Menon, White, Glaser, & Reiss, 2002). This data suggests that there is a gene-dosage effect, which affects brain functioning and anatomy to a variable degree.

Potential Clinical Applications of MRI

While the traditional aim of neuroimaging was to establish what has often been described as the 'neural correlates' of various mental disorders (e.g. to determine the location and magnitude of changes in brain structure or function in patients vs. controls), traditional techniques offer limited applicability in the clinical setting. This is largely due to the fact that conventional analytical approaches are based on comparisons of 'mean' values between groups (e.g. averaged mean image intensity) so that no assumptions can be made with regards to individuals. Furthermore, traditional approaches do not have any predictive value.

Recent advances in analytical techniques however now make it possible to utilize complex, multivariate biological data in order to make a prediction. In the context of brain imaging, these techniques are also known as 'brain-reading' or 'brain-decoding' methods and belong to a broad group of techniques collectively known as *machine learning*. The basic idea of machine learning is to train a computer algorithm to identify a complex pattern of data that can then be applied on new individuals to make a prediction. Training usually occurs in a well-characterized sample by finding a boundary or 'hyperplane' that best discriminates between different classes (e.g. patients vs. controls). Once the classifier is 'trained', it can then be used to predict group membership of a new test example (e.g. a new individual with currently unknown group membership). A key feature of pattern classification is their potential to detect global, complex and multimodal patterns of brain abnormalities that cannot be efficiently identified with univariate methods (e.g. general linear model). This makes machine learning particularly suited in the search for brain-based disease biomarkers.

So far, machine learning has been applied to a variety of diseases including Alzheimer's diseases (AD), schizophrenia, depression and autism (Ecker et al., 2010). The search for biomarkers is of particular importance for conditions such as ASD, which are currently being diagnosed using behavioural criteria exclusively.

The behavioural diagnosis is, however, often problematic—particularly in adults where coping strategies developed over the lifespan might mask symptoms. Furthermore, the neurobiology of the condition is complex and unlikely to be caused by a single gene or a single brain region. Thus, future ASD biomarkers for psychiatric conditions need to be multivariate and complex as well, encompassing data from different aspects of biology as well as genetics.

Most of the applications of neuroimaging biomarkers are diagnostic (e.g. identifying patients from controls using MRI data). However, pattern classification approaches hold promise not only assisting diagnosis but also for predicting treatment outcomes and disease progression (e.g. responders vs. nonresponders). For example, it has been demonstrated that classifiers are sufficiently sensitive to separate patients with AD and mild cognitive impairment (MCI) from neurotypicals (reviewed in Kloppel et al., 2011). These classifiers can then be used to identify those individuals that will develop MCI and AD in the future. Also, it was shown in depression that structural MRI features are predictive of patient's clinical response to antidepressant medication at an accuracy of 89 % (Costafreda, Chu, Ashburner, & Fu, 2009). Thus, machine learning will have important clinical applications in the future and allow for more specifically tailored treatment strategies.

The MRI-assisted diagnosis of certain conditions IS of particular interest in ID, as the conventional diagnostic assessment is often problematic due to impaired cognitive functioning. For example, it is difficult to distinguish between general LD and low-functioning individuals with ASD. The existence of imaging biomarkers would therefore be invaluable to assist the conventional diagnostic process. Thus, while the search for complex biomarkers is still in its infancy, the availability of new analytical techniques with high exploratory power and predictive value offers promising new ventures into finding a biomarker whose complexity equals the aetiology and phenotype of many psychiatric conditions. If successful, such a biomarker (or a set of biomarkers) might one day prove invaluable in diagnosing and treating mental health conditions.

Conclusions

Over the last 2 decades, neuroimaging has proven an invaluable tool to investigate brain functioning and anatomy in a wide range of disorders, including genetic and psychiatric conditions with ID. In the future, such imaging biomarkers might also be used in the clinical setting in order to assist the conventional clinical diagnosis and/or to predict response to treatment and intervention. Although large clinical trials are yet required to explore the sensitivity and specificity of these techniques in the clinical setting, neuroimaging may provide an invaluable tool in the future towards a more individual approach to treatment and personalized intervention. Currently, these techniques are extensively being explored in the research setting and are increasingly more often used in clinical practice. While it is unlikely that neuroimaging techniques will make the need for conventional neuropsychological and quantitative psychiatric, physical and metabolic examinations obsolete, they can provide important additional information when the diagnostic process is complicated, such as in individuals with ID. In addition, techniques such as MRS are important research tools to not only investigate the neurobiology of psychiatric conditions but also to provide new pharmacological targets, which may be used to develop treatments. Finally, neuroimaging may also be used to exclude other neurological conditions prior to the conventional diagnosis and should therefore be applied routinely in any psychiatric assessment.

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Epilepsy

7

Robert Winterhalder and Howard Ring

Introduction

Seizure disorders can compromise cognitive, behavioural and emotional functioning in people with ID through a variety of mechanisms. These may include processes related to the aetiology of the seizure disorder itself or its management, or to the physiological, psychological or interpersonal consequences of seizure activity. Such is the importance of antiepileptic drug (AED) effects on cognition that this topic merits particular attention. The relationship between epilepsy and psychiatric disturbance is also important as well as being complex and variable. However, as is the case elsewhere in the healthcare of people with ID, careful and thorough history taking, which must include the gathering of information from those who know the individual well, generally provides sufficient information to provide the basis of an understanding of the presenting issues and the development of a management plan. It is important to remember, however, that changes in seizure frequency in response to therapeutic interventions frequently take weeks to months to become apparent and so ongoing monitoring following treatment is essential. In terms of the nature of potential therapeutic interventions, management of cognitive and psychiatric symptoms using psychosocial and pharmacological approaches may also improve seizure control (although it is important to remember that some psychotropic agents may lower seizure threshold, increasing risk of seizures), whilst improvements in seizure management are likely to reduce psychiatric and behavioural pathology. In terms of the evidence base that clinicians can draw upon when called to manage psychiatric and cognitive disorders in people with ID and epilepsy, though most of the published literature refers to the general population with epilepsy, there is now increasing research that includes people with ID.

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Cognitive Dysfunction in Epilepsy

Findings from Studies of the General Population with Epilepsy

Factors affecting neuropsychological functioning include the aetiology of the ID and of the epilepsy, epilepsy syndrome, age at onset, seizure type and frequency and duration of seizure disorder. There are various mechanisms in epilepsy which can lead to cognitive impairment including structural lesions, neuronal loss, ictal and subictal activity, neuronal dysfunction, AED side effects and comorbid psychiatric (especially mood) disorders. The impact on cognitive function

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also depends on whether the epilepsy arises from a localised focus or is generalised, and the precise location of an epileptogenic focus. For instance, damage to the hippocampus is likely to cause a disproportionate impairment in memory function. It remains equivocal whether neuropsychological impairments may deteriorate over time and what factors are important in their development and progress (Baker, 2010).

The key neuropsychological functions most commonly affected are psychomotor speed, vigilance, memory and mood (Thompson & Duncan, 2005). Left-hemisphere seizures are associated with impaired verbal function, whereas right-hemisphere seizures show visuoperceptual, visual memory and constructional impairments (Lezak, Howieson, & Loring, 2004).

Cognitive impairment may be more debilitating than the actual epileptic seizures (Aldenkamp, 2006). Neuropsychological impairments have been identified in patients with epilepsy at the time of diagnosis, before commencing AEDs, or after only a few seizures, which suggest that the underlying lesion, and not AEDs, is responsible (Taylor et al., 2010). Verbal memory performance was affected more in newly diagnosed adult patients with left temporal lobe epilepsy (TLE) than in those with chronic TLE. However, there was no further deterioration in verbal memory performance in the newly diagnosed group during the 5-year follow-up period. Memory impairment was not associated with the aetiology of epilepsy or hippocampal volumes, but was associated with early onset of epilepsy and secondary generalisation in newly diagnosed patients. This suggests that the memory problems observed in patients with chronic epilepsy cannot be attributed solely to AED side effects or to the cumulative effects of recurrent seizures (Aikia, Salmenpera, Partanen, & Kalviainen, 2001). However, direct effects of seizure activity on memory functioning have also been demonstrated (Dodrill, 2004).

Focussing specifically on memory, the degree of impairment in epilepsy correlates with pathological and neuroimaging abnormalities, age of onset of epilepsy, seizure frequency and lifetime number of seizures. Left TLE is associated with greater deficits in verbal memory, whilst right TLE, to a lesser extent, is associated with impairment of non-verbal memory. Subjective complaints of memory impairment do not correlate with objective measures—standard neuropsychological testing of memory typically assesses the ability to retain new information over a delay of half an hour. In a recent review article (Butler & Zeman, 2008), three forms of memory impairment associated with epilepsy were reviewed with a particular emphasis on TLE. In transient epileptic amnesia (TEA), the ictal event is a transient amnesia, whilst other cognitive functions remain intact, and the patient also commonly complains of inter-ictal memory problems, although performance on standard tests of memory is usually normal. TEA was not usually associated with clinically detectable focal brain lesions, but when present they involved the medial temporal lobes. In accelerated long-term forgetting (ALF), patients learn and retain information normally but there is an unusually rapid rate of forgetfulness over the next few days or weeks. Contributory mechanisms included clinical/subclinical seizure activity and structural or other underlying brain pathology. However, AED side effects or adverse psychosocial factors were not implicated. In *remote memory impairment*, there is patchy loss of autobiographical memories extending back over many years. Contributory mechanisms were similar to those for ALF (Butler & Zeman, 2008). Whilst this article does not refer specifically to the ID population, these memory problems may be present particularly in patients with milder forms of ID.

Low mood may lead to memory impairment, at least in part due to reduced motivation and poor attention. The most common comorbid mood disorder associated with epilepsy is depression (Kanner, 2007). Low mood may be an iatrogenic side effect of AEDs, although there are other possible mechanisms including endocrine or metabolic effects, seizure-related mood change including post-ictal depression, psychological response to having epilepsy, societal stigma and incidental comorbid psychiatric disorder.

In a study addressing neuropsychiatric morbidity in focal epilepsy, a significant association

between depressive symptomatology and non-lesional focal epilepsy was observed (Adams et al., 2008). There were no significant differences in prevalence of neuropsychiatric disorders between groups with TLE and those with extra-temporal lobe epilepsy. This may suggest that this observation is due to defects in a diffuse epileptogenic area (Adams et al., 2008). Focal lesions may completely interrupt a neuronal connection at the site of the lesion, whereas diffuse pathology may result in more widespread disruption of, for example, frontolimbic pathways.

Findings from Studies of People with ID and Epilepsy

As the degree of ID increases, so does the likelihood of epilepsy—whilst 5 % of patients with mild ID have epilepsy, this increases to 50 % in those with profound levels of ID. Epilepsy and ID may also be caused by the same underlying disorder. Less common associations include head trauma (accidental or deliberate) leading to epilepsy, and severe uncontrolled epilepsy, particularly in infancy and childhood, interfering with neurocognitive development. The above are set against a background in which both conditions are also associated with low socio-demographic status and, particularly in the past, institutionalisation.

Cognitive functioning in patients with ID and epilepsy is influenced by several factors. Greater severity of ID, longer bedtime periods, poor sleep efficiency, seizure frequency and AED polytherapy have been found to be significant predictors of day time arousal and attention during inter-ictal states in people with epilepsy and severe ID. This study was particularly impressive as it not only studied individuals with severe ID but included ambulatory and sleep EEG monitoring. Inter-ictal cognitive function appeared to be mediated by a combination of organic, circadian (sleep/wake), clinical and pharmacological factors (Espie et al., 1999).

Age of seizure onset, particularly before 5 years, is a significant risk factor for eventual low IQ. Increased tonic-clonic seizure frequency is associated with reduced IQ test performance. Additionally, individual tonic-clonic seizures may produce attentional slowing in the post-ictal phase for up to 36 h (Aldenkamp & Bodde, 2005).

Epileptic encephalopathies such as severe myoclonic epilepsy of infancy (Dravet syndrome), Lennox–Gastaut syndrome and West syndrome are all associated with significant ID. Whilst cognitive impairment often occurs in cryptogenic and symptomatic epilepsy syndromes, it can also be associated with idiopathic generalised epilepsy. Significant impairment in attention has also been reported in patients with childhood absence epilepsy (Levav, 1991). Impairment of frontal lobe function affecting cognitive skills such as planning, reasoning, flexibility and focussing of attentions has also been reported in juvenile myoclonic epilepsy (Devinsky et al., 1997).

Neurocognitive Side Effects of Antiepileptic Drugs

Findings from Studies of the General Population with Epilepsy

All current AEDs may be associated with impairment of neuropsychological functioning. Sodium valproate, carbamazepine, lamotrigine and gabapentin are less likely, and phenytoin and phenobarbitone more likely, to cause neurocognitive impairment. The newer drugs topiramate and zonisamide may also cause significant effects on working memory (Baker, 2010), verbal IQ, verbal fluency and learning, and dysphasia and anomia have also been observed (Lodhi & Agrawal, 2012).

Cognitive side effects may be due to the GABAergic properties of some AEDs. Sodium channel blockers have the least cognitive side effects and may have a positive effect on learning and psychomotor speed. Lamotrigine also has an anti-glutamatergic effect which may promote learning and memory (Lodhi & Agrawal, 2012). Levetiracetam may have positive effects on cognition—in particular, improved reaction times, tapping rate of non-dominant hand and memory for simultaneously presented words (Neyens, Alpherts, & Aldenkamp, 1995).

Findings from Studies of People with ID and Epilepsy

There are numerous difficulties in assessing cognitive side effects of AEDs in patients with ID and epilepsy. These include the heterogeneity of seizure syndromes, differing aetiologies of ID, the presence of pre-existing cognitive deficits, impaired communication, comorbid medical (including neurodevelopmental) and psychiatric disorders and polypharmacy.

Older AEDs, e.g. phenytoin and barbiturates, have been associated with encephalopathy, cognitive impairment and cerebellar syndrome; clonazepam and clobazam have been associated with drowsiness, reduced concentration, hyperactivity and irritability. A series of reviews, which are summarised below, of more frequently currently used AEDs in ID included topiramate, lamotrigine, gabapentin, carbamazepine and valproate.

Topiramate has been associated with a tendency to neuropsychiatric side effects, e.g. somnolence, psychomotor slowness, nervousness, confusion and word-finding difficulties, when used in polytherapy, which reduced when it was used in monotherapy. However, caution over its use has been recommended if there is a history of psychosis, being underweight or personal/family history of renal stones (Kerr & Merrick, 2000).

A review of lamotrigine highlighted that side effects such as diplopia or dizziness may lead to distress and behavioural disturbance, particularly when co-administered with carbamazepine, through a pharmacodynamic interaction. Lamotrigine by treating subtle or overt seizures may lead to 'release phenomena' as patients become more alert and active, with behavioural problems developing. Most authors suggest an improvement in behaviour in ID through these alerting and mood-elevating properties (Besag, 1998). Lamotrigine had a more favourable cognitive profile compared to topiramate, which affected cognitive function, particularly attention and verbal function (Aldenkamp, De Krom, & Reiif, 2003).

Carbamazepine demonstrated minimal cognitive or behavioural side effects. Adverse behaviour seemed more frequent in people with brain damage, particularly those with pre-existing abnormal behaviour. Overall there were improvements in cognitive functioning. Cognitive effects may sometimes be secondary to hyponatremia (Waisburg & Alvarez, 1998). Eslicarbazepine significantly reduces this risk.

A study in children with refractory partial seizures treated with gabapentin revealed behavioural side effects were significantly more common in ID patients and patients under the age of 10 years. There was no relationship between the severity of ID and occurrence of side effects. Nine percent of subjects had positive behavioural effects and appeared happier and brighter (Mikati et al., 1998).

Significant experience has been gained with valproate in the treatment of West syndrome and Lennox–Gastaut syndrome—cognitive dysfunction was reported as a side effect (Fris, 1998).

A Cochrane review concluded that the majority of studies addressing behavioural exacerbations were unhelpful due to lack of or unreliable measures, although when measured there was little obvious negative impact on behaviour (Beavis, Kerr, & Marson, 2008). Several studies were identified which addressed cognitive issues. Gabapentin showed a significant improvement in co-operation and restlessness. There were significant differences between gabapentin and lamotrigine in communication, co-operation and restlessness, but both were similar in reducing levels of challenging behaviour (Crawford, Brown, & Kerr, 2001). There was a trend towards significance in improvement of the ELDQOL (Epilepsy and Learning Disabilities Quality of Life) behaviour subscale score for patients on topiramate compared to placebo (Kerr, Baker, & Brody, 2005). Lamotrigine was superior to placebo in benefitting behaviour and alertness even without concomitant seizure reduction (Eriksson, Nergardh, & Hoppu, 1998). Drowsiness, fatigue, anxiety, aggression and 'personality disorder' were amongst the side effects noted for topiramate (Sachdeo et al., 1999).

It is important to remember that AED-related adverse effects on a range of physiological processes may have implications for cognitive functioning. A study of 281 patients with ID receiving

AEDs found significant correlations between hepatic, dermatological, respiratory and electrolyte disturbances and cognitive impairment (Sipes et al., 2011). This emphasises that when using AEDs in people with ID, who may be unlikely or unable to draw clinicians' attention to possible adverse effects directly, overall physical state should be kept under review.

Psychiatric Disorders in Epilepsy: Prevalence, Aetiology and Presentation

Prevalence

Over the years there has been much debate as to whether rates of psychopathology are increased in those with epilepsy compared to the rest of the population. For those with epilepsy and an IQ in the normal range, the consensus appears to be that whilst rates of comorbid psychiatric illness are not raised in those whose epilepsy is well controlled, in the minority whose epilepsy is treatment refractory, there is an evidence of increased rates of psychiatric morbidity. In the population with ID, the situation is more complicated and has been less researched. Relevant factors to consider are the observations that rates of psychiatric symptomatology are in general increased in people with ID, as is the prevalence of epilepsy. It has also been reported in several studies that epilepsy in those with ID is more likely to be treatment refractory than epilepsy in the rest of the population. Hence, whilst it might be predicted that psychiatric comorbidities are more prevalent in those with epilepsy and ID than in those with epilepsy and an IQ in the normal range, it is more difficult to make definitive statements about the relative frequencies of such comorbidities in those with ID and epilepsy compared to those with ID alone.

However, several community-based studies have produced estimates of the prevalence of some psychiatric comorbidities in adults with ID and epilepsy. In a Scottish population, Espie et al. (2003) reported affective or neurotic disorder in 29 % of their sample and a psychotic disorder in 10 %. Ring, Zia, Lindeman, and Himlok (2007), in a study of adults with ID in one region of England, observed affective disorder in 26 % and possible psychosis in 11 %. In both these studies the rates of psychosis observed were lower than the 24 % reported in a Japanese population by Matsuura et al. (2005). This wide range of prevalence of psychosis may in part relate to differences in the diagnostic processes and definitions employed in these studies and also to differences in participant ascertainment. However, even the lower figure of 10 % is appreciably greater than the 4 % rate of clinically diagnosed psychotic disorders identified by Cooper, Smiley, Morrison, Williamson, and Allan (in a community sample of adults with ID but no epilepsy) (2007a). Likewise, the rates of affective disorder that have been reported in adults with ID and epilepsy (26–29 %) are appreciably higher than the rate of 7 % reported by Cooper et al. (2007a).

Up to a point, such discussions over the reasons for differences in prevalence rates between studies in those with ID resemble equivalent discussions regarding studies in the general population. However, unlike the rest of the population with epilepsy, but in common with those who have ID but no epilepsy, it has been reported that prevalence of psychopathology in adults with ID and epilepsy varies according to IQ (Adachi et al., 2002; Ring et al., 2007). The basis for such a relationship is likely to include both practical difficulties in identifying psychiatric symptoms in those with more severe LD, for instance, related to communication difficulties as well as conceptual issues regarding the role of intellectual and biological brain development in determining the nature of psychopathology. For instance, in patients with LD but no epilepsy, Holden and Gitlesen (2004) reported increased rates of both depression and psychosis in those with moderate compared with those with severe or profound LD and concluded that prevalence of psychiatric illness decreases with increasing severity of LD. More recently, however, in a community-based epidemiological study, Cooper, Smiley, Morrison, Williamson, and Allan (2007b) did not find a relationship between prevalence of depression and severity of LD.

Aetiology and Presentation of Psychopathology in People with ID and Epilepsy

Discussion of this topic is limited by the generally poor understanding of the aetiology of psychopathological symptoms in the population at large. In the absence of definitive genetic or biochemical explanations of psychopathology, consideration of aetiological mechanisms is likely to be speculative, albeit often arising out of careful clinical observation and investigation. Processes that have been suggested as potentially underlying associations between epilepsy and psychopathology include direct and developmental consequences of epileptic brain activity on inter-ictal and post-ictal psychiatric states, AED effects and the psychosocial consequences of living with epilepsy.

A seizure has generally been considered as an event defined in time with a start and a finish and between individual seizures a period of time of anything from a minute or less to a number of years. The stages in which seizure-related phenomena are considered are the prodrome, the aura, the ictus itself and the post-ictal period—though not all seizures or all individuals will manifest each of these stages. There is some evidence that psychopathological states emerging at different stages in this progression may have differing aetiologies and different emotional symptoms have been described in association with different stages in the evolution and dissolution of observable seizures.

Prodromes may present as distinct changes in mood or behaviour before an epileptic event. During a prodrome—which may vary in length between a few minutes and perhaps a day or more—symptoms of depression and irritability have been well described. Conversely, seizure auras, generally reported as occurring in people with focal onset epilepsy, often with a focus in the temporal lobe, may be associated with a range of brief (generally lasting for less than a minute) psychic phenomena, including visual, olfactory or other sensory experiences, or an altered emotional state—most often a brief feeling of intense fear. In those with adequate communication skills, it may be possible for them to explain what is going on and for those around them to provide explanation and possibly management to reassure or perhaps reduce these experiences. However, in people with more severe ID, the diagnosis of such experiences may be much more problematic, leading to difficulties in offering appropriate management, possibly with the resulting generation of more behavioural manifestations of distress.

During the seizure (also known as the ictus) itself, if this takes the form of a generalised seizure, particularly if accompanied by tonic-clonic movements or characterised by dropping to the ground, the diagnosis should be clear and the individual will not exhibit other behavioural symptoms during the ictus. However, in other types of seizure, particularly those with focal onset, a variety of motor or vocal manifestations may be confused by observers for apparently purposeful actions. In addition, during some focal seizures, particularly if these are sustained in duration or occurring in clusters with limited recovery between individual seizures, individuals may experience symptoms of confusion, irritability or psychosis which may be difficult to diagnose as epileptic in nature.

The period after a seizure—often termed the post-ictal period, lasting from minutes to several hours or occasionally longer—is relatively frequently associated, in those with IQ in the normal range, with symptoms of depression. Post-ictal depression is often relatively brief—usually persisting for no more than a couple of hours—and is often experienced as intrusively sad. Strong feelings of unhappiness, 'feeling awful' and crying are reported, rather than the 'empty, flat' feelings described during inter-ictal states of depression. In some people post-ictal depressive states may be associated with suicidal thoughts. Whilst generally of short duration, occasionally post-ictal mood disturbances may last for days or weeks. In those with limited communication skills, it may be difficult to diagnose such a state but being aware of it as a possibility should assist in developing a differential diagnosis in the presence of more sustained periods of post-ictal disturbances of behaviour.

Behavioural, affective and psychotic symptoms may also emerge during the interval between seizures (the inter-ictal period). In these circumstances, there may well be no direct biological link between brain seizure activity and apparent psychopathology. There is a large research literature, mostly from the population with IQ greater than 70, describing the effects of living with epilepsy on social and emotional state. Many of the relevant factors, including adverse effects of AED treatment, social isolation, overprotective care by families or other carers and social stigma, are also highly applicable to the lives of people with ID and epilepsy. These factors should always be considered by clinicians when considering psychopathology presenting in somebody with epilepsy.

In addition, however, in some individuals, there are occasions in which inter-ictal symptoms of depression, irritability or psychosis—lasting anywhere from days to weeks—may develop in association with a reduction in seizure frequency or temporary cessation of all seizures, possibly with associated reduction in epileptiform EEG if this is measured at that time. Such relationships have been described as 'alternative psychosis' or 'forced normalisation' (Krishnamoorthy, Trimble, Sander, & Kanner, 2002). Views differ as to possible explanations of these apparently reciprocal relationships between epilepsy severity and emergence of psychopathology. Whilst some experts consider them coincidence or to arise because the individual and those around them are no longer pre-occupied with the epilepsy, others consider that there may be a mechanistic association between reduction in seizure activity and the development of psychiatric symptoms. Regardless of the basis of the observed relationship, it is sometimes noted that the abnormal mental phenomena resolve as seizures re-emerge and the EEG returns to a more epileptiform pattern, as in the case reported by Krishnamoorthy et al. (2002).

Finally, of particular interest are non-epileptic seizures which may be psychogenic or non-psychogenic. The ID population is at increased risk of both epilepsy and psychiatric disorder. The diagnosis of psychogenic non-epileptic seizures (PNES) in ID can be difficult due to the presence of stereotypies, behavioural disorders, etc. In addition, deep-seated frontal lobe seizures may present with paroxysmal episodes of bizarre behaviour, which may not be recognised as being epileptic in origin.

In a comparison study between patients with and without ID who experienced PNES, higher proportions of the ID group had epilepsy as well as PNES and were also taking AEDs at the time of diagnosis of PNES. Fewer patients with ID had a history of sexual abuse. A higher proportion of the ID group had previous pseudostatus and immediate situational or emotional triggers prior to their attacks. There were trends towards more men in the ID group and a longer delay between onset of PNES and diagnosis (Duncan & Oto, 2008).

In a comprehensive review of PNES, ID was considered a predisposing, and low IQ a perpetuating, aetiological factor (Reuber, 2005). Whilst patients with mild ID may be overrepresented, PNES should not be confused with stereotypic behaviours seen in more severely affected ID patients (Russell, 2006).

To summarise the psychiatric phenomena that occur at the different stages of seizure manifestation, whilst irritability occurs in all stages, the subjective experience of sadness (depression) is less common during the aura or the seizure itself, but much more common during the post-ictal period. Psychotic phenomena, though less common and often relatively brief and fragmentary, may occur at any of these stages. However, symptoms of elevated mood (such as elation, grandiosity and disordered thinking associated with flight of ideas) occur only rarely at any stage.

Management of Cognitive Dysfunction in Epilepsy

This depends on the aetiology of the neuropsychological impairment—improved seizure control and reduced sub-ictal activity may lead to significant improvement in cognitive functioning. In the case of iatrogenic side effects, reducing the dose or withdrawing the offending AED

may be sufficient. Treatment of accompanying low mood or anxiety, with an antidepressant drug (e.g. an SSRI) or other anxiolytic, may also improve neuropsychological performance.

Several AEDs have accompanying mood-stabilising properties, and pregabalin is licensed as an anxiolytic. Vagal nerve stimulation is not only an effective treatment in epilepsy but is also recognised as a treatment option in depression, with consequent improvement in related memory problems. Epilepsy surgery may also improve cognitive functioning including memory.

Assessment and management of comorbid cognitive dysfunction requires a multidisciplinary approach. Where possible, baseline measures of cognitive function and mental state are invaluable for the early detection of developing cognitive (and psychiatric) problems. Repeat investigations (e.g. serial EEGs) and cognitive/psychiatric assessments will help guide treatment decisions and quantify responses to treatment.

Management of Psychiatric Disorders in Epilepsy

Consideration of how psychiatric symptoms are manifest in relation to different aspects of a person's epilepsy provides some guidance to management approaches. In all cases, current AED therapy should be considered from the viewpoints of whether it is causing psychiatric or behavioural side effects (Schmitz, 2006) as well as the extent to which it is controlling the epilepsy. Some AEDs may cause behavioural effects at therapeutic doses whilst others may lead to symptoms including confusion and irritability if doses are too high. Symptoms arising in the context of seizure prodrome, aura or ictus, or brief periods of post-ictal depression may be managed by optimising seizure control. However, more sustained behavioural or affective disturbances in the context of sustained post-ictal depression or psychosis, or inter-ictal psychopathology are likely to require specific psychotropic and non-psychotropic interventions. It is important to consider both direct effects of any prescribed agents on seizure threshold as well as risks of drug interactions with concomitant AEDs.

Pharmacological Management

Depression has been associated with AEDs such as the barbiturates, topiramate and phenytoin. Underlying depression and anxiety symptoms may be exacerbated by levetiracetam, whilst psychotic symptoms, albeit rare, have been reported with topiramate, levetiracetam and zonisamide (Ettinger, 2006). Rationalisation of the patient's antiepileptic regime may be sufficient.

It is also important to consider the possible seizureogenic effects of psychotropic drugs. Alper, Schwartz, Kolts, and Khan (2007) compared seizure incidence amongst active drug and placebo groups in psychopharmacological clinical trials between 1985 and 2004 in the United States against the background of published rates of unprovoked seizures in the general population. Neither first-generation (traditional) neuroleptics nor tricyclic antidepressants, with the exception of clomipramine, were included in this study. Amongst the neuroleptics, clozapine, olanzapine, and to a lesser extent quetiapine were associated with an increased seizure incidence. Aripiprazole, ziprasidone and risperidone did not increase seizure frequency.

The incidence of seizures was significantly lower amongst patients on antidepressant medication compared to placebo. Antidepressants included citalopram, escitalopram, fluoxetine, paroxetine, sertraline, mirtazapine, duloxetine and venlafaxine. Anxiolytic drugs included buspirone. An increased seizure incidence was observed however for clomipramine.

The more recent antidepressants may have antiepileptic effects, given the association between epilepsy and depression. The inhibitory neurotransmitter gamma-aminobutyric acid (GABA) plays a significant role in both depression and epilepsy. Post-ictal psychosis is associated with a family history of mood disorder (Alper et al., 2007). Postmortem examination of depressed patients reveals hippocampal atrophy (Stockmeier et al., 2004), which is commonly seen in epilepsy series. Antidepressants increase hippocampal cell proliferation and neurogenesis. Imaging studies report common deficiencies of serotonergic transmission in depression and

epilepsy, and in animal models, antidepressant drugs typically elevate extracellular serotonin in the hippocampus and cerebral cortex which is associated with an anticonvulsant effect.

Traditional neuroleptics such as trifluoperazine and haloperidol, together with sulpiride, are indicated in the treatment of psychotic disorders (although the management of forced normalisation may simply require reducing the dose of the relevant AED). Chlorpromazine should be avoided where possible. Amongst the atypical neuroleptics, risperidone is considered reasonably safe. The SSRI group of antidepressants and moclobemide are considered good choices for the treatment of depressive and anxiety disorders. Tricyclics should generally be avoided and care is recommended when using lithium (Taylor, Paton, & Kapur, 2012).

Several AEDs also have psychotropic properties. Lamotrigine and vagal nerve stimulation have antidepressant actions. Carbamazepine, valproate, lamotrigine and possibly oxcarbazepine have mood-stabilising properties. Pregabalin is also licensed for the treatment of generalised anxiety disorder, whilst gabapentin and tiagabine also have anxiolytic benefits.

Non-pharmacological Management

Approaches include appropriate education and training for carers together with psychoeducation for the individual if appropriate. A recent Cochrane review (Ramaratnam, Baker, & Goldstein, 2011) assessed whether psychological approaches to the treatment of epilepsy could lead to a better quality of life and/or reduce seizure frequency. Because of methodological deficiencies and the limited number of patients studied, there was insufficient evidence to support the use of psychological treatments. Two trials of cognitive behavioural therapy (CBT) were effective in reducing depression in people with epilepsy and a depressed affect, whilst a third was not. Two trials of CBT found improvements in quality of life scores. The use of relaxation combined with behaviour modification was beneficial for anxiety and adjustment in one study. One study using EEG biofeedback improved cognitive and motor functions in individuals with a greater seizure reduction. No studies designed to address psychological interventions in patients with epilepsy and ID were identified—indeed ID was often an exclusion criteria.

Whilst the aetiology of mental health problems including depression and anxiety is likely to be multifactorial, an understanding of their epilepsy may help patients cope with this disorder. A video-based training package has been designed for people with ID (Paul, 1996). There is evidence to suggest that this type of approach can increase a patient's understanding of their seizure disorder (Clark, Espie, & Paul, 2001). This may in turn empower the patient and reduce any related depressive or anxiety features they may have.

Self-induction of seizures is a behaviour which may be observed in children and people with ID, possibly as a learned avoidance response or because some aspects of the seizures themselves may be subjectively experienced as intrinsically rewarding. A behavioural approach, e.g. distraction techniques or avoidance of reinforcement, is indicated, together with a broader review of the individual's environment, activities, sensory needs, communication and general health to exclude, for example, sensory impairment which may be contributing to the behaviour.

Neuropsychiatry of Epilepsy: Special Populations

Autism and Pervasive Developmental Disorders

When considering comorbidities between autism and other conditions, including epilepsy and ID, it is important to bear in mind that, as commented on by Berg and Plioplys (2012), in some circumstances the identification of features of autism does not necessarily imply that a diagnosis of autism is also present, particularly given the move away from a narrow definition of classical autism to a broader range of symptoms and traits in the context of an autism spectrum that may

include people with more severe as well as people with less severe impairments than might previously have been considered for such a diagnosis.

Nevertheless, there is good evidence that autism and epilepsy occur together more often than would be expected by chance. When the nature of this relationship is looked at more carefully, it has been determined that rates of such co-occurrence are indeed raised in children who along with their epilepsy also have ID or additional neurological problems, with 16 % of such children also having a pervasive developmental disorder, but co-occurrence rates are not raised in children with uncomplicated epilepsy (Davies, Heyman, & Goodman, 2003). Supporting the same conclusion that disturbances of brain development are a risk factor for having both epilepsy and autism, it has been reported that in those with autism and ID, up to 21 % may have epilepsy whilst in people with autism but no ID epilepsy rates were 8 % (Amiet et al., 2008).

With respect to the question of whether any particular seizure type is more likely to be associated with autism, whilst many seizure types have been reported in those who also have autism, it has been suggested that the relationship is strongest with seizures of focal onset with a focus in the temporal lobe (Matsuo, Maeda, Sasaki, Ishii, & Hamasaki, 2010). Currently whilst some reports suggest that epilepsy tends to predate the development of autism, other studies find the reverse, and there is no evidence that epilepsy can result in the development of autism. Rather, individuals ascertained on the basis of epilepsy are more likely to also have features of autism if they have younger onset of seizures, lower IQ and more severe epilepsy whilst those ascertained on the basis of autism who are also found to have epilepsy are more likely to have evidence of more severe autism, including more cognitive, motor and language deficits (Tuchman, Moshe, & Rapin, 2009). Hence it appears more likely that both autism and epilepsy, when they co-occur, arise from some primary disturbance of brain development that also results in a low IQ. However, as pointed out by Stafstrom, Hagerman, and Pessah (2012), the heterogeneous aetiologies of autism and epilepsy make it unlikely that a single common mechanism could explain seizure predisposition in both disorders.

Down Syndrome

In down syndrome (DS) a tri-phasic (Pueschel, Louis, & McKnight, 1991) distribution of seizure frequency, during infancy, early adulthood and in later life (50 years +), has been suggested. Others suggest a bimodal distribution with peak incidence during infancy/childhood and a second peak in later life (Veall, 1974).

In the early-onset group, infantile spasms in particular are more frequent than in the general population (Goldberg-Stern et al., 2001). The cause of seizures is often unknown but may in some cases be due to head injuries, underlying congenital cardiac disorders or recurrent infections (Stafstrom, Patxot, Gilmore, & Wisniewski, 1991). The second peak in early adulthood could be due to ongoing myelination in the central nervous system, whereas the significant increase in late-onset epilepsy is due to Alzheimer's disease.

Later in the dementia process as epilepsy develops, myoclonic seizures are frequently seen—Late Onset Myoclonic Epilepsy in DS (LOMEDS) (Möller, Hammer, Oertel, & Roenow, 2001). This seizure disorder resembles another type of epilepsy—Unverricht-Lundborg disease, whose gene is also located on chromosome 21, which suggests that LOMEDS may be a distinct type of epilepsy caused by excess genes on the third chromosome 21.

Valproate and levetiracetam are the drugs of choice, although cognitive side effects including sedation may be a problem even at relatively low doses, given the patient's already comprised cognitive reserve. Anecdotal commentaries in the literature (Scheepers, 2010) suggest that early intervention with acetylcholinesterase inhibitors may have a prophylactic effect on the development of myoclonic seizures in LOMEDS. If an evidence base develops to support this observation, this will add a further clinical dimension to decision making around when acetylcholinesterase inhibitors should be withdrawn.

Conclusion

Patients with epilepsy and ID are at increased risk of developing additional cognitive dysfunction and psychiatric disorder. Either disorder may further compromise the quality of life of individuals already coping with long-term disabilities. Because of diagnostic overshadowing, it is possible that such additional complications may go unrecognised and untreated. It is therefore important that clinicians working in services for people with ID are aware of these comorbid disorders and routinely assess patients for their presence. The complex relationship between cognitive functioning, epilepsy, AEDs, psychiatric disorder and psychotropic medication requires an understanding of the various mechanisms that may be operating in any particular individual in order to formulate an appropriate management plan.

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Behavioral Phenotypes and Genetic Syndromes

8

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Behavioral Phenotypes

The term behavioral phenotype refers to a characteristic pattern of social, linguistic, cognitive, and motor observations consistently associated with a biological/genetic disorder (O'Brien & Bevan, 2011). Understanding the genotype associated with behavioral phenotypes (and vice versa) is imperative because it may lead to new or more specific treatment options for mental or behavioral disorders in this and other populations. It may also help us to better understand typical behavior. However, genotype–phenotype relationships are often quite complex, and although many behavioral phenotypes are driven by the core genetic anomaly associated with the ID (i.e., loci of major effect), the effects of multiple genetic modifiers and other genes may also be relevant. Gene–environment interactions are likely to have an important role in the behavioral presentation of patients with ID, although these interactions remain poorly understood.

The behavioral phenotypes that are particularly of interest in ID syndromes are extreme phenotypes and those that are rare or absent in the general population—such as non-volitional self-injury associated with Cornelia de Lange and Lesch–Nyhan syndromes (see below). Behaviors with "high penetrance" (i.e., occurring in most people with the syndrome) such as the abnormal eating behavior associated with Prader–Willi syndrome (PWS) are also of interest, as well as differences between syndromes in the presentation of disorders such as autism spectrum disorders (ASD). Detailed exploration of within-syndrome phenotypic variation has been used to show that certain genetic causes of PWS are more likely to be associated with psychotic mood disorder than the others (Boer et al., 2002; see also below).

Researchers often employ cross-syndrome comparisons to elicit subtle variations in phenotypic expression, such as the differences between syndromes in autistic disorder symptomatology (Bruining et al., 2010; Oliver, Berg, Moss, Arron, & Burbridge, 2011). Developmental psychologists and neuroscientists are interested in the cognitive phenotypes associated with genetic syndromes and cross-syndrome comparisons have helped to gain a better understanding of the relative cognitive strengths and weaknesses of many syndromes such as Down syndrome (DS) and Williams syndrome (e.g., Annaz, Karmiloff-Smith, Johnson, & Thomas, 2009) as well as the development of cognition during childhood.

Behavioral phenotypes are likely to be dynamic, changing during development and aging. People with intellectual disabilities are living longer and clinicians are now more familiar

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with the trajectories of behavioral and physical phenotypes over the life course of individuals with ID. The rates of disorders such as autism spectrum conditions may change over the developmental period, and the presentation of some of the features of behavioral and especially cognitive phenotypes associated with specific syndromes often changes with aging (O'Brien & Bevan, 2011). A classic example is the behavioral and cognitive phenotype associated with Down syndrome (DS), which changes with aging and with the development of Alzheimer's disease (see below for more details). It is important that clinicians are able to anticipate the impact of aging on behavior, ability, and mental health and that they are aware of the likely future medical complications.

Advances in Genetic Diagnosis

Intellectual disability is highly heritable, and the estimated proportion that can be accounted for by genetic factors (chromosomal, monogenic, or multifactorial) is increasing (Baker, Raymond, & Bass, 2012). Until recently G-banded karyotyping, supplemented by other tests such as telomere screening and fluorescence in situ hybridization (FISH) has been the mainstay of the genetic work-up for people with intellectual disability. At this level of analysis abnormalities associated with common syndromes (such as Down syndrome and fragile X syndrome [FXS]) are detected in up to 20 % of people with intellectual disability (Battaglia, Bianchini, & Carey, 1999).

Over the last decade technological advances have driven forward the field of molecular genetics. The application of array comparative *genomic hybridization* (aCGH) and more recently next-generation sequencing has led to the identification of many new genetic loci contributing to ID and psychiatric disorders. aCGH is used to comprehensively test for submicroscopic deletions and duplications, known as *copy number variants* (CNVs), and is becoming the first-line genetic test for the investigation of unexplained childhood developmental delay and adult intellectual disability. Next-generation sequencing has proved a hugely powerful tool for the study of the genetics of human disease and its application in the clinic holds much promise for the near future.

The extent to which the human genome can tolerate sequence variation is only now becoming fully apparent. For example, CNVs are common across the genome and it would appear generally benign. Consequently establishing the pathogenicity of individual genetic differences can be a complex task (Baker et al., 2012).

Several new intellectual disability "syndromes" have emerged in the last few years, but the full extent of the associated phenotypes and behavioral issues remains to be established. Genetic findings in intellectual disabilities and psychiatric disorders are indicating common biological pathways with an aggregation of genetic changes in genes related to synaptic components and neurotransmitter systems (Collins & Sullivan, 2013). This raises the possibility of identifying treatments that can be applied across syndromes.

Recurrent CNVs

22q11deletion syndrome and Williams syndrome (caused by a deletion on 7q) are well-known examples of CNV syndromes. The widespread application of aCGH has led to the identification of several other ID-associated recurrent CNVs at 1q21.1, 3q29, 10q22-q23, 15q11.2, 15q13.3, 15q24, 16p11.2, 16p12, 16p13.11, 17q12, and 17q21.3 (Cooper et al., 2011; Mefford, Batshaw, & Hoffman, 2012).

Microdeletions or duplications at 16p11.2 occur in more than 3 in 10,000 people, and they present with a variable phenotype. Interestingly, some of the phenotypic features are mirrored depending on whether the person has a deletion or duplication, for example, obesity is associated with 16p11.2 deletion, while underweight is associated with 16p11.2 duplication (Jacquemont et al., 2011). Though both deletions and duplications have been associated with autism and schizophrenia, there is some evidence that 16p11.2 deletion is more associated with autism, while the duplication is more associated with schizophrenia (St Clair, 2013). Imbalances at 15q11.2 (within a

region prone to frequent deletion and duplication events) are the next most common, associated with diverse neurodevelopmental phenotypes, including autism (Burnside et al., 2011). Other CNVs occur less frequently.

Phenotypic variability which crosses traditional diagnostic boundaries is emerging as a common theme—the same CNV may be associated with different neurodevelopmental disorders such as ID, epilepsy, and autism or even schizophrenia (St Clair, 2013). While penetrance is generally high for these CNVs, it is not always complete. Unaffected carriers have been observed for some of the loci—notably being 15q11.2 and 16p13.11. This further hinders syndrome definition and prognostic value. Researchers are currently trying to understand the cause of the phenotypic variability and reduced penetrance. A "two-hit model" has been proposed with manifesting carriers being likely to carry a second large CNV (Girirajan et al., 2010).

Monogenic Causes of ID and Rare Mutations

A very large number of monogenic causes of ID have been identified via linkage and DNA sequence analysis in families with multiple cases of intellectual disability. The X chromosome has been systematically sequenced in families with X-linked intellectual disability (X-ID) which resulted in the discovery of multiple recurrent monogenic causes, in addition to known loci such as the FMR1 site associated with FXS, and detailed screening will reveal a mutation in nearly half of families with clear X-linked pedigrees (Baker et al., 2012).

Next-generation sequencing is now proving a powerful research tool to further elucidate the genetic architecture of ID. Mapping strategies where whole exome sequencing has formed a pivotal component have been highly successful in the identification of novel dominant de novo and recessive mutations (Najmabadi et al., 2011; Vissers et al., 2010). Furthermore a recent study was able to identify possible etiological changes in around half of the people with idiopathic ID using a whole exome-sequencing approach (Rauch et al., 2012). However the expense of sequencing and limitations in interpreting results mean that this is not yet suitable for widespread clinical use.

Common Behavioral Phenotypes

Angelman Syndrome

Angelman syndrome was first described in 1965 by Dr Harry Angelman, an English pediatrician who reported a group of children with severe intellectual disability and characteristic facial appearance, unstable gait, jerky movements, and a happy demeanor. Current estimated prevalence is 1 in 40,000 births.

Angelman syndrome is caused by a variety of genetic abnormalities affecting the expression of the UBE3A gene at chromosome 15q11-13. The UBE3A gene encodes information for a protein called ubiquitin-protein ligase. This protein regulates excitatory synapse development by controlling the degradation of proteins involved in the function of glutamate receptors. Disruption in this pathway is thought to contribute to the neurological picture and cognitive dysfunction that occurs in Angelman syndrome (Greer et al., 2010).

The majority of people with Angelman syndrome (75 %) have a maternally derived deletion of 15q11-13, and the rest have either *uniparental disomy* of chromosome 15, impaired *imprinting* of the maternal copy of 15q11-13, or other small deletions of the UBE3A gene (DYSCERNE, 2010). Some phenotypic differences between cases of uniparental disomy and deletions have been reported. People with Angelman syndrome due to uniparental disomy tend to present with a milder phenotype than those with deletions and also have a lower incidence of seizures, less severe ataxia, and fewer dysmorphic features (Veltman, Ellen, & Bolton, 2005). Depending on the underlying genetic mechanism, recurrence rates of Angelman syndrome may be as high as 50 % and genetic counseling is generally advised.

Typical features of the syndrome include deep-set eyes, broad smiling mouth, prominent

chin, microcephaly, hypotonia, jerky movements, or unsteady walking (puppetlike gait). Seizures are present in most cases and often start in early childhood. A variety of types of seizures may be observed with atypical absences and myoclonic seizures being particularly prevalent. Nonconvulsive status epilepticus is also common. Specific EEG abnormalities have been described in children and adults with Angelman syndrome with commonest finding prolonged high-amplitude rhythmic 2–3 Hz activity, predominantly over the frontal regions, with superimposed interictal epileptiform discharges (Laan & Vein, 2005; Van Buggenhout & Fryns, 2009).

Most people with Angelman syndrome have severe to profound intellectual disability and significant speech and language impairment. A happy demeanor and a tendency to burst out laughing are thought to be typical of the syndrome with hyperactivity, short attention span, and sleep problems equally common. Behavioral disorders can be present to a significant proportion of people with Angelman syndrome. It is not clear if such behavioral problems depend more on the specific genotype than the coexisting intellectual disability. Aggressive behaviors are reported in nearly 6–10 % of children with the syndrome; however, the frequency of such behaviors decreases with increasing age (Horsler & Oliver, 2006; O'Brien & Bevan, 2011; Veltman et al., 2005).

In adulthood people with Angelman syndrome display less hyperactivity, excitability, and an improved ability to concentrate. Sleep pattern also improves by age while, in contrast, seizures tend to persist in adult life. The most common psychiatric condition affecting adults with Angelman syndrome is anxiety (Clayton-Smith, 2001).

Fragile X Syndrome

FXS is the most commonly inherited form of intellectual disability and an identified cause of some cases of autism. It occurs in approximately 1 in 4,000 males and 1 in 8,000 females. More than 99 % of people with FXS have a mutation in the X chromosome's long arm (position Xq27.3) resulting in loss of function of the FMR1 gene. This is caused by an increased number of CGG trinucleotide repeats—usually more than 200 (normal range 5–40) and also by abnormal *DNA methylation* of the FMR1 gene. Other types of mutations of the FMR1 gene may cause FXS including deletions. Cases of FXS *mosaicism* have also been reported.

The FMR1 gene is responsible for the production of the fragile X mental retardation 1 protein (FMRP) involved in the development of synapses in the brain. The abnormally expanded CGG repeats "silence" the FMR1 gene and prevents the production of FMRP. There is evidence that this results in defects in synaptic structure and plasticity and is responsible for imbalanced glutamatergic signaling pathways (Antar, Afroz, Dictenberg, Carroll & Bassell, 2004).

FMR1 alleles are divided according to the number of CGG repeats to normal (5–44), intermediate (45–54), premutation (55–200), and full-mutation (>200) alleles. The severity of phenotype in FXS appears to depend on the allele type and number of CGG repeats but also on the degree of *DNA* methylation and presence of mosaicism.

Both increased CGG repeats and methylation changes in FMR1 can be detected by genetic tests including *PCR* and *FISH*. The choice of test and diagnostic method depends on the type of suspected mutation. FXS is an example of a genetic disorder that shows anticipation, i.e., more severe presentation in successive generations in some families.

Males with an *FMR1* full mutation can have a characteristic appearance which becomes more apparent with aging. It includes protruding ears, large head, long and narrow face, prominent forehead and chin, flexible fingers, and large testicles after puberty. Females may have a milder phenotype. Epileptic seizures develop in about 15–20 % of males and about 5 % of females with FXS. Adults with FMR1 premutation are at increased risk of two medical disorders: (a) fragile X-associated primary ovarian insufficiency (FXPOI) occurring only in females and (b) fragile X-associated tremor/ataxia syndrome (FXTAS). FXTAS can affect

both males and females in later life and is characterized by progressive cerebellar ataxia and tremor (Saul & Tarleton, 1998).

Full FMR1 mutation in males usually causes moderate to severe intellectual disability while about 50 % of females with full FMR1 mutation have intellectual disability; however, females are usually less severely affected. The behavioral characteristics of FXS include ADHD symptoms, social anxiety, behavioral disorders, gaze avoidance, and repetitive activities or movements. Sensory integration dysfunction and self-injurious behavior, including hand biting and scratching, may also be present.

ASD coexists in nearly 25 % of full FMR1 mutation cases of FXS. Premutation carriers also have an increased risk for ASD and ADHD (Hagerman et al., 2009; Hatton et al., 2006). Hyperactivity and impulsivity improve with increasing age but attention deficits and inhibitory control problems tend to be more persistent (Cornish, Turk, & Hagerman, 2008).

Hyperarousal, overactivity, cognitive problems, anxiety, repetitive behaviors, and hypersensitivity to tactile stimuli have all been linked to excessive glutamate excitatory neurotransmission according to the glutamatergic signaling pathways theory of FXS (Bear, Huber, & Warren, 2004; O'Brien & Bevan, 2011). This has led to hope that agents with glutamate antagonist properties could provide the first targeted treatment for FXS. A few pilot open-label studies of such agents (fenobam, riluzole) in adults with FXS have not provided any conclusive evidence of effectiveness or safety as yet; however, clinical research in this area is ongoing (Berry-Kravis et al., 2009; Erickson et al., 2011).

Prader–Willi Syndrome

The incidence of PWS is reported to be 1 in 15–29,000 births. PWS can result from various mechanisms that affect the expression of paternally inherited genes in the q11.2–q13 region of chromosome 15. Common such mechanisms include paternal microdeletion, maternal *uniparental disomy*, and *imprinting defect* on the paternally inherited chromosome. Over 99 % of cases of PWS can be diagnosed with a simple test, DNA methylation analysis (Cassidy & Driscoll, 2009; Whittington et al., 2001).

Symptoms of PWS such as infantile lethargy and hypotonia usually become apparent soon after birth. These symptoms can cause severe feeding problems that lead to failure to thrive in babies with PWS. This is followed by the onset of intense food-seeking behaviors, insatiable appetite, and hyperphagia (overeating) with resulting obesity and associated medical problems.

Hyperphagia, a hallmark of the behavioral phenotype of PWS, is thought to be the result of hypothalamic deficiency (Veltman et al., 2005); however, other hypotheses have also been explored. Ghrelin (a hormone secreted by the stomach that stimulates appetite) concentrations are elevated in people with PWS suggesting that ghrelin may contribute to hyperphagia. De Waele et al. (2008) conducted a 56-week prospective, randomized, cross-over trial to see if an appetite suppressant, a long-acting octreotide (Oct), decreased ghrelin concentrations, body mass, appetite, and compulsive food-seeking behavior in adolescents with PWS ($n=9$). Their findings suggest that although Oct treatment was effective in causing a prolonged decrease in ghrelin concentrations, it did not improve body mass or appetite in people taking part in the study. Other features of PWS that suggest a hypothalamic dysfunction include hypogonadism, short stature, and abnormal body temperature regulation. People with PWS often display other typical characteristics such as small hands and feet, loss of skin color, and a narrow face.

Most people with PWS function within the mild–moderate intellectual disability range. Particular strengths in reading, long-term memory, and visual memory have been reported. Characteristic behaviors include temper tantrums, skin picking, and compulsive behaviors such as hoarding. Behavioral problems tend to worsen in adolescents and young adults but subside over time (Whittington & Holland, 2010). ASDs and ADHD are reported to be common and of early onset. Mood fluctuation, anxiety, and depression are commonly seen as well as high

rates of psychosis (5–10 %), especially in people with the maternal uniparental disomy subtype of PWS (Cassidy & Driscoll, 2009; O'Brien & Bevan, 2011).

Several phenotypic differences with regard to cognitive abilities, mental health, and behavior have been reported such as that uniparental disomy cases display fewer of the facial characteristics, have higher IQ, and fewer tendencies to compulsive behaviors than cases due to deletions but may be more prone to psychosis and affective disorders in later life. People with PWS due to deletions are reported to have relative strength in specific visual perceptual skills (Veltman et al., 2005), but further research in genotype–phenotype correlations in people with PWS is needed.

Tuberous Sclerosis

Tuberous sclerosis or tuberous sclerosis complex (TSC) is caused by mutations in one of the *tumor suppressor genes*, TSC1 or TSC2 on chromosomes 9q34.3 or 16p13.3. Almost 80 % of affected patients have a new mutation, and 20 % have inherited a TSC gene mutation from a parent (*autosomal dominant inheritance*). Approximately 1 in 6,000 people is affected by the syndrome (Holmes, Stafstrom, & The Tuberous Sclerosis Study Group, 2007).

TSC is characterized by the development of benign tumors, also called "hamartomas," in many organ systems including the brain, skin, lungs, heart, and the kidneys. The development of hamartomas in the central nervous system causes the most disabling symptoms such as developmental delay, intellectual disability, mental disorders, and epileptic seizures. Epileptic seizures can present in almost 90 % of affected people and are often difficult to control due to limited response to anticonvulsant medications.

Molecular genetic testing has provided evidence for additional *locus heterogeneity* in people with TSC. *Sequence analysis* of TSC1 and TSC2 identifies a mutation in about 85 % of people with a definite diagnosis of TSC, and 15 % of persons with TSC have no identifiable mutation. In terms of genotype–phenotype correlations, it is known that TSC2 mutations produce a more severe phenotype than TSC1 mutations.

About half of people with TSC have cognitive impairment across the whole spectrum of intellectual disability from mild to profound. Language delay, restlessness, aggression, and self-injurious behavior are often observed. ASD and ADHD can be diagnosed in up to 50 % of cases. Anxiety, bipolar and unipolar depression is also common in adults with TSC (Holmes et al., 2007; O'Brien & Bevan, 2011). A comprehensive mutation analysis of TSC1 and TSC2 in a cohort of 150 unrelated patients with TSC and their families found a higher frequency of intellectual disability in persons with a mutation in TSC2 (Jones et al., 1999). Autistic spectrum disorder and infantile spasms are also more frequently associated with a TSC2 mutation (Northrup, Koenig, & Au, 1999; 2011).

Delineation of the TSC biochemical signaling pathway suggested mTOR inhibition as a potential therapy, and subsequent trials of rapamycin analogs (sirolimus and everolimus) demonstrated reduction of the tumors associated with TSC (Davies et al., 2008). Current trials are evaluating the effect on the cognitive problems of the syndrome.

Williams Syndrome

Williams syndrome has a prevalence of up to 1 in 7,500 and polygenic etiology. It is caused by a hemizygous deletion of about 28 genes on the long arm of chromosome 7 (Meyer-Lindenberg, Mervis, & Berman, 2006). The syndrome follows autosomal dominant pattern of inheritance. Genes involved in the production of protein elastine are thought to be defective resulting in recognizable physical characteristics and connective tissue pathology affecting the vascular and nervous systems.

Babies with Williams syndrome have feeding problems including reflux and vomiting. Short stature, sunken chest, and dysmorphic facial features such as flattened nasal bridge, small nose, prominent lips, open mouth, and

widely spaced teeth are characteristic findings. Cardiovascular symptoms are common but vary in severity and may include hypertension, aortic stenosis, and pulmonary artery stenosis. Hypercalcemia (high blood calcium level) is also common and can lead to seizures and muscular rigidity (Morris, 1999, 2006).

People with Williams syndrome have developmental delay and their IQ is usually about 50–60 but they also present with unusual strengths in language and a unique "social phenotype." Typical personality traits include being very friendly, trusting strangers, and having a strong interest in music and musical abilities (Meyer-Lindenberg et al., 2006; Rosner, Hodapp, Fidler, Sagun, & Dykens, 2004). The sociable personality and friendliness of some people with Williams syndrome combined with poor social judgments and deficits in socially adaptive skills can make them particularly vulnerable to abuse and exploitation.

Common psychiatric and behavioral problems include hyperactivity, distractibility, and obsessive behaviors and preoccupations. Anxiety and phobias with onset in childhood and depression in later life have also been associated with Williams syndrome (O'Brien & Bevan, 2011).

Meyer-Lindenberg et al. (2006) presented a review of neural mechanisms in Williams syndrome as an example of genetic influences on cognitive abilities and behavior. They reported on studies that have identified severe deficits in visuospatial construction and a relative strength in short-term auditory memory and language. Imaging studies documented reduced grey matter volume and depth in the intraparietal sulcus (area important for visuospatial constructive function) while functional studies have shown that the amygdala of people with Williams syndrome is less active to threatening faces with evidence of increased activity to threatening nonsocial stimuli. In their review the above authors also reported findings suggesting impaired connectivity between the amygdala and the orbitofrontal cortex, which may reflect different genetic developmental trajectories of neural systems for social and nonsocial fear in people with Williams syndrome.

Down Syndrome

People with Down syndrome have an extra copy of a region of chromosome 21 (Fig. 8.1). In 92 % of cases the syndrome is due to full trisomy 21. Mosaicism, which occurs when only some cells have trisomy 21, presents in 2–3 % of people with Down syndrome. In 3–5 % of all cases the cause of the chromosomal abnormality is an unbalanced translocation of chromosome 21 which in some instances may be familial (O'Brien & Bevan, 2011). The risk of having a child with Down syndrome increases with advancing maternal age.

Children with Down syndrome are born with decreased muscle tone. Physical features include short stature, excess skin at the nape of the neck, flattened nose, single crease in the palm of the hand, small ears and mouth, and upward slanting eyes as well as short hands with short fingers. Common medical problems include congenital cardiac abnormalities, gastrointestinal problems, cataract, underactive thyroid, hip and hip problems (risk of dislocation). There is also an increased risk for certain types of leukemia in young children, while solid tumors are relatively rare in adults.

Although people with Down syndrome can have variable cognitive abilities, most function within the mild–moderate intellectual disability range. Stancliffe et al. (2012) reported that 68 % of people with Down syndrome had mild or moderate intellectual disability in a large sample ($n = 1{,}199$) drawn from national surveys of adults using specialist services in 25 US states. There are reports of particular strengths in visual spatial tasks and visual memory. Poor attention, impulsivity and repetitive behaviors have been reported as part of a behavioral phenotype. In comparison to people with intellectual disability of other etiology, people with Down syndrome are thought to be more socially competent and their social skills tend to improve with age (Wishart, 2007).

In addition to the well-known association between Down syndrome and early onset Alzheimer's disease, higher rates of depressive, anxiety, and obsessive–compulsive disorder have been reported. Lower rates of psychosis have been documented and depressive disorder is

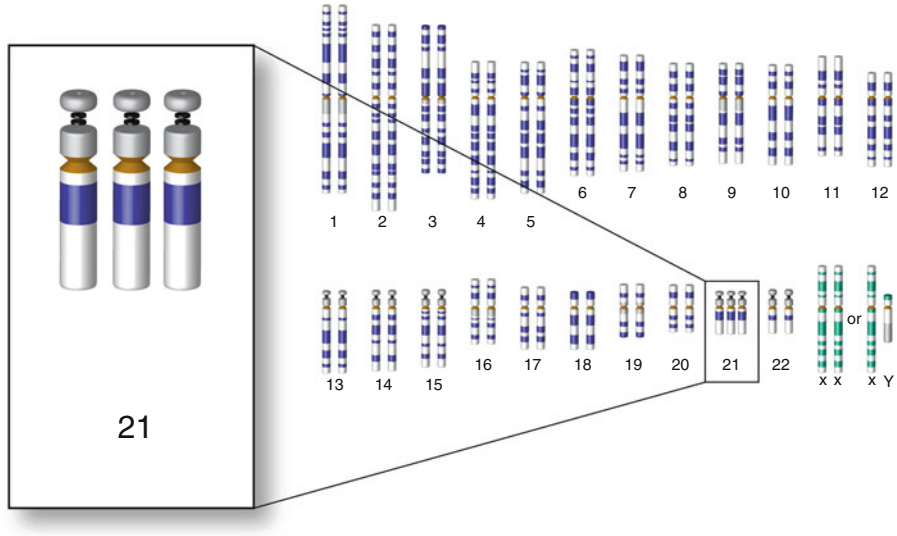

Fig. 8.1 Trisomy 21. Courtesy: National Human Genome Research Institute, US National Institutes of Health. National Human Genome Research Institute. "Talking Glossary of Genetic Terms." Retrieved November 18, 2012, from http://www.genome.gov/glossary/

often present at the early stages of dementia (O'Brien & Bevan, 2011). Age-specific prevalence rates for dementia in a sample of people with Down syndrome ($n = 201$) have been reported by Prasher (1995) as 9.4 % for ages 40–49 years, 36.1 % for 50–59 years, and 54.5 % for 60–69 years.

Beta amyloid is a component of the amyloid plaques involved in the pathogenesis of Alzheimer's disease. The APP gene (responsible for the production of amyloid precursor protein) is located on the long arm of chromosome 21. Triplication and overexpression of this gene in people with Down syndrome has been thought to confer high risk for dementia. However, as in many other examples in this chapter, the genotype–phenotype relationship is likely to be complex and is likely to include environmental influences. To explain the wide variation in age at onset of Alzheimer's disease in this population, Schupf (2002) reviewed genetic and host factors for dementia and concluded that factors which influence beta amyloid levels, (rather than overexpression of APP) may better account for the observed differences in age at onset.

Cornelia de Lange Syndrome

Cornelia de Lange syndrome is associated with abnormalities on chromosomes 5, 10, and X, with the majority of cases due to spontaneous mutations. Its incidence is estimated at 1 in 10,000–30,000. Although there is evidence of some degree of correlation between the genotype and phenotype, it does not seem to be sufficient to accurately predict developmental or clinical outcomes (Wulffaert et al., 2009). Several features of the physical phenotype have been described such as small stature, dysmorphic facial characteristics, heart defects, abnormal limb formation, cleft palate, kidney and gastrointestinal disorders, and seizures.

Levels of cognitive impairment vary from mild to profound intellectual disability with expressive language being generally poor. Common behavioral problems include self-injury and there are reports of features of ASD such as self-absorption, reduced social reciprocity, and repetitive and compulsive behaviors. Other features include hyperactivity and poor attention.

There is some evidence that problems with socially adaptive functioning and behavioral disorders increase with age (Basile, Villa, Selicorni, & Molteni, 2007; Wulffaert et al., 2009).

Oliver, Arron, Sloneem, and Hall (2008) attempted to clarify the behavioral phenotype of the syndrome with specific reference to ASD features. They assessed a group of 54 individuals with intellectual disabilities due to Cornelia de Lange syndrome on measures of ASD and behavioral problems and compared them with 46 individuals with intellectual disability alone. Even though both groups presented the same levels of nonspecific behavioral problems, severe ASD features and levels of compulsive behavior were significantly higher in the syndrome group (32.1 %) than the comparison group (7.1 %). Self-injurious behavior is frequently cited as part of the behavioral phenotype of Cornelia de Lange syndrome; however, a recent case–control study (Oliver, Sloneem, Hall, & Arron, 2009) showed that clinically significant self-injury was not more prevalent in the syndrome group. Also the way that self-injurious behavior manifested in the Cornelia de Lange syndrome group was not different from that seen in the control group with non-syndromic intellectual disability.

16p11.2 Microdeletion Syndrome

This syndrome is caused by a microdeletion of a small piece of chromosome 16 with an estimated prevalence of approximately 3 in 10,000. People with 16p11.2 deletion syndrome are missing a sequence of about 600 kb at the area p11.2 of chromosome 16. This deletion affects one of the two copies of the chromosome. Therefore the inheritance is autosomal-dominant; however, most cases are not inherited and the chromosomal abnormality appears de novo. In inherited cases, other family members may be affected and genetic counseling may be offered.

There is no characteristic pattern of dysmorphic features in 16p11.2 deletion, but several studies reported various dysmorphic features. There is a higher than average risk for seizures and hypotonia has been noted in a minority of cases. Obesity and aortic valve abnormality may also be part of the phenotype (Shinawi et al., 2010). As noted earlier, some people with the deletion may have no identified physical, cognitive, or behavioral abnormalities.

The cognitive abilities of persons with 16p11.2 microdeletion range from mildly impaired to normal intellectual ability. Language delay is common with expressive language more affected than receptive language. Hanson et al. (2010) screened 11 individuals with 16p11.2 microdeletion for ASD with the use of structured assessments such as the Autism Diagnostic Observation Schedule and the Autism Diagnostic Interview. Three (27 %) met full diagnostic criteria for ASD with six (55 %) exhibiting ASD features but not reaching diagnostic threshold. Atypical language, deficits in social skills, and socially adaptive behavior were common.

An increased frequency of 16p11.2 microdeletion among individuals with ASD and a psychiatric or language disorder, as compared to controls, was reported by Weiss et al. (2008). Psychiatric conditions identified included schizophrenia, bipolar disorder, ADHD, and anxiety disorders. The authors concluded that microdeletion at 16p11.2 carries substantial susceptibility to autism and may account for approximately 1 % of cases of ASD and also represents a highly penetrant cause of developmental disability.

22q11.2 Deletion Syndrome

This syndrome is caused by the deletion of a small region of chromosome 22 at the location q11.2. It has prevalence of 1 in 4,000. There are several presentations, based on different symptom grouping, including DiGeorge syndrome (first described in 1968 by pediatrician Angelo DiGeorge) and velo-cardio-facial syndrome. The condition can be diagnosed by FISH and aCGH. aCGH can better identify the extent of the microdeletion and characterize any missing genes. 22q11.2 deletion syndrome is inherited in an autosomal dominant pattern; however, about 90–95 % of cases are due to a new deletion of 22q11.2 and not inherited by an affected parent.

The physical manifestations of 22q11 deletion syndrome are highly variable. Common features include congenital heart disease, cleft palate, defects in the palate, and mild dysmorphic facial features. Recurrent infections are also common due to problems with the T-cell-mediated immunological response often because of the absent or hypoplastic thymus gland. Parathyroid glands abnormalities and low levels of parathyroid hormone and hypocalcemia may be present, and kidney abnormalities, hypothyroidism, and low platelet levels have also been reported.

Ousley, Rockers, Dell, Coleman, and Cubells (2007) described cognitive and behavioral difficulties in people with 22q11 deletion syndrome including delayed motor and language development, ID of various severity, impaired spatial reasoning, ADHD, ASD, mood disorders, and schizophrenia spectrum disorders.

The association of the syndrome with schizophrenia is widely reported in the literature and microdeletions in chromosomal region 22q11.2 are thought to be associated with a 20- to 30-fold increased risk of schizophrenia (Horowitz, Shifman, Rivlin, Pisante, & Darvasi, 2005). Conversely, people with schizophrenia have an 80-fold increased prevalence of the microdeletion in comparison to the general population (Karayiorgou et al., 1995) with more recent linkage and association studies' findings also suggesting a schizophrenia susceptibility loci at the 22q chromosomal region (Harrison & Owen, 2003). Baker and Skuse (2005) compared a group of adolescents and young adults with 22q11 deletion syndrome with an age and IQ-matched control group. There were more psychiatric disorders and overall psychopathology in those with the syndrome including ADHD, depression, and anxiety. Almost half members of the syndrome group reported psychotic experiences.

Lesch–Nyhan Syndrome

This syndrome is an example of an X-linked recessive disorder caused by a mutation of a gene on the long arm of the X chromosome. About a third of all cases arise de novo and do not have a family history of Lesch–Nyhan syndrome. It affects about 1 in 380,000 and it is a disorder of purine metabolism involving defects in the production of hypoxanthine–guanine phosphoribosyltransferase (HGprt). HGprt is an enzyme that has a vital role in purine metabolism. Complete deficiency of the enzyme results in marked accumulation of xanthine and uric acid which leads to the development of gout and renal symptoms. Different mutations of the gene associated with Lesch–Nyhan syndrome result in varied levels of HGprt enzyme activity and a spectrum of disease characteristics. Although both the gene and HGprt enzyme have been studied, it is not known if similar mutations result in similar phenotypes. Sampat et al. (2011) discusses potential mechanisms for genotype–phenotype correlation and discordance in people with Lesch–Nyhan syndrome. Typical features include hyperuricemia, gout, nephrolithiasis, dystonia, chorea, and spasticity. As the condition initially resembles athetoid cerebral palsy, most affected children are initially misdiagnosed as having cerebral palsy (Nyhan, O'Neill, Jinnah, & Harris, 2010).

Jinnah et al. (2006) studied motor and movement disorders in a group of Lesch–Nyhan syndrome patients ($n=44$) who presented with severe action-related dystonia and baseline hypotonia and compared their findings with a review of 122 previous reports ($n=254$). Although other extrapyramidal symptoms have also been described, they have not been as prominent as dystonia which makes this symptom a likely phenotypic marker.

Since the first description of the syndrome by Lesch and Nyhan (1964), several studies have reported a strong association with self-injurious behavior including cases of severe injuries caused by biting of the fingers, hands, lips, and cheeks (Robey, Reck, Giacomini, Barabas, & Eddey, 2003). The frequency and severity of self-injurious behavior has been linked to physical or emotional stress (Anderson & Ernst, 1994), and more adverse outcomes have been associated with early onset of the self-injurious behavior. Almost all affected individuals have intellectual disability in the moderate to severe range.

Conclusion

Genetic research into human disease has been undergoing a revolution in recent years and our understanding of genetic influences on ID and other mental disorders is rapidly advancing. An increasing number of genetic variants are being found to be associated with ID and the relationship between ID and other mental disorders is beginning to be clarified. There are now several examples of variants which are associated with a range of neurodevelopmental disorders such ID, autism, and schizophrenia. Concurrently the detailed study of behavioral phenotypes is improving our understanding of the typical presentation of intellectual disability syndromes.

The proportion of people classified as having idiopathic ID is gradually declining, and as more individuals with a specific genetic change are identified, the phenotypic description of that syndrome will improve. However specific diagnoses are still relatively rare and may even be unique to an individual or family, which limits the prognostic utility of the diagnosis. Furthermore, genotype–phenotype relationships are complex and often difficult to interpret.

The coming challenge will be to translate our increased understanding of the genetics of ID into an increased understanding of the pathological mechanisms which end in impaired cognition and behavioral and emotional disturbance. It is hoped that through mechanistic insight new targets for treatment will emerge.

Glossary

Allele: Alternative version of a specific gene responsible for variations in characteristics such as blood type.

Copy number variations (CNVs): Abnormal number of copies of sections of the DNA on certain chromosomes. It is usually caused by submicroscopic structural rearrangements such as deletions, duplications, or translocations.

Comparative genomic hybridization (CGH): Method for analysis of CNVs in a part of DNA based on microarray technology.

Dominant: Alleles that determine the phenotype seen in a heterozygote.

DNA methylation: Addition of a methyl group to DNA that may affect the expression of genes. An epigenetic mechanism.

DNA sequencing: Process of determining the order of nucleotides containing four bases [(G) guanine, (A) adenine, (T) thymine, and (C) cytosine] in a segment of DNA.

Epigenetic: Any factor which influences the phenotype but is not part of the genotype.

FISH (fluorescence in situ hybridization): Technique developed in the early 1980s to detect the presence or absence of specific DNA sequences on chromosomes.

Genetic linkage map: A map of positions of known genes on the chromosomes.

Genomic imprinting: A natural process where certain genes are expressed or silenced (without alteration of their genetic code) depending on whether they are inherited from the mother or the father. Genetic conditions associated with imprinting defects include Angelman and Prader–Willi syndromes.

Genome-wide association studies: Examination of the genome for common variants in different individuals usually focusing on associations between SNPs and major diseases.

Karyotype: Photographed chromosomes arranged by size.

Linkage: The tendency of genes close together on the same chromosome to be inherited together.

Linkage disequilibrium: The presence of certain combinations of alleles of closely linked genes on the same chromosome more often than it would be expected by chance.

Locus heterogeneity: This occurs when mutations in genes at different chromosomal loci cause the same phenotype.

Mosaicism: Presence of two or more populations of cells with different genotypes in one organism. It results from a mutation during development affecting only a subset of the organism's cells.

Polymerase chain reaction (PCR): Technique for copying strands of DNA in order to increase the available amount for study and analysis.

Recessive: A gene that is masked in the presence of a dominant allele.

Sequence analysis (also called "gene sequencing" or "sequencing"): Lab technique by which the nucleotide sequence for a specific segment of DNA is determined.

Single-nucleotide polymorphisms (SNPs): A variant DNA sequence in which the base of a single nucleotide has been replaced by another (e.g., cytosine replaced by thymine).

Tumor suppressor gene: A gene that protects a cell from transforming to a cancerous cell. When this gene is mutated and loses or reduces its function, the cell can progress to cancer, usually in combination with other factors.

Uniparental disomy: Inheritance of both copies of a chromosome from one parent.

X-linked trait: Trait that is caused by mutation of a gene located on the X chromosome.

X-linked recessive disorder: A disorder due to a mutation in a gene on the X chromosome that causes the disease traits to be expressed in all males or females who are homozygous for the gene mutation.

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Psychological and Social Factors

9

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Introduction

Consistently high rates of psychiatric and behavioral disorders have been reported in incidence and prevalence studies of youth and adults with Developmental and Intellectual Disabilities (DD/ID, i.e., Deb, Thomas, & Bright, 2001; Smiley et al., 2007). This has led to increasingly more research and clinical attention being paid in better understanding mental health difficulties and associated psychopathology in this population in the last 20–30 years, with a small number of excellent comprehensive publications emerging in this field (i.e., Bouras & Holt, 2007; Odom, Horner, Snell, & Blancher, 2007; this handbook). Despite major advances over recent decades in assessing, diagnosing, and treating comorbidity in DD/ID, many limitations in our knowledge and understanding persist. A particular "gap" appears to be in conceptualizing such psychopathology from a psychosocial perspective and in integrating psychosocial and biological perspectives towards an integrated understanding of the precipitating, predisposing, maintaining, and protective factors associated with psychopathology in DD/ID.

There is little doubt that genetic and biological factors are strongly implicated in the increased rates of psychopathology in people with DD/ID (see, e.g., Chapters 6, 7 and 8 in this handbook). At the same time, it is also becoming increasingly clearer that unidimensional biological perspectives are somewhat limited in helping us better understand emotional and behavioral difficulties associated with DD/ID. Increased rates and range of psychosocial stressors precipitating or maintaining challenging behaviors and psychiatric problems have been consistently documented in this population; thus, a biopsychosocial model is likely to be more appropriate and helpful in considering the complex relationships between biological, psychological, and social factors in the development and maintenance of psychopathology (see also O'Hara, 2007).

It is thought that psychosocial adversity may be implicated in increased psychopathology in this population via a number of possible pathways (Matson & Sevin, 1994). Individuals with DD/ID are reported to experience more adverse life events and social experiences associated with emotional and behavioral difficulties (such as abuse, trauma, discrimination, and life transitions) compared to the non-ID population (i.e., Reiss & Benson, 1985). Thus, increased rates of psychopathology could be due to increased exposure to negative life events and experiences.

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At the same time, individuals with DD/ID may develop more comorbid psychiatric conditions or behavioral problems as they are likely to have reduced coping skills and resources to deal with both normative and nonnormative life stressors compared to the general population (i.e., Tonge & Einfeld, 2003; Weisz, 1990).

This chapter will critically explore and synthesize the literature on psychosocial factors that have been identified as likely to be implicated in the development or maintenance of psychopathology in DD/ID. A developmental framework is adopted and thus current knowledge on psychosocial factors is presented for children, youth, adults, and older adults with DD/ID when available. In addition, Bronfenbrenner's socio-ecological model (1977) is used as a structural framework and thus psychosocial factors are presented at the individual, family, and social levels. Emphasis is placed on findings from longitudinal prospective studies, studies exploring multiple psychosocial factors, and recently published systematic reviews or meta-analyses where available.

Individual Factors Associated with Psychopathology in Individuals with DD/ID

Age

A number of research studies consistently document persistently high and stable rates of psychopathology in this population compared to individuals without DD/ID across the life span (i.e., de Ruiter, Dekker, Verhulst, & Koot, 2007; Emerson & Einfeld, 2010; McCarthy & Boyd, 2001; Tonge & Einfeld, 2003). However, some evidence suggests that the overall rate of emotional and behavioral difficulties may lessen with age (i.e., Chadwick, Kusel, Cuddy, & Taylor, 2005; de Ruiter et al., 2007; Einfeld et al., 2006). In a review of 26 studies of psychopathology in youth with ID, Witwer and Lecavalier (2008) found that a small number of empirical studies have consistently reported a negative association between age and specific types of behavioral disturbances, with hyperactivity and conduct disorder symptoms being more common in younger youth with ID. However, findings have generally been inconsistent, possibly as a consequence of complex interactions between age and other variables (such as gender, level of functioning, and behavioral phenotypes), which are not considered in the majority of studies. Although longitudinal studies across the life span are rare in this field, there is little evidence, to date, of age-related effects on the development of psychopathology among individuals with DD/ID.

Gender

In studies of children and adolescents with DD/ID, reported gender differences in psychopathology have been inconsistent, with some studies reporting males with ID as more likely to have higher problem behavior scores (specifically disruptive and hyperactive symptoms), girls to have more depressive symptoms in adolescence, or no gender differences (see Witwer & Lecavalier, 2008 for review). In a prospective, longitudinal epidemiological study of 578 youth with ID followed up from 12 to 23 years using the Developmental Behaviour Checklist (DBC), Einfeld et al. (2006) reported that gender appeared to have a small but significant effect on the *course* of psychopathology. Thus, boys showed greater decreases in overall problem behaviors than girls over time, and girls with severe ID showed more increases in anxiety problems compared to girls with mild ID and boys with mild and severe ID. Similarly, Emerson, Einfeld, and Stancliffe (2011) reported that male gender was uniquely associated with the *persistence* of conduct difficulties at age 6/7 and 8/9 in a large nationally representative sample of children with cognitive delays assessed at age 4/5.

In studies of adults with DD/ID, some researchers have suggested that women may be more vulnerable to psychopathology (i.e., Cooper, Smiley, Morrison, Williamson, & Allan, 2007). Specifically, they may be more likely to be diagnosed with affective disorders (i.e., Lunsky, 2003), dementia, or adjustment reaction disorders, while men may be more likely to be diagnosed with personality

disorders (Tsakanikos, Costello, Holt, et al., 2006). Although there is some emerging evidence that gender may play a role in explaining some of the individual differences in the types, course, or persistence of psychopathology in individuals with ID, it remains unclear whether gender plays as important a role as it does in typically developing individuals or, if it does, how it may affect the prevalence, prognosis, and predisposing or protective factors implicated in psychopathology in this population.

Level of Intellectual Functioning

Increased rates of comorbid psychopathology have been consistently reported in studies of individuals with DD/ID across all levels of intellectual and adaptive behavior functioning (i.e., Cooper & Bailey, 2001; Hemmings, 2007). However, whether psychiatric and behavioral difficulties are more prevalent as the severity of DD/ID increases is less clear. Some studies have reported higher rates in individuals with milder ID (i.e., Cowley et al., 2004; Holden & Gitlesen, 2004; Iverson & Fox, 1989; Jacobson, 1990; Taggart, Taylor, & McCrum-Gardner, 2010); others have found higher rates in people with more severe ID (i.e., Cooper et al., 2007; Cooper & Bailey, 2001; Smiley et al., 2007).

The lower rates of psychiatric comorbidity reported in some studies in people with severe IDs could to some extent be a reflection of the challenges in diagnosing such disorders in this population. Due to atypical presentation and reliance on behavioral symptoms reported by carers, diagnosis of a number of disorders, such as schizophrenia or anxiety, can be particularly difficult. However, difficulties in identifying psychiatric symptoms in people with severe ID may not fully explain the inconsistent findings with regard to the association between psychopathology and level of ID. Some researchers have reported similar rates of *behaviorally expressed* overt psychiatric symptoms in adults with moderate, severe, and profound disabilities (i.e., Holden & Gitlesen, 2004; Reiss, 1988). One would expect that if symptom detection accounted for the differences in symptomatology, people with severe and profound ID would score higher on nonverbal and overt symptoms compared to verbal or internalizing symptoms, but this pattern has not been consistently reported. However, it is also possible that the measures developed for assessing psychopathology in this population may not be sensitive enough for people with no or very limited verbal ability.

In one of the very few prospective longitudinal studies in the field using the DBC, Tonge and Einfeld (2003) examined psychopathology in young people with ID over 9 years. They suggested there may be differences in the *patterns* of associated difficulties according to the level of ID. Thus, individuals with mild ID showed more disruptive or antisocial difficulties, while those with moderate or severe ID presented with more self-absorbed, social relating, self-injurious, and stereotyped behavioral problems (see also Dekker & Koot, 2003a; Dekker, Koot, van der Ende, & Verhuslt, 2002; Witwer & Lecavalier, 2008 for related findings). Similarly, de Ruiter et al. (2007), also using the DBC, found a small but significant difference in specific problem behaviors (self-absorbed, social relating), with youth with moderate and severe ID showing higher stability and persistence compared to individuals with mild ID. These studies highlight the importance of investigating longitudinal differences in relation to levels of ID, not only in rates but also in patterns of psychopathology.

Although studies have so far produced mixed findings in relation to the role of the level of ID in psychiatric comorbidity, severe ID has been consistently and strongly associated with challenging behaviors (i.e., self-injurious, aggressive, stereotyped, or inappropriate sexualized behaviors; i.e., Dawson, Matson, & Cherry, 1998; O'Brien & Pearson, 2004; Reese, Richman, Belmont, & Morse, 2005). It is important to note, however, that there are a number of theoretical and methodological challenges in attempting to distinguish challenging behaviors from psychiatric symptomatology. A number of researchers have cautioned that there may be a considerable overlap between the two (i.e., Holden & Gitlesen, 2008; Kearney & Healy, 2011; Tsiouris, Cohen,

Patti, & Korosh, 2003) with self-injurious behaviors, in particular, being associated with affective symptoms (Hemmings, Gravestock, Pickard, & Bouras, 2006). Nevertheless, in a recent study of individuals with ID and ASD, challenging behaviors were independent of comorbid psychopathology (McCarthy et al., 2010).

Physical and Health Factors

Physical factors associated with increased psychopathology in DD/ID include higher specific genetic conditions, increased physical health problems, and the increased use of medication in this population (see also Estia Centre, 2006).

Genetic syndromes and developmental disorders, such as Autism Spectrum Disorders (ASD), fragile X, Down syndrome, and Prader-Willi syndrome, are often comorbid with DD/ID, and their presence has been consistently identified in the research literature as positively associated with higher rates of particular mental health or behavioral presentations (Dykens, Hodapp, & Finucane, 2000; Hodapp & Dykens, 2004; Skuse & Seigal, 2008). For example, ASDs have been associated with an increased risk of anxiety and depression (i.e., Davis et al., 2011), Prader-Willi syndrome with particular eating difficulties (i.e., Clarke, Boer, Chung, Sturmey, & Webb, 1996), and Down syndrome with an increased risk of dementia (see Chapter 8 in this handbook for a more detailed review of genetic syndromes and psychopathology in DD/ID).

People with DD/ID are also more susceptible to physical health problems often associated with their conditions. It is not clearly understood how these impairments impact on the emotional and behavioral well-being of people with DD/ID; however, it is thought that they may do so in ways similar to the general population. For example, youth with ID who have more physical health symptoms or chronic physical impairments have been found to be at higher risk for both concurrent and future new psychopathology (i.e., Taggart et al., 2010; Wallander, Dekker, & Koot, 2006). However, a small number of studies of adults with DD/ID with and without physical disabilities have reported that being mobile and not having severe physical disabilities was independently associated with mental ill-health (Cooper et al., 2007; Smiley et al., 2007), suggesting that physical disabilities may not be as important a risk factor as initially considered.

Communication, Sensory, Social, and Adaptive Behavior Functioning

Communication and language impairments may contribute to vulnerability to psychopathology in individuals with DD/ID, as people with such difficulties may find it difficult to express themselves and their needs and may instead engage in both internalizing and externalizing behaviors to communicate distress. Thus, psychiatric symptoms or challenging behaviors may serve the function of communication or emotional expression. Such difficulties may also place people with DD/ID at higher risk for exposure to negative life events, social isolation, discrimination, and abuse. However, the only studies to explore communication impairments independently of level of functioning have suggested that when level of functioning is controlled for, communication impairments are not independently associated with mental health difficulties (i.e., Cooper et al., 2007).

People with sensory impairments and DD/ID may also be at greater risk of mental health problems, via a pathway of increased social isolation, limited opportunities to access community resources, more restricted adaptive behavior repertoires, or an increased sense of loss (Estia Centre, 2006, see also Matson & Sevin, 1994). So far, however, this field of enquiry has received little attention. Few studies have attempted empirically to disentangle the independent effects of sensory and communication impairments over and above those of level of ID and these report no relationship between hearing impairment and mental ill-health (Cooper et al., 2007; Smiley et al., 2007).

Deficits in social and interpersonal skills and adaptive behavior are a defining aspect of DD/ID and affect many areas of functioning. Social skill and daily living adaptive behavior deficits have

been reported to predict DSM-IV disorders in children with ID 1 year later (Dekker & Koot, 2003b). Poorer pro-social behaviors have also been independently associated with increased prevalence of conduct difficulties in children with cognitive delays (Emerson et al., 2011). Studies of adults, too, have found that individuals with DD/ID who present with high levels of psychiatric psychopathology score significantly lower on measures of social skills (i.e., Kearney & Healy, 2011). Finally, a very small number of studies, mostly multiple case or small group studies, have reported initially encouraging results for adults with mild ID following social problem-solving skills training in reducing psychopathology and challenging behaviors (i.e., Anderson & Kazantzis, 2008; Nezu, Nezu, & Arean, 1991). However, most studies in adults with DD/ID have been cross-sectional or case reports and as such cannot establish whether social skills deficits predispose individuals with ID/DD towards higher rates of psychiatric comorbidity or vice versa.

Individual Psychological Factors

In individuals with ID with more severe intellectual impairments, it has proved very challenging to measure individual psychological factors known to be implicated in risk and resilience in the general population. There are particular problems in assessing areas such as self-awareness, self-esteem, problem-solving abilities, coping behaviors, or attributions of events, which require self-report and often complex metacognitive abilities. For this reason, few attempts have been made to explore the role of such factors in the development and maintenance of psychopathology in people with ID, with the exception of a small number of studies of people with mild to moderate ID (i.e., Dagnan & Sandhu, 1999; Hartley & MacLean, 2005, 2008; Reiss & Benson, 1985). These studies have generally shown that factors that are implicated in psychopathology in the non-ID population are also implicated in psychological distress in people with DD/ID. For example, Dagnan and Sandhu (1999) showed that adults with mild ID tend to compare themselves unfavorably to others and often feel inadequate. Other researchers have reported that they are more likely than their peers without ID to attribute failures to internal and stable factors and others' actions as hostile in ambiguous interpersonal situations resulting in increased affective or aggressive symptoms (i.e., MacMahon, Jahoda, Espie, & Broomfield, 2006; Pert, Jahoda, & Squire, 1999).

Coping difficulties accounted for a significant portion of the variance explaining psychological distress in a study of 127 adults with mild ID (Hartley & MacLean, 2008), with problem-focused coping being negatively correlated with psychological distress. Similar findings of lower emotion-orienting coping and higher self-esteem predicting higher life satisfaction and lower anxiety in a small mixed sample of youth with disabilities or chronic illnesses have also been reported (Dahlbeck & Lightsey, 2008). Findings from the limited research in this field strongly indicate that how individuals with DD/ID perceive, interpret, and cope with stressors may be important factors in conceptualizing and treating psychopathology in this group.

Family Factors

Attachment

Insecure attachment has been identified as a risk factor for the development of later psychopathology in individuals without DD/ID, and higher rates of insecure attachment relationships between children and youth with ID and their carers compared to those without ID have been reported in some studies (i.e., Atkinson et al., 1999; Ganiban, Barnett, & Cicchetti, 2000; Muris & Maas, 2004; Schuengel & Janssen, 2006). The challenges in forming and maintaining secure and long-lasting attachment relationships are likely to be a result of complex interactions between individual (cognitive and behavioral challenges associated with DD/ID),

parenting (i.e., increased stress, higher level of parenting skills, and sensitivity required), and social variables (i.e., multiple carers in group care, frequent changes of support staff, increased risk for abuse, and maltreatment; see Perkins, Holburn, Deaux, Flory, & Vietze, 2002; see also Janssen, Schuengel, & Stolk, 2002; Schuengel & Janssen, 2006). Although there are clear implications from incorporating attachment theory in conceptualizing and treating psychopathology in people with DD/ID (i.e., see Schuengel, Sterkenburg, Jeczynski, Janssen, & Jongbloed, 2009 for a multiple case design study providing exciting preliminary evidence that children with severe ID respond better to intervention delivered by a therapist who has developed a positive attachment relationship with them), few studies have been carried out investigating attachment styles and their relationship with psychopathology in adults with DD/ID. Clegg and Lansdall-Welfare (1995) and Clegg and Sheard (2002) reported a positive correlation between insecure attachment style and challenging behavior in adults with severe intellectual disabilities, but they only used a single question to measure attachment.

Two recent studies have investigated attachment behaviors in relation to challenging behavior or mental health in adults with ID. Controlling for level of functioning and the presence of ASD and using support staff as informants, De Schipper and Schuengel (2010) found a negative relationship between secure attachment to day-care staff and irritable, lethargic, and stereotyped challenging behaviors in their sample of 156 adults with moderate and severe ID. Larson, Alim, and Tsakanikos (2010) also found negative links between secure attachment, specific challenging behaviors (i.e., refusing medication), and depression in adults with mild to moderate ID using self and carer reports. Although limited, findings in this area support the attachment-psychopathology hypothesis (Janssen et al., 2002) in this population and suggest that promoting secure attachment relationships could thus be a target for the prevention and intervention of mental health and challenging behavior problems in DD/ID (see also Sterkenburg, Janssen, & Schuengel, 2008; Schuengel et al., 2009).

Family Composition

A single-parent home has been associated with increased psychopathology in children and youth with ID (Taggart et al., 2010; see also Witwer & Lecavalier, 2008 for a review of psychosocial risk markers and correlates of psychopathology in children with DD/ID), while living with both parents has been associated with less psychopathology in children with DD/ID (Koskentausta, Iivanainen, & Almqvist, 2007; Taggart et al., 2010). It remains unclear whether earlier history of separation, divorce, or single parenthood in their family is similarly associated with higher rates of mental health and behavioral problems in adults with DD/ID. The only study to explore this relationship (Smiley et al., 2007) found that a history of parental divorce in childhood, as reported at initial assessment, independently predicted problem behaviors at a second assessment 2 years later in their prospective cohort study of 651 adults with mild to profound ID. However, the direction of this relationship needs to be better established, as most studies have been cross-sectional and have not explored the complex relationship between family composition, other variables of psychosocial deprivation, and DD/ID over time.

Family Functioning and Parenting

Consistently with findings in the general population, a small number of cross-sectional studies of young people with DD/ID have shown strong associations between child psychopathology and the use of inconsistent and punitive parenting strategies (i.e., Emerson, 2003; Emerson & Hatton, 2007). Dysfunctional family functioning has also been found to be associated with or to predict future youth psychopathology independently of family SES in longitudinal studies (Dekker & Koot, 2003b; Tonge & Einfeld, 2003;

Wallander et al., 2006). In a prospective, longitudinal study of Australian children, Emerson et al. (2011) found that angry/harsh and inconsistent parenting experienced by children with cognitive delays at age 4/5 independently predicted (with the highest odds ratios together with male gender) both the prevalence and persistence of conduct difficulties at ages 6/7 and 8/9 years old. Their findings suggest that children with intellectual disabilities may be more likely than typically developing peers to show persistent conduct difficulties when exposed to multiple environmental risks, including harsh and inconsistent parenting. A small number of intervention studies targeting parenting skills and strategies in managing young children with developmental disabilities also report improvements in child behavior (i.e., Sanders, Mazzucchelli, & Studman, 2004). These findings suggest that less effective parenting may act as a precipitating factor in psychopathology among children with DD/ID. However, we could not identify any studies investigating parenting in relation to psychopathology in adults with DD/ ID.

There are two issues that are important to discuss here. Firstly, in most of the above studies, when earlier child psychopathology at initial assessment was controlled for, the predictive strength of family factors either decreased significantly at later timepoints or disappeared entirely. For example, Dekker and Koot (2003b) found that disruptive disorders in youth with ID were best predicted by inadequate daily living skills, social incompetence, high physical symptoms, and parental referral to mental health services. However, after controlling for initial child problem behaviors, only social incompetence remained significant; family factors were no longer significant. This suggests that individual and biological child factors may be stronger predictors of later psychopathology than family factors, although more research is needed to investigate this possibility. Secondly, despite the prospective longitudinal nature of these studies, the causal role of these factors cannot be determined with certainty, as it is difficult to establish precedence or to control for the possible accumulating effects of individual child or child disability factors relating to parental mental health and family functioning prior to the start of the studies.

Parental Stress and Psychopathology

Parental psychopathology has been independently associated with increased rates of emotional and behavioral difficulties in children and young people with DD/ID (Dekker & Koot, 2003b; Tonge & Einfeld, 2003; Wallander et al., 2006). Youth with ID and additional emotional or behavioral problems are more likely to have parents with mental health or substance abuse problems, than youth with ID without such difficulties (Taggart et al., 2010). Families of people with DD/ID frequently face additional challenges in their lives (i.e., increased financial burden, caring for their child with DD/ID well into adulthood, complex roles as carers), and increased stress has often been reported in the literature in parents of children and adults with DD/ID. Reviewing the literature on parenting stress in families of people with ID is beyond the scope of this chapter (but see Hill & Rose, 2010; Miodrag & Hodapp, 2010 for recent reviews). A smaller number of cross-sectional studies examining parenting stress as a variable implicated in comorbid emotional and behavioral difficulties in this population have associated maternal stress with internalizing and externalizing symptoms in children and youth with ID (Baker, Blacher, Crnic, & Edelbrock, 2002; Hastings, 2002; Hastings, Daley, Burns, & Beck, 2006). Hastings et al. (2006) provided some initial evidence for the temporal precedence of maternal stress in their 2-year follow-up longitudinal study. They found that earlier maternal stress predicted later child externalizing problem scores, but also that earlier child problem scores predicted later maternal stress, indicating a bidirectional relationship between maternal stress and child psychopathology in youth with DD/ID (see also Gray et al., 2011). Although there are strong conceptual reasons to assume that parental stress can both lead to and is an outcome of psychopathology in DD/ID, the investigation of the direction and processes involved in this interaction remains limited and deserves considerably more focus.

Socio-Ecological Factors

Culture, Ethnicity, and Ethnic Minority Status

In children with DD/ID, Emerson, Robertson, and Wood (2005) reported that teachers' ratings of emotional and behavioral needs of black children were much higher than their ratings of South Asian children, although it is not clear whether this reflected actual higher problems or biased perspectives by teachers (Littlewood, 2006). It is apparent, too, that ethnicity is likely to affect access to mental health services for young people and adults with DD/ID, with people from South Asian communities accessing services least (Raghavan & Waseem, 2007). Emerson and Einfeld (2010) also reported significant cross-national variation in the association between family socioeconomic circumstances (SECs), DD/ID, and emotional and behavioral difficulties in preschool children with DD/ID in Australia and the UK. They found higher rates of psychopathology associated with higher risk of exposure to adverse SECs to a much larger extent in the UK than Australia. These studies collectively point out that it is important for future research to explore the possible moderating effects of culture, nationality, or ethnic minority group membership in the relationship between risk factors and psychopathology in DD/ID.

To our knowledge, only one published study has specifically investigated the role of ethnicity in the prevalence of mental health problems and psychopathology in adults with DD/ID. Tsakanikos, McCarthy, Kravariti, Fearon, and Bouras (2010) analyzed rates of ICD-10-based psychiatric diagnoses (including schizophrenia and ASD, depressive disorders, anxiety disorders, personality disorders, dementia, and adjustment reaction disorders) in all adults with ID referred between 1984 and 2004 to a specialist mental health service for adults with ID in South London, UK ($N = 804$). Participants were grouped in White, Black, and other nonwhite ethnic minority groups. Schizophrenia and ASD were significantly overrepresented in the ethnic minority groups. Adults with ID from ethnic minority groups were also less likely to live in supported housing placements and were younger when diagnosed with comorbid psychiatric disorders. In their review of the impact of ethnicity for people with LD and mental health difficulties, McCarthy, Mir, and Wright (2008) urged researchers to explore the influence of ethnicity on the presentation of mental health problems in DD/ID and to investigate whether the approaches that have been successfully used in improving access to mental health services for people without DD/ID can also benefit people with DD/ID and their families. Practitioners also need to be mindful of the cultural and spiritual/religious needs of individuals with DD/ID and their carers, which may often be ignored or misinterpreted resulting in increased vulnerability to mental health problems.

Socioeconomic Circumstances

There is a strong and consistent association between having mild ID and lower SEC (i.e., McDermott, Durkin, Schupf, & Stein, 2007), and a well-established link between socioeconomic adversity and psychopathology in the general population (i.e., Green et al., 2005; Marmot & Wilkinson, 2006). Dekker and Koot (2003b) reported that rates of psychopathology were higher in children with ID and lower levels of SEC compared to those of higher SEC; people with ID are also more likely to be exposed to socioeconomic risk factors than people without disabilities (Emerson & Hatton, 2007). A number of largely cross-sectional studies have also reported that lower family income and poverty, living in households with no paid employment or in deprived neighborhoods, and poorer parental educational qualifications have been associated with higher rates of psychopathology in youth and adults with DD/ID (Emerson & Hatton, 2007; Emerson et al., 2005, 2011; Taggart et al., 2010). Only one prospective longitudinal study has identified earlier poor socioeconomic environments as predictors of differential pathways of persistent behavioral difficulties over time in children with cognitive delays as compared to children with ID not exposed to these poorer SEC (Emerson et al., 2011).

Overall, however, the relationship between SEC and psychopathology in individuals with ID/DD has not been well established with some studies finding no relationship (i.e., Dekker & Koot, 2003b; Tonge & Einfeld, 2003; Smiley et al., 2007). It is highly probable that the few and inconsistent findings reported in this area to some extent are accounted for by the fact that studies reporting links between SEC and psychopathology often do not control for other mediating variables (such as parental mental health, employment levels, education, family composition).

Living Arrangements

Residential placements for adults with ID have been found to be associated with psychopathology, with those living in settings other than family homes being at higher risk (Cooper et al., 2007; Taggart et al., 2010) and those living with family carers at lower risk (Smiley et al., 2007). This association, however, may weaken with age (Lifshitz, Merrick, & Morad, 2008). In a sample of 750 first-time referrals to a specialist mental health service for people with ID and controlling for level of ID and age, Chaplin et al. (2010) found that the risk for diagnosis of psychiatric disorders (specifically schizophrenia spectrum and personality disorders) was significantly higher in individuals who lived independently compared to those who lived with their families or in supported housing. The opposite pattern was found for anxiety disorders. However, psychopathology may also influence type of residence. For example, an additional diagnosis of personality disorders in people with ID may make family life or supported accommodation arrangements more challenging to maintain (Tajuddin, Nadkarni, Biswas, Watson, & Bhaumik, 2004; Tsakanikos et al., 2010). Alternatively, the type and quality of residential care may influence psychopathology in this vulnerable population via exposing the individual to additional stressors precipitating additional psychopathology or through differential care pathways affecting referral rates for psychiatric or behavioral disorders (Johnson, Griffiths, & Nottingham, 2006; Tsakanikos et al., 2010). Tsakanikos, Bouras, Costello, and Holt (2006) reported that the most frequent life event reported in their study of exposure to life events in adults with ID was "moving house or residence." Although causation cannot be assumed, these findings indicate the importance of carefully considering the quality of residential provision for individuals with ID, as it is likely that this will affect their emotional well-being and mental health.

Clearly, the relationship between the social and economic factors reviewed above and psychopathology in DD/ID is complex. Future studies need to explore the independent and relative contribution of these factors in the development and maintenance of psychopathology in prospective longitudinal studies. Similarly, the possible impact of DD/ID on social adversity needs to be better understood. What is also needed is a better understanding of the impact of lower SES in individuals with varying levels of ID and in individuals with different phenotypic profiles.

Life Events and Traumatic Experiences

A number of studies have been published in this area examining (a) whether individuals with DD/ID experience higher rates as well as different types of life event stressors specific to ID compared to the general population or (b) whether experiencing negative life events (including bullying, trauma, discrimination, abuse, separation, loss, and bereavement) is associated with increased psychiatric and behavioral problems in adults with DD/ID. Typically, life event studies in DD/ID have been mostly cross-sectional, retrospective, and based simply on the presence/absence of such events over a specified period.

Coe et al. (1999) interviewed mothers and teachers of 88 children with ID, half of whom had Down syndrome, and showed a significant association between life events and children's scores on the Revised Behavior Problem Checklist. In a secondary analysis of data collected by the 1999 Office of National Statistics survey of the *Mental Health of Children and Adolescents in Great Britain* study (Meltzer,

Gatward, Goodman, & Ford, 2000), Hatton and Emerson (2004) compared lifetime experience of negative life events and psychopathology in 264 children with ID and 10,040 children without ID using primarily parental report. Youth with DD/ID were more likely to experience a greater number and range of adverse life events compared to the non-ID group, including parental financial crisis; death of parent, sibling, or grandparent; parental separation; and serious illness requiring hospitalization. Some of the variance in life events between the two groups was explained by family poverty and deprivation. In addition, the accumulation of two or more adverse life events substantially increased the odds ratio for emotional or conduct disorders in the ID group. Taggart et al. (2010) compared 155 young people aged 11–19 with ID and emotional/behavioral difficulties (EBD) to 94 youth with ID without additional EBD as reported by teachers. Although some of the differences between the two groups disappeared when level of ID was controlled for, the EBD group was more likely to experience higher number of life events (including parental court appearance; removal into state care; reconstituted family and new parental figure; exposure or allegation of verbal, physical, and sexual abuse; being bullied; and having troubles with the community), suggesting that increased life events are associated with increased psychopathology. Taken together, these findings provide initial evidence for the relationship between life events and emotional and behavioral problems in youth with DD/ID.

Several studies of adults with DD/ID provide evidence of higher rates of negative life events compared to the general population (i.e., Owen et al., 2004) and of an association between negative life events and comorbid psychopathology (i.e., Cooper et al., 2007; Dagnan, & Waring, 2004; Hamilton, Sutherland, & Iacono, 2005; Hulbert-Williams, Hastings, Crowe, & Pemberton, 2011; Smiley et al., 2007; for reviews see Hulbert-Williams & Hastings, 2008; Wigham, Hatton, & Taylor, 2011). Some studies have identified a specific link between adverse life experiences and affective disorders (i.e., Cooper, Melville, & Morrison, 2004; Hastings, Hatton, Taylor, & Maddison, 2004; Hatton & Emerson, 2004; Owen et al., 2004; Taggart et al., 2010). Some also suggest a small effect of life events on challenging behaviors and behavioral problems (Hatton & Emerson, 2004; Owen et al., 2004), although no relationship with psychotic symptoms has so far been established (Cooper et al., 2007; Hastings et al., 2004; Owen et al., 2004). Few of these studies have attempted to control for variables potentially moderating the relationship between life events and psychopathology, such as age, gender, or level of functioning (see Hulbert-Williams & Hastings, 2008 for a more detailed discussion on this issue), but in those that have the added contribution of life events to the statistical models remained significant (Cooper et al., 2007; Esbensen & Benson, 2006; Hamilton et al., 2005; Hastings et al., 2004; Owen et al., 2004;). Even fewer researchers have employed a prospective longitudinal design to show the temporal association between the two variables—that is, life events as a risk factor preceding increases in psychopathology (i.e., Esbensen & Benson, 2006; Monaghan & Soni, 1992). These studies have provided such temporal evidence with generally small to medium effect sizes, but data on earlier life events were in most cases collected retrospectively. Causal attributions still cannot be made, as none of the studies examined both possible temporal directions to clearly establish the direction of the observed associations. Thus far, the evidence appears to strongly suggest that life events are likely to affect the well-being of people with DD/ID, although various definitions and operationalizations of life events have made it difficult to pull studies together and to draw conclusions on the rates, types, and effects of life events in psychopathology of people with DD/ID. In addition, the majority of studies have focused simply on the presence or absence of life events—only Esbensen and Benson (2006) have made an effort to assess the relationship between the perceived impact of these events (i.e., positive, negative, or little influence) and psychopathology.

Another pertinent issue is whether it is conceptually and clinically useful to distinguish between traumatic and normative life events and to examine whether these have potentially differential impacts on the psychopathology of individuals with DD/ID. Although it is often difficult to distinguish between traumatic experiences and life events (Martorell & Tsakanikos, 2008), Martorell et al. (2009) have attempted to distinguish between traumatic experiences (i.e., sudden death of close relative or friend, accident, assault or stalking, sexual abuse or assault, being in a natural disaster) and "normative" life events (i.e., death of close relative, moving residence, relationship breakup, serious illness, and problems with friends or relatives) in assessing their relationship with psychopathology in people with mild and moderate ID. Both traumatic experiences and life events increased the odds of a mental health disorder, but when entered together in the model only traumatic experiences remained significant predictors, providing some initial support that traumatic experiences may play a more important role in psychopathology in ID than life events. The authors, however, acknowledged that more research is needed to establish this, as data regarding traumatic experiences were lifetime compared to life events which were in the last 1 year only. Measures of these experiences may be to a large extent overlapping. In efforts to explore possible specific effects of trauma on emotional well-being and behavior of people with DD/ID, a small number of studies have identified associations between experiences of trauma and increases in stereotypical, aggressive, irritable, and challenging behaviors and reduced self-care—these are effects typically not considered in general population measures of the effects of trauma (see Wigham et al., 2011 for a review).

The only study that explored whether single or multiple exposure to life events was associated with different psychopathology presentations was the retrospective study by Tsakanikos, Bouras, Costello, et al. (2006). Using the Life Events list of the Psychiatric Assessment Schedule for Adults with Developmental Disorders Checklist (PAS-ADD Checklist; Moss et al., 1998) and psychiatric diagnoses based on ICD-10 criteria made by clinicians, they examined life events during the last 12 months and psychiatric disorders in 281 men and women with ID consecutively referred to a specialist mental health service. Single exposure to life events was associated with being female, schizophrenia, personality disorders, and depression. Multiple exposure to life events was associated with personality disorder, depression, and adjustment reaction. It may be particularly informative to investigate the role of multiple/cumulative negative life events, as it can be useful to explore whether some cutoff point for the number and severity of such events can be established as a way to screen for people with DD/ID who may be at higher risk for mental health or behavioral disorders (Martorell & Tsakanikos, 2008).

It is likely that the relationship between life events and psychopathology in people with DD/ID shares at least some similarities with that in the general population and that this relationship is complex and moderated by other factors. For instance, Owen et al. (2004) showed that males with DD/ID, those with higher adaptive behavior functioning and those living in staffed accommodation experienced more negative life events (see also Hamilton et al., 2005; Hastings et al., 2004). Patti, Amble, and Flory (2005) also reported that older adults with Down syndrome experienced more negative life events compared to adults with DD/ID but without Down syndrome. Hamilton et al. (2005) suggest that the strength of the association between life events and emotional/behavioral disturbances may vary by type and level of disability, but most of the research in this field has reported findings irrespective of subgroup. Understanding the direction and dimensions of the relationship between life events and psychopathology in different subgroups of people with DD/ID needs to be a priority in future research.

Finally, some studies have specifically addressed specific life event/trauma types and we briefly discuss loss and bereavement, abuse, bullying, and stigma/discrimination, as people with DD/ID have been identified as more vulnerable in experiencing these compared to the general population.

Loss and Bereavement

In a review of emotional, psychiatric, and behavioral responses to bereavement in people with ID, Dodd, Dowling, and Hollins (2005) concluded that loss and bereavement is followed by increases in anxiety and depression symptoms and deterioration or alterations in usual patterns of behavior or functioning (see also MacHale & Carey, 2002). In an earlier study, Hollins and Esterhuyzen (1997) compared 50 adults with ID who had lost a parent in the previous 2 years and a matched non-bereaved comparison group and found significantly higher rates and cases of psychopathology in the bereaved group. A follow-up of the same sample approximately 5 years later found that most of the participants in the bereaved group no longer met "caseness" criteria for psychopathology, indicating that the increase in psychopathology was precipitated by the experience of bereavement and declined thereafter (Bonell-Pascual et al., 1999). In one of the few studies in which individuals with ID themselves were interviewed, Harper and Wadsworth (1993) reported a number of disruptions in their lives following the death of a loved one, including anger, anxiety, depression, and sleep disruptions. These findings suggest a potential causal role of bereavement as a life event in the development of psychopathology in this population. Large-scale prospective longitudinal studies using agreed diagnostic criteria and valid assessment measures to establish the nature, severity, and effects of the experience of bereavement in individuals with ID over time are clearly needed in this field of enquiry.

Abuse and Maltreatment

Govindshenoy and Spencer (2006) systematically reviewed the literature on population-based cohort studies of children (<18 years) with physical and intellectual disabilities that reported on the links between abuse and disability. Only four studies met their inclusion criteria (Brown, Cohen, Johnson, & Salzinger, 1998; Sidebotham, Heron, & ALPSAC Study Team, 2003; Spencer et al., 2005; Vizcarra, Cortes, Bustos, & Munoz, 2001), indicating that the evidence for the links between abuse, disability, and psychopathology in children and youth with DD/ID remains weak. Three of these studies reported an association between abuse and increased emotional and behavioral problems in children with disabilities, but this association lacked systematic empirical evidence. At least in relation to children and youth with DD/ID, there is insufficient good quality evidence to support the commonly held view that they are at increased risk of abuse and neglect and that this experience is implicated in increased emotional and behavioral problems.

In a 2-year prospective study of all the registered adults with ID in the wider Glasgow area, Smiley et al. (2007) also found the experience of known abuse, neglect, or exploitation during adult life to be associated with later episodes of mental ill-health. Sequeira and Hollins (2003) reviewed 25 studies published between 1974 and 2001 exploring the psychological effects of sexual abuse in people with learning disabilities. The majority of these reports were retrospective, case studies, lacking in comparison groups or any operational definition or systematic assessment of sexual abuse, and relied on parent or staff anecdotal report of abuse, thus considerably limiting the validity of the findings. Nevertheless, most reported post-abuse difficulties similar to those found in the non-ID population, including post-traumatic stress disorders (PTSD), withdrawal, aggressive behaviors, inappropriate sexualized behaviors, anger, poor self-esteem, anxiety, low mood, alcohol abuse, self-harm, and nightmares (see also Matich-Maroney, 2003 and O'Callaghan, Murphy, & Clare, 2003).

Bullying

Few researchers have explored bullying towards and by individuals with DD/ID and its relationship with psychopathology, but existing data with children and adults with disabilities shows that they are at high risk of being bullied and/or of bullying others (i.e., Bramston, Fogarty, & Cummins, 1999; Dickson, Emerson, & Hatton, 2005; Kaukiainen et al., 2002; Norwich & Kelly, 2004).

In typically developing youth and adults without ID, the associations between direct and relational bullying and emotional and behavioral difficulties are well documented (i.e., Wolke, Woods, Bloomfield, & Karstsdt, 2000). It is thus surprising that we could not identify any studies focusing on bullying and psychopathology in either youth or adults with DD/ID in the existing literature, with the exception of a recent anti-bullying intervention study for adults with ID reporting lower rates of victimization in the treatment but not the control groups post-intervention and at 3-month follow-up (McGrath, Jones, & Hastings, 2010). Mishna (2003) notes that bullying and learning disabilities may pose a risk for social, emotional, and behavioral difficulties, but at present our empirical investigations in this area are very limited and our hypotheses regarding the possible role of bullying in psychopathology are largely drawn from research not specific to DD/ ID.

Stigma and Discrimination

Although the concept of stigma has received considerable theoretical and empirical attention in relation to psychiatric disorders, fewer efforts have been made to understand how stigma may be implicated in psychopathology in DD/ID. Partly, this could be due to the complexity of the construct of stigma (which incorporates negative attitudes and stereotypes, prejudice, and behavioral reactions), as well as the different types of stigma (including public, self, and family; see Werner, Corrigan, Ditchman, & Sokol, 2012 for an excellent recent review of stigma in DD/ID). Among people with different types of disabilities, those with intellectual disabilities are among the most stigmatized (i.e., Hernandez, Keys, & Balcazar, 2000). Many individuals with DD/ID are aware of the stigma associated with their disabilities and experience emotional pain as a result of this (see Beart, Hardy, & Buchan, 2005 for a review). Their families also report significant distress often associated with stigmatizing experiences (i.e., Mak & Cheung, 2008). However, it may be more relevant to investigate links between self-stigma and psychopathology, the persons' own experiences and their internalization of stigma which may lead to lower self-esteem, more negative feelings, and behavioral withdrawal or avoidance (Corrigan & Watson, 2002). Stigma and discrimination may also impact psychopathology via being a barrier in the access to appropriate services for people with DD/ID (Gill, Kroese, & Rose, 2002). There are currently very few reliable and valid measures to assess self-stigma in DD/ID, particularly in individuals with moderate to profound disabilities (Werner et al., 2012), which may explain the limited research initiatives in this area.

General Discussion and Recommendations for Future Research

Attempting to pull together the current research findings on psychosocial variables in psychopathology in DD/ID is problematic for a number of reasons largely relating to (a) poor methodologies; (b) diverse samples, definitions, and measurements of key constructs; and (c) largely one-dimensional and unidirectional investigations of the relationships between the variables under study.

The majority of the studies reviewed in this chapter have been cross-sectional, and although associations between psychosocial factors and psychopathology in DD/ID were reported with some confidence, the temporal precedence or potential causal mechanisms cannot yet be established (Kraemer et al., 1997; see also Witwer & Lecavalier, 2008). Furthermore, most of the studies relied on retrospective psychosocial information, obtained from informants' memories or clinical case records. Group sizes are typically small and participants were often recruited from both community and clinical samples, which may result in different rates of comorbid difficulties and contribute to the inconsistent findings in relation to the patterns or strengths of identified relationships. Many of the samples have been self-selected or convenience samples and have been poorly described, with inadequate

information on levels of adaptive functioning or intellectual impairment and lacking in any operational definition of DD/ID. Prospective longitudinal studies from early childhood to adulthood with large representative population-based and well-characterized samples are rare. However, these are essential in order to examine the independent and combined contributions of several psychosocial factors in the development and maintenance of psychopathology in people with DD/ID over time (for promising recent efforts see de Ruiter et al., 2007; Einfeld et al., 2006; Emerson et al., 2011; Gray et al., 2011; Smiley et al., 2007; Wallander et al., 2006).

Measurement limitations are also notable. Measuring and diagnosing psychopathology in DD/ID is challenging, as existing diagnostic criteria may not be appropriate for this group. Diagnostic criteria developed specifically for people with DD/ID may result in different and non-comparable rates of psychopathology and different studies have used a range of systems to assess and diagnose psychiatric disorders (see Chapters 3 and 4 of this handbook for more on assessment of psychopathology in DD/ID). For example, using screening checklists or rating scales, some researchers have reported rates of psychiatric symptoms, while others have used diagnostic tools and clinical interviews by specialist professionals to report rates of formally diagnosed psychiatric disorders (i.e., Tsakanikos, Bouras, Costello, et al., 2006). Instruments such as the PAS-ADD Checklist (Moss et al., 1998) have attempted to overcome some of these challenges, but difficulties remain. A further issue in this field is whether challenging behavior constitutes a symptom of other psychiatric comorbidity or is a psychiatric disorder in itself (see Hemmings, 2007; McCarthy et al., 2010 for further discussion). Moreover, general population measures of life events may not capture the range or types of life events experienced by people with intellectual disabilities (e.g., see Bramston et al., 1999 in relation to group living; Hollins & Esterhuyzen, 1997 in relation to dependency on carers). In addition, most researchers have relied on proxy informants including parents and staff to provide information and very few have included self-reports by the individuals with DD/ID themselves. Memory, proximity of relationship, and social desirability biases are inherent problems when relying on other informants, and future research should aim to include multiple sources of information including the individuals with DD/ID themselves (for good examples of this, see Cooper et al., 2007; Smiley et al., 2007). Measures need to be validated to be applicable to people with DD/ID and a consensus needs to be reached as to the types of life events to be included in assessing risk of psychopathology.

To date, research on psychosocial factors related to psychopathology in DD/ID has generally relied on correlational investigations of a small number of factors. The nature, frequency, severity, or duration of individual, family, and social adversity factors has rarely been measured. Although there was initially a need for such studies in order to establish some tentative links between particular psychosocial variables and psychopathology in this population, we now need to move towards exploring more complex conceptual models and using more sophisticated methodological and statistical designs that focus on the mechanisms through which psychosocial and DD/ID variables influence and are influenced by psychopathology over time. Such investigations should be based on available theoretical frameworks and empirical evidence drawn from the existing research reviewed here as well as available frameworks in the non-ID population (see an example by Woolfson, 2004). However, concepts or frameworks adopted from the non-ID population may or may not apply to individuals with DD/ID who present with a wide range of abilities, experiences, and environments. Even if etiological factors implicated in psychopathology of people with DD/ID are similar to those of the general population, their onset, duration, course, and prognosis may be different for individuals with DD/ID and comorbid difficulties (Konarski & Cavalier, 1982). At the same time, the fact that people with DD/ID have higher rates of psychopathology compared to people without DD/ID suggests that there may be either a higher

risk of exposure to the same factors or additional variables associated with having DD/ID which put these people at higher risk of comorbid psychopathology. The evidence reviewed in this chapter suggests that a number of psychosocial factors implicated in psychopathology in the general population may interact with intellectual disability to account for the increased rates of psychopathology in this population through complex pathways that still require systematic investigation. Similarly, there appears to be some evidence that some factors more specific to DD/ID may also be implicated (i.e., relationship with carers, group living, social participation, stigma towards disabilities). We urgently need better to understand these in order to develop more informed, appropriate, and useful preventative, primary, and tertiary care services for individuals with DD/ID and comorbid psychiatric, emotional, and behavioral difficulties and their families/carers.

Clinical Implications and Recommendations

Children, youth, and adults with DD/ID are at increased risk of various types of psychopathology, and this risk is persistent across the life span. The present review of psychosocial factors likely to be implicated in the onset and maintenance of comorbid conditions has highlighted that a range of individual, family, and socio-ecological factors need to be considered. Although the effects of the relationships identified so far in the literature appear to be of a small to medium effect size, a thorough clinical assessment of psychosocial past and current factors in the individual's life is crucial in better understanding the person's difficulties and in developing effective individualized treatments.

Clinicians need to obtain as accurate information as possible with regard to the individual's social and family past and present circumstances (including current and past social circumstances, transitions and major life changes, support networks, opportunities for work and activities, family history and relationships, parenting and carers' psychopathology, living arrangements, interpersonal and other losses, bullying, victimization, and abuse; see also O'Hara, 2007 for a comprehensive review of multimodal assessment for mental health problems in people with ID). Life events are likely to act as precipitating factors and thus should specifically be assessed. Clinicians need to make efforts to obtain this information from the person themselves as well as other informants and to explore not only the presence of these potentially factors but also their impact on the person with DD/ID. These psychosocial variables need to be incorporated into a dynamic formulation of the individual's presenting difficulties which should include a thorough understanding of the biological, genetic, and developmental factors also likely to be implicated.

Regardless of whether psychosocial adversities are the cause or consequence of psychopathology in individuals with DD/ID (and they are likely to be both), it seems clinically and socially important to assess and monitor these variables and to provide continuous support to the individual and their systems with a focus on promoting well-being, decreasing risk, and increasing resilience. It is currently acknowledged and accepted that with appropriate adaptations a number of evidence-based psychosocial interventions and supports for people with developmental disabilities (mostly with a focus on cognitive, behavioral, or environmental strategies) can result in clinically significant benefits for many individuals, their families, and carers (Hurley, 2005). In terms of intervention planning and research, Emerson and Einfeld (2010) strongly urged towards public efforts that seek to reduce the population-level prevalence of psychopathology in individuals with DD/ID. Policy interventions aiming to reduce family poverty, improve parental mental health and well-being, improve access to and quality of health care, and train carers to recognize and respond to psychological distress are also likely to have direct and indirect effects on psychopathology of people with DD/ID (see also Hatton & Emerson, 2004).

Summary and Conclusions

In this chapter, we aimed to critically review the available literature on psychosocial factors likely to be implicated in the development and maintenance of comorbid psychiatric, emotional, and behavioral difficulties in individuals with DD/ID. We report on research findings relating to individual (age, gender, level of functioning, individual psychological factors), family-related (attachment, parenting, parental stress and mental health, family composition), and socio-ecological (ethnicity, SECs, living arrangements, and life events) factors that have so far received theoretical and empirical attention in this population. Several of these variables can help better explain individual differences in mental health and psychopathology in DD/ID, but studies so far have been limited by cross-sectional, small-scale correlational designs focusing on one or two factors only.

It is now time consistently to adopt a developmental psychopathology framework in our efforts to better understand the psychosocial factors that may increase, decrease, or maintain psychopathology in individuals with DD/ID. Such an approach will help us better understand how and why certain individuals with DD/ID recover, while in others comorbid problems persist (Cicchetti & Cohen, 1995). The emphasis needs to be in bridging psychosocial and behavioral genetics research (Emerson & Hatton, 2007). Risk factors need to be better understood, as do protective factors (such as secure attachments, supportive relationships with carers and teachers, achievements, social support, friendships and social networking, and community participation), which have largely so far been ignored (Foundation for People with Learning Disabilities, 2003).

Despite the existing limitations and challenges, our understanding of psychosocial variables in relation to psychopathology in DD/ID has improved substantially over recent years. Addressing some of the challenges discussed here and coordinating larger-scale longitudinal research and policy efforts in the near future can further advance our knowledge and help create psychosocial environments that are healthier and more protective against comorbid psychopathology for individuals with DD/ID across the life span.

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Part III

Co-morbid Psychopathology

Schizophrenia Spectrum Disorders 10

Colin Hemmings

Introduction

Historical Context

It has long been thought that the relationship between intellectual impairment and psychosis may provide important clues to the aetiology of psychosis in general (Offord & Cross, 1971). Kraepelin (1919) described *pfropfschizophrenie* as being psychosis literally 'engrafted' onto *idiocy* and accounting for up to 7 % of cases of *dementia praecox*. He believed that *pfropfschizophrenie* was an early onset and severe form of psychosis in pre-existing ID. Kraepelin also believed that a schizophrenia-type psychosis beginning in the first decade of life could itself cause 'mental defect' (Heaton-Ward, 1977). For a long time after however most researchers believed that the joint occurrence of both conditions in the same individual was merely a chance combination (Heaton-Ward, 1977). There was accordingly a lack of research interest in the study of co-morbid ID and schizophrenia (Turner, 1989). More recently there has been much renewed interest in the context of much better recognition and understanding of mental disorders in people with ID.

Nomenclature

The term *schizophrenia spectrum disorder* (SSD) is widely accepted to include the schizophrenias as well as schizoaffective, persistent delusional and schizoptypal disorders. The evidence base regarding the epidemiology, risk factors, neuropathology, psychopathology, management and prognosis of SSDs in people with ID will be reviewed. This chapter will focus (whenever possible) on SSDs in people with ID as opposed to the less specific category of (non-affective) psychosis, which is perhaps more diagnosed in clinical practice and thus has often been used instead of SSDs in the research literature.

Epidemiology

Diagnosis

SSDs in people with ID are generally more difficult to detect and diagnose than in the general population. Sometimes a specific diagnosis of an SSD in people with ID is impossible. 'First rank' (Schneider, 1959) positive symptoms of schizophrenia are conceptually complex and thus often difficult to reliably assess in people with ID

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(Hucker, Day, George, & Roth, 1979). There may be difficulties distinguishing true delusional beliefs and hallucinations from self-talk, fantasies and talking to imaginary friends, which can be developmentally appropriate for a person with ID (Heaton-Ward, 1977; Hurley, 1996). People with ID often 'remember' conversations and think out loud about them. They might not realise that this is 'socially inappropriate'. Develop-mentally appropriate features may become more prominent and exaggerated when under stress (Hellendoorn & Hoekman, 1992). Formal thought disorder may also be difficult to elicit in people with ID for it can often be difficult anyway to follow the thread of their speech. People with ID are also often hypersensitive to how they are perceived by others and this can often be grounded in real-life experiences of being mocked and rejected (Hurley, 1996). This hypersensitivity may be mistaken for delusional persecutory thinking. It has also been suggested that people with ID are more likely to be able to be persuaded of the falseness of the delusional beliefs. It may be more important to observe for their readiness to restate their beliefs when not so subject to suggestibility (Hurley, 1996). Apparent 'negative' symptoms can also be misattributed to a psychotic disorder when they may be due to many other factors such as institutionalisation, over-sedation, lack of environmental stimulation and severe cognitive impairment (James & Mukherjee, 1996). It may be difficult to discriminate psychotic symptoms from baseline features of ID, a diagnostic problem which Sovner and Hurley (1986) termed 'baseline exaggeration'. However, the difficulty of differentiating psychotic symptoms and features related to a person's ID might also lead to the problem of 'diagnostic overshadowing' (Reiss, Levitan, & Zyszko, 1982) and psychosis therefore being actually under-diagnosed in this service user group.

One of the particular problems with diagnosing schizophrenia in people with ID is the difficulty of differentiating it from autism (Bakken, Friis, Lovoll, Smeby, & Martinsen, 2007; Bradley, Lunsky, Palucka, & Homitidis, 2011; Helevershou, Bakken, & Martinsen, 2008; Palucka, Bradley, & Lunsky, 2008). Sometimes it may be virtually impossible to be definitive as to whether a person has autism instead of, or as well as, schizophrenia unless an adequate developmental history is available (James & Mukherjee, 1996). Unusual preoccupations held rigidly by people with autism may be particularly difficult to differentiate from delusions. Flattened and incongruent affect, poor non-verbal communication and poverty of speech are commonly seen in autism as well as schizophrenia and so also lead to diagnostic difficulties. The term 'autism' itself was actually coined by Bleuler (1911) to describe social withdrawal in schizophrenia. He included autism as one of the four core characteristics of schizophrenia. Kolvin (1971) though clearly later showed that the pervasive developmental disorder of childhood autism was distinct from schizophrenia and other psychoses. There has been growing research activity into the associations between ID, autism and psychopathology including symptoms of SSDs (Matson & Shoemaker, 2009). It still remains unclear as to whether the risk of SSDs in people with ID and autism is greater than in non-autistic individuals with ID.

Diagnostic Criteria

Research has been hampered by inconsistencies in diagnostic criteria and rating instruments used (Sturmey, 1999). A major reason for the inconsistency of approaches has been the continued doubts about the usefulness of standard diagnostic classification systems in diagnosing mental disorders in those with more severe ID. The major difference in the various approaches used has been whether researchers have used standardised criteria or criteria modified for people with ID. For example, Hucker et al. (1979) modified the Research Diagnostic Criteria of Feighner et al. (1972) in their study of schizophrenia in people with ID and thus included features such as 'behaves as if hallucinated' as evidence of the disorder. Nor have definitions of the term 'intellectual disabilities' been consistent. However, recent improvement in consistency of criteria used in studies of mental disorders in people with ID has allowed greater comparison between them. The specific problems of diagnosis of schizophrenia

in people with ID have improved in recent years with the development of standardised assessment instruments for this service user group (e.g. Hatton et al., 2005). In people with mild ID and good verbal skills, it seems that the standard classification systems can be used with reliability and validity (Dosen & Day, 2001). By analogy, ICD and DSM criteria for schizophrenia can be reliable when used with children with typical IQ as young as 7 years old (Green, Padron-Gayol, Hardesty, & Bassiri, 1992). The linguistic competence of a normal 7-year old is similar to an adult with moderate ID (Melville, 2003). The developmental perspective is helpful conceptually, as complex delusions and hallucinations are unusual in young children but become more frequent in older children and children with a higher IQ (Watkins, Asarnow, & Tanguay, 1988).

Most of the research literature on SSDs in ID has focused on people with mild ID (IQ 50–70). Reid (1972) influentially argued that it is impossible to diagnose schizophrenia in people with limited verbal communication. In practice, this means below an IQ level of around 45. A person with more severe ID has limited verbal expression and ability to employ abstract reasoning, which are essential for the manifestation of the complex, subjective experiences of the language-based symptoms of schizophrenia. Sovner (1986) has described this phenomenon as 'intellectual distortion'. Sovner and Hurley (1986) also argued that 'cognitive disintegration', or psychotic-type reactions to stress, occurred in people with ID because of their limited coping abilities. Higher incidences of SSDs have been found in those with mild ID compared with more severe ID (Bouras & Drummond, 1992; Cowley et al., 2004). It remains unclear whether this reflects reality or whether it merely reflects the problems in detection of psychosis in people with more severe ID. Most researchers probably share the view of Heaton-Ward (1977) that a person's degree of insight and touch with reality cannot be fully assessed if they do not have a reasonable level of intelligible communication. However the study of Einfield et al. (2007) showed that at least there was good inter-rater reliability among experienced clinicians about the clinical diagnoses of psychosis in people with ID.

Prevalence and Incidence

There have been long-standing difficulties with establishing true incidence and prevalence rates of SSDs in people with ID. Taking into consideration all of the foregoing problems in accurately diagnosing SSDs in people with ID, epidemiological studies have converged around point prevalence rates of around 3 % and this figure is now widely accepted (Deb, Thomas, & Bright, 2001; Fraser & Nolan, 1994). This prevalence rate is around three times higher than in the general population although still thought to be a possible underestimate of the true figure (Turner, 1989). In a population-based cohort study of people with ID, Cooper et al. (2007) found the 2-year incidence of non-affective psychoses to be 1.4 % and for the first episode incidence to be 0.5 %.

There are three major possibilities for the increased prevalence rates of SSDs in people with ID (Blackwood, Thiagarajah, Malloy, Malloy, & Muir, 2008). First, a lowered IQ could be a premorbid sign of schizophrenia (Gunnell, Harrison, Rasmussen, Fouskakis, & Tynelius, 2002). Second, cerebral abnormalities caused by genetic factors or in utero may cause both ID and SSDs. This has been described as the 'common aetiology' hypothesis (David, Malmberg, Brandt, Allebeck, & Lewis, 1997). Third, low IQ could lead to incorrect social assumptions and thus increased risk of symptoms such as delusions and hallucinations. David et al. (1997) showed that low intellectual ability in itself is a risk factor for schizophrenia and other psychoses, although their study, like the vast majority in schizophrenia research, excluded those with premorbid ID.

Risk Factors

Genetics

Kallman (1938) influentially reported no increase in ID in the relatives of people with schizophrenia and vice versa. This study concluded that the co-morbid state was a chance occurrence of two unrelated conditions and this contributed to a period of subsequent decreased research interest into co-morbid ID and schizophrenia (Turner, 1989).

However Kallman's study did not consider the co-morbid probands and, when Doody, Johnstone, et al. (1998) did so, they found that both the prevalence of ID and schizophrenia were increased in the relatives of the co-morbid probands. This supported the idea that a highly familial form of schizophrenia might occur in the ID population. Doody, Johnstone, et al. (1998) also showed that there was a high rate of chromosomal variants in karyotype testing in the co-morbid group. Cytogenetic anomalies, which are often associated with intellectual impairment, may be associated with an earlier age of onset of schizophrenia (Bassett et al., 1998). Schizophrenia and ID in association have been reported with a wide range of chromosomal abnormalities (Blackwood et al., 2008). Psychotic symptoms are sometimes associated with Prader-Willi syndrome (15q11-13 partial deletion), particularly the maternal disomy form (Boer, Holland, Whittington, Butler, & Webb, 2002). High rates of schizophrenia have been reported in adults with velo-cardio-facial syndrome (VCFS), which is associated with a deletion on chromosome 22 and is often associated with ID (Green et al., 2009). In addition, 2 % of service users with schizophrenia exhibit this 22q11 deletion but 6 % of the early onset cases do so (Usiskin et al., 1999). Increased rates of dysmorphic features, cognitive impairment and structural brain abnormalities have led to the proposal of 22q11 deletion as a model for a neurodevelopmental subtype of schizophrenia (Bassett, Chow, & Weksberg, 2000). However many studies of VCFS and psychosis have excluded people with ID.

Obstetric Complications

O'Dwyer (1997) found that in people who develop ID and schizophrenia, the number of obstetric complications increases, compared to those matched for age, sex, severity of ID and epilepsy. These complications included abnormally long labour, dysmaturity, pre-eclamptic toxaemia and maternal episiotomies, all of which may give rise to anoxic cerebral trauma, which the authors concluded was the major aetiological factor predisposing people with ID to develop psychoses. Hucker et al. (1979) studied 24 people with ID and schizophrenia and 40 control subjects. They found higher proportions of people with impaired hearing, gestation below 36 weeks and low birth weight in the group with schizophrenia.

Epilepsy

Deb et al. (2001) and Doody, Johnstone, et al. (1998) found that those with schizophrenia and ID were more likely to have epilepsy compared to those with schizophrenia of typical intelligence. These findings would be expected given that epilepsy is more common in people with ID (Bowley & Kerr, 2000). However, Cowley et al. (2004), Cooper et al. (2007) and Arshad et al. (2011) found in their studies that people with epilepsy and ID had a lower incidence of schizophrenia or non-affective psychosis than in people with ID only. These somewhat conflicting findings from research to date are perhaps unsurprising as epilepsy provides further complications in the diagnosis of SSDs in people with ID. For example, it can be difficult to differentiate temporal lobe epilepsy from schizophrenia. Ring, Zia, Lindeman, and Himlock (2007) found that psychoses were more common in people with ID and epilepsy when there had been no seizures for the preceding 3 months. The phenomenon of 'forced normalisation' has been described in relation to the schizophrenia-like psychoses of epilepsy (Trimble, 1996) whereby a reduction in seizure activity following the introduction of anticonvulsant agents is associated with an exacerbation of psychosis.

Other Risk Factors

Meadows et al. (1991) and Fraser and Nolan (1994) suggested that schizophrenia might have an earlier age of presentation in people with ID. One possible explanation given was that the existence of schizophrenia in the context of ID is more likely to produce a more severe illness

with earlier appearance of psychotic symptoms (Meadows et al., 1991). However, there is also the possibility that the presence of schizophrenia increases the likelihood of an individual being diagnosed as having ID, and vice versa (Sanderson, Doody, Best, Owens, & Johnstone, 2001). There can sometimes also be a delay in presentation of schizophrenia in people with ID due to the difficulties of detection, including the suggestion that the onset of psychosis may sometimes be less noticeable in people with ID (James & Mukherjee, 1996). However, Doody, Johnstone, et al. (1998) found no significant differences in the ages of first symptoms of schizophrenia, consultation, admission or diagnosis between subjects with both mild ID and schizophrenia and matched controls with schizophrenia alone. This was despite the fact that co-morbid subjects had more psychopathology and duration of admissions than the controls.

Doody, Thomson, Miller, and Johnstone (2000) reported that women with ID generally developed schizophrenia later than men with ID, who were also less likely to have a family history of the disorder. They suggested that gender differences in schizophrenia might be more pronounced among the co-morbid population where early cognitive impairment can herald the onset of a more severe form of onset. Cowley et al. (2004) found higher prevalence of SSDs with older age, whereas Cooper (1997) found no differences in the prevalence of schizophrenia between younger and older adults with ID. Ethnicity has also been associated with SSDs in the ID population in the UK, with a higher prevalence in those referred to services being from ethnic minority groups than would have been expected by chance (Chaplin, Thorp, Ismail, Collacott, & Bhaumik, 1996; Cowley et al., 2004; Tsakanikos, Bouras, Costello, & Holt, 2009). Sensory impairments are more common among people with ID (Carvill, 2001). It might be expected that sensory impairments would increase the risk of SSDs in people with ID, but there have been few studies to confirm this. Cooper et al. (2007) found that visual impairment was independently associated with psychosis in adults with ID.

Neuropathology

In the classification systems of DSM-IV (American Psychiatric Association, 1994) and ICD-10 (World Health Organisation, 1992), it is stipulated that schizophrenia should not be diagnosed in the presence of 'overt' brain disease, which is more common in people with ID (Hagberg & Kyllerman, 1983). This complicates the study of neuropathology in individuals with both schizophrenia and ID. A recent case-controlled study of volumetric cerebral magnetic resonance reported that in terms of brain structure, people with ID and schizophrenia resemble those with schizophrenia more closely than those with ID (Sanderson, Best, Doody, Owens, & Johnstone, 1999). These findings were confirmed by further studies (Moorhead et al., 2004; Sanderson et al., 2001). Sanderson et al. (1999) suggested that the higher frequency of schizophrenia in ID was due to the greater tendency of people with schizophrenia to develop cognitive deficits and that within the ID population there may be people whose deficits result from undiagnosed schizophrenia. These authors support the view that co-morbid schizophrenia and ID may represent an early onset and severe form of schizophrenia, rather than ID complicated by psychosis (Moorhead et al., 2005; Spencer et al., 2007). It could be then that a subset of adolescents with ID have a form of schizophrenia that has not yet made itself manifest. Hence Johnstone et al. (2007) looked at schizotypal traits in adolescents with ID and used these to screen for those likely to develop schizophrenia at a later date. Welch et al. (2010) found that within a sample of people with ID those with schizotypal traits showed reduced amygdala volume on magnetic resonance imaging scanning compared to those with ID alone.

Psychopathology

Presentation

Disagreement remains as to what extent the presentation of SSDs is different in people with and

without ID. Most of the literature suggests that there are no *substantial* differences in the symptoms of schizophrenia between people with ID and people of normal intelligence (Hucker et al., 1979; Moss, Prosser, & Goldberg, 1996; Reid, 1972). Meadows et al. (1991) suggested that there was no difference in the presentations of symptoms of schizophrenia between those with and without ID although there was still a trend for less overt psychopathology in 13 of the 16 symptoms. Bassett et al. (2003) also found no significant differences in the schizophrenia phenotype between service users with and without ID. However other studies have found differences in presentation, especially a tendency for less florid positive symptoms of schizophrenia in those with ID. For example, Linnaker and Helle (1994) found that those diagnosed with schizophrenia with the Psychopathology Instrument for Mentally Retarded Adults (PIMRA) had fewer delusions, flatter affect and more incoherent speech than people with schizophrenia of normal intelligence, especially persecutory delusions and formal thought disorder. Bouras et al. (2004) showed that those with ID and SSDs showed more observable and fewer self-reported symptoms than people with SSDs alone.

Turner (1989) described the symptoms of schizophrenia in people with ID as 'often shallow or banal'. Because of their reduced opportunity to engage in normal life experiences and social opportunities, the delusions of people with ID, when they occur, may be relatively simple. For example, there is little attempt to interpret strange subjective phenomena and secondary elaboration of abnormal perceptions or odd beliefs producing complex delusional systems rarely occurs (Reid, 1989). Complex hallucinations such as voices giving as running commentary in the third person or discussing the person between themselves are also uncommon. Sovner (1986) described this phenomenon as an example of 'psychosocial masking' of symptoms in people with ID. James and Mukherjee (1996) found that the presenting complaints of schizophrenia in people with ID were very often a decline in functioning, deterioration of skills and social withdrawal, rather than hallucinations and delusions. Reid (1993) suggested that hebephrenic, paranoid and catatonic subtypes of schizophrenia could all be seen in people with ID. However, most clinicians believe that it is less often possible to sub-classify schizophrenia in people with ID than in those with more typical IQ.

Positive and Negative Symptoms

Although researchers in the field of psychosis in ID have used the categorisation of psychotic symptoms into 'positive' and 'negative' (Andreasen, 1982), the validity of these in the ID population remains unclear (Melville, 2003). There have again been some apparently conflicting findings; for example, Bannerjee, Morgan, Lewis, Rowe, and White (2001) and Cherry, Penn, Matson, and Bamburg (2000) emphasised positive symptoms in the presentation of schizophrenia in people with ID. Doody, Johnstone, et al. (1998) and Bouras et al. (2004) found 'negative' symptoms to be more common in people with ID and SSDs than in SSDs alone. Moss et al. (1996) showed negative symptoms to have low specificity for schizophrenia in adults with ID. They found that auditory hallucinations seemed easiest to elicit in people with ID and schizophrenia, followed by thought disorder, and then by delusions relating to replacement of will.

'Behavioural' Symptoms

Some have argued that there may be possible behavioural manifestations or 'behavioural equivalents' of mental disorders such as psychosis in people with ID (Dosen, 2005). Associations have been found between psychotic symptoms and problem behaviours in people with ID, but their relationship remains unclear (Myrbakk & von Tetzchner, 2008). Certainly some behavioural problems respond to antipsychotic medication suggesting that they might have been caused or exacerbated by underlying psychosis that is difficult (if not impossible) to diagnose clearly using standard diagnostic criteria. It has been argued that people with ID and psychosis may

present with other atypical features. For example, hysterical type symptoms such as over-breathing, pseudo-fits, gait disturbance and Ganser states may be more frequent in people with ID and psychosis (Dosen & Day, 2001; Hucker et al., 1979; Reid, 1972). Some schizophreniform psychoses also present acutely with apparent changes in consciousness (Reid, 1993). Turner (1989) reported that disturbed and aggressive behaviour, bizarre rituals and 'hysterical' behaviours are relatively more common in ID and may complicate the presentation of psychosis in ID. People with ID and schizophrenia may also present with social withdrawal, fearfulness and sleep disturbance (Eaton & Menolascino, 1982; Myers, 1999). The 'DC-LD' guide (Royal College of Psychiatrists, 2001) suggested that early signs of a psychotic illness could be new problem behaviours (especially when odd or bizarre or uncharacteristic for the person), or an increase in frequency or severity of pre-existing behaviours.

Behaviour that might suggest auditory hallucinations could be the person with ID shouting back apparently at people not present when this has not been their previous behaviour. Similarly, suspiciousness, blunted or incongruent affect and social withdrawal not previously part of the person's personality and behaviour could also be suggestive of a schizophreniform-type psychosis. Non-verbal evidence for possible psychosis by necessity becomes of greater diagnostic significance in the more severely intellectually disabled. It has been claimed that that it is possible using observations and non-verbal communication to diagnose schizophrenia in people with more severe ID. Both Eaton and Menolascino (1982) and Cherry et al. (2000) claimed behaviour characteristic of schizophrenia could be identified in people with severe ID. Heaton-Ward (1977) also argued that for some people with more severe ID, their emotional lability, noisy outbursts and disorganised, purposeless activity, including aggression, destructiveness and self-mutilation, could be considered 'psychotic'. However, he also recognised that it would be impossible to determine whether such individuals were demonstrating lack of insight and loss of sense of reality, which are fundamental to psychosis. Turner (1989) noted the persistent comment on catatonia in the literature. Hucker et al. (1979), Eaton and Menolascino (1982) and Heaton-Ward (1977) all remarked upon the high proportion of their study samples of subjects with ID and schizophrenia who showed catatonic features. The relationship between catatonia and schizophrenia in people with ID is complex and still needs further clarification.

Cognitive Functioning

An episodic psychotic illness may be associated with a reduction in transient functioning to the mild or even moderate level of ID (Russell & Tanguay, 1981). A chronic psychosis may, however, lead to a prolonged or even permanent reduction in intellectual and social functioning. Sometimes those with chronic schizophrenia are thus mistakenly diagnosed as also having ID because of deterioration in intellectual functioning. There has been little specifically published though in the literature of neuropsychological findings in people with ID and schizophrenia. Heaton-Ward (1977) found a 20 % fall in IQ in some of his service users with ID and schizophrenia over time. Doody, Johnstone, et al. (1998) found that their co-morbid subjects were more likely to have impairment of episodic memory, which may affect their compliance with treatment. They also had greater impairment of theory of mind on second-order tests than subjects with schizophrenia and normal pre-morbid IQ (Doody, Gotz, Johnstone, Frith, & Cunningham Owens, 1998). Rowe, Rudkin, and Crawford (2000) found that there was an increased rate of mixed handedness among people with co-morbid ID and schizophrenia compared to controls. Mixed handedness is taken as an index of diminished cerebral dominance or laterality, and thus these findings also supported the idea of a neurodevelopmental hypothesis of schizophrenia. Weinberger (1987) has suggested that people with ID and schizophrenia form a subgroup of schizophrenia with a neurodevelopmental aetiology. Doody, Johnstone, et al. (1998) found evidence consistent with the neurodevelopmental

hypothesis in their study of co-morbid ID and schizophrenia. For example, they found that their co-morbid subjects had more 'soft' neurological signs than controls. They suggested that there is a form of schizophrenia that manifests in childhood with cognitive impairment prior to the onset of psychotic symptoms.

Management

The principles of treatment of schizophrenia in people with ID are essentially similar to those without ID. There have been a few studies that have suggested that treatment with antipsychotics in ID is broadly similar in efficacy with no significant risk in side effects (e.g. Craft & Schiff, 1980; Reid, 1972). Shedlack, Hennen, Magee, and Cheron (2005) found that in those with ID and SSDs, there was substantial improvement in social withdrawal following treatment with both typical and atypical antipsychotic medication. Some studies have reported beneficial effects (and sometimes fewer side effects) with atypical antipsychotics (e.g. Advokat, Mayville, & Matson, 2000; Shastri, Alla, & Sabaratnam, 2006; Williams, Clarke, Bouras, Martin, & Holt, 2000). However Matson and Mahan (2010) cautioned that a long-term perspective on atypical antipsychotic use in people with ID is still not yet possible in the same way as it is with typical antipsychotic medications. It is still not clear whether people with ID and SSDs, or at least a subgroup of them, are more likely to be treatment-resistant. Clozapine is efficacious and well tolerated in people with ID and co-morbid mental illness including schizophrenia (Antonacci & de Groot, 2000). Varghese and Banerjee (2010) reported in their audit of their community teams that of seven of their treatment-resistant service users, only two had been considered for clozapine. They noted the additional difficulties in prescribing clozapine for clinicians treating people with ID.

Clarke (2001) argued that people with brain pathology are more likely to develop tardive dyskinesia than the general population. Sachdev (1992) found that 34 % of those people with ID receiving antipsychotic medication in a long-stay institution had tardive dyskinesia. Gingell and Nadararajah (1994) also suggested that people with ID might be at greater risk of developing movement disorders when taking antipsychotic medication. However, the evidence for increased risk of tardive dyskinesia due to antipsychotic medication in people with ID and SSDs relative to those with SSDs alone is not yet conclusive. For example, Gualtieri, Shroeder, Hicks, and Quade (1986) found no evidence that ID increased the risk. One possible reason for this discrepancy in findings is that differentiating between movement disorders and medication side effects is again more difficult in service users with ID. Rogers, Karki, Bartlett, and Pocock (1991) showed that people with more severe ID also have high rates of motor disorders not attributable to antipsychotic medication. The presence of muscle tone abnormalities and stereotypies including tics, mannerism and self-stimulatory behaviours can all mask the presence of antipsychotic-induced movement disorders (Shedlack et al., 2005). One reason is the increased propensity towards tardive dyskinesia in people with ID relative to those with normal IQ (Cohen, Khan, Zheng, & Chiles, 1991; Fodstad et al., 2010; Rao, Cowie, & Matthew, 1987). However none of these studies have conclusively answered the complex question of whether people with ID and SSDs are even more likely to develop tardive dyskinesia as a result of being treated with antipsychotic medication than people with SSDs but without ID would be. It has also been suggested that the risk of neuroleptic malignant syndrome may also be possibly increased in people with ID on antipsychotic medication (Boyd, 1992).

Many clinicians have reported that the optimal dosage levels of antipsychotic medications in clinical practice appear to be lower in service users with ID than in those with normal IQ (Menolascino, Ruedrich, Golden, & Wilson, 1985). Problems such as possible exacerbation of pre-existing cognitive impairment, the lowering of seizure thresholds and the reduced ability to communicate symptoms and side effects can make the prescribing of antipsychotics for people with ID and SSDs even more difficult than

in people with SSDs without ID. There is a consensus that antipsychotic prescribing often needs to be instigated and increased more cautiously than in service users without ID. Duggan and Brylewski (1999) argued that there was still insufficient evidence to judge the efficacy of antipsychotics when used specifically in people with ID. The use of antipsychotics to treat SSDs in this service user group therefore is still largely based on extrapolation from the evidence base of general adult psychiatry. There remains a need for randomised controlled trials of the efficacy of antipsychotic medication in people with ID and SSDs. Electroconvulsive therapy (ECT) is sometimes used in the treatment of SSDs (e.g. in catatonic states), but there has been little published regarding its use as treatment of SSDs specifically in people with ID (Chanpattana, 1999).

There have often been criticisms of prescribing antipsychotic medication for people with ID and SSDs given the dearth of specific evidence for 'biological' treatments, but there is also not a great deal of evidence regarding the use of psychosocial (including family) interventions. Crowley, Rose, Smith, Hobster, and Ansell (2008) conducted a preliminary study of using two psycho-educational groups for eight people with the dual diagnoses of psychosis and ID. Participants were reported to have had their knowledge increased about psychosis, medication, early signs of relapse and the role of stress. Behavioural techniques have been used to improve the social skills of individuals with psychosis and ID (Hatton, 2002). The application of either individual or family cognitive-behaviour therapy (CBT) approaches has not been systematically evaluated adequately as yet in people ID who have psychosis. A case series of CBT for five service users with psychosis and mild ID has been reported (Haddock, Lobban, Hatton, & Carson, 2004). This included two cases in which family interventions were also integrated into the individual CBT. Taylor, Lindsay, and Willner (2008) have argued that it is unjustified to not make available psychotherapy based on cognitive-behavioural approaches for people with ID and psychopathology.

Prognosis

It has long been established that in service users with schizophrenia there is a relationship between pre-psychotic IQ and disease prognosis (Offord & Cross, 1971). There has been very little published, however, into the long-term prognosis in people with ID and schizophrenia. There have not been either any published studies of the latency period or prodrome before presentation of psychotic symptoms in this service user group. Most clinicians believe that the course of schizophrenia in people with ID will tend to be more severe than in people with normal IQ. However, again the literature is scant and findings have not always been consistent. For example, Reid (1972) reported that the psychoses ran a more benign course in people with ID, especially in those with more severe ID.

Bouras et al. (2004) showed that in a group of people with ID and SSDs, matched for duration of illness, there were greater functional disabilities compared to a group with SSDs attending a general mental health outpatient clinic. The authors suggested that the increased prevalence of observable psychopathology might lead to increased risk of stigma and social isolation for those with SSDs who also had ID. The higher rate of negative symptoms seen also might contribute to social withdrawal and isolation and raised the question of whether those with ID as well as SSDs might have progressed more rapidly to the chronic deficit state of SSDs. The authors suggested that people with ID are more debilitated by SSDs than those without ID and may thus need additional input. Doody, Johnstone, et al. (1998) found that service users with dual diagnosis of schizophrenia and ID had fewer psychiatric admissions but for longer periods of time, and that at point of discharge they needed more support than service users with schizophrenia alone. With some outcome parameters, such as total time in hospital and offending behaviour, there was no evidence that the comorbid group were more impaired than the schizophrenia control group. However they also found that those with both conditions received

more community supports than control subjects with schizophrenia alone. Doody et al. (2000) showed that in people with co-morbid ID and schizophrenia, males with an early age of onset and no known family history of either ID or schizophrenia were more likely to require care and treatment in a high-security hospital. Chaplin et al. (2006) also found that people in their study with lower IQ (including those with borderline low IQ as well as those with ID) and schizophrenia had a lower quality of life, more severe psychotic symptoms and reduced functioning compared to those with schizophrenia and higher IQ.

SSDs in people with ID cause a great deal of human suffering, carer burden and also significant service and care costs. Hemmings, O'Hara, et al. (2009) found that the SSDs were overall the largest group of diagnoses for people with ID admitted to both a specialist and generic mental health units in their study in South London. In Taiwan, Lai, Hung, Lin, Chien, and Lin (2011) have reported that a large proportion of inpatient admissions and associated financial costs were accounted for by a coexisting diagnosis of schizophrenia in people with ID. For community services, focus group studies (Hemmings, Underwood, & Bouras, 2009a) and a Delphi study (Hemmings, Underwood, & Bouras, 2009b) have helped to identify a consensus of what needs to be provided for this disadvantaged dually diagnosed service user group. There is some evidence that outcomes may be better when people with both ID and psychosis are treated by specialist rather than generic services (Raitasuo, Taiminen, & Salokangas, 1999).

Conclusion

The evidence base regarding SSDs in people with ID remains limited. There is some evidence though to suggest that people with ID and SSDs may have even more complex needs than service users with SSDs of typical intelligence and may be harder to treat. We are not likely to see any clear-cut relationships between ID and SSDs when ID is itself on a spectrum with the general population. Although in some individuals with ID the development of SSDs may be by chance, it seems that for others there may be direct and indirect links. The relationship between IQ and schizophrenia is complex, as lower IQ may cause vulnerability to the development of schizophrenia or be an early manifestation of the disorder (Offord & Cross, 1971). Many believe that co-morbid ID and schizophrenia may often represent a severe form of schizophrenia with poorer outcomes (Doody, Johnstone, et al., 1998) and both arising from the same genetic aetiology. The majority of studies of SSDs have excluded people with premorbid ID but is important that further research does not unless there are compelling reasons to do so. Studying SSDs in people with ID might not only improve diagnosis and treatment but also help to identify those with ID who will go on to develop these severe mental illnesses. It may also provide one important way forward in the understanding of the SSDs in the general population. Research across the biopsychosocial spectrum is needed and this must include developing our knowledge on the best service provision and outcomes for this dually disadvantaged group of service users and their careers.

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Mood and Anxiety Disorders

11

Angela Hassiotis, Kerstin Stueber, Bini Thomas, and Lauren Charlot

Diagnostic Features

Depressive disorders are characterised by a sustained depressed mood and loss of interest or pleasure which represents a change from previous functioning (American Psychiatric Association, 2000). Associated features include depressed mood or loss of interest or pleasure in usual activities, increased irritability, weight loss/gain, disturbed sleep, psychomotor agitation/retardation, fatigue, difficulties in concentration and recurrent thoughts of death/suicide/suicidal ideation.

The key feature of anxiety disorders is apprehension and worry that is disproportionate to the circumstances. "The focus of anticipated danger may be external or internal" (American Psychiatric Association, 2000). Emotional symptoms include constant worrying, intrusive thoughts about situations that may be a source of anxiety, inability to tolerate uncertainty and the need to know what is going to happen in the future. Often there are accompanying behavioural symptoms such as inability to relax or enjoy quiet time, difficulty in focusing on activities, putting things off and avoiding situations that may be seen as difficult to manage. Physical symptoms may include feeling tense, muscle tightness or body aches or nausea.

Obsessive–compulsive disorder (OCD) is characterised by obsessions, that is, unwelcome thoughts and impulses that are difficult to dismiss or control and compulsions or repetitive actions that cannot be resisted easily. Compulsions tend to be rare in people with intellectual disability as may require a degree of abstract thinking, e.g. fear of contamination of hands (compulsive hand-washing) or safety (repetitive checking of whether the door is locked). More frequently seen in this patient group are ordering, hoarding, telling, asking or rubbing. Specific sensory impairments need to be taken into account too: a blind person with OCD is more likely to engage in compulsions involving touch (e.g. repetitive tapping or ripping of clothes).

Post-traumatic stress disorder (PTSD) is defined as a delayed or protracted response to a stressful event or situation (of either brief or long duration) of an exceptionally threatening or catastrophic nature, which is likely to cause pervasive distress in almost anyone. It has been widely recognised that people with ID experience interpersonal violence and crime (Focht-New, Clements, Barol, Faulkner, & Service, 2008).

Use of diagnostic criteria such as the International Classification of Diseases version 10 (ICD-10) and the Diagnostic and Statistical Manual (DSM-IV-TR) may lead to underascertainment of mental disorders, particularly in

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adults with moderate to profound intellectual disability as several of the domains require verbal communication (Cooper, Melville, & Einfeld, 2003). This limitation led to the development of the Diagnostic Manual-Intellectual Disability (DM-ID; Fletcher, Loschen, Stavrakaki, & First, 2007) in the USA and the Diagnostic Criteria-Learning Disability (DC-LD; Royal College of Psychiatrists, 2001) in the UK. Both of these manuals purport to address the restrictions posed by the main classification systems in the diagnosis of mood disorders in this population. DM-ID provides detailed descriptions and amended criteria for individuals across the range of intellectual disability and the DC-LD is complementary to ICD-10 (World Health Organization, 1994) for adults with moderate to profound intellectual disability. The adaptations have been necessary as certain symptoms are expressed differently in persons with intellectual disability, for example, unconventional means of self-harming behaviour such as running in front of cars may represent suicidal intent. Other similar observations are reflected in the choice of core symptoms: irritable mood in DM-ID, "sufficiently severe or frequent symptoms of anxiety to cause suffering to the individual" in DC-LD.

Ascertainment of compulsions and obsessions depends on information that is usually communicated verbally by the affected individual. However, it may be difficult to clarify whether the thoughts are derived from the person's own mind or to establish attempts to suppress the thoughts in a person with intellectual disability. Certain repetitive behaviours in patients with severe or profound intellectual disability are not considered to constitute symptoms of OCD (e.g. pacing, stealing, demanding attention).

Epidemiology

Several studies have examined the epidemiology of affective disorders in adults with intellectual disability but may not distinguish between rates of different types of such disorders. Lund (1985) studied 302 individuals from the Danish National Service for the Mentally Retarded Register. He reported prevalence rates of affective disorder of 0.8 % in females and 2.4 % in males and similar rates for neurosis. Though the diagnoses were made according to the MRC handicaps, behaviour and skills schedule (Wing, 1980), the author does not describe the ascertainment criteria for either category. The study showed a trend for the prevalence rate of affective disorder to increase with increasing age and the reverse for neurosis. The range of intellectual disability in the sample ranged from borderline to profound. Van Schrojestein Lantman de-Valk et al. (1997), in an epidemiological survey of 1,020 individuals across the range of intellectual disability, found that prevalence of affective disorders increased with increasing age (approximately 3 % in the 20–29 age group up to 8.3 % in the 60–69 age group). In the same sample, incidence rate peaked in the 40–49 age group at 4.9 % and attenuated to about 3 % in older age groups. Deb, Thomas, and Bright (2001) studied a random sample of 101 adults with intellectual disability living in the community and known to adult social and healthcare providers. A two-stage diagnostic interview based on versions of the Psychiatric Assessment Schedule for Adults with Developmental Disability (PASADD; Moss et al., 1993; Prosser et al., 1998) was employed. Depression was diagnosed in 3.3 %, anxiety disorder in 7.8 % and OCD 1.1 %. Cooper, Smiley, Morrison, Williamson, and Allan (2007a) carried out an epidemiological survey of 1,023 adults across the range of intellectual disability. They reported point prevalence rates for all affective disorders of 6.6 % based on clinical diagnosis. However, these rates reduced further depending on the diagnostic criteria used (5.7 % by DC-LD, 4.8 % by ICD-10-DCR and 3.7 % by DSM-IV-TR). Point prevalence of depression was 3.8 % and of all anxiety disorders approximately 4 % (Cooper, Smiley, Morrison, Williamson, & Allan, 2007b; Reid, Smiley, & Cooper, 2011). Generalised anxiety disorder was the most common (1.7 %) followed by agoraphobia (0.7 %).

A number of studies support the evidence that suicidality occurs in patients with intellectual disability although it is less frequent than in the general population and presents differently

(Hurley, Folstein, & Lam, 2003; Lunsky, 2003; Walters, 1990; Walters, Barrett, Knapp, & Borden, 1995). However, an increased rate of suicidal behaviour in people with borderline intellectual functioning that was linked to younger age and socioeconomic disadvantage was demonstrated by the use of data of the third Adult Psychiatric Morbidity Survey (Hassiotis, Tanzarella, Bebbington, & Cooper, 2011). Presence of suicidal acts and ideation was found in at least half of all admissions of persons with intellectual disability in a Canadian study (Burge et al., 2002).

The point prevalence of OCD ranges from 0.7 % using clinician diagnosis to 0.2 % using ICD-10-DCR or DSM-IV-TR diagnostic criteria (Cooper et al., 2007a, 2007b). However higher prevalence of 2.5 % has been described in the past (Cooper & Bailey, 2001). Ryan (1994) examined 310 persons with moderate to severe intellectual disability who presented to a consultation service in Colorado using DSM-III-R criteria for diagnosis of PTSD. All service users had suffered significant abuse or trauma. 16.5 % of them met the criteria for PTSD.

Comparison with the General Population

Such comparisons are marred by the variance of methodology and classification systems in the normal population and in intellectual disability. However, Smiley (2009) compared incidence rates in people with ID and in the normal population using standardised incidence ratios (SIR). The findings suggest that common mental disorders are as common in people with ID as they are in the normal population (SIR 1.09, 95 % CI 0.75–1.52) based on findings from Singleton and Lewis (2003). Regarding depression, SRI of 0.81 was calculated (95 % CI 0.55–1.16). Using a study by Bijl, De Graaf, Ravelli, Smit, Vollebergh, and Netherlands Mental Health Survey and Incidence Study (2002), the SRI is 0.88 (95 % CI 0.55–1.35). Although the implication is that common mental disorders including anxiety, depression and OCD are as common in people with intellectual disability as in the normal population, we should treat the conclusion with caution given the significant methodological differences in the studies.

Aetiology

As with most psychiatric illness, the aetiology of mood disorders is considered to be multifactorial involving biological, psychological and social factors.

Biological Factors

Little is known about the biological underpinnings of mood disorders in individuals with intellectual disability. There is a reported association between intellectual disability itself and the diagnosis of depressive disorder (Hurley et al., 2003). Cerebral changes can be acquired through an injury in the developmental period, through self-injury (e.g. head banging) or the disorder directly impacting on neurotransmitter pathways, e.g. reduced levels of serotonin, GABA, taurine and dopamine have been demonstrated in the frontal cortex of fetal brains with Down's syndrome (Whittle, Sartori, Dierrsen, Lubec, & Singewald, 2007).

More commonly, there may be additive effects from multiple vulnerabilities common to people with intellectual disability. These may include limited or reduced neural redundancy, abnormalities in protein functions that derail complete wiring of neural circuits important to threat assessment and emotional regulation and absence of or markedly reduced neural tissue. Such abnormalities in brain structure are likely to impact on functioning globally (as in syndromes associated with agenesis of the corpus callosum) (Jou et al., 2011).

Specific Genetic Syndromes and Mood Disorders

Increasingly, studies are revealing a variety of complex genetic contributions to the occurrence of ID. Understanding that a child or adult may be

prone to experiencing certain medical problems can facilitate earlier diagnosis and treatment of these conditions. The knowledge that an individual has a specific genetic syndrome can enhance differential diagnosis of a co-occurring psychiatric disorder especially when the diagnostic picture is confusing. Individuals with Down's syndrome (DS) appear to be prone to depression, anxiety, OCD and Alzheimer's disease. Depression, however, may not occur more often among individuals with DS when compared with peers with other aetiologies of ID (Walker, Dosen, Buitelaar, & Janzing, 2011). Mantry et al. (2008) found that when people with DS do have mental health concerns, the most frequent diagnoses are depression and dementia (diagnosed in 5.2 % of a population-based sample). Of interest has been the observation that individuals with DS rarely experience bipolar disorder (Collacott, Cooper, & McGrother, 1992; Craddock & Owen, 1994; Stavrakaki, Antochi, & Emery, 2004). "Obsessional slowness" has been particularly described in people with Down's syndrome (Charlot, Fox, & Friedlander, 2002).

Another syndrome that has received a great deal of recent research attention is velocardiofacial syndrome or VCF also known as 22q11 deletion syndrome. About 30 % of individuals with VCF also have ID. Individuals affected by VCF are 25 times more likely than individuals without the syndrome, to develop one or more of a number of neuropsychiatric conditions (Shprintzen, 2008) most commonly psychosis in as many as 30 % of adults. Other psychiatric symptoms commonly include depression and anxiety (Jolin, Weller, & Weller, 2011). Investigations have demonstrated that a core defect may be related to the effects of the microdeletion on the COMT gene, having effects on catecholaminergic neurotransmission (Gothelf, Frisch, Michaelovsky, Weizman, & Shprintzen, 2009).

Fragile-X or FRA-X, the most common genetic cause of ID, has also been studied extensively in recent years. Individuals with FRA-X often suffer from anxiety disorders and may be diagnosed with ADHD during childhood.

Prayer–Willis syndrome (PWS) arises from three possible pathways, all of which result in a lack of expression of paternally inherited genes on chromosome 15, in the q11.2–q13 region. These include paternal microdeletion, maternal unapparent disomy and imprinting defect. Individuals with PWS have been described as displaying a particular behavioural phenotype and have risk for developing OCD and challenging behaviours (Cassidy & Driscoll, 2009).

In Table 11.1, a number of syndromes associated with ID and mood disorders are listed (genetic and others, i.e. fetal alcohol syndrome). The mental health disorders that have been reported to occur at elevated rates in association with these syndromes are provided. For each, a key reference is listed that will provide an overview of the syndrome and associated mental health issues. Of note, for many syndromes, studies examining behavioural phenotypes and liability for psychiatric syndromes are frequently conducted on small samples because the disorders are rare or under-diagnosed (Delorme et al., 2010). More recent studies have demonstrated that there is much variability in phenotypic expression across many of these (Mitchell, 2011).

Psychological Factors

Cognitive factors which have been implicated in the onset of depressive illness are negative social comparison, low self-esteem (Dagnan & Sandhu, 1999) and poor self-concept (Benson & Ivins, 1992). Such cognitions may be due to stressful early experiences and increased risk for trauma, abuse and neglect. Impaired cognitive skills due to intellectual disability may impact adversely on adaptive coping skills and may lead to poor social skills and "learned helplessness". The role of life events and traumatic experiences as predictors of psychopathology in people with intellectual disability is not yet clear as the impact of the two types of experiences has not been studied sufficiently including the paucity of research investigating factors that may promote resilience in this population (Lunsky & Canrinus, 2005; Martorell & Tsakanikos, 2008; Sequeira, Howlin, & Hollins, 2003). Negative life events show high correlation with depressive symptoms as do, surprisingly,

Table 11.1 Mood disorders and genetic conditions

Syndrome	Reported associations with psychiatric syndromes/symptoms	Suggested reading
Down's syndrome	Depression, OCD, anxiety, obsessional slowness	Walker JC, Dosen A, Buitelaar JK, Janzing JG. Depression in Down syndrome: A review of the literature. Research in Developmental Disabilities, 2011, 32: 1432–1440
Fragile X	ADHD, anxiety	Cordeiro L, Ballinger E, Hagerman R, Hessl D. Clinical assessment of DSM-IV anxiety disorders in fragile X syndrome: prevalence and characterization. Journal of Neurodevelopmental Disorders 2011, 3: 57–67
Velocardiofacial syndrome (VCF)	Childhood: ADHD, anxiety and depression Adults: Schizophrenia/psychosis, bipolar disorder	Jolin EM, Weller RA, Weller EB. Occurrence of affective disorders compared to other psychiatric disorders in children and adolescents with 22q11.2 deletion syndrome. Journal of Affective Disorders, 2011, 136: 222–228
Neurofibromatosis NF1	Anxiety, depression	Maalouf FT, Mia Atwi CH, Boustan R-MN. Psychiatric comorbidities in common genetic disorders with physical disability. Pediatric Health, 2010, 4: 591–601
Tuberous sclerosis	Anxiety, depressed mood	Prather P, de Vries PJ. Behavioral and cognitive aspects of tuberous sclerosis complex. Journal of Child Neurology, 2004, 19: 666–674
Rubinstein–Taybi syndrome (RTS)	Depression	Verhoevena WMA, Tuiniera BS, Kuijpersa HJH, Eggera JIM, Brunnerd CHG. Psychiatric profile in Rubinstein-Taybi syndrome: a review and case report. Psychopathology 2010, 43: 63–68
Myotonic dystrophy type 1	Depression, anxiety	Winblad S, Jensen C, Mansson J, Samuelsson L, Lindberg C. Depression in myotonic dystrophy type 1: clinical and neuronal correlates. Behavioral and Brain Functions, 2010, 6: 1–7
Phenylketonuria (PKU)	Depressed mood, generalised anxiety, phobias	Brumm VL, Bilder D, Waisbren SE. Psychiatric symptoms and disorders in phenylketonuria. Molecular Genetics and Metabolism, Supplement Phenylketonuria: Psychology and the Brain 2010, S59–S63

positive life events (Esbensen & Benson, 2006). Further investigation of those with and without depression showed that number of life events and perception of them as negative in the preceding 4 months predicted severe depressive episode. A study which investigated the relationship between life events exposure and psychiatric problems in adults with ID found some evidence of increased rates of affective/neurotic symptoms in 1,000 persons with intellectual disability using the PASADD checklist (Hastings, Hatton, Taylor, & Maddison, 2004). Recent life events have been

shown to be significant independent predictors of anxiety in addition to lack of daytime activities (Reid et al., 2011). Female gender, smoking and presence of primary care appointments (Cooper et al., 2007b) are independently associated with depression. However, as there are discrepancies in the methodologies used by more recent and earlier studies, such associations require further elucidation.

Social Factors

A relationship between low levels of social support and depression in people with intellectual disability has been reported. Other factors such as poor diet, unemployment, lack of exercise, limited choice, labelling or stigmatisation, being rejected or teased by peers, having limited access to social experiences or being socially isolated, reduced access to employment, poor arrangements of care, loss of friends and family and lack of confiding relationships have all been postulated to increase vulnerability to develop a depressive disorder (Bernal & Hollins, 1995; Hastings et al., 2004; Hatton & Emerson, 2004; McGillivray & McCabe, 2007; Owen et al., 2004; Tsakanikos et al. 2007).

Informants may inadvertently underestimate the importance of stressful life events on the lives of individuals with ID (Charlot et al., 2007). A key concern is that transient and reasonable reactions to stress should not be viewed as representing acute psychiatric disorder, e.g. an individual referred for psychiatric admission when reacted with crying and pacing following a diagnosis of cancer.

Diagnostic Instruments

There are several informant as well as self-reported screening and diagnostic instruments for depressive disorders in people with intellectual disability (for exhaustive reviews please see Hermans & Evenhuis, 2010; Hermans, Van der Pas, & Evenhuis, 2011; Perez-Achiaga, Nelson, & Hassiotis, 2009). Below we summarise some of the most commonly used:

Informant-based

- Psychiatric Assessment Schedule for Adults with a Developmental Disability Checklist (PAS-ADD Checklist; Moss et al., 1998): The PAS-ADD Checklist is used in the screening of individuals with an intellectual disability for mental health problems. It contains an affective/neurotic disorder subscale. It may be the most appropriate tool to be used by non-psychiatrists (Hermans & Evenhuis, 2010). A more detailed version is the mini PAS-ADD (Prosser et al., 1998) which contains a subscale for depression and one for anxiety.
- Reiss Screen for Maladaptive Behaviour (RSMB; Reiss, 1988). The RSMB is an informant-rated screening scale. It contains an alphabetic listing of 38 key symptoms of psychopathology accompanied by definition and examples. RSMB is organised into eight scales. Each item is scored on a three-point scale based on the functioning of the person during the last 2 months. The instrument is aimed for clinician use and has an internal consistency of 0.8–0.92 for the total scale and has a test–retest validity of 0.75 (Stavrakaki & Lunsky, 2007).

Self-reported

- Glasgow Depression Scale for people with Learning Disability (GDS-LD; Cuthill, Espie, & Cooper, 2003): The GDS-LD is a screening tool, may be used for monitoring progress and contributing to outcome appraisal. It consists of 20 items which are based on DC-LD, DSM-IV, ICD-10 and other questionnaires. It is used for individuals with mild and moderate intellectual disability and the authors report good test–retest reliability ($r=0.97$) and internal consistency ($\alpha=0.90$). A cut-off score of 13 yielded 96 % sensitivity and 90 % specificity
- Glasgow Anxiety Scale for people with intellectual disability (Mindham & Espie, 2003): A self-reported scale with 27 items which has been developed for people with intellectual disability. The scale has a good test–retest reliability ($r=0.95$) and internal consistency ($\alpha=0.96$) and was well correlated with the Becks Anxiety Inventory ($\rho=0.75$).
- Zung self-rating anxiety scales (Lindsay & Michie, 1988): This scale was developed for

the general population and has subsequently been adapted for people with intellectual disability. The self-administered test has 20 questions. Each question is scored on a scale of 1–4 (none or a little of the time; some of the time; good part of the time; most of the time). There are 15 questions worded toward increasing anxiety levels and five questions worded toward decreasing anxiety levels.

Clinical Considerations in the Assessment and Management of Mood Disorders in Adults with Intellectual Disability

People with Intellectual Disability Have Diminished Capacity to Self-Report Internal Mood States

First, the determination that an individual is experiencing alterations in affect is often highly reliant on their capacity to self-report internal states. Individuals with intellectual disability are challenged in this regard, and may not describe their emotions accurately or reliably (Ross & Oliver, 2003; Tsakanikos et al., 2006). It has been noted that much of the information used to identify the presence of symptoms of psychiatric disorders comes from the reported observations of caregivers (Charlot et al., 2007; Hurley, 2007; Tsakanikos et al., 2006). Caregivers, teachers, parents and other informants may inadvertently underestimate the extent of concerns such as depression and anxiety and focus on externalising behavioural concerns (Charlot, Doucette, & Mezzecappa, 1993; Edelbrock, Costello, Dulcan, Kalas, & Conover, 1985; Moss, Prosser, Ibbotson, & Goldberg, 1996). Hurtig et al. (2009) found that the self-report of adolescents with high-functioning autism or Asperger's syndrome contained more references to internalising symptoms like anxiety than those of their parents. This is critical when assessing individuals with neurodevelopmental disorders as the additional tendency to engage in aggressive acts when distressed can lead to inaccurate diagnoses. Individuals with intellectual disability frequently present for mental health assessment primarily because of aggression to self, others or property and do not usually self-refer. There can be pressure to treat aggression itself without attention to other clinical clues that might point to the identification of a mood problem (Tsiouris, 2010). Accurate diagnosis is key to identification of effective treatments and clinicians should be careful to resist pressures to treat aggressive behaviours "non-specifically" and to focus on establishing whether or not a psychiatric disorder is present. As suggested by many experts, this will require a comprehensive multidisciplinary assessment, the use of multiple sources of information and likely more time than usually allocated in practice with individuals without intellectual disability (Cooray & Bakala, 2005).

Individuals with ID Represent a Very Broad, Heterogeneous Population

A key issue in determining whether or not a person is experiencing a mood disorder is to establish whether or not current behaviour represents a departure from usual behaviour and from what might be expected for a person of this individual's age, culture, gender, education and other factors (Tsakanikos et al., 2006). For example, an individual with severe intellectual disability might present at the Emergency Department because of assaults to caregivers, yelling and running back and forth. The history may reveal the individual has been having the same outbursts since age 2, always in response to specific stressful events, and the reason for the current visit is that the individual is now too large for family members to manage the behaviour at home. Consider a person with an autism spectrum disorder and intellectual disability having an increase in agitation, repetitive behaviours, increased motor restlessness and not sleeping through the night. If the individual has never slept through the night in their life, the fact that he or she is not sleeping presently is not a new or acute "psychiatric" symptom. Parents, siblings and long-term caregivers as well as records of past evaluations may reveal key information regarding what is usual behaviour for the individual being assessed (Charlot et al., 2007). Defining what is

"abnormal" requires understanding of what should be expected behaviour and abilities of a person with a particular developmental profile. For example, usual fears change over the course of development. Fears of a monster in the closet might be usual early in development but would seem pathological in an adolescent. Among individuals who have more "immature" neurocognitive profiles, "magical thinking", concrete thinking and lack of coherence in communications may give the appearance of psychotic thinking (Hurley, 1996). Also, such tendencies may be exaggerated under stress. Clinicians are advised to take a developmental perspective to avoid inadvertent misidentification of developmentally expected behaviours as signs of psychopathology when they are not.

The Manifestation of Core Symptoms of Mood Disorders May Be Altered by Developmental Features

Population heterogeneity is linked to a second challenge for the clinician, which is the need to consider how symptoms of depressive and anxiety syndromes may present differently based on developmental features (Harris, 1998). Research has shown that children without intellectual disability are likely to show some variations in the nature of symptoms they display when anxious or depressed. One example is the finding that children with depression may present with psychomotor agitation, tantrums and irritable mood more often than depressed adults (Timimi, 2004; Trad, 1987). Very young depressed children are less likely to describe suicidal ideas or hopelessness, symptoms that require a certain level of baseline cognitive capacity to experience. Individuals with intellectual disability may have neurodevelopmental and cognitive profiles that parallel peers who are younger but neurotypical, which may affect how depression or anxiety is displayed (though the core features likely remain the same). Given expressive communication deficits, and developmental considerations, clinicians may need to ask caregiver's targeted questions that elicit full descriptions of clinical features to ensure that a mood disorder is not missed (Adams & Oliver, 2011).

Medical Problems and Medication Side Effects and Other Life Events May Cause Distress and Provoke Apparent Psychiatric Illness

Differential diagnosis of mood and anxiety disorders in individuals with intellectual disability is critical. Individuals may present with symptoms of depression or anxiety, for example, when they are ill (Charlot et al., 2011). In some cases, poor sleep, irritable mood and severe psychomotor agitation provoked by pain or physical distress can, on the surface, look like mania. It is particularly important that clinicians base diagnoses on the full history of the presenting problem and careful considerations of what other phenomena or conditions might better explain the clinical picture (Charlot et al., 2007). Additional findings such as the age of onset, clinical course, coincidental occurrences (introductions of new medications, onset of illnesses, major life events) and family history can enhance diagnostic accuracy in the face of diminished self-report data. Reliance on checklists or a simple accounting of symptoms may be especially misleading in this patient population. For example, depression and dementia share a number of common symptoms, with key differences relying less on phenomenology (dysphoric mood, anhedonia, difficulty focusing attention, poor sleep) than on clinical course and history (insidious vs. circumscribed onset). As noted, missed or under-treated medical problems and medication side effects may provoke a clinical picture that mimics mental illness including constipation, urinary retention and infections, reflux disease, dental pain, akathisia, extrapyramidal symptoms, sedation, seizures and numerous other physical conditions (Charlot et al., 2011; Espie et al., 2003; Gardner & Whalen, 1996; Gunsett, Mulick, Fernald, & Martin, 1989; Kennedy, Juarez, Greenslade, Harvey, & Tally, 2007; Kwok & Cheung, 2007; McDermott et al., 1997; O'Reilly, 1995; Valdivinos, Caruso, Roberts, Kim, & Kennedy, 2005).

Aggression and Other Externalising Behaviours May Occur as "State-Dependent" Features of Mood Episodes

An important consideration when assessing individuals with intellectual disability is the relationship between aggression and other externalising problem behaviours and mood disorders. A number of studies have demonstrated that mood disorders may be comorbid with aggression (Charlot et al., 1993; Hurley, 2007; Meins, 1995; Reiss & Rojhan, 1993). However, the relationship is not "diagnostically specific", and research does not support the use of "behavioural equivalents" (Charlot, 2005; Hemmings, Gravestock, Pickard, & Bouras, 2006). It is possible that externalising behaviours may be related to dysphoric mood (Lowry, 1998). If a person with intellectual disability presents for evaluation just because they have severe aggression does not mean they have an externalising disorder (Charlot, 2005). The patient may in fact be depressed or anxious. Individuals with intellectual disability suffering from anxiety, mania, PTSD and health problems causing pain or discomfort all have elevated likelihood of becoming aggressive. Similarly, in a person with apparent obsessive or compulsive behaviours alternative diagnoses may include the rituals seen in autism spectrum disorders, stereotypies as well as tics (Bodfish et al., 1995).

Charlot et al. (1993) argued that aggression acts as a "final common pathway" for distress in individuals with limited behavioural repertoire. Like neurotypical children, individuals with intellectual disability will often engage in negative behaviours rather communicate distress. A recent investigation by Hayes, McGuire, O'Neill, Oliver, and Morrison (2011) showed that individuals with severe and profound intellectual disability with low mood were more likely to display challenging behaviours than peers who did not have such difficulties. From a behavioural perspective, dysphoric mood (sadness, anxiety, irritability) may render demands more aversive and increase the person's efforts to escape them (Lowry & Sovner, 1992). The elevated risk for aggression in this setting is "state dependent". Similarly, anxiety may drive an increase in "attention seeking", when the individual is trying to gain caregiver assistance to reduce the sources of distress. It is important to recognise that individuals with intellectual disability, by definition, have abnormal brain substrates and are less likely to be capable of regulating their dysphoric mood states.

Management

The main focus of management is to treat the underlining cause that has been identified by a comprehensive multidisciplinary assessment (e.g. addressing sensory impairment, medical condition, physical problems, poly-pharmacy within a developmental perspective) (Al-Sheikh & O'Hara, 2008; Morin, Rivard, Cobigo, & Lepine, 2010). However, it is important not to underestimate the possible benefit of changes to the environment (living situation, family dynamics, occupation, stimulation) and the need for increasing staff/family carer awareness of mental health issues and training in ways in which they may be able to support the individual.

Before the commencement of any treatment, be it pharmacological or psychological, all measures should be taken to ensure that the individual's capacity has been assessed and where possible to obtain consent for the treatment plan. Verbal and written/visual information on the likely benefits and disadvantages of each mode of treatment, including potential side effects associated with medication, must be provided to the individual and depending on the circumstances, to paid carers and family.

The medication of choice for depression or OCD is specific serotonin reuptake inhibitors. These medications can also be used in depression that may be comorbid with anxiety disorders. Acute anxiety may be relieved with benzodiazepines but care should be taken to avoid longer-term use with the attendant problems of dependence. Also, medications should not be stopped abruptly as there is the risk of withdrawal symptoms which may resemble those of the original disorder. Overall, pharmacotherapy should

be carried out in a systematic fashion with slow titrations and detailed observations of any symptoms to avoid unnecessary side effects or alleviate known side effects in a timely fashion (Santosh & Baird, 1999).

In the UK, the National Institute for Health and Clinical Excellence (NICE) guidelines for the management of mood disorders (2011) suggest that people with mild intellectual disability should receive the same interventions as those offered to adults in the general population (Hassiotis et al., 2011). Further consultation with a specialist health professional in intellectual disability may aid the implementation of an appropriate treatment plan.

Regarding treatment of PTSD, NICE (2005) does not recommend routine debriefing following the traumatic event and suggests trauma-based psychological treatment only if symptoms persist 3 months after the traumatic event, as the first line of treatment. The clinical evidence suggests people with intellectual disability with PTSD will benefit from all different forms of psychotherapeutic methods as general population (McCarthy, 2001). A variety of such approaches have been adopted such as exposure therapy and imagery rehearsal therapy (Lemmon & Mizes, 2002; Stenfert Kroese & Thomas, 2006). A recent trial of eye movement desensitisation and reprocessing (EMDR) in four people with intellectual disability and PTSD (Mevissen, Lievegoed, & De Jongh, 2011) demonstrated that PTSD symptoms had decreased and the gains were maintained at 3 months to 2.5-year follow-up. In addition there was improvement in depressive symptoms and social and adaptive skills.

Willner (2005) reviewed the effectiveness of psychotherapeutic interventions for people with intellectual disability and concluded that the available literature supports the use of several types of psychological therapies for people with mild intellectual disability. Feasibility studies to investigate the effectiveness and costs associated with such interventions are under way (Hassiotis, Tanzarella, et al., 2011).

The Improving Access to Psychological Therapies report (IAPT, 2009) prepared by the National Health Service, UK, recommends that people with intellectual disability should have equal access to psychological therapies. It recommends reasonable adjustments such as providing materials in easy-to-read format, offering appointments at convenient times for the individual and his/her carer, longer duration of the sessions and engagement with primary healthcare and community specialist intellectual disability services to improve the availability and efficiency of service delivery.

Prevention

Given the increased vulnerability of people with intellectual disability to develop mental disorders, including those affecting mood, stronger focus needs to be placed on strategies which include:

(a) Person-centred planning: In a longitudinal analysis of the impact and cost of person-centred planning for people with intellectual disability in England, it was concluded that after introduction of person-centred planning, positive changes were found in the areas of social networks, contact with family, contact with friends, community-based activities, scheduled day activities and choice (Robertson et al., 2006). All these factors are known to contribute toward maintaining stable mental health.

(b) Promotion of healthy lifestyle: It has been well recognised that there is a common interface between physical health conditions and mental illness in people with intellectual disability. It is therefore recommended that family carers, social care providers, primary care practitioners and specialist psychiatric teams should all play an important role in recognising ill health and navigating access to the appropriate health services that people with intellectual disability need (Kwok & Cheung, 2007).

(c) Staff training: It is recommended that family carers and support staff should have proactive attention to loss and alertness for identification of abuse in people with intellectual disability. Active support training is an effective strategy for empowering staff to better

support people with an intellectual disability to be meaningfully engaged in daily activities. Implementation of active support was associated with improvements in service user engagement in domestic tasks and decreased depression levels (Riches et al., 2011).

Conclusion

Depressive and anxiety disorders including OCD and PTSD are frequently seen in individuals with intellectual disability but may be poorly defined and underascertained. Clinicians must be alert to the ways in which illness may present in people with intellectual disability and should strive to complete as full an assessment as possible of potential causes of onset. There is now an emerging body of work around appropriate diagnostic instruments and effective treatments that can be employed in the ascertainment and managements of mood disorders. However, further research is necessary in order to define service models and interventions that are based on best evidence to benefit service users and their carers.

Major research efforts are currently underway aimed at better explaining the significant degree in phenotypic variation so often found among individuals with the same genetic anomalies. Investigators are hoping to characterise the nature and extent of environmental influences, the effects of multiple adverse events and the impact of additive gene effects on phenotypic outcomes. Advances may point the way for more targeted treatments, and opportunities to alter the extent of morbidity associated with gene defects, including those that confer risk for mood and anxiety disorders (Hagerman, Hoem, & Hagerman, 2010).

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Personality Disorders and Intellectual Disability

12

Andrew Flynn

Introduction

The study of personality can be thought of as proceeding along two threads. The first concerns the relationship between the person and mental illness. Here personality stands as the emotional and behavioural baseline from which illness or disorder emerges and the backdrop against which its nature, course and progression may be understood. It comprises aspects of vulnerability and resilience, genetically determined or acquired through experience, that may predispose to illness in the face of stressors and which may make recovery, either naturally or through treatment, more or less likely. In this way personality represents a powerful organising principle in clinical reasoning and within the narratives that patients and professionals construct about illness and disability. From this perspective, personality and illness are maintained as distinct but interrelated ideas, one a focus for explanation and understanding, the other a target for treatment and prognosis.

Personality may itself become the focus of attention. There are occasions when interpersonal conflicts, dysfunctional attitudes, aggressiveness, self-destructive tendencies and periods of intense emotional reactivity become seen not as signs of illness but manifestations of a *personality disorder*. Interest moves away from the domain of symptoms and signs to that depth psychology and with it a figurative move from the psychiatrist's office to the psychotherapist's couch and with it from the language of physiology to that of relationships and, perhaps more significantly, from a discourse of affliction to one of personal responsibility.

Although it is easy to present these two threads as separate, the ground between them has become progressively less distinct as operationalised criteria for the diagnosis of personality disorders were established in DSM-III 3 decades ago. The personality disorders are increasingly thought of not just as conditions of psychological and social development but as complex neurodevelopmental disorders.

In this chapter we will review the concept of personality disorder as it applies to people with intellectual disability. In contrast to the general literature, research in this field remains small and unsystematic. Indeed, there is evidence that clinicians hold quite diverse views on its value as a diagnosis and how best to respond to the undoubtedly highly challenging problems that it poses. Some (Moreland, Hendy, & Brown, 2008) have questioned its fundamental validity and utility and cautioned strongly against its adoption. The issue at stake, it seems, is not whether we can *identify* personality disorder in people with intellectual disability so much as understanding the *value* in doing so.

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Studying Personality in People with Intellectual Disability

In their prelude to one of the few empirical studies of personality in people with intellectual disability, Zigler, Bennett-Gates, Hodapp, and Henrich (2002) argue that an emphasis on cognitive deficits had created an assumption, at least amongst theoreticians, that "the behaviours of people with mental retardation [*sic*] are solely the products of their cognitive deficits", with motivational, affective or personality aspects largely neglected.

In contrast to a convergence of mainstream personality research around what are increasingly considered the five fundamental trait dimensions of *Neuroticism*, *Extraversion*, *Openness*, *Agreeableness* and *Conscientiousness* (Costa & McCrae, 1992), Zigler has proposed an alternative set of dimensions to capture the motivational aspects of behaviour in people with intellectual disability: *positive reaction tendency* (PR; interest in interaction with a supportive other), *negative reaction tendency* (NR; initial wariness towards new people), *expectancy of success* (ES; anticipatory style in the face of a novel task), *outerdirectedness* (O; looking to others for clues to finding solutions to problems) and *effective motivation* (EM; taking pleasure in solving a novel problem).

The 37-item EZ-Yale Personality Scale (EZPQ) is a measure based on these dimensions and designed to help promote motivational research in people (primarily children) with intellectual disabilities. It is given as a self-report questionnaire to teachers and caregivers and asks primarily about aspects of problem-solving style in educational and related settings with prompts such as "*Child is constantly seeking attention and praise*" (PR), "Child isolates himself" (NR) and "*Child is confident*" (ES).

The EZPQ is able to discriminate amongst youngsters with intellectual disability based on socio-economic status, educational placement and experiences of academic success and failure. However, beyond its reliability and validation studies, the EZPQ has so far found little use in either research or clinical practice and remains untested against other personality models.

Its development does, however, raise some interesting points about personality research in people with intellectual disability. The first is a methodological one: to what extent is it possible to translate the defining trait features of, say, the NEO-PI-R (the measuring instrument for the conventional five factor model), into terms that can be applied to people with significant intellectual disability? This translational problem occurs in many structured psychometric scales used in intellectual disability, and in particular those for assessing psychopathology (Flynn & Gravestock, 2010). For example, in one study comparing self-report using modified versions of the well-known Spielberger State-Trait Anger Expression Inventory and Kovacs's Children's Depression Inventory (itself derived from the Beck Depression Inventory) with key informant and clinical interview as ways of assessing psychopathology in adults with intellectual disability, Bramston and Fogarty (2000) found unexpectedly low agreement between the scales and the clinical assessment. Because of literacy issues, the self-report items were necessarily changed so that they could be read out by an investigator, with statements (e.g. "I am quick tempered") converted into questions ("Are you quick tempered?") and allowance for checks in understanding, clarification and, it must be presumed, a degree of interpretation. This inevitably raises questions of method such as the extent to which an item can be reframed before it moves away from its original meaning (does feeling "upset", say, amount to the same thing as feeling "low"?) and the extent to which a validated instrument remains robust if its format is changed, even a small degree.

The second issue is more philosophical, or even ethical, in nature: in short, how far down the IQ scale does the concept of personality run? Does it work as an idea for instances of severe and profound intellectual disability in the same

way as for those with milder impairment? This is a hard question to resolve satisfactorily, turning as it does on our standards for inferring complex motivations (including hopes, wishes and belief-systems) as lying behind overt behaviour. High amongst these sits agency and, with it, moral responsibility.

Finally, there is the matter of attribution. Although many readers of Zigler's work might find his contention that personality is disregarded a provocative one, it is certainly the case that health and social care services for people with intellectual disabilities tend much more towards environmental factors as the determinants of behaviour than they do "internal" ones.

It is worth noting that intelligence itself has a trait-like structure, one with no sizeable association with any other personality dimension (Brebner & Stough, 1995). However, there is good evidence for links between personality variables and performance on intellectual tasks that are situation-dependent. For example, after being confounded by an intractable task, "mastery-orientated" children (typified by persistence and a positive outlook on challenge, captured, perhaps by the "ES" scale on the EZPQ) outperform equally intelligent children who are "helpless" (negative self-image after failure, lacking persistence) on subsequent solvable tasks (Chiu, Hong, & Dweck, 1994). Personality function may thus, under certain conditions, make an individual more likely to perform in the intellectual disability range. On occasion it may even seem to suit the individual to do so, again for motivational reasons, perhaps finding it easier to relate to a less able peer group with fewer expectations of success or for other psychologically defensive reasons, a phenomenon that has been termed "secondary handicap" (Jones, Harrison, & Ball, 2008; Sinason, 1986). Cooper and Collacott (1995) describe a particularly dramatic example in their case study of a young woman with "histrionic" personality disorder and above average intelligence who presented with what the authors termed a "pseudo-learning disability", a presumed manifestation of a defensive psychological need to be identified and accepted within a less able peer group.

Overview of Personality as a Psychiatric Condition

In 1923, Kurt Schneider published *Psychopathic Personalities*, in which he defined ten pathological, and discrete, personality types. Although he appealed to the underlying dimensional nature of character, his system was otherwise unsystematic and empirical; that is, it was based on clinical observations of temperamental and behavioural characteristics that seemed to cluster together without speculation about the underlying nature of the variations resulting in their formation (Standage, 1979). Whilst recognising personality disorder as warranting treatment, Schneider nonetheless maintained a clear distinction from mental illness, a conceptual separation that remains to the present day. Indeed, his scheme remains the basis for the identification and classification of the personality disorders.

In both systems the personality disorders are conditions of adulthood although many of their features will have clear childhood precursors or continuities. The most well-known example is the continuity of severe conduct disorder in childhood progressing, in some individuals, to the later offending behaviour and emotional disturbances of adult antisocial personality disorder.

Despite their presentation as discrete conditions, it is unusual to find "pure forms" of any of the subtypes, comorbidity or co-occurrence being the rule rather than the exception both in general adult (Dolan, Evans, & Norton, 1995) and intellectual disability (Flynn, Matthews, & Hollins, 2002) populations. Although DSM-IV (APA, 1994) as described in Table 12.1 attempts to accommodate this by grouping the personality disorders into "clusters" (A, the *solitary*; B, the *flamboyant*; and C, the *anxious*) in research studies, as in clinical experience, even these have indistinct boundaries, one of a number of enduring controversies concerning personality disorder diagnosis. For example, sensitivity to rejection (which may itself be formulated as a trait and which has been extensively studied in its own right as a risk factor for depressive illness) appears in borderline, avoidant and dependent personality disorder, although

Table 12.1 Personality disorder categories from DSM-IV (APA, 1994)

Personality disorder	Summary of principal clinical features	Cluster
Paranoid	Suspiciousness, sensitive to slights, grudge-bearing	A
Schizoid	Solitariness, disinterest in relationships, aloof	A
Schizotypal	Magical thinking, superstitiousness, social anxiety	A
Antisocial	Intolerance of frustration, aggression, irresponsible, callousness	B
Borderline	Emotional intensity and mood instability, anger and rage-prone, fear of abandonment, identity disturbance, repeated self-harm, dissociative episodes	B
Histrionic	Theatricality, emotional shallowness, suggestibility	B
Narcissistic	Grandiosity, self-centeredness, arrogance, disregard for the feelings of others	B
Avoidant	Feeling inferior, fear of becoming involved because of worries about rejection	C
Obsessive-compulsive (anankastic)	Cautiousness, overly conscientious, moralistic, rule-bound, pedantic	C
Dependent	Submissiveness, reassurance-needing, anxious preoccupation with being left alone	C

the responses to real or perceived relationship failures may be somewhat different. DSM-5 will maintain the categorical model and criteria for the ten personality disorders as described in Table 12.1 but will have a separate section to encourage the further study of specific traits in diagnosing personality disorders. This may also be a useful approach in studies looking at the diagnosis and management of personality disorders in people with intellectual disability.

Studying Personality Disorder in People with Intellectual Disability

The first studies of personality disorder as a distinct diagnostic entity in intellectual disability were undertaken on an inpatient population in Scotland in the 1980s when Ballinger and Reid (1987) employed the recently developed "generic" personality assessment tool, the Standardised Assessment of Personality (SAP; Pilgrim & Mann, 1990). The SAP was chosen as an informant-based semi-structured interview that bypassed the communication problems associated with intellectual disability. The only requirement of informants was that that should have known subjects well (i.e. across a range of situations) for a lengthy period (approximately 5 years). Approximately 50 % of subjects in this study met criteria for "explosive personality disorder", the predecessor of "emotionally unstable" personality disorder in ICD-10 and related to the cluster B disorders of DSM. When followed up these subjects were amongst the last to be discharged from the hospital, a very basic demonstration of validity.

A subsequent study, again using the SAP, confirmed the apparently high prevalence of personality disorder in people with intellectual disability, this time in contact with services in the community (Khan, Cowan, & Roy, 1997). However, in another large UK community survey, using a method of clinical case-note identification of patients with personality disorder (Naik, Gangadharan, & Alexander, 2002), the investigators could find only 29 cases out of a sample of 430, with the diagnosis confined to a subgroup with mild or borderline intellectual impairment. This study, as much one of the *clinical perception* as it is of "objective" epidemiology, throws some light on psychiatric diagnostic practice. In particular it picks up on an issue we have already encountered: should the diagnosis of personality disorder be restricted to the group of people with intellectual disability for whom the idea of personality is most easily conceived? That is, is it appropriate as a diagnosis for only the *mildest* presentations of intellectual disability?

As we have already seen, there is no straightforwardly principled answer to this question. But, still, these findings certainly resonate with usual clinical practice and it was, too, the position adopted by Flynn et al. (2002) in their survey of another specialist inpatient population.

This study employed a mixed design to try and shed further light on validity and also aspects of diagnostic practice. Using the SAP this study once more found a high prevalence of personality disorder: this time as many as 92 % of patients meeting the threshold for at least one type of personality disorder, with "dissocial" and "emotionally unstable" especially prevalent (53 % and 50 %, respectively) though with "paranoid" (64 %) the most common of all.

In contrast to previous work, allowance was made for comorbid personality disorders with the result that the mean number of personality diagnoses per patient was 3.4, broadly in line with observations in the general adult psychiatric population (e.g. Dolan et al., 1995). When an arbitrary level of 3 or more concurrent diagnoses as indicative of "severe personality disorder" was used in subsequent analyses, the prevalence rate fell to 38 %.

The authors drew special attention to their observation that fewer than half of these "severe" subjects had diagnoses of personality disorder made by their psychiatrist. This was not to say, they concluded, that clinicians were missing the diagnosis or were necessarily making any error in their assessments. It pointed, instead, to the possibility that diagnosis depended on considerations other than just objective behaviours or emotional states. For example, when asked for thoughts on this diagnostic discrepancy, one psychiatrist commented on the lack of useful therapeutic guidance that the diagnosis offered and, in particular, the negative connotations that the diagnosis of personality disorder carried, with the possible risk of becoming a self-fulfilling prophesy and prejudicing discharge.

Aetiology and the Validity of Personality Disorder in Intellectual Disability

The best-established risk factor for the personality disorders (though principally for the cluster B conditions) in the general population is neglect or abuse in childhood (Herman, Perry, & Kolk, 1989). Although some children, perhaps because of a complex set of personal and social-learning factors collectively conferring "resilience", may escape later psychiatric complications, many will later suffer with mood (including bipolar) disorders, be vulnerable to psychotic episodes, fall into repeated offending or develop the pattern of severe emotional dysregulation characteristic of borderline personality disorder. Such early pathogenic experiences may be overrepresented in people with intellectual disabilities (Sequeira & Hollins, 2003), in particular those with milder degrees of cognitive impairment with its stronger association with general indices of social deprivation.

In the Flynn et al. (2002) study already described, the arbitrarily set research threshold of "severe personality disorder" showed a significant association with case records of early adversity, including abandonment into care, physical and sexual abuse and care proceedings for neglect, an association that persisted regardless of *clinician* diagnosis of mental illness, personality disorder or other developmental conditions such as autism. In other words, certain types of disturbance (notably aspects of impulse control and general emotional dysregulation) were associated with early adversity even if the patient never received a diagnosis of a personality disorder.

Though far from conclusive, the construct of personality disorder in intellectual disability shares at least some validity-relevant features with the general population. However, aetiology (social, genetic or otherwise) is only one aspect of the arguably more pertinent idea of clinical *utility*. Though it is possible to identify patients who might be candidates for a diagnosis of personality disorder, studies of prognosis and response to intervention remain lacking despite their recent rapid growth in the mainstream literature.

Absence of validity and a clear script for therapeutic action may be one reason for ambivalence amongst clinicians. However, aspects of the interpersonal functioning of patients typically thought of as "personality disordered" may be another.

The Interpersonal Dimension of Personality Disorder

> When a patient gets better it is a most reassuring event for his doctor or nurse … But those who recover only slowly or incompletely are less satisfying. Only the most mature of therapists are able to encounter frustration of their hopes without some ambivalence towards the patient; with patients who do not get better, or who even get worse despite long devoted care, major strain may arise. (Tom Main, *The Ailment*, 1957).

In his famous study published as *The Ailment*, the psychiatrist, psychoanalyst and coiner of the term "therapeutic community" Tom Main (1911–1990) wrote about his experience of working with a particularly difficult group of patients in an anonymous specialist psychotherapeutic hospital. These patients, he noted, had been referred following long careers of unsuccessful treatment and a variety of psychiatric diagnoses varying from "severe hysteria and compulsive obsessional traits to depressive and schizoid character disorder". Although he does not use the term, most of these patients would nowadays attract a diagnosis of borderline personality disorder (Stein, 2011). Borderline personality disorder has probably received more research and therapeutic attention than any of the other personality disorders (Dahl, 2008) and, in the minds of many clinicians, represents the archetypical form of severe personality dysfunction. What in Main's view united these patients was an ability to bring about complex and conflicting emotional reactions in professional caregivers, feelings that would sway between a kind of heroic optimism, through a compelling need to respond (no matter how ineffectively) to expressions of distress, to, ultimately, frustration, rejection and feelings of professional failure. In his analysis these emotional responses themselves become part of the disorder, forming part of a cycle of interaction that has the capacity to maintain dysfunction. The patient brings about emotional changes in others that, in turn, rekindle symptoms in the patient and so on. At its worst such responses may be implicated to some of the fatal outcomes seen for such patients in hospital care through a process of "malignant alienation" (Watts & Morgan, 1994). As we shall see, attending to this aspect of borderline personality disorder is an important aspect of its treatment, both with medication and psychotherapy.

The capacity of personality disorder to induce antipathy in psychiatrists was shown in a famous and regularly cited vignette study by Lewis and Appleby (1998). Two groups of experienced psychiatrists were asked to comment on a case vignette of a difficult patient. In one group, however, reference to the diagnosis of personality disorder was included with the result that respondents were more likely to rate the patient's behaviours as manipulative, under voluntary control, not part of an "illness" and attention-seeking in nature. Unsurprisingly, such patients were not regarded positively as candidates for treatment. Indeed, the sense of nihilism and therapeutic rejection that the mere mention of personality disorder seemed able to induce in clinicians led the authors to conclude that the diagnosis should be abandoned. However, the fact that such an ambivalent response can be induced at all is of special interest.

A recent variation of this study, this time focusing on intellectual disability, surveyed the responses not only of specialist psychiatrists but also behaviour therapists and direct cares staff working in the Netherlands (Van den Hazel, Didden, & Korzilius, 2009). Whilst not expressing the same sense of negativity as general psychiatrists of a decade ago, these clinicians still regarded the presence of a personality disorder, even when no other aspect of it was specified, as having negative therapeutic and prognostic implications.

Personality Disorder and Mental Illness

As we have already seen, there is extensive co-occurrence amongst the personality disorders themselves with a significance that remains a

matter for debate. We have also seen that there are important connections between some neurodevelopmental conditions and personality disorder, again with their significance unclear.

However, to date there has been far greater interest in general in the relationship between personality disorder and mental illness or, in the DSM system, Axis II and Axis I. Extensive comorbidity of this type is as well-established a phenomenon as that amongst the personality disorders themselves. For example, recurrent depressive illness and dysthymia have been shown to have high rates of association with personality disorder diagnoses. The relationship between personality disorder and mental illness is usually understood in a number of ways.

Personality Disorder Function as a Risk Factor for Mental Illness

Social factors in interaction with personality vulnerabilities (which in some may reach the level of personality disorder) play a central role in the onset and maintenance of a number of important psychiatric conditions, including for people with intellectual disability (Hartley & MacLean, 2009). Patients' personality disorder may act in ways that alienates them from sources of social support at times of stress, expose themselves through impulsivity to potentially traumatic events or bring about crises as a result of interpersonal conflict. At other times their interpersonal style may undermine the efforts of others to help. In their study of the relationship between exposure to life events and psychiatric problems in people with intellectual disability (Tsakanikos, Bouras, Costello, & Holt, 2006), it was found that the occurrence of multiple life events, in particular "dependent" ones (i.e. events that may have been brought about directly through the person's own conduct), were especially marked amongst patients who had received a personality disorder diagnosis.

Another example might be substance misuse, which although understudied in intellectual disabilities is a well-known complication of personality disorder in the general population. The connections are again complex, however, with both social (e.g. acceptance by a peer network) as well as personal aspects involved. There is evidence, for instance, from clinical and laboratory studies that challenges to self-esteem in vulnerable individuals (typically those high in self-consciousness) generate an experience of emotional distress that increases the use of alcohol, if it is available, as a soothing measure (Baumeister, 1997). Other measures of impulsivity are similarly increased in such circumstances. The ability to tolerate distress without resorting to harmful coping methods is the aspect "ego function" often most obviously impaired in borderline and antisocial personality disorder patients. In interviews with people with intellectual disabilities who misuse substances (Taggart, McLaughlin, Quinn, & McFarlane, 2007), participants spoke of substances both in terms of a way of "fitting in" and helping to cope with the distress of traumatic experiences, including physical, emotional and sexual abuse.

Personality Disorder May Make a Mental Illness Harder to Recognise

The problem of "diagnostic overshadowing" may apply here much as it does for people with intellectual disability more generally. It is easy to miss a major depressive episode in the context of a lifelong tendency to emotional instability or chaotic relationships. In some cases of posttraumatic stress disorder ensuing complex emotional disturbance, substance misuse and relational difficulties may take on the quality of a personality disorder (e.g. Lonie, 1993) as may attention-deficit hyperactivity disorder (ADHD; Philipsen et al., 2008).

Personality Disorder May Make a Mental Illness Harder to Treat

This could be because of special aspects of their underlying "neurobiology" as well as the problems helping such individuals access and make effective

use of medical and psychological therapies. This has been shown, for example, for comorbid depressive illness (Newton-Howes, Tyrer, & Johnson, 2006).

Personality Disorder May Represent a Case of Misattribution or Misdiagnosis

Whilst schizoid and anankastic personality disorders may well be forms of autism known by another name (Anckarsater et al., 2006; Bejerot, 2007), the most lively debate here has been over the boundary between affective illness and borderline personality disorder. This is perhaps not surprising as recognition of a "bipolar spectrum" has become increasingly popular and acquired a media profile (Chan & Sireling, 2010), both conditions sharing affective instability and impulsivity as important features and with evidence that bipolar disorder is overrepresented in the families of people diagnosed with borderline personality (e.g. Akiskal, Hirschfeld, & Yervanian, 1983; Perugi & Akiskal, 2002). From this perspective, mislabelling a complicated or atypical mood disorder as a personality disorder risks not only denying patients access to potentially effective forms of pharmacotherapy but also the care and concern due to those who, despite their relational challenges, are "ill".

This last point is telling and its implications are worth considering with important considerations for clinical practice and in the interpretation of research. For philosophers of psychiatry (Fulford, 1994), it turns less on a question of facts than of values. Reviewing the nature of care-eliciting behaviour in psychiatry, Henderson (1974) concluded that, aberrant and ultimately self-defeating though they might seem, dysfunctional expressions of distress nonetheless fulfil the "phylogenetically ancient" function of bringing others closer. In doing so, however, patients are perceived as violating the terms of the "sick role", the implicit clinician–patient contract that underpins the delivery of the traditional medical model of health care. Although not the sole factor, the perception of emotional distress as "abnormal illness behaviour" may partly dictate whether a patient is seen as suffering from a mental illness or a personality disorder when there is a potential choice between these on symptomatic grounds.

In his important review of the relationship between personality disorder and mental illness, Kendell (2002) concluded that, because the term "mental illness" has no universally agreed meaning, it is impossible to decide the issue as to whether or not the personality disorders *really are* mental illnesses or something separate. Furthermore, whatever historical divides there may have been are being steadily eroded by both clinical, genetic and neuroscientific research.

Personality Disorder or Challenging Behaviour?

Although this debate over boundaries has concentrated on mental illness, within the field of intellectual disability, the one with challenging behaviour, the literature for which is rather more substantial, becomes as relevant.

There is evidence from a number of studies (Hastings & Remington, 1994) that staff responses play a role in the development and maintenance of challenging behaviour. The complex determinants of these responses include aspects of the behaviour itself, personal factors in caregivers (such as depressive symptoms) and perceived peer and organisational support. Although research looking for particular patterns of negative emotional responses towards challenging behaviour has produced conflicting results, one study notable for using a mixed hypothetical and "real instance" method and with specific reference to clients with *milder* degrees of intellectual impairment (Wanless & Jahoda, 2002) demonstrated substantially more negative feeling towards episodes of challenging behaviour experienced in real life than those portrayed in hypothetical vignettes, with this negativity associated with the view that clients were *in control* of their behaviour.

That experience and understanding play an important part in such responses was demonstrated by Hastings, Tombs, Monzani, and

Boulton (2003) in a comparison of the self-rated emotional responses students and experienced staff to videos of self-injurious behaviour, this time in someone with severe intellectual disability. Negative emotions were more frequently endorsed in the former group, particularly in videos designed to convey "attentional maintenance" of self-injury. Of course, how such reactions play out in practice remains less certain, but there seems little doubt that under some circumstances care staff are pushed towards frames of mind reminiscent of those in *The Ailment*.

Significantly, the motivation for clients with intellectual disability to satisfy complex attachment-based (and, hence, personality-like) relational needs through challenging behaviour remains largely unexplored. In a preliminary study of 54 school-leavers with moderate and severe intellectual disability, Clegg and Sheard (2002) found that a third showed challenging behaviours that, in the view of caregivers, related to "over-investing in one or a few relationships which became a source of jealousy" and which, on the basis of qualitative comments, had the quality of insecure attachment. Here once more we encounter the limitations of personality attribution in mild as opposed to more severe intellectual disability. To over-infer complex motivational attributes in someone with severe intellectual disability may be as problematic as to negate it in those with milder cognitive impairment.

The Treatment of Personality Disorder in Clinical Practice

However we frame personality disorder—as a form of mental illness, an attachment disorder, an extension of a neurodevelopmental condition or otherwise—it remains difficult to treat and the problems associated with it are usually long lived, recurrent and, for borderline and antisocial personality disorder, associated with high morbidity and mortality. Clinicians become easily frustrated and the teams in which they work may unwittingly find themselves under the influence of destructive interpersonal forces. The opportunity to refer such patients to specialist personality disorder services is welcomed by generic mental health services. There is a growing trend towards such specialist services in the UK following the publication of the key policy paper "Personality Disorder: No Longer a Diagnosis of Exclusion" and treatment guidelines for borderline personality disorder (National Institute for Health and Clinical Excellence [NICE], 2009) and, subsequently, antisocial personality disorder.

Pharmacological Treatment of Personality Disorder

Despite being so regularly deployed in the management of borderline personality disorder especially, medication is perhaps the least well conceptualised of the available treatments both in research and in clinical practice. Is treatment aimed at the disorder itself, component aspects of it (e.g. affective instability) or at the comorbidities (such as major depression) that so often accompany it? Alternatively, if we accept the proposal that cases of personality disorder are, in fact, forms of affective disorder or untreated ADHD, then the proposed remedy becomes, arguably, uncontroversial. Moreover, clinical trials for more severely disturbed patients can be difficult to conduct over long follow-up periods, and placebo-response rates are often reported as high.

A recent systematic review of the treatment of borderline personality disorder has gone some way towards unpicking a few of these possibilities (Lieb, Vollm, Rucker, Timmer, & Stoffers, 2010), at least for the general psychiatric population. For example, whilst benefit could be demonstrated across several symptom domains (interpersonal pathology, impulsivity and self-harm, affective dysregulation) for the atypical antipsychotic aripiprazole as well as a number of anticonvulsants (principally sodium valproate, topiramate and lamotrigine), an effect for olanzapine was confined to affective symptoms. Surprisingly there was little evidence to support the use of SSRIs unless there was a clear major depressive episode. Finally, no medications could

yet demonstrate efficacy for the symptoms of abandonment anxiety, inner emptiness, identity disturbance or dissociation.

Aside from the difficulties of interpreting the evidence, prescribing for patients with personality disorder is a complex matter for other reasons. For example, some patients seem to form psychological attachments to medication as tokens of relationships with prescribers in a manner analogous to transitional objects (Book, 1987). Attempts to limit medication or to withdraw it can result in emotional deterioration when such actions are perceived as a form of abandonment or a prelude to such abandonment. At other times prescribed medication may be used to overdose and, risk aside, damage the clinician–patient relationship further. Finally, care staff may find themselves resorting to medication to help manage their own negative feelings towards the patient, preferring sedation to emotional demand and exhaustion. A final passage from *The Ailment* is both instructive and cautionary in this regard:

> For a time I studied the use of sedatives in hospital practice, and discussed with nurses the events that led up to each act of sedation. It ultimately became clear to me and to them that, no matter what the rationale was, a nurse would give a sedative only at the moment when she had reached the limit of her human resources and was no longer able to stand the patient's problems without anxiety, impatience, guilt, anger or despair.

Psychological Interventions

Dunn and Bolton (2004) describe a case of a man with mild intellectual impairment and a history of violent threats and other problems who had been admitted to a secure forensic unit. His behaviour at first worsened, staff construing his problems in terms of attention-seeking and due to his intellectual disability. Reactive responses appeared to do little other than reinforce bouts of threatening conduct. However, making a positive diagnosis of borderline personality disorder, the authors claim, allowed the staff team to understand his behaviour in terms of psychological defence against poor self-image, to reflect on the ways in which they responded to this, to recognise and acknowledge harmful "splits" that had emerged between them as a team and for him to begin a slow recovery.

Psychotherapy forms the cornerstone of the treatment of personality disorder. Dunn and Bolton outline a multifaceted and pragmatic model of treatment drawing on several psychological traditions. The act of diagnosis, they argue, allows this to be achieved with a degree of coherence that would not otherwise have been possible and for which the framing as "challenging behaviour" would have been inadequate. The approach they describe is otherwise surprisingly "non-specialist" in many respects.

The NICE guideline for borderline personality disorder emphasises just this sort of multimodal approach (NICE, 2009). In practice, however, the special needs of this patient group make it both hard to achieve and easy to undermine and have led to the development of several structured, manualised and increasingly evidence-based treatment programmes that, despite differences in theoretical origin, try to ensure consistency and effectiveness in practice. Two have excited special interest over the past decade: dialectical behavioural therapy, derived from the cognitive-behavioural tradition, and mentalisation-based therapy (MBT) with roots in psychoanalysis, attachment theory and social neuroscience.

MBT is an individual and group treatment extending over an 18-month period. The model proposes that the "core" psychological deficit in personality disorder is an inability to accurately understand the behaviour of the self and others in terms of "intentional" mental states, that is, as arising from thoughts, feelings, desires and wishes. Borderline patients are notably prone to inflexible or "black and white" thinking, with rapid switches in their perceptions about the motives or intentions of others. These are often based on misattributions of innocent behaviours or adopting an excessively egocentric perspective in their interpersonal dealings. At times this can be extreme and form a trigger for self-harm, harmful impulsive acts or displays of intense anger. MBT makes extensive use of notions of empathic reasoning and, through repeated practice in therapy, learning how to reflect on

instances of self-destructive behaviour from the perspectives of other people.

MBT has been the subject of two important randomised controlled trials (Bateman & Fonagy, 1999, 2009). Results are impressive, at least in the hands of MBT's developers, with substantial and sustained improvements in self-harm, depressed mood, hospital admission and use of psychotropic medication. There is evidence that improvement continues even during the post-treatment follow-up period, not just in terms of behavioural regulation and diminished use of hospital admission but also in positive affect.

In a recent replication study (Bateman & Fonagy, 2009), MBT was compared with an *enhanced* control condition (as opposed to the more usual, and often rather minimal, "treatment as usual") termed structured clinical management, a group and individual treatment schedule but designed for non-specialist clinicians working in a general psychiatric settings. Both MBT and the control treatment showed significant positive effects, though with MBT the more effective of the two. The authors draw considerable encouragement from this: non-specialist "best practice" treatment can be an effective option where a specialist service is not available. However, the latter, perhaps because of closer adherence to a coherent guiding theory, has important additional benefit and may be suited to more complex or severe cases.

MBT so far remains untested with adults with intellectual disability where, perhaps because of its more obvious behavioural influences and use of some non-verbal techniques, there has been greater interest in DBT. DBT incorporates Zen principles of mindful awareness of emotions alongside more traditional behavioural analysis of behaviours as well as social skills building. Like MBT it is carried out through closely supervised group and individual work. The model postulates the "invalidating environment" in which the child's developmental need for empathic emotional responsiveness from caregivers is repeatedly frustrated as the cause of borderline and related personality disorders. Once established, this pattern continues into adulthood with symptoms provoked by repeated experiences of psychological invalidation in the course of interpersonal encounters. Most abusive environments are invalidating in this context, but the model helps understand why not all cases of borderline personality disorder are associated with frank trauma, with more subtle forms of inappropriate responsiveness (or "misattunement") able to exert a similarly harmful effect on some especially vulnerable children (e.g. Lyons-Ruth, 2003).

Evidence for the effectiveness of DBT remains primarily for deliberate self-harm in women, with its impact in other domains less certain. In a recent randomised trial comparing DBT with an enhanced non-specialist psychodynamically informed control treatment (McMain et al., 2009), DBT, whilst effective, failed to show superiority. A small number of pilot DBT projects using modified versions of the manual have been described in intellectual disability services (Baillie et al., 2010; Charlton & Dykstra, 2011) though information on outcomes is still awaited.

Other established psychotherapies have been modified for working specifically with borderline patients though not, to date, for people with intellectual disability. Cognitive Analytic Therapy (CAT), for example, has developed a specific treatment manual (Ryle, 1997) and has been subject to a recent randomised controlled trial (Chanen et al., 2008). It has, though, remained a relatively brief, individual treatment and perhaps because of this has yet to emerge as superior to routine clinical care.

Conclusion

The argument presented here is but one way of looking at diagnosing personality disorder in people with intellectual disabilities. In doing so we have not only had to consider matters of methodology but also the interpersonal aspects of this collection of conditions, and in particular the ways these may play on clinical judgments.

Although borderline personality disorder, or at least aspects of it, seems to improve substantially in the general population with time and the avoidance of a fatal outcome (Zanarini, Frankenburg, Hennen, & Silk, 2003), it is nonetheless true that the personality disorders are

enduring in nature. In fact, it is partly by definition that they must be so. It is, though, felt as a *different kind* of enduring to that of the chronic or treatment-refractory mental illnesses or persistent "challenging behaviour". The origins of this difference in perspective, as one driven by "countertransferential" reactions to certain types of care-seeking behaviour, have been an important concern for us here.

Beyond this, no one knows the outlook for people with intellectual disability who have a personality disorder. There are clues in the general literature about what might help and what may make things worse, many of which can be observed in care settings. We do know that however the mental health needs of this group of people are labelled (atypical mood disorder, complex challenging behaviour, social care issue), chronic and severe emotional problems tend to remain so for considerable periods despite the efforts of professionals to treat or support them. To the extent that we are aware of our own reactions to these problems, particularly when these reactions involve withdrawal or an impulse to displace them elsewhere, then we might say that we are dealing with a "true" case of personality disorder.

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Dementia

13

André Strydom and Amanda Sinai

Introduction

Over the years, significant improvements in health and social care for people with intellectual disability have led to a dramatic increase in the life expectancy of this population. For example, the mean life expectancy of people with intellectual disability in the 1930s was estimated at 18.5 years, which increased to 66 years in the 1990s (Braddock, 1999). These improvements in life expectancy have been particularly striking in the Down syndrome population—it has been estimated that the survival of Down syndrome babies with additional congenital birth defects increased from 0 to 18 years or more in the early 1990s (Yang, Rasmussen, & Friedman, 2002) and that the number of people with Down syndrome over 40 years old has doubled in Northern European countries since 1990 (De Graff et al., 2011). There is therefore now a much larger proportion of people with intellectual disability living into older adulthood, which is associated with increased rates of age-related conditions, including dementia. It is important to remember that the impact of debilitating conditions such as dementia reaches much further than the individual affected and may have social, financial and emotional consequences for families, carers and other social support systems, including regional and national level healthcare and social welfare systems.

Although many of the causes and features of dementia in people with intellectual disabilities are similar to that of the general population, there are some differences which will be highlighted in this chapter. Predisposing factors may include genetic risk factors, for example, triplication of genes such as APP in people with Down syndrome. Other factors may also play a role in predisposing or protecting people with intellectual disability to dementia. People with intellectual disabilities may have lower baseline cognitive function and therefore lower cognitive reserve. This may lead to earlier development of symptoms during the course of neurodegenerative disorders, such as dementia, as the disease rapidly overcomes low cognitive reserve. Comorbidities, such as head injury or epilepsy, may also influence the presentation. People with intellectual disability have higher rates of obesity (de Winter, Bastiaanse, Hilgenkamp, Evenhuis, & Echteld, 2012), which is a risk factor for Alzheimer's disease and dementia in general (Ballard et al., 2011). However, they are less likely to be smokers, which may therefore be a protective factor for people with intellectual disability.

As a defining characteristic of dementia is a decline in cognitive function, making a diagnosis of dementia in a person with an intellectual

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disability may be particularly challenging, especially if their previous level of cognitive functioning has not been well documented. As well as this, other features may also be more common in dementia in people with intellectual disability, such as the early presentation of frontal lobe-related features in Alzheimer's disease in people with Down syndrome. Although assessment and management of dementia in people with intellectual disability is largely the same as that in the general population, there are also some additional considerations that need to be taken into account when assessing and managing dementia in people with intellectual disability; these will also be discussed in more detail in this chapter.

Box 13.1 Some of the Main Types of Dementias

- Alzheimer's disease
- Vascular dementias
- Lewy body dementias
- Fronto-temporal dementias (e.g. Pick's disease)
- Dementias related to other conditions (e.g. dementia in Huntington's disease, dementia in HIV)
- Treatable dementias (e.g. hypothyroidism, vitamin B12 deficiency, hypercalcaemia, syphilis)
- Genetic syndromes associated with dementia presenting at an early age (e.g. Sanfilippo syndrome, Cockayne syndrome)

Definition

Dementia is a term used to describe a group of brain disorders which are characterised by a decline in memory and other cognitive functions, which are severe enough to lead to impairments in activities of daily living. Dementia is usually chronic and progressive. The definition of dementia is given in the international classification systems, ICD-10 (World Health Organisation, 1993) and DSM-IV (American Psychiatric Association, 2000). In order to make a diagnosis, both classification systems require development of a decline in memory and other cognitive functions. DSM-IV requires that these should be sufficiently severe to cause impairment in social or occupational functioning; ICD-10–DCR does not define this as a separate criterion. ICD-10 requires an additional criterion of change in behavioural or emotional functioning and also requires that symptoms are present for at least 6 months (World Health Organisation, 1993). DC-LD is a diagnostic classification system based on ICD-10, which has been designed for use in adults with moderate to profound intellectual disability, which also includes a definition of dementia that emphasises the importance of a change from a premorbid level of functioning (Cooper, Melville, & Einfeld, 2003).

There are several types of dementias, which have different aetiologies and clinical features. Box 13.1 lists some of the different types of dementias.

In the general population, DSM-IV criteria have been found to be more inclusive compared to ICD-10 criteria (Erkinjuntti, Ostbye, Steenhuis, & Hachinski, 1997). This is also the case in people with intellectual disabilities (Strydom, Livingston, King, & Hassiotis, 2007). Both ICD-10 and DSM-IV criteria showed substantial inter-rater reliability ($\kappa > 0.68$) and high specificity (~95 %) but were less stable than in the general population, and some caution is advisable in those with more severe intellectual disabilities or additional sensory disability (Strydom et al., 2013).

The DSM-5 workgroup has proposed a new "neurocognitive disorder" category which contains three broadly defined syndromes—delirium, minor neurocognitive disorder (similar to the current concept of MCI) and major neurocognitive disorder (dementia)—while known aetiologies are to be coded as subtypes (http://www.dsm5.org/proposedrevision/Pages/NeurocognitiveDisorders.aspx). Memory decline may not be an absolute requirement for a diagnosis of neurocognitive disorder. These changes could improve the ability to diagnose dementia

in individuals with intellectual disabilities but may also affect the number of people who are diagnosed.

Epidemiology

Most epidemiological studies of dementia in those with intellectual disability report separate rates for dementia in people with Down syndrome and those without Down syndrome, as there are much higher rates of dementia in people with Down syndrome.

Estimated prevalence rates of dementia in people with intellectual disability without Down syndrome aged 65 and over vary from 9 % in a US longitudinal study (Zigman et al., 2004)—which was found to be within the range for adults without intellectual disability—to more than 18 % in UK studies which is approximately 3 times higher than the general population prevalence rates (Cooper, 1997; Strydom et al., 2007). This variation between studies may be due to differences in study design as well as differences in diagnostic classification systems used (Strydom et al., 2010).

As in the general population, Alzheimer's disease has been shown to be the most prevalent type of dementia in people with intellectual disability (12 % prevalence in people with intellectual disability aged 65 and over) (Strydom et al., 2007).

Epidemiology of Dementia in People with Down Syndrome

Dementia is common in older people with Down syndrome and occurs at a younger age. Although prevalence rates vary, the prevalence of dementia in adults with Down syndrome aged between 40 and 49 has been found to range from approximately 6 % in a cross-sectional study in Ireland (Tyrrell et al., 2001) to approximately 10 % in a UK cross-sectional study (Holland, Hon, Huppert, Stevens, & Watson, 1998). Between 50 and 59 years of age, 30–40 % of people with Down syndrome are estimated to have a diagnosis of dementia (Holland et al., 1998; Tyrrell et al., 2001), while estimates in those aged 60 and older vary between 26 and 77 % (Coppus et al., 2006; Visser, Aldenkamp, vanHuffelen, Kuilman, & Overweg, 1997). Prevalence rates in these oldest adults may be lower compared to younger groups due to increased mortality of those with dementia (Coppus et al., 2006).

Incidence of dementia in people with Down syndrome continues to increase with age, to 13.3 per 100 person years in those aged 60 and older (Coppus et al., 2006). Although risk of dementia in people with Down syndrome has not been found to be associated with gender, the age at onset of dementia in women with Down syndrome has been found to be correlated with the age of onset of menopause (Cosgrave, Tyrrell, McCarron, Gill, & Lawlor, 1999).

It is often thought that the prognosis of Alzheimer's disease in people with Down syndrome is worse than in the general population but there are no comparison studies. One follow-up study in an institutionalised sample of people with Down syndrome found that the survival period from onset of dementia to death was 3.5 years, with a mean age at death of 59.3 years (Margallo-Lana et al., 2007).

Some other rare genetic syndromes are also associated with intellectual disability and symptoms of dementia. These include Sanfilippo syndrome (mucopolysaccharidosis III) (Moog et al., 2007). Cockayne syndrome is related to degeneration of multiple tissues, resulting in multiple problems including premature ageing and dementia (Rapin et al., 2006). Very few studies have investigated rates of dementia in other genetic syndromes in older adults with intellectual disability. Screening for Sanfilippo syndrome and other metabolic disorders should be considered in people with intellectual disability and symptoms of dementia, particularly those with early onset (Verhoeven et al., 2010).

Alzheimer's Disease

Alzheimer's disease is the commonest type of dementia, diagnosed in 60 % of dementia cases. It is characterised by an insidious onset and decline

in episodic memory and learning often with subsequent gradual progression of symptoms.

Pathogenesis

Alzheimer's disease is associated with the appearance of amyloid plaques and neurofibrillary tangles in the brain. The amyloid cascade hypothesis suggests that amyloid deposits trigger neuronal cell dysfunction and death in the brain, although this hypothesis continues to be revised and challenged (Ballard et al., 2011). Brain atrophy is seen in Alzheimer's disease and early changes may be seen in the medial temporal lobes (Tartaglia, Rosen, & Miller, 2011); however, people with intellectual disabilities such as those with Down syndrome may have medial temporal lobe atrophy even without any symptoms of dementia (British Psychological Society & Royal College of Psychiatrists, 2009).

Amyloid precursor protein (APP) is one of the proteins associated with the production of amyloid. The gene that codes for APP is located on chromosome 21 and is therefore triplicated in people with Down syndrome (Trisomy 21)—this is thought to be one of the main reasons for the increased risk of dementia in people with Down syndrome. However, not all adults with Down syndrome develop the symptoms of dementia, even though they may have neuropathological changes. Interestingly, it has been shown that a diagnosis of dementia in people with Down syndrome is more closely related to the density of neurofibrillary tangles rather than the density of amyloid plaques (Margallo-Lana et al., 2007). It is still not clear whether there are any specific preventative factors in those people with Down syndrome who do not develop dementia, and risk is likely to be related to a combination of factors, including genetic and environmental factors. Several other genes on chromosome 21 are also implicated in the development of Alzheimer's disease in people with Down syndrome, including superoxide dismutase 1 (SOD1), S100 β, β-site APP cleaving enzyme (BACE) and DYRK1A (see Lott & Dierssen, 2010 for a review).

The variant of gene apolipoprotein E (APO-E) allele 4 is known to be associated with an increased risk of developing Alzheimer's disease. APO-E also influences the development and progression of Alzheimer's dementia in people with Down syndrome (Prasher et al., 2008). Mutations in APP and other genes, including presenilin 1 and presenilin 2, are associated with familial Alzheimer's disease, and genome-wide association studies have suggested several new Alzheimer's disease susceptibility genes (see Bertram, 2011 for a review).

Other risk factors for Alzheimer's disease include lifestyle factors such as obesity, physical activity and alcohol intake (Ballard et al., 2011). Obesity related to reduced physical exercise, poorer awareness of healthy eating and certain medications is more common in people with intellectual disability (de Winter et al., 2012).

Clinical Features

Alzheimer's disease often presents as a slow decline in cognitive functioning. This includes a decline in memory problems as well as other cognitive functions, such as speech and language skills, praxis and executive functioning skills, resulting in impairments in functional skills.

Alzheimer's Disease in People with Down Syndrome

Alongside the increased rate, Alzheimer's disease in people with Down syndrome is also known to present at a younger age. There are also differences in the clinical presentation of Alzheimer's disease in people with Down syndrome.

Like the general population, memory changes are an important symptom in dementia in people with Down syndrome. Cognitive functions have been shown to decline sequentially, with different cognitive functions being lost at different stages of the dementia (Devenny, Krinsky-McHale, Sersen, & Silverman, 2000).

Although forgetfulness and confusion are common early symptoms, frontal lobe-related features, including language problems, loss of interest, social withdrawal and emotional and behavioural changes, also present early in dementia in people with Down syndrome (Deb, Hare, & Prior, 2007). Adults with Down syndrome may meet the criteria for a dementia of frontal type before they progress to meeting the criteria for Alzheimer's disease (Ball, Holland, Hon, et al., 2006). When behavioural and emotional changes in people with Down syndrome and dementia were compared with a group of people with Alzheimer's disease from the general population, although the group with Down syndrome had fewer behavioural problems and delusions, they were more physically active (Temple & Konstantareas, 2005). As behavioural excesses and behaviours which have an impact on staff have been shown to be factors related to referral for dementia assessments (Adams et al., 2008), it is important not to forget to consider a diagnosis of dementia in individuals who develop behavioural deficits such as withdrawal.

The later stages of dementia in Down syndrome are often associated with mobility issues and falls and myoclonic jerks and epilepsy (Prasher, 1995). These are specific features that should be addressed and managed accordingly when reviewing people with Down syndrome and dementia.

Alzheimer's Disease in Other Adults with Intellectual Disabilities

Although the clinical presentation of Alzheimer's disease in people with intellectual disability who do not have Down syndrome is not very different from the general population, dementia in this group may be more difficult to recognise, especially in the early stages, as it may be difficult to differentiate any changes in cognitive function from baseline. This emphasises the importance of baseline cognitive assessments in people with intellectual disability and asking about premorbid cognitive function during assessment. A recent study found that when informants were asked about initial presenting symptoms, the majority reported general deterioration of functioning as the initial symptom (50 %). Emotional or behavioural change was reported by 15 % of informants. Memory or other cognitive changes were not as commonly identified as initial presenting symptoms (Strydom et al., 2007).

By end-stage Alzheimer's disease, including that in people with Down syndrome, people may be fully dependant on others for all aspects of their care, including washing and dressing, feeding, toileting and mobility.

Vascular Dementia

In vascular dementia a stepwise decline may occur, with periods of relative stability and periods of rapid decline. There may be some areas of cognitive function that are relatively well preserved. Focal neurological signs may be present.

Pathogenesis

Vascular dementia is caused by vascular insults causing damage to the brain. Vascular insults can include one or several major strokes/cerebrovascular accidents (CVAs) or a series of smaller infarcts (multi-infarct dementia). A CVA (related to thrombosis, embolism or haemorrhage) may lead to damage to one specific area of the brain and subsequent impairment of function in that area. Multi-infarct dementia may result in multiple smaller areas of the brain being affected, which may present in a more insidious manner.

Cardiovascular factors increase the risk of developing vascular dementia. These include cardiac problems, such as atrial fibrillation, and vascular problems, such as carotid artery stenosis. Factors that increase cardiovascular risk include hypertension, hypercholesterolaemia, diabetes and family history of cardiovascular problems. People with intellectual disability may have more of these risk factors, due to genetic or lifestyle reasons. Other risk factors, such as smoking, may be less common in people with intellectual disabilities.

Clinical Features

The presentation of vascular dementia varies according to the area of the brain affected. As it is caused by a vascular insult, as well as a decline in cognitive functions, there may also be physical deficits, such as reduced power and/or reduced sensation. According to the areas of the brain affected, some functions may be relatively spared. Cases of vascular dementia or those of mixed aetiology are likely to be underreported, and it is important to remember that the two conditions can coexist. Mixed vascular dementia and Alzheimer's disease have been described in Down syndrome (Collacott, Cooper, & Ismail, 1994), but this is likely to be relatively uncommon.

Other Dementias

Although less common, it is important to differentiate other dementias as the pathogenesis, clinical features and subsequent management of the different dementias can be very different.

Lewy Body Dementia

In Lewy body dementia, deposits of Lewy body proteins (an abnormal aggregate of proteins including alpha-synuclein) are found in nerve cells in the brain. Alongside symptoms of a progressive dementia, features of Lewy body dementias may include fluctuating cognition, visual hallucinations (often seeing people or animals) and features of Parkinsonism such as rigidity, resting tremor and a slow, shuffling gait.

Although there is very little in the recent literature about people with intellectual disability and Lewy body dementias, in one cliniconeuropathological survey, 2 out of 23 people with Down syndrome were found to have Lewy bodies in the substantia nigra as well as brain changes related to Alzheimer's disease (Raghavan et al., 1993). One case study reports a case of an older person with Down syndrome, dementia and Parkinsonism and who had cortical Lewy bodies in addition to brain changes related to Alzheimer's disease (Bodhireddy, Dickson, Mattiace, & Weidenheim, 1994). A significant proportion of dementia cases (approximately 7.7 % of those aged 65 or older) met the criteria for Lewy body dementia in a dementia survey of older adults with intellectual disability who did not have Down syndrome (Strydom et al., 2007).

Fronto-temporal Dementias

Fronto-temporal dementias are a group of dementias related to damage to the frontal and/or temporal regions of the brain. Neuroimaging may show brain atrophy in affected regions. Amongst other things, the frontal lobes are important centres for personality and behaviour. One fronto-temporal dementia is Pick's disease, which can present with personality or behavioural changes, including disinhibition. Memory may be relatively well preserved in the early stages. Dementia in people with Down syndrome often initially presents with frontal lobe-related features and then progresses to more typical features of Alzheimer's disease (Ball, Holland, Hon, et al., 2006; Holland, Hon, Huppert, & Stevens, 2000). Fronto-temporal dementias may also occur in people with intellectual disability who do not have Down syndrome (Strydom et al., 2007).

Treatable Dementias

Some physical conditions can lead to presentation of symptoms of dementia. They are important to consider, as, if treated appropriately, the symptoms may be reversed. Causes of treatable dementias include hypothyroidism, vitamin B12 deficiency, hypercalcaemia and syphilis. Hypothyroidism is common in people with Down syndrome and it is recommended that all people with Down syndrome have their thyroid levels checked regularly (Down's Syndrome Medical Interest Group, 2001). Dementia may also be associated with other neurological disorders such as Parkinson's disease and Huntington's disease.

Assessment

Assessment of dementia in people with intellectual disability is similar to assessment in the general population; however there are additional factors that are also important to consider. As a diagnosis of dementia relies on a cognitive decline, it is not possible to make this diagnosis without considering the individual's baseline functioning.

The US National Taskforce on Intellectual Disabilities and Dementia Practices has recently published recommendations and a National Action Plan (National Task Group on Intellectual Disabilities and Dementia Practice, 2012). The British Psychological Society and Royal College of Psychiatrists have published guidance on assessment, diagnosis and treatment of dementia in people with intellectual disability (British Psychological Society & Royal College of Psychiatrists, 2009).

Information about baseline function can be obtained through a number of sources, including the person with intellectual disability, their relatives or carers and any professional workers involved. If a professional health or social worker has been involved, it is useful to speak to them and/or look at case records to obtain more information about their previous level of function. An IQ score or a previous cognitive or functional assessment can help contribute to an understanding of baseline functioning. Other factors, such as level of educational attainment, employment, accommodation and previous level of support required, can also give important information about the person's previous level of function.

Assessment and management of dementia in people with intellectual disability should be multidisciplinary, involving a range of professionals, so that both health and social care needs can be considered and managed in an integrated manner. Local health, social care and nongovernmental services should work together and with relatives, carers and the individual affected in order to tailor care according to the individual's needs and wishes.

History of Presentation

Features to consider during history taking include short- and long-term memory problems; other cognitive functions such as language, visual-spatial skills and executive functioning; activities of daily living; personality change; and any behavioural problems. Feeding problems, sleep problems, incontinence and mobility problems will have an impact on the individual's day to day life and the care that they will require and it is worth specifically asking about these. It is important to consider the time of onset of symptoms, duration and rate of progression, as well as the different symptoms which characterise the different types of dementia, in order to differentiate between them. A structured dementia history such as the CAMDEX-DS informant interview may be helpful (Ball, Holland, Huppert, Treppner, & Dodd, 2006).

Also important is family history of dementia. In people with Down syndrome, it is especially important to ask about falls, myoclonic jerks, swallowing problems and seizures.

Risk issues must be considered and may include self-neglect, safety concerns such as leaving taps or gas/electric cookers on and challenging behaviours, such as wandering, inappropriate behaviour such as removing clothes and aggression. Neglect and elder abuse are also potential risk issues and should always be considered.

Box 13.2 lists some of the differential diagnoses to consider when making a diagnosis of dementia.

Examination

An assessment of dementia is not complete without a physical and mental state examination, although the professional(s) who conduct this may vary according to local service structure. Physical examination will help differentiate between different types of dementia and will therefore help tailor appropriate management.

Box 13.2 Some of the Differential Diagnoses of Dementia in People with Intellectual Disability

Physical Health Problems
- Hypothyroidism
- Vitamin B12 deficiency
- Hypercalcaemia
- Syphilis
- Infection

Intercranial Pathology
- Epilepsy
- Normal pressure hydrocephalus
- Space occupying lesions

Mental Health Problems
- Depression
- Psychosis

Sensory Problems
- Hearing impairment
- Visual impairment

Psychological Factors
- Recent loss or bereavement

Social Factors
- Recent change of environment
- Elder abuse

Physical examination should include:
- General physical examination—looking for signs of treatable causes of dementia (e.g. hypothyroidism)
- Cardiovascular assessment—looking for any cardiovascular risk factors
- Neurological assessment—looking for any focal neurological signs, features of Parkinsonism and late-stage symptoms such as myoclonus and primitive reflexes
- Gait assessment—looking for any risk of falls, footwear and foot hygiene
- Clinical screen for hearing and vision problems

Investigations

Recommended physical investigations include:
- Blood tests—including full blood count, urea and electrolytes, thyroid and liver function tests, calcium levels, B12 and folate levels, blood sugar and lipid profile.
- Tests for sensory problems (hearing and vision) if indicated.
- ECG (electrocardiogram)—particularly if there are any cardiovascular features or if considering the use of medication.
- EEG (electroencephalogram)—if there are symptoms suggestive of epilepsy or myoclonus.
- Neuroimaging—brain scans may be difficult for some people with intellectual disability to tolerate. Also, neuroimaging may not be so helpful for looking at subtle brain changes in this population. For example, people with Down syndrome have smaller brains and even those without dementia may have medial temporal atrophy, which is often seen in Alzheimer's disease. Neuroimaging is therefore recommended if there are suggestions of structural intercranial pathology, e.g. space-occupying lesion or normal pressure hydrocephalus (British Psychological Society & Royal College of Psychiatrists, 2009). Although MRI is preferable to CT, CT is easier to tolerate and newer methods may provide more sensitive results. Single-photon emission computed tomography (SPECT) scanning can help clarify the diagnosis and distinguish between types of dementia (Deb et al., 1992). A dopamine transporter SPECT scan (DaTscan) is particularly useful when Lewy body dementia is suspected, while positron emission tomography (PET) of cerebral glucose metabolism could be useful to assist in the diagnosis of fronto-temporal dementia (Tartaglia et al., 2011).

Blood tests for syphilis serology and/or HIV should only be conducted if there are risk factors or the clinical picture is suggestive of the condition (National Institute for Health and Clinical Excellence, 2006).

Assessment Tools

There are several assessment tools that can be used for assessing dementia in people with intellectual disability. These are either screening tools or formal diagnostic assessment tools. However as yet there are no screening tools that can be specifically recommended on the basis of sensitivity and specificity for dementia in the intellectual disabilities

population. Most screening tools require assessments at repeated intervals in order to assess change from baseline functioning. It is therefore recommended that one of these tools is undertaken before the onset of dementia, particularly for people with Down syndrome, in order to have a baseline for future assessments to be measured against.

See Box 13.3 for some of the screening and assessment tools that can be used when assessing dementia in people with intellectual disability.

Box 13.3 Assessing Dementia in People with Intellectual Disability: Some Assessment Tools (British Psychological Society & Royal College of Psychiatrists, 2009)

Screening Tools (Completed with Informants)
- Dementia Questionnaire for People with Learning Disabilities, DLD—previously DMR (Evenhuis, Kengen, & Eurlings, 2007)
- Dementia Screening Questionnaire for Individuals with Intellectual Disabilities, DSQIID (Deb, Hare, Prior, & Bhaumik, 2007)—adapted and termed the Dementia Screening Tool in the USA
- Adaptive Behaviour Dementia Questionnaire, ABDQ (Prasher, Holder, & Asim, 2004)

Assessments of Neuropsychological Function
- CAMCOG-DS—Neuropsychological part of the CAMDEX-DS (Ball, Holland, Huppert, et al., 2006)
- Neuropsychological Assessment of Dementia in Adults with Intellectual Disabilities, NAID (Crayton, Oliver, Holland, Bradbury, & Hal, 1998)
- Severe Impairment Battery, SIB—for use in people with lower levels of cognitive function (Saxton, McGonigle, Swihart, & Boller, 1993)
- Test for Severe Impairment, TSI (Albert & Cohen, 1992)—for use in people with severe cognitive impairment. This has been shown to be used in people with Down syndrome without floor or ceiling effects (Tyrrell et al., 2001)

Detailed Informant Interviews
- CAMDEX-DS informant interview (Ball, Holland, Huppert, et al., 2006)

Assessments of Daily Living Skills and Functioning
- Assessment of Motor and Process Skills, AMPS (Fisher, 2006)
- AAMD Adapted Behaviour Scales, ABS, 1974 revision (Nihira, Foster, Shellhaas, & Leyland, 1974)
- Adaptive Behaviour Assessment System-II, ABAS-II (Harrison & Oakland, 2003)
- Vineland Adaptive Behaviour Scales: Second Edition (Sparrow, Cicchetti, & Balla, 2007)

Test Batteries
- Test Battery for Dementia (Burt & Aylward, 2000)—this incorporates several standardised neuropsychological tests and informant interviews
- Arizona Cognitive Test Battery, ACTB (Edgin et al., 2010)—this is a battery of neuropsychological tests designed for use in people with Down syndrome, although it has not yet been used in older adults or for assessment of dementia

Management

Like assessment, the management of dementia should take a multidisciplinary and person-centred approach and take into account the individual's needs and wishes. It is helpful to use a biopsychosocial approach, in order to consider biological, psychological and social interventions that may be required. Biological interventions include consideration of medications, which

may vary according to the type of dementia. Psychological and social interventions are relevant in the management of all types of dementia and should be considered in all cases. A person's needs are likely to change as their dementia progresses. Management, including medication, psychological and social interventions, will need to be reviewed and revised according to these changes in need.

Biological and Pharmacological Management

Acetylcholinesterase inhibitors (AChEIs) increase the levels of acetylcholine in the brain. They include donepezil, rivastigmine and galantamine. They are not disease-modifying drugs, and although they have been shown to improve cognitive functioning, activities of daily living and behaviour in people with Alzheimer's disease for a period of time (Birks, 2006), they do not reverse the illness.

There are few studies investigating AChEIs in people with intellectual disabilities. One open-label study examining donepezil in people with Down syndrome and Alzheimer's disease found significantly less decline in global functioning and adaptive behaviours over 2 years in the group taking donepezil (Prasher, Adams, & Holder, 2003). Prasher, Fung, and Adams (2005) found less decline in global functioning and adaptive behaviours over 24 weeks in people with Down syndrome and dementia taking rivastigmine compared to those who were untreated. The authors suggest that this may not have reached statistical significance due to small sample size (Prasher et al., 2005).

AChEIs can be used in people with intellectual disability and Alzheimer's disease. They can also be used in the management of behavioural or psychological symptoms in people with Alzheimer's disease or Lewy body dementia where non-pharmacological methods have been unsuccessful (British Psychological Society & Royal College of Psychiatrists, 2009) but are not recommended for use in vascular dementia. When considering a trial of an AChEI, clinicians should weigh up the risks and benefits as AChEIs have a number of side effects and may not always be tolerated by some people with intellectual disability. Clinicians should follow the same guidelines for initiating and monitoring AChEIs as in the general population; however it is important to be aware that people with intellectual disability may have increased susceptibility to side effects; it is therefore advisable to start at a low dose and titrate up cautiously.

Before an acetylcholinesterase inhibitor is started, blood pressure, pulse, an ECG and relevant blood tests (if not recently taken at assessment) should be conducted, as well as a baseline assessment of cognitive function which can be used to track changes. Cognitive function and behavioural, functional and global assessments should then be reviewed at regular intervals (National Institute for Health and Clinical Excellence, 2006).

Memantine is an NMDA-receptor antagonist, which has been found to have a beneficial effect on cognition, activities of daily living and behaviour in people with moderate to severe Alzheimer's disease and a very small effect on cognition in people with mild Alzheimer's disease (McShane, Sastre, & Minakaran, 2006). In Britain, it is currently used in adults with moderate to severe Alzheimer's disease, including those with intellectual disability. A recent randomised double-blind controlled trial of memantine in adults with Down syndrome aged 40 and older found no significant difference in cognition or function between those taking memantine and those taking placebo over 52 weeks (Hanney et al., 2012). A subgroup with clinical dementia did not benefit either.

Treatment of vascular dementia in particular includes reduction of cardiovascular risk factors. Medication may include antihypertensives, anticholesterol agents, antiarrhythmics and/or anticoagulation as appropriate. Other interventions may include treatment of associated symptoms related to dementia, such as antiepileptic medication for people with comorbid epilepsy or antidepressants for depression. Antiepileptic medication is useful in the management of myoclonus in people with Down syndrome.

Adequate pain management is also an important consideration. It has been shown that a systematic approach to pain management in people with moderate to severe dementia living in nursing homes can not only reduce pain but also significantly reduce agitation (Husebo, Ballard, Sandvik, Nilsen, & Aarsland, 2011).

Behavioural problems should be managed using a biopsychosocial approach, including psychological and environmental approaches, and may include consideration of medication (Royal College of Psychiatrists, British Psychological Society, & Royal College of Speech and Language Therapists, 2007). Physical interventions may be required to support physical problems, such as mobility, feeding or continence. This may require involvement of other health professionals, including occupational therapy and physiotherapy, speech and language therapy, nursing and psychology.

Psychological, Environmental and Social Management

Structured group cognitive stimulation programmes are recommended for people with mild to moderate dementia (National Institute for Health and Clinical Excellence, 2006); however these may need to be adapted for people with intellectual disability. Very little research has been conducted looking at psychological interventions for people with intellectual disability and dementia (Courtenay, Jokinen, & Strydom, 2010).

One observational tool that is being used in people with intellectual disability and dementia is Dementia Care Mapping (Brooker & Surr, 2005), which involves observation of the person with dementia and their environment to develop a "map" which can be used to aid person-centred care (British Psychological Society & Royal College of Psychiatrists, 2009).

Social interventions are important to consider in the management of dementia, in particular, consideration of the person's environment, staff training (Janicki, 2011) and carer support.

It is important to ensure that the environment is comfortable, appropriate and safe. A caring-in-place model is preferred to avoid moving a person away from an environment that they are familiar with, which may contribute to confusion and further cognitive decline. It is therefore preferable that modifications or adjustments are made to the individual's current environment. This may include providing further support for relatives and carers, including emotional support, further training as well as physical supports such as hoists, bathroom adaptations or alarms (e.g. to alert carers when the person is at the door or having a seizure). In the case where a person's environment is not suitable for them, careful consideration of new accommodation should include consideration of physical and emotional needs and the individual's beliefs and wishes and taking account of their prognosis.

End-of-Life Care

Dementia is a terminal disease and at present there is no curative treatment available. However, decline in cognitive function may progress slowly, which may provide time for the individual, together with relatives and carers to consider their wishes for their care in the later stages of the disease. Factors that may be important to consider include social-, financial- and health-related issues. It is important to remember the principles of considering a person's best interests and using the least restrictive option when making decisions for people who lack capacity to make a decision for themselves.

Conclusion

Dementia is becoming a major cause of morbidity and mortality in older people with intellectual disability. Alongside this, there have been substantial developments in the understanding of the pathology, clinical features and management of this debilitating condition. Research in the field of intellectual disability and dementia continues

to develop and ranges from genetic and molecular research to observational studies and randomised controlled trials. Hopefully, a further understanding of dementia in people with intellectual disability will contribute to both an improvement in the health and care of people with intellectual disability as well as contributing to an understanding of dementia in the general population.

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Substance Misuse

14

Laurence Taggart and Eddie Chaplin

Introduction

A review of existing literature regarding ID and substance use and abuse highlighted three clearly identifiable areas:

- Legal substances: caffeine, tobacco and alcohol
- Illicit drugs: cannabis, cocaine
- Overuse of prescribed prescription medicines

There are a number of licit and illicit substances that can be misused or abused, which can have a broad range of effects (i.e. physiological, psychological and social); these can also impinge upon their families and communities. These include tobacco, alcohol, illicit drugs and overuse of prescribed medication, although other 'drugs' such as caffeine can also be classified under the umbrella term of 'substances'. However, this chapter will exclude caffeine. This chapter will use the definition of substance abuse as in the DSM-Classification System (American Psychological Association): 'the hazardous consumption of alcohol, illicit drugs and/or over use of prescribed medications which has been proven to be harmful to the persons' physical, psychological, interpersonal and social health within a-month period'. Aspects of the definition relating to role obligation, legal implications and hazardous tasks were deemed to be less relevant for people with ID (Taggart et al., 2006) when cross-referenced with Diagnostic Criterion for People with Learning Disabilities (DC-LD) (Royal College of Psychiatrists, 2001) (see Box 14.1).

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Prevalence Rates

Smoking

Prevalence rates across a number of western countries are expected to reduce, as many countries are putting measures in place to reduce smoking (e.g. bans in public places), because of the adverse health effects associated with it. As a result smoking has been banned in a number of NHS premises and also by organisations that provide housing for people with ID within the UK. The last 20 years have provided contrary evidence as to the extent of smoking; some studies report young people with ID are less likely to use tobacco, alcohol and cannabis than their non-disabled peers (McCrystal et al., 2007). However in a review of studies McGillicuddy (2006), the author, found identical rates among high school students (Gress & Boss, 1996) and higher rates between 11- and 15-year olds, 14 % ID against 8 % of their non-disabled peers (Emerson & Turnbull, 2005).

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Box 14.1 ICD-10 Diagnostic Guidelines for Dependence Syndrome

A definite diagnosis of dependence should usually be made only if three or more of the following have been present together at some time during the previous year:

(a) A strong desire or sense of compulsion to take the substance
(b) Difficulties in controlling substance-taking behaviour in terms of its onset, termination or levels of use
(c) A physiological withdrawal state (see F1x.3 and F1x.4) when substance use has ceased or been reduced, as evidenced by the characteristic withdrawal syndrome for the substance, or use of the same (or a closely related) substance with the intention of relieving or avoiding withdrawal symptoms
(d) Evidence of tolerance, such that increased doses of the psychoactive substances are required in order to achieve effects originally produced by lower doses (clear examples of this are found in alcohol- and opiate-dependent individuals who may take daily doses sufficient to incapacitate or kill non-tolerant users)
(e) Progressive neglect of alternative pleasures or interests because of psychoactive substance use, increased amount of time necessary to obtain or take the substance or to recover from its effects
(f) Persisting with substance use despite clear evidence of overtly harmful consequences, such as harm to the liver through excessive drinking, depressive mood states consequent on periods of heavy substance use or drug-related impairment of cognitive functioning; efforts should be made to determine that the user was actually, or could be expected to be, aware of the nature and extent of the harm

*Narrowing of the personal repertoire of patterns of psychoactive substance use has also been described as a characteristic feature (such as a tendency to drink alcoholic drinks in the same way on weekdays and weekends, regardless of social constraints that determine appropriate drinking behaviour).

*It is an essential characteristic of the dependence syndrome that either psychoactive substance taking or a desire to take a particular substance should be present; the subjective awareness of compulsion to use drugs is most commonly seen during attempts to stop or control substance use. This diagnostic requirement would exclude, for instance, surgical patients given opioid drugs for relief of pain, who may show signs of an opioid withdrawal state when drugs are not given but who have no desire to continue taking drugs.

*The dependence syndrome may be present for a specific substance (e.g. tobacco or diazepam), for a class of substances (e.g. opioid drugs) or for a wider range of different substances (as for those individuals who feel a sense of compulsion regularly to use whatever drugs are available and who show distress, agitation and/or physical signs of a withdrawal state upon abstinence).

Includes: chronic alcoholism, dipsomania, drug addiction

Adapted from the ICD-10 Clinical Diagnosis Manual (www.who.int/substanceabuse/terminology/ICD10ClinicalDiagnosis.pdf, p. 5)

Alcohol Use and Abuse

Most studies cite that alcohol use in both teenagers and adults with ID is lower compared to their non-ID peers, although Burgard et al. (2000) in the first systematic review of the literature on this topic reported rates to be equal for the non-ID population. However, the growth of ‘substance abuse’ in both the non-ID and psychiatric populations

appears to be mirrored in a growing trend for people with ID to also abuse alcohol and illicit drugs. Overall the prevalence of alcohol abuse in people with ID appears to be lower when compared with both the non-ID and psychiatric populations (McGillicuddy, 2006). UK studies suggest a 1 % prevalence rate (Cooper, Smiley, Morrison, Williamson, & Allan, 2007; Taggart et al., 2006), whereas figures for USA populations range from 0.5 to 2.5 % (Slayter, 2010; Sturmey, Reyer, Lee, & Robek, 2003). Such discrepancies centre upon methodological problems concerning:

- Operational definitions of use and abuse
- How such use and misuse is reported (i.e. self-report, proxy informant reports)
- Whether the person is known to ID services or not known to services

Evidence is inconclusive as to whether teenagers with ID abuse alcohol more than their non-disabled school peers. Some studies indicate that having an ID is a precursor for developing a substance abuse disorder (Beitchman, Wilson, Douglas, & Adlaf, 2001). However this relationship is unclear as poor academic attainment, behavioural problems, low self-esteem and low socio-economic status are all associated with having a substance abuse disorder in teenagers without ID but also frequently found in young people with ID as well, whereas other studies report that having an ID is a deterrent from developing a substance abuse disorder. In reviewing the evidence pertaining to 'substance-related disorders' in adults with ID, it is difficult to conclude any agreement between these studies as to the exact prevalence of alcohol misuse. However, what is reported across many of these studies is that current prevalence rates of substance abuse among people with ID are an underestimation of the true figure which is much higher (Cocco & Harper, 2002).

Illicit Drug Use

Figures for illicit drug misuse also indicate overall lower prevalence rates compared with alcohol abuse (Christian & Poling, 1997, Gress & Boss, 1996, McGillicuddy, 2006). This use of illicit drugs in people with ID is also associated with where the person lives, in terms of both neighbourhood and with whom, age, local culture, ethnicity, availability and personal history (i.e. role models) (Snow, Wallace, & Munro, 2001; Sturmey et al., 2003). Taggart et al. (2006), in a study of 67 people with ID reported to have a substance abuse disorder in N. Ireland, found alcohol was reported to be misused by all of the users; of these 67 users, 13 (19.4 %) were also reported to be misusing other substances such as illicit drugs and/or overusing prescribed medications, including cannabis, prescribed medications (i.e. paracetamol, ibuprofen, diazepam, kepak (i.e. painkiller)), Ecstasy, amphetamines, solvents and cocaine. One participant was also described to be addicted to 'gambling machines'. The informants reported that for those 13 users misusing both alcohol and a combination of illicit drugs and/or prescribed medications, they were significantly more likely to be 30 years and below, to be male and to live independently.

Psychopathology and Substance Abuse

As with the mainstream psychiatric population, substance misuse is a growing problem leading to mental health problems. Similarly, Didden, Embregts, van der Toorn, and Laarhoven (2009) have also reported that substance abuse in people with ID also leads to severe emotional and behavioural problems.

Chaplin, Gilvarry, and Tsakanikos (2011) explored the clinical case notes of 115 consecutive new referrals to specialist mental health services for adults with ID in South-East London. The author's found alcohol was the most frequently used substance (80 %) followed by cannabis (28 %) and cocaine (12 %). Approximately 15 % of patients had a history of substance use; however only 8 % were currently using substances. Predictors of substance abuse included being male, having a mild level of ID, those with a forensic history and having schizophrenia. Logistic regression analyses revealed that those with a forensic history were about 5 times more likely to have current substance use problems. Illicit substance use was about 3 times more likely among people with schizophrenia spectrum disorders.

Offenders and Substance Abuse

Plant, McDermott, Chester, and Alexander (2011) examined the clinical case notes of 74 adults with ID in attending a community forensic ID services in England; they found alcohol was the most common substance being used, followed by cannabis, then cocaine, stimulants and opiates. Over a third of the sample was currently abusing such substances leading up to the index offence. Men and women were equally found to be abusing alcohol and cannabis. The authors found that those diagnosed with a personality disorder and a history of violent offences was more likely to predict those with a substance dependence disorder. Likewise, there is growing evidence to show that offenders with a mild ID were also found to have a comorbid psychiatric condition such as bipolar disorder, schizophrenia and personality disorder (Alexander, Bullman, & Plant, 2011; Alexander et al., 2010).

Figures for the number of people with ID in prisons vary between 0.5 and 13 % depending upon the methods utilised (Barron, Hassiotis, & Banes, 2002; Fazel, Xentiditis, & Powell, 2008; Herington, 2009; Mannynsalo, Putkonen, Linberg, & Kotilanien, 2009; McGillivray & Moore, 2001; Sondenaa, Rasmussn, Palmstierna, & Nottestad, 2008). Chaplin et al. (2011) found that those with a forensic history were about 5 times more likely to have current substance use problems. Hassiotis et al. (2011) undertook a study psychiatric morbidity in prisoners with ID across England and Wales. The authors reported high rates of psychiatric disorders particularly psychosis, attempted suicide and cannabis dependence/frequent user. Lifetime drug use and alcohol dependence were similar in non-ID prisoners. Hassiotis and colleagues found that there was a relationship between the ID prisoners, cannabis use and psychosis. Likewise, Moore et al. (2007) in their systematic review found there was a correlation between cannabis dependence and schizophrenia in the non-ID population. Klimecki, Jenkinson, and Wilson (1994) examined the influence of substance use on recidivism for people with ID who had already served a sentence, as part of a wider study. From the sample of 60, (IQ 65–75) 45.1 % of first offenders had a history of substance use, with the proportion rising to 87.5 % by the fourth offence. Of the sample the majority had a comorbid mental disorder, and of those with substance use and mental disorder, 50 % had reoffended within 1 month of release.

Risk Factors

There are a number of complex and interacting intra- and interpersonal risk factors that lead people to develop a substance abuse disorder (see Box 14.2). Many of these core risk factors have also been found to be in the non-ID population who have been found to abuse a range of substances. There is also growing research exploring the factors that predispose young people with and without ID to engage in alcohol abuse and illicit drug use. These include biological, low self-esteem, social isolation, personality, mental health, behaviours, perceived environment and social environment. On the other hand, there is some literature pertaining to factors that protect young people from abusing such substances such as perceived closeness to parents and perceptions of parental expectations for school (Blum et al. 2001). Gress and Boss (1996) in the USA identified several characteristics that might be related to why young people with LD turn to misuse of alcohol and illicit drugs. They included the inability:

- To establish self-identity
- To develop social attachment
- To project affective social images within their own peer groups
- An inability to experience immediate gratification of beliefs or desires

Taggart, Huxley, and Baker (2008) in a review of the literature on substance abuse in people with ID reported that as a greater number of people with ID are supported to live in a variety of accommodations in the community and given greater freedom, these individuals may be exposed to greater social stressors. This exposure may lead to a greater use of alcohol and illicit drugs as a coping mechanism/stress reliever

Box 14.2 Identified Risk Factors for Substance Misuse in People with ID

Intrapersonal variables

- Having a borderline to mild learning disabilities
- Being young and male
- Having a specific genetic condition
- Adolescents with conduct disorders, ADHD and antisocial personality disorders
- Compromised tolerance to drugs
- Coming from an ethnic minority group
- Coexistence of a mental health problem
- Low self-esteem
- Disempowerment
- Inadequate self-control/regulatory behaviour
- Impulsivity
- Cognitive limitations (illiteracy, short attention span, memory deficits, poor problem-solving skills, tendencies to distort abstract cognitive concepts, over-compliant dispositions)
- Poor insight into the effects and negative consequences of substance use
- Lack of knowledge of the toxic interactions with psychotropic medications compounded by the use of illicit drugs
- Frustration

Interpersonal variables

- Living in the community with low levels of supervision
- Poverty
- Parental alcohol-related neuropsychiatric disorders
- Presence of negative role models with punitive child management practices
- Family dysfunction
- Negative life events (e.g. neglect, abuse, bereavement)
- Unemployment
- Limited educational and recreational opportunities
- Excessive amounts of free time
- Deviant peer group pressure
- Limited relationships/friends
- Lack of meaning in life
- Lack of routine
- Loneliness
- Desire for social acceptance/method for 'fitting in'

(Baker, 2007; Barnhill, 2000; McGillicuddy & Blane, 1999; Rimmer et al., 1995; Sturmey et al., 2003; Taggart, Mc Laughlin, Quinn, & Mc Farlane, 2007). Furthermore, as more people with ID live in facilities, including family homes, with minimal supervision than ever before, they will therefore have greater access to readily available cash to obtain such substances. The person with an ID may then see alcohol and illicit drugs as a method of 'fitting in' and 'socialising' with one's non-disabled peers, adopting an identity that is consistent with non-ID populations. This process of 'fitting in' may compensate for a lack of social skills/supports/friendships/relationships, isolation and frustrations frequently described by people with ID for many years (Taggart et al., 2007). These are individuals who have also been found to have low self-esteem, inadequate self-control, impulsivity and poor social and communication skills further suggesting a population who may be susceptible to developing substance-related problems as they lack the adaptive skills to protect against misusing substances (Baker, 2007; Clarke & Wilson, 1999; Gress & Boss, 1996, Stavrakaki, 2002, Sturmey et al., 2003; Taggart et al., 2007).

In-depth self-report studies exploring the reasons why people with ID use and abuse substances have been viewed; most studies have used third-party proxy reports (i.e. those of staff). Taggart et al. (2007) using a series of 1-1 interviews explored the lives of 10 adults with ID in N. Ireland who were found to have a substance abuse disorder into the reasons why they abused alcohol and/or drugs and what impact this behaviour had on them. One overarching theme of the reasons for such misuse was labelled as 'self-medicating against life's negative experiences'. This was divided into two subthemes: firstly,

'psychological trauma'. This included multiple deaths of close family members, 'long-term physical, emotional and financial abuse' and also 'sexual abuse'. The second subtheme focused on the person with ID 'social distance from the community'. Many of the people with ID spoke of their lack of companionship; of having no friends, whether non-disabled or disabled; and the loneliness of living by oneself. Others recounted stories of being exploited by their 'drinking peers', people who they drank with on the streets, in the pubs and in their own homes. Overall, a disturbing trend that could be heard throughout many of the interviews was that of isolation and loneliness. Loneliness has been reported in previous studies that have explored why people with ID abuse alcohol (Baker, 2007; Christian & Poling, 1997). Degenhardt (2000) has argued that this motive of loneliness might be more important for a person with ID because of the greater social isolation they often experience due to stigma, limited access to non-disabled friends and lack of social skills.

As within the non-disabled population, substance abuse also affects the physical and mental health of people with ID, leading to behavioural and social difficulties. Some authors highlight that alcohol and illicit drugs exacerbate the affects of these substances in people with ID (Baker, 2007; Burgard et al., 2000; Taggart et al., 2008; Westermeyer et al., 1996).

Assessment

There is difficulty recognising the signs and symptoms of substance abuse disorder in people with ID. Slayter and Steenrod (2009) reported that some of the behavioural problems displayed by people with ID can mask some of the substance-related problems often observed. Assessment and treatment requires an individual multimodal approach geared towards relapse prevention, which aims to motivate the person to change their attitudes and behaviour to substance use (American Psychiatric Association, 1994). When undertaking a comprehensive assessment of a person with ID with substance-related problems, care must be undertaken to collate various information from multiple informants in order to ensure the accuracy of the data collected. The sharing and testing of this information will help formulate a working hypothesis of the possible underlying reasons for predisposing, precipitating and/or maintaining the substance abuse. On the basis of this information, a clinical diagnosis can be made and a multielement intervention package then developed.

Interviewing the Person with ID

Undertaking a clinical interview with a person with ID is the first step in the assessment process, yet this stage may be fraught with a number of challenges for the clinician when attempting to collate a history of the pattern and reasons for substance abuse and related problems. Miller and Whicher (2009) identified a number of these difficulties:

- Cognitive and communication difficulties will make substance use history taking and recall difficult.
- Problems with temporal recall may make answering questions about what the person drank and their drinking patterns further difficult.
- Numeracy problems will make calculating the number of drinks and units drank challenging.
- A lack of knowledge about alcohol may mean the person cannot give accurate answers and might not even know that certain drinks contain alcohol (i.e. alcopops).
- The person may not know that their problems are related to abusing substances and therefore will need to be educated about this before they understand help is required about their substance use.

Miller and Whicher (2009) offer clinicians advice on key questions to ask when collecting an alcohol history of a person with ID (see Box 14.3). When interviewing the person with ID, the clinician must be aware of the risk and vulnerability factors, the existing comorbidities and the person's level of ID and communication difficulties.

Box 14.3 Taking an Alcohol History from Person with ID

- Speak with the person without a carer present; reassure interview confidentiality.
- Ask what they drink and then ask how many they had at any one time (i.e. the most).
- Then ask if this is usually what they drink and if not ask how it is different.
- Ask to see the bottle or can they show you pictures of what they drink.
- Ask about drinking in the previous week.
- Then ask is this a usual week.
- Establish past events that are significant to them and that they can remember and then ask them to recall whether they were drinking then. Try to establish changes in drinking over time and periods of abstinence.
- Ask their view on why they drink.
- Ask do they think drinking is good or bad, and ask them to explain their answer.
- Ask whether family and friends around them drink. Have people around them said anything to them about their drinking?
- Ask are they the same or different when they drink. Ask them to explain.
- Ask about whether they have been recently angry, had arguments or got into trouble with police or hurt themselves. If they said yes, ask was this when they were drinking.
- Ask has their memory got better or worse when drinking.
- Ask about physical withdrawal symptoms (i.e. tremor, sweating, nausea particularly on waking) then psychological symptoms (i.e. anxiety).
- In asking about cravings, give examples from the person's history.
- Ask what they have spent their money on in the last week; is this a usual week expenditure?
- Ask them what they know about alcohol (i.e. effects, safe limits); then ask do they think their drinking is OK or too much.
- Ask if they stop drinking will their life be the same of different. If different, ask them to explain.
- Ask do they want help with their drinking.

Adapted from Miller & Whicher (2009, p. 108)

Clinical Examination

Clinical examination can sometimes be helpful in the detection of chronic alcohol abuse. Clinical screening procedures have been developed for tremor of the hands, the appearance of blood vessels in the face and changes observed in the mucous membranes (e.g. conjunctivitis) and oral cavity (e.g. glossitis) and elevated liver enzymes. Several laboratory tests are useful in the detection of alcohol misuse. Serum gamma-glutamyl transferase (GGT), carbohydrate-deficient transferrin (CDT), mean corpuscular volume (MCV) of red blood cells and serum aspartate amino transferase (AST) are likely to provide, at relatively low cost, a possible indication of recent excessive alcohol consumption. It should be noted that false positives can occur when the individual uses drugs (such as barbiturates) that induce GGT or has hand tremor because of nervousness, neurological disorder or nicotine dependence (Babor, Higgins-Biddle, Saunders, & Monteiro, 2001).

Clinical Tools

There are a number of mainstream addiction clinical screening tools that have been developed to assess nicotine, alcohol specific or other drugs. Some are designed for self-administration and others for use in clinical interviews.

Although these have not been validated for the ID population, with some minor amendments to the wording and reading these aloud to the person, they could be easily used with this population (Miller & Whicher, 2009). Three examples of these clinical tools that could be read alongside a person with ID include:

- *Fagerström test for nicotine dependence*: The Fagerström Test for Nicotine Dependence (Heatherton, Kozlowski, & Frecker, 2001) is a standard instrument for assessing the intensity of this physical addiction (see Box 14.4). The Fagerström test helps medical practitioners document the indications for prescribing medication for nicotine withdrawal. The higher the Fagerström score, the more intense is the patient's physical dependence on nicotine. Higher scores indicate that treatment of withdrawal symptoms, usually with nicotine replacement therapy, will be an important factor in the patient's plan of care.
- *AUDIT (alcohol use disorder identification test)*: The Alcohol Use Disorder Identification Test (Babor et al., 2001) describes how to use it to identify persons with hazardous and harmful patterns of alcohol consumption. The AUDIT was developed by the World Health Organization as a simple method of screening for excessive drinking and to assist in brief assessment. It can help in identifying excessive drinking as the cause of the presenting illness. It also provides a framework for intervention to help hazardous and harmful drinkers reduce or cease alcohol consumption and thereby avoid the harmful consequences of their drinking. The AUDIT may be administered either as an oral interview or as a self-report questionnaire (see Box 14.5).
- *DAST (drug abuse screening test)*: The purpose of the DAST is (1) to provide a brief, simple, practical but valid method for identifying individuals who are abusing psychoactive drugs and (2) to yield a quantitative index score of the degree of problems related to drug use and misuse (Yudko, Lozhkina, & Fouts, 2007). DAST scores are highly diagnostic with respect to a DSM diagnosis of psychoactive drug dependence. It obtains no information on the various types of drugs used or on

Box 14.4 The Fagerström Test for Nicotine Dependence

Score each of the following questions (the scores are given in brackets):

1. How soon after you wake up do you have your first cigarette?
   A. Within 5 min (3)
   B. 6–30 min (2)
   C. 31–60 min (1)
   D. After 60 min (0)
2. Do you find it difficult to refrain from smoking in places where it is forbidden, e.g. in church, the library and the cinema?
   A. Yes (1)
   B. No (0)
3. Which cigarette would you hate most to give up?
   A. The first one in the morning (1)
   B. All others (0)
4. How many cigarettes do you smoke per day?
   A. 10 or fewer (0)
   B. 11–20 (1)
   C. 21–30 (2)
   D. 31 or more (3)
5. Do you smoke more often during the first hours after waking than during the rest of the day?
   A. Yes (1)
   B. No (0)
6. Do you smoke even if you are so ill that you are in bed most of the day?
   A. Yes (1)
   B. No (0)

Now add up your score.

7–10 points = highly dependent on nicotine

4–6 points = moderately dependent on nicotine

Less than 4 points = less dependent

The higher your score, the more likely you are to have withdrawal symptoms if you give up smoking, and the withdrawal symptoms are likely to be stronger.

Taken from http://www.patient.co.uk/health/Smoking-How-Addicted-Are-You?.htm

Box 14.5 Audit: The Alcohol Use Disorders Identification Test Guidelines for Use in Primary Care

1. How often do you have a drink containing alcohol?

Never	Monthly or less	Two to four times a month	Two to three times a week	Four or more times a week

2. How many drinks containing alcohol do you have on a typical day when you are drinking?

1 or 2	3 or 4	5 or 6	7 to 9	10 or more

3. How often do you have six or more drinks on one occasion?

Never	Less than monthly	Monthly	Weekly	Daily or almost daily

4. How often during the last year have you found that you were not able to stop drinking once you had started?

Never	Less than monthly	Monthly	Weekly	Daily or almost daily

5. How often during the last year have you failed to do what was normally expected from you because of drinking?

Never	Less than monthly	Monthly	Weekly	Daily or almost daily

6. How often during the last year have you needed a first drink in the morning to get yourself going after a heavy drinking session?

Never	Less than monthly	Monthly	Weekly	Daily or almost daily

7. How often during the last year have you had a feeling of guilt or remorse after drinking?

Never	Less than monthly	Monthly	Weekly	Daily or almost daily

8. How often during the last year have you been unable to remember what happened the night before because you had been drinking?

Never	Less than monthly	Monthly	Weekly	Daily or almost daily

9. Have you or someone else been injured as a result of your drinking?

No	Yes, but not in the last year	Yes, during the last year

10. Has a relative or friend or a doctor or other health workers been concerned about your drinking or suggested you cut down?

No	Yes, but not in the last year	Yes, during the last year

Taken from http://whqlibdoc.who.int/hq/2001/who_msd_msb_01.6a.pdf

the frequency or duration of the drug use. There is a question regarding multiple drug use, and some of the types of problems caused by drug use/abuse in the following life areas are surveyed: family relationships, social relationships, employment, legal and physical (medical symptoms and conditions) (see Box 14.6). A brief examination of the individual item responses indicates the specific life problem areas.

Taggart (2011) highlights that other screening tools should be used to collect data on the person's baseline behaviours in four core areas by

Box 14.6 The Drug Abuse Screening Test (DAST)

The following questions concern information about your involvement with drugs. Drug abuse refers to (1) the use of prescribed or 'over-the-counter' drugs in excess of the directions and (2) any non-medical use of drugs. Consider the past year (12 months) and carefully read each statement. Then decide whether your answer is YES or NO and check the appropriate space.

Please be sure to answer every question.

YES NO

1. Have you used drugs other than those required for medical reasons?
   ___ ___
2. Have you abused prescription drugs?
   ___ ___
3. Do you abuse more than one drug at a time?
   ___ ___
4. Can you get through the week without using drugs (other than those required for medical reasons)?
   ___ ___
5. Are you always able to stop using drugs when you want to?
   ___ ___
6. Do you abuse drugs on a continuous basis?
   ___ ___
7. Do you try to limit your drug use to certain situations?
   ___ ___
8. Have you had 'blackouts' or 'flashbacks' as a result of drug use?
   ___ ___
9. Do you ever feel bad about your drug abuse?
   ___ ___
10. Does your spouse (or parents) ever complain about your involvement with drugs?
    ___ ___
11. Do your friends or relatives know or suspect you abuse drugs?
    ___ ___
12. Has drug abuse ever created problems between you and your spouse?
    ___ ___
13. Has any family member ever sought help for problems related to your drug use?
    ___ ___
14. Have you ever lost friends because of your use of drugs?
    ___ ___
15. Have you ever neglected your family or missed work because of your use of drugs?
    ___ ___
16. Have you ever been in trouble at work because of drug abuse?
    ___ ___
17. Have you ever lost a job because of drug abuse?
    ___ ___
18. Have you gotten into fights when under the influence of drugs?
    ___ ___
19. Have you ever been arrested because of unusual behaviour while under the influence of drugs?
    ___ ___
20. Have you ever been arrested for driving while under the influence of drugs?
    ___ ___
21. Have you engaged in illegal activities in order to obtain drug?
    ___ ___
22. Have you ever been arrested for possession of illegal drugs?
    ___ ___
23. Have you ever experienced withdrawal symptoms as a result of heavy drug intake?
    ___ ___

(continued)

Box 14.6 (continued)

24. Have you had medical problems as a result of your drug use (e.g. memory loss, hepatitis, convulsions, bleeding)?

    ___ ___

25. Have you ever gone to anyone for help for a drug problem?

    ___ ___

26. Have you ever been in a hospital for medical problems related to your drug use?

    ___ ___

27. Have you ever been involved in a treatment programme specifically related to drug use?

    ___ ___

28. Have you been treated as an outpatient for problems related to drug abuse?

    ___ ___

Scoring and interpretation: A score of '1' is given for each YES response, except for items 4, 5 and 7, for which a NO response is given a score of '1'. Based on data from a heterogeneous psychiatric patient population, cutoff scores of 6 through 11 are considered to be optimal for screening for substance use disorders. Using a cutoff score of 6 has been found to provide excellent sensitivity for identifying patients with substance use disorders as well as satisfactory specificity (i.e. identification of patients who do not have substance use disorders). Using a cutoff score of <11 somewhat reduces the sensitivity for identifying patients with substance use disorders but more accurately identifies the patients who do not have a substance use disorders. Over 12 is definitely a substance abuse problem. In a heterogeneous psychiatric patient population, most items have been shown to correlate at least moderately well with the total scale scores. The items that correlate poorly with the total scale scores appear to be items 4,7,16, 20 and 22.

Taken from http://www.drtepp.com/pdf/substance_abuse.pdf

other informants; this can further help identify the interplay between these risk factors and any underlying medical or mental health problems that will need to be addressed:

- Physical health (i.e. OK Health Checklist)
- Functioning ability (i.e. Adaptive Behaviour Scale)
- Challenging behaviours (i.e. Aberrant Behaviour Checklist)
- Mental health (i.e. PAS-ADD Checklist or Interview)

Treatment Strategies

People with ID who misuse substances sometimes can be 'unwilling' or 'uncooperative' to fully engage in a range of assessment procedures and treatments further complicating the delivery, maintenance and success of such treatment packages (Mc Laughlin, Taggart, Quinn, & Mc Farlane, 2007). Huxley et al. (2007) highlighted that this 'uncooperativeness' should not be interpreted as being of poor motivation, as sometimes it is the case that a person's lack of cooperation is due to their lack of understanding of their care plan or treatment. A number of explanations for these difficulties in engagement centres upon this population's ID and associated cognitive deficits (i.e. slower learning, communication issues, illiteracy, short attention span, memory deficits, low self-esteem, inadequate self-control/regulatory behaviour) (Degenhardt, 2000; McGillicuddy, 2006; McGillicuddy & Blane, 1999; McGillivray & Moore, 2001; Sturmey et al., 2003). These individuals have also been found to have lower levels of knowledge regarding the effects of abusing excessive amounts of alcohol and drugs (McGillivray & Moore, 2001). Many researchers and clinicians argue that any successful treatment package must be adapted to reflect the learning style of people with ID.

Therefore, given the complex aetiology of the acumination of risk factors and the multivariate context, then there is no one treatment intervention that can be used exclusively to target the person with a substance abuse disorder. Likewise, previous theories that believed abused substances to have a

homogenous pathology and similar personality traits have now been dispelled, and more recent research illustrates a population to have a broad diversity in such individuals (Miller & Brown, 1997). Therefore, treatment programmes or packages must be clearly underpinned by a holistic framework that targets the person, family and community. One suggestion is to adapt a bio-psychosocial model that has been utilised by in a report to the WHO on mental health in people with ID. This is in comparison to the medical/biological model that has frequently been employed across medicine, psychiatry and nursing that has traditionally viewed substance abuse as organic and based within the person. This alternative model taken from a health psychology perspective provides a holistic proactive approach to understanding how substance abuse develops but also how such problems can be prevented, if possible, in this population.

Any group work/therapy can also become a stressor and increase anxiety and frustration for a person with an ID, and the person with ID may struggle to keep up with the non-ID patients. Therefore, any treatment programme and written materials must be tailored to the individual's:

- Developmental level
- Cognitive strengths
- Communication ability
- Level of comprehension
- Literacy skills
- Concentration span
- Substances being used/abused
- Patterns of substance use/abuse
- Access to substances
- Attitudes to substances
- Triggers to abusing substances
- Underlying problems
- Behaviours associated when using and abusing substances

Smoking Cessation Programmes

Despite the growing prevalence rates of smoking among people with ID (Emerson & Turnbull, 2005; Rimmer et al., 1995; Robertson et al., 2000), few studies have been undertaken to reduce or eliminate smoking within this population. Of those that have been completed, they are methodologically weak with small sample sizes, no control groups and no long-term follow-ups, therefore making generalisations to other settings difficult.

Tracy and Hosken (1997) undertook a small smoking education programme for 11 adults with ID in a community setting in Australia; they found that on completion of the programme, half of the participants quit smoking. Chester, Fatima, and Alexander (2011) undertook a smoking cessation programme with an emphasis on health education and nicotine replacement within a forensic hospital for 48 adults with ID in England. The authors reported that one third of these smokers stopped smoking and over half reduced the number of cigarettes smoked. Singh et al. (2011) conducted a mindfulness-based smoking cessation programme for a man with ID who had smoked for 17 years. The programme focused upon three areas: (1) intention, (2) mindful observation of thoughts and (3) mediation. Three months after completion of the programme, the man stopped smoking and at a 12-month and 3-year follow-up points the man was still abstaining from smoking.

Alcohol Treatment Programmes

There is some debate as to the goal of treatment for those who abuse alcohol, total abstinence or controlled drinking. Degenhardt (2000) has argued for total abstinence as a more appropriate treatment goal for people with ID who abuse alcohol. Total abstinence is clear and had only 'one rule', no drinking, whereas controlled drinking has numerous rules about what and when to drink and this can be a difficult concept to understand for a person with ID.

In conjunction with the individual with ID, it is important to establish goals that they would like to achieve. This process will help gain an understanding of the person's knowledge and also reasons/motivation for drinking. This information will identify key priority areas to be identified by the person with ID (i.e. trauma, family issues, loneliness, offending behaviours) and

allow structure to be built into the treatment programme thereby promoting a person-centred approach to the therapeutic process (Taggart et al., 2008). These person-centred targets can act as evaluation and progress markers and further motivate the person with ID to monitor their own improvement.

There are a number of robust alcohol treatment programmes that have been developed for the non-ID population; however some of these interventions have been adapted for people with ID although no systematic evaluations of these have been conducted: (Some of the treatments below will be used in the treatment of other substances as well as alcohol. This is indicated where applicable.)

- *Detoxification:* The first step in any treatment plan for those individuals dependent upon alcohol is to detoxify both physically and psychologically from the alcohol. For many people with ID, they will need support from trained health personnel, normally within an inpatient facility particularly where there are other comorbid conditions (i.e. physical and mental health problems).
- *Psychopharmacology:* There are various medications used depending on the substance used. Some work by reducing craving, e.g. bupropion for smoking and naltrexone for alcohol, while others such as disulfiram (Antabuse) work on the metabolism to produce unpleasant and potentially dangerous side effects, e.g. nausea and dizziness, if the person consumes alcohol during treatment. Other approaches are to use a 'less harmful' substitute medication to wean individuals off, e.g. methadone for heroin, or to take the substance causing addiction in a safer form such as nicotine replacement as a smoking cessation aid.
- *Education programmes:* Brown and Coldwell (2006) developed an alcohol awareness programme for adults with ID within a medium-secure hospital in England; this programme was adapted from a mainstream alcohol programme. The 8-week programme, 1 h per week, aimed to increase the knowledge of the participants regarding the positive and negative effects of using alcohol, and therefore using alcohol in a safe and controlled pattern. A variety of methods were employed and these included videos, quizzes, work files, assignments, posters, group work, discussions, observations, role plays, drink tests and practical visits to the pub. Hospital staff were trained within this programme and then they had to deliver the programme to the participants. This paper described the development and delivery of the programme.
- *Modifications of AA and 12-step programme:* The 23 steps, originated by Alcoholics Anonymous, is the spiritual foundation for personal recovery from the effects of alcoholism, not only for the alcoholic but also for their friends and family in Al-Anon Family Groups. Many members of the 12-step recovery programmes have found that these steps were not merely a way to stop drinking, but they became a guide towards a new way of life. This programme may suit people with a borderline/mild ID but they will need support to work through this programme (Miller & Whicher, 2009).
- *Skills training:* Building upon existing, and developing new, skills to manage everyday life including pressures and stresses can prevent relapse and a return to abusing alcohol. These skills include:
  - Social skills training focusing on expressing emotions, eye contact, responding to criticism, giving and receiving complements and developing coping and refusal skills and self-monitoring skills promote interpersonal communication, appropriately responding to criticism and engaging in realistic role plays (Mattick & Jarvis, 1993; Paxon, 1995)
  - Communication skills training
  - Refusal skills training (i.e. saying 'no', suggesting an alternative) (Witkiewitz et al., 2012)
  - Structured problem solving
- *Behavioural and cognitive approaches:* This can be used for alcohol and other substances and involves assertiveness skills, distinguishing between positive and negative role models within substance abuse situations.

Cognitive behavioural therapy (CBT) is one such treatment that works by challenging dysfunctional cognitions and processes that maintain alcohol and other substance use. The treatment works by working on the antecedents to and consequences of the behaviour and increases coping skills during and between sessions through homework. Sessions focus on problem-solving and coping skills, e.g. managing thoughts and cravings, planning for emergencies such as drink/drug refusal. Associated issues are also addressed such as anger management, negative cognitions and being able to self-disclose and express negative feelings (in an appropriate way) to prevent escalation. They may also be taught listening skills. Homework involves planning how to handle one such situation, and then actually trying out the skills in it, with positive behaviours that are likely to prevent relapse promoted (Kadden, 2002). There is little evidence of outcomes for CBT in this population to treat substance use. In a non-ID population of people with schizophrenia, an integrated treatment programme of CBT, motivational interviewing and family therapy lasting 9 months reported a significant improvement in general functioning than routine care for a non-ID maintained at the end and 3 months posttreatment for those followed up ($n=30$). The programme also reported a reduction in positive symptoms and in symptom exacerbations and an increase in the percent of days of abstinence from drugs or alcohol over the 12-month period from bias.

- *Motivational interviewing:* Is used in both alcohol and other substances, using an empathetic approach designed to motivate the person to change by taking their perspective as to the pros and cons of their substance use by exploring their goals and associated ambivalence towards it (American Psychiatric Association, 2006). Mendel and Hipkins (2002) used motivational interviewing techniques in assisting 7 people with ID and alcohol-related problems within a forensic hospital in England through the stages of change (Prochaska & Diclemente 1986). Hospital staff were trained in this group and they ran a series of 1-h workshops. A variety of measures (pre- and post group) demonstrated increases in clients' motivation, self-efficacy and determination to change their drinking behaviour.
- *Relapse prevention programme:* This type of programme focuses on self-regulation of thinking and feeling, accepting past relapses, identifying the causes of relapse and learning to prevent and interrupt relapses.

Drug Treatment Programmes

Within all schools today, young people receive information and education on the dangers of illicit drugs. While debate surrounds the effectiveness of this type of education as many teenagers experiment with illicit drugs, some authors have also questioned the efficacy of this type of education for young people with ID. Snow et al. (2001) in a review of drug education for young people with ID question how such cognitively based alcohol and drug education programmes be equitable for this population. Such programmes do not utilise material in a format that is accessible to young people with cognitive deficits and communication difficulties, as well as relying on group education. Snow and colleagues highlighted a number of factors that needed to be considered with respect of delivery of such material:

- A greater degree of flexibility is required given each person's cognitive and communication challenges.
- Timing and placement of the alcohol and drug education has to be considered within the person's developmental stage.
- The mainstream delivery style of current alcohol and drug programmes is based upon cognitive instructions and interactions (i.e. information transfer, role plays, debate, discourse about one's own values and experiences) in a group setting; such modalities may not be suitable learning environments for people with ID.

- People with ID may have greater difficulty in generalising knowledge and skills acquired in one setting to another setting.
- The issue of prescribed medications (i.e. anti-convulsants, analgesics) may need particular attention in schools (e.g. side effects, intra-contradictions with alcohol, correct dosages, signs of toxicity).
- Students with ID may require more simplification, repletion and opportunities for mastery than their non-ID counterparts in mainstream settings.

Prevention of Substance Abuse

There has been some discussion about the benefits of the current public health prevention and education programmes targeting the cessation of smoking, drinking in moderation and deterrence of illicit drug use within schools and in local communities across some countries for teenagers and adults with ID. However, the impacts of such national campaigns have not been examined. Many people with ID will struggle to understand the importance and implications of such written campaigns and programmes, as a result of their cognitive and communication limitations (Burtner, Wakham, McNeal, & Garvey, 1995). Snow et al. (2001) found that children and teenagers with ID were often excluded from school smoking, alcohol and drug education programmes and other preventative measures. McCrystal et al. (2007) argue that as a result of a lack of a strong evidence base in substance abuse in young people and adults with ID, this has had series implications for policy initiatives such as targeted smoking, alcohol and drug prevention programmes in schools and at the community level. As many countries are supporting people with ID to engage in their local communities and mainstream activities and services today, this is also an opportunity to educate and inform people with ID about the consequences of substance use and abuse. However, such opportunities are not being availed off and people with ID are not being educated about substance use and abuse (McGillicuddy, 2006).

Service Provision

Evidence shows there is little if any coordinated joint service provision for people with ID who have substance-related problems/substance disorder. Confusion exists between and within the various ID and addiction agencies on the strategies currently in place to assess and treat people with ID. Referral routes are rarely in place. Addiction services appear to impact negatively on adults with ID, further diminishing their self-esteem and confidence and helping to create the familiar revolving door between treatment services or the criminal justice system. Many staff within the addiction services, like many other mainstream health organisations, reported that the limited cognitive abilities of people with ID (i.e. communication skills, reading ability, information retention, social skills) were barriers to engaging in therapeutic individual and group work (Lottman, 1993; Mc Laughlin et al., 2007).

There is growing research to illustrate that staff working within addiction services do not have the appropriate education, training and confidence to work with adults with ID who present with substance-related problems and this also resulted in such personnel not engaging in therapeutic interventions (Aman, Geeta, & Burrow, 1995; Lottman, 1993; McLaughlin et al., 2007; Vandernagel et al., 2011). Mc Laughlin et al. (2007) in N. Ireland undertook a series of thirteen 1-1 semi-structured interviews with a range of health professionals who worked in ID and addiction services to discover their experiences of caring for people with ID who abused substances. Although small numbers of people with ID presented to both types of services, many more people with ID used ID services for support, rather than their local addiction services. While the numbers may be relatively small, the challenges this client group posed were very distressing in relation to their physical, emotional and social health costs. The addiction professionals reported a lack of education in working with this doubly disadvantaged population. Moreover, policies and guidelines were absent to guide staff to work collaboratively with their colleagues in ID services.

Vandernagel, Kiewik, Buitelaar, and DeJong (2011) from 86 case reports from staff in different ID services in Holland (IQ 50–85) reported that substance users had various psychosocial problems and that staff were poorly equipped to meet the users' needs, citing the low take-up of and poor access of mainstream treatment programmes.

Implications for Practice

There are a number of areas that need to be targeted to reduce the levels of substance use and address its harmful consequences. Although there has been an increase in local population-based research, there is a weak evidence base still in its infancy on what treatments are accessed by people with ID or reports as to their effectiveness. Many of those with ID that access mainstream drug services may not be known to ID services. To reduce levels of substance abuse, there is a greater role for mental health promotion targeted specifically to people with ID, along with the need for early intervention needs to be addressed, by addressing individual risk factors (Hawkins, Catalano, Miller, 1992), from school age onwards (see http://www.nottinghamdrugaware.co.uk/). There is a need for clinical pathways for people with ID and substance use to provide interfaces between ID, mental health and substance use services to achieve joined-up care, through consultation and cross-agency collaboration (Vandernagel et al., 2011). With improvements in the development of and access to services should come guidance, specifically on assessment and treatment issues specific to people with ID, that reflect the level and complexity of dependence and individual comorbidities and consider a range of issues that may be present including maladaptive coping skills, cognitive impairment, adverse reaction and reduced tolerance to substances.

Implications for Future Research

The robust-structured smoking, alcohol and drug interventions developed to improve the biomedical, psychosocial and self-management strategies for non-disabled people with substance abuse disorders have not been applied to people with ID. Taggart et al. (2007, 2008) reported that there was a lack of appropriate information on smoking, alcohol and drug use, and little emotional support was being offered to this population to address their underlying mental health issues. People with ID rarely accessed structured education programmes for smoking, alcohol and drug use that were routinely offered to the non-ID, further illustrating ongoing inequality and disadvantage. These programmes have neither recognised nor addressed the specific challenges posed by this population's cognitive deficits, communication difficulties and low levels of literacy skills and learning styles (McGillicuddy, 2006; Slayter, 2008; Snow et al., 2001; Taggart et al., 2008).

There is also a lack of attention for carer support and also negative attitudes held among many healthcare professionals in engaging with people with ID (Greenhalgh, 2009). There have been a small number of smoking, alcohol and drug educational initiatives targeted at this population. However, none of these resources robustly developed and have been formally evaluated for people with ID (Slayter, 2008; Snow et al., 2001; Taggart et al., 2008). There are a number of conceptual models to show that people with ID can successfully develop new knowledge, skills and therefore change their lifestyle behaviours (Wilson & Goodman, 2011). Taggart et al. (2008) highlighted the importance of addressing the person's intrinsic motivation, to want to change their behaviour and to engage in self-help, possibly by using the techniques of motivational interviewing rather than enforcing motivation from external sources. Other successful techniques include offering both group and one-to-one sessions with more time flexibility, based on repetition, greater use of kinesics learning and role-play scenarios, and also utilising family and paid carers to support the person with ID to maintain the behaviour changes over time. Wilson and Goodman (2011) in England found that adults with mild/moderate ID and comorbid physical health conditions (i.e. diabetes, arthritis, hypertension) could successfully participate in chronic disease self-management programmes

if such programmes are modified; the programme also improved this population's access to healthcare. Likewise robust smoking, alcohol and illicit drug programmes could be modified for people with ID and also other vulnerable groups with literacy problems.

Conclusion

Substance use in ID is a relatively new area of study, which has come about as people have taken their place in society as equal citizens. With independence of people with ID come exposure, peer pressure and choices. Risk factors mirror those in the main society. In terms of assessment and treatment, there is little evidence of specific services that target this group; although the Netherlands has treatment regimes, many of the service users would be considered not to have a learning disability in other countries given the high IQ cutoff. In terms of assessment and treatment, there are a number of established strategies used. However there is little evidence of the effectiveness or use of different approaches in people with ID. Whatever approach is used, care should be made to make the process inclusive and accessible as a partnership. The challenge is to move towards prevention through effective education programmes and strategies, starting at school. To do this the issue needs to be highlighted as we educate people to make positive choices concerning their lifestyle.

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Part IV

Common Clinical Conditions

Attention-Deficit Hyperactivity Disorder (ADHD)

15

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Background

Attention-deficit hyperactivity disorder (ADHD) is a neurodevelopmental syndrome, associated with developmentally inappropriate and impairing levels of inattentive, impulsive and hyperactive behaviour, that are often accompanied by neurocognitive deficits. The relationship between ADHD and intellectual disability (ID) has been recognised for a long time. ADHD is currently acknowledged as a common comorbidity for individuals with ID (Buckley et al., 2006). Studies have shown a higher prevalence of ADHD in children and adults with ID than in the general population (Lambert, Hartsough, Sassone, & Sandoval, 1987; Peterson, Pine, Cohen, & Brook, 2001). More specifically, the coexistence of hyperactivity and ID has long been noted, but the validity of the diagnosis of ADHD in people with ID has only recently attracted increased attention (Antshel, Phillips, Gordon, Barkley, & Faraone, 2006).

Historically the common roots of ADHD and ID can be traced in some classical texts. AF Tredgold (1908) described an incipient brain damage that causes latent behavioural and learning difficulties. The symptoms were thought to be improved by both medication and environmental manipulation. Goldstein (1936) studied brain injured adults in World War 1 and described a behavioural neurological syndrome similar to ADHD. Ebaugh (1923) reported that brain insults other than mechanical trauma, such as encephalitis, may cause hyperkinesis, fatigability, explosive behaviour and inattention. In the 1940s the term minimal brain damage was used, and in the 1960s it was replaced with minimal brain dysfunction.

Clinical Presentation

The clinical presentation of ADHD in people with ID has not been widely described (Sevin, Bowers-Stephens, & Crafton, 2003). The phenomenology of ADHD and ID has been studied much more in children and adolescents than adults although there are several accounts of the typical presentation of ADHD in adults (Asherson, 2005; Kooij et al., 2010; Weiss, Murray, & Weiss, 2002). The presentation of mental disorders in people with ID is often atypical so that establishing psychiatric diagnoses in this population can be more difficult. With regard to ADHD, however, there is evidence that children with ID and ADHD exhibit some of the classical symptoms of the disorder such as fidgeting, off-task behaviours and poor selective attention, compared with their ID peers without ADHD (Handen, McAuliffe, Janosky, Feldman, & Breaux, 1998; Pearson, Yaffee, Loveland, & Lewis, 1996).

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There is some research evidence in children to suggest that the clinical presentation of ADHD is similar across individuals with different levels of intellectual ability. For example, Fee et al. found that children with ID and ADHD show a pattern of ADHD symptoms similar to that of average intelligence children diagnosed with ADHD (Fee, Matson, & Benavidez, 1994). Similarly, Ishii et al. found no statistically significant difference in the presentation of DSM-IV symptoms of ADHD in children with ID compared with children of average intelligence (Ishii, Takahashi, Kawamura, & Ohta, 2003). Simonoff et al. found a negative linear relationship between ADHD symptoms and IQ in adolescents with mild ID (Simonoff, Pickles, Wood, Gringras, & Chadwick, 2007).

In a longitudinal study of ADHD among 5–8-year-old children with and without ID, Niece et al. found that ADHD was over 3 times as prevalent in the ID group as in children with typical development (Neece, Baker, Blacher, & Crnic, 2011). They also found that although the total number and relative frequency of ADHD symptoms endorsed was similar in the two groups, across time children in the ID group had higher levels of both inattentive and hyperactive-impulsive symptoms. They also observed a tendency for the diagnosis of ADHD to emerge earlier and to be more stable in the ID group (Neece et al., 2011). In another study, Baker et al. found that among children who met criteria for ADHD at age 5, groups with and without ID did not differ significantly in the number of inattentive, hyperactive-impulsive or total ADHD symptoms endorsed (Baker, Neece, Fenning, Crnic, & Blacher, 2010).

Both ADHD and ID are independently associated with increased lifelong rates of psychiatric morbidity. For example, adults with ID have increased rates of affective disorder (Richards et al., 2001), while adults with ADHD have increased rates of affective disorders, anxiety disorders and substance use disorders (Kessler et al., 2006). Thus, co-occurrence of ID and ADHD increases the risk of other psychiatric morbidity in general and behavioural disorders in particular. When compared to peers with ADHD and average intellectual ability, children with ADHD and ID are more likely to have comorbid separation anxiety disorder and behavioural disturbance in adolescence and have more restrictive educational placements (Aman, Armstrong, Buican, & Sillick, 2002; Aman, Kern, McGhee, & Arnold, 1993; Lambert et al., 1987). A follow-up study of 51 people with ADHD and moderate to borderline ID found that many continued to have behavioural problems and take prescribed medication at 1–5 years follow-up (Handen, Janosky, & McAuliffe, 1997). Bullying and violent behaviour have been found to be associated with hyperactivity in adolescents with ID (Reiter, Bryen, & Shachar, 2007). A study of medication for disruptive behaviour in children with ID reported high rates of comorbid ADHD (Aman et al., 2002).

A recent British study indicated that prevalence, sex distribution and presentation of comorbid psychiatric disorders may differ in patients with ID from those of the general population (Tonge & Einfeld, 2000). However, in this study, ADHD diagnosis was not considered for any patient although a significant proportion of patients did present with behavioural problems. People with ID may have a poorer prognosis when they present with mental health problems in general (Tonge & Einfeld, 2000). In the case of ADHD, people with ID may present decreased ability to develop coping strategies partly because of their cognitive deficits. A recent retrospective study of adults with ADHD found increased overall severity of ADHD in those with ID (Xenitidis, Paliokosta, Rose, Maltezos, & Bramham, 2010). This is consistent with follow-up medication studies that concluded that ADHD persistence into adulthood is related with IQ in people with ID (Lambert et al., 1987; Peterson et al., 2001).

Epidemiology

Reported prevalence of ADHD in children and adults with LD varies widely. This variability may be accounted for by the range of definitions and diagnostic criteria used for LD, in addition to the factors contributing to the variability of ADHD prevalence rates in children and adults in general.

In the non-ID population, the average worldwide prevalence for ADHD is around 5 % in children (Polanczyk, de Lima, Horta, Biederman, & Rohde, 2007), although in the UK a slightly lower estimate of 3.6 % was reported in a national child and adolescent mental health survey (Ford, Goodman, & Meltzer, 2003). In adults the prevalence is estimated to be around two thirds of the childhood rate (Faraone, Biederman, & Mick, 2006) and in the range of 2–4 % (Fayyad et al., 2007; Simon, Czobor, Bálint, Mészáros, & Bitter, 2009). Estimated rates of ADHD in children and adults with ID vary widely, ranging from 4 to 42 %, depending on the target population, sampling methodology, the severity of ID and the context in which each study was conducted. Emerson performed a secondary analysis of the 1999 Office for National Statistics survey of the Mental Health of Children and Adolescents in Great Britain and estimated the prevalence of ADHD as eightfold increased in children with ID compared to children without ID (Emerson, 2003). Lindbland et al. screened for neurodevelopmental and neuropsychiatric disorders and found that about 50 % of children with mild ID had clinically significant levels of ADHD[51]. One US study suggested that at least 15 % of individuals with severe and profound levels of ID may meet diagnostic criteria for ADHD, even when mental age has been taken into account (Fox & Wade, 1998). Estimates of prevalence are higher from studies which only measure target symptoms, such as hyperactivity, poor concentration and impulsivity (Dekker & Koot, 2003; Fox & Wade, 1998; Hardan & Sahl, 1997). In adults, prevalence rates of 15 % are commonly quoted, and an *ADHD positive* rate of 16.9 % was reported in a small sample of adults with ID over a wide range of severity (La Malfa, Lassi, Bertelli, Pallanti, & Albertini, 2008).

Diagnosis of ADHD in ID

The validity of the ADHD diagnosis in ID population is still however a subject of debate. A frequent question when investigating ADHD in people with ID is whether ADHD is the same disorder in an ID versus non-ID population. We know, for example, that genetic studies find a 6-times higher rate of copy number variants (submicroscopic chromosomal deletions and duplications) in those with ADHD compared to controls, which contrast with only a twofold higher rate within ADHD subjects within the normal IQ range (Williams et al., 2010). Furthermore, ADHD can occur as a comorbid syndrome in a range of known genetic neurodevelopmental disorders. It is possible, however, that higher scores for ADHD symptoms in children and adults with ADHD and ID occur simply as a by-product of their globally lower cognitive functioning (Gjaerum & Bjornerem, 2003; Reiss & Valenti-Hein, 1994; Tonge et al., 1996). This is not unexpected given that ID is often accompanied by deficits in attention and executive functions and that these abilities correlate with intelligence. Lower IQ in childhood is associated with a reduced capacity for impulse control and an increase of psychiatric abnormalities especially ADHD (Antshel et al., 2006; Simonoff et al., 2007). A negative correlation between IQ and ADHD symptoms severity was found in adults across the intellectual ability range (Xenitidis et al., 2010). Similarly, in a population twin study of 5-year-old children, a moderate negative association of IQ with ADHD was observed (−0.3). Genetic model fitting showed that 86 % of the covariation between ADHD symptoms and IQ, and 100 % of the covariation between ADHD diagnosis and IQ, was the result of shared genetic influences (Kuntsi et al., 2004).

It has been suggested that it is not possible to diagnose ADHD with certainty in individuals with an IQ below 85 (Buchmann, Gierow, Reis, & Haessler, 2011). The lower the IQ, the higher the probability that it is impulsivity rather than attention deficit that contributes to the clinical picture (Buchmann et al., 2011). On the other hand, in some cases overactivity, inattention and associated deficits of executive function may impair general performance and contribute to lower IQ scores (Ek, Westerlund, Holmberg, & Fernell, 2011). There is, however, some evidence that specific cognitive deficits exhibited by children and adults with ADHD and ID are

different from those of those with ID but without ADHD (Handen et al., 1998; Pearson et al., 2000). Studies have shown individuals with ID and comorbid ADHD to exhibit more severe neuropsychological deficits in comparison with those who have ADHD without ID, particularly with regard to executive function deficits (Lazar & Frank, 1998; Seidman et al., 2006; Seidman, Biederman, Monuteaux, Doyle, & Faraone, 2001). Other studies that have focused more directly on aspects of attention, including selective and sustained attention, suggest that the combination of ADHD and ID have a greater detrimental effect on these domains than either condition alone (Pearson et al., 1996; Robins, 1992; Tarnowski, Prinz, & Nay, 1986). Rose et al. showed that general intellectual functioning has a substantial effect on attentional and response inhibition performance. Deficits in these domains are greater in adults with ADHD and ID compared to ADHD without ID. However, these deficits remained even after the contribution of intelligence was controlled for in the analysis (Rose, Bramham, Young, Paliokostas, & Xenitidis, 2009). Other studies also showing a similar symptom picture and developmental course of ADHD symptoms for young children and adults with and without ID (Baker et al., 2010; Neece et al., 2011; Simonoff et al., 2007; Xenitidis et al., 2010) offer further support for the validity of the ADHD diagnosis regardless of intellectual functioning level. This conclusion has important implications for children with ID and ADHD who may benefit from treatments for ADHD.

Prognosis of ADHD in ID

ADHD symptoms tend to improve over time owing to both brain and psychosocial maturation. While this is the case for most children with ADHD, with a degree of *catching up* with their peers in some cases, in many others the improvements parallel the changes in the normal population without ADHD so that significant case–control differences in symptoms and impairments remain (Biederman et al., 2012; Taylor, Chadwick, Heptinstall, & Danckaerts, 1996). It is less clear whether the same degree of symptom change is seen in people with comorbid ID and ADHD. For example, it is not known whether individuals with ADHD and ID, compared to those with ADHD alone, have a qualitatively different disorder or more severe form of the same disorder. It seems likely that individuals with ID have a reduced ability to develop adaptive skills to manage ADHD symptoms, leading to greater persistence of impairments. Overall, the course of ADHD in adults with ID is an area with little empirical data available but an area of emerging research.

In a retrospective study of adults with ADHD, based on informant rated DSM-IV rating scale scores for adulthood and childhood symptoms, Xenitidis et al. found that the mean total ADHD symptom scores for adulthood were significantly higher in the ID compared to the non-ID group, indicating greater overall severity of the condition in those with ID (Xenitidis et al., 2010). This difference was even more pronounced for childhood total score. Thus, it was concluded that not only do people with ID have more severe levels of ADHD symptoms but also these symptoms show a lower tendency to improve over time compared with non-ID peers.

Treatment Response

There is some evidence that ADHD can be successfully treated in people, mainly children, with ID (Handen, Feldman, Lurier, & Murray, 1999; Pearson et al., 2003). Studies of the treatment of adults with ADHD and ID are very scarce. However studies in adults in the average IQ range show a similar range of effect size to those seen in children (Kooij et al., 2010). The main treatment approaches are pharmacological with stimulants (methylphenidate, dexamfetamine) and non-stimulants (e.g. atomoxetine, bupropion). Similar to ADHD in the general population, methylphenidate is considered to be an effective treatment for both cognitive and behavioural symptoms in ADHD in people with ID. Randomised controlled trials (RCTs) conducted mostly in children suggest a favourable response with moderate adverse effects. However, the rate

of response in people with ID may be reduced compared to non-ID population, while the rate of adverse effects may be increased. One retrospective study of patients treated with stimulants in a clinic specialised in developmental disabilities has reported good results (Jou, Handen, & Hardan, 2004).

Amfetamine, prescribed either as the D-isomer (dexamfetamine) or as a mixture of L- and D-ISOMERS (mixed amfetamine salts), has been shown to be as effective as methylphenidate for the treatment of ADHD. Although there are some case studies reporting its efficacy in people with ID and comorbid ADHD, in a systematic review of RCTs, only one study met inclusion criteria and this concluded that amfetamine had adverse effects but no significant beneficial effect on symptoms of ADHD compared to placebo (Hagerman, Murphy, & Wittenberger, 1988; Thomson, Maltezos, Paliokosta, & Xenitidis, 2009a).

Atomoxetine is a noradrenaline reuptake inhibitor which is generally recommended as a second-line treatment for ADHD. There is no literature about RCTs of atomoxetine conducted in children or adults with ID and ADHD. There is therefore a lack of data for the use of atomoxetine as a treatment option in this population. Potential benefits of atomoxetine include simple dosage regime suitable for patients with poor compliance, stable 24-h effects on ADHD symptoms, no drug abuse potential and reduced exacerbation of symptoms such as anxiety and sleep problems.

It is estimated that just under 1/3 of people with ID are prescribed antipsychotic medications for psychiatric disorders or challenging behaviours such as aggression and self-injury (Valdovinos et al., 2002). A systematic review, however, found little evidence to support their use in the context of challenging behaviour (Brylewski & Duggan, 2004). A prescribing survey in the USA reported far greater rates of antipsychotic prescribing compared to stimulants in children with intellectual and developmental disabilities (Lott et al., 2004). In a 4-week, single-blind, parallel-group trial, in children with moderate mental retardation and ADHD, risperidone was associated with greater reductions in ADHD total score. There was a significant weight reduction in the methylphenidate group and a weight gain in the risperidone group (Ghuman et al., 2009). However, in another Cochrane Review there was no RCT-based evidence that risperidone is effective for the treatment of ADHD in people with ID (Thomson, Maltezos, Paliokosta, & Xenitidis, 2009b).

Psychotropic medication prescribing in this population can only be based on open-label studies or extrapolation from research in people with other developmental disorders such as autism or disruptive behaviour disorders. However, these studies have not investigated specifically people with ID. Thus, caution is recommended regarding the applicability of these findings. There is a paucity of RCTs investigating the efficacy of pharmacotherapy in the treatment of ADHD in people with ID. The fact that available clinical evidence is based mostly on small open-label trials or retrospective studies is a severe obstacle in developing guidelines for the pharmacological management of ADHD in this population. Good quality, independent research is required to determine the efficacy and safety of pharmacological treatments of ADHD in people with ID. The comorbidities present in people with ID represent a further diagnostic challenge and may influence both the types of treatment offered and its efficacy.

In addition to pharmacotherapy, psychological interventions, mainly along the lines of behavioural therapy, cognitive behavioural therapy and psycho-education, are widely available for children and adults with ADHD. However, there is a scarcity of reports about evidence-based psychological treatments for ADHD in the population of people with ID (Knouse, Cooper-Vince, Sprich, & Safren, 2008; Safren, 2006).

Conclusions and Implications for Service and Research Development

The diagnosis of ADHD in the general population, especially adults, is often met with scepticism. The applicability of diagnostic criteria in people with ID has been questioned. However,

current research evidence indicates that ADHD symptoms increase with lower IQ. The validity of diagnosis of ADHD is supported by evidence.

The current findings indicate that children and adults with ID are at increased risk for the development of ADHD and that they are more likely to have an earlier onset, more persistent course and be associated with higher levels of impairment.

Despite this, it appears that ADHD is diagnosed less frequently in this group. The phenomenon of 'diagnostic overshadowing', the attribution of symptoms of a superimposed psychiatric condition to underlying ID, may explain this in part. Lack of awareness of clinicians and a tendency to avoid stigmatisation may be alternative explanations.

Individuals with ADHD and ID face a 'double vulnerability' because their impairment arises from both the deficits imposed by ADHD and those associated with ID (Pearson et al., 1996). People with ID have increased mental and physical health needs and requirement for social support. Thus, identification of the contribution of ADHD symptoms to the overall impairment is likely to be important as ADHD symptoms can be managed both with pharmacological and psychosocial interventions, which in turn may lead to functional improvements and increased quality of life. Specifically with regard to behavioural impairment, it is likely that when ADHD coexists with ID its symptoms, especially hyperactive/impulsive ones, contribute in varying degrees to the genesis and maintenance of challenging behaviour.

The increasing identification and treatment availability of ADHD has important implication in service development at both local/regional and national level. At present time there is a small number of outpatient services and an even smaller number of inpatient units offering a specialist assessment and treatment service. As public and clinicians' awareness increases and research evidence accumulates, it is likely that specialist services will be increasingly supplemented and even replaced by routine services at a more local level, in both the public and the independent sector. In any case the accurate assessment and effective treatment of ADHD in this group of people will require an active ongoing collaboration between clinicians, managers and (mental health and learning disabilities) commissioners.

With regard to research, the majority of the evidence for assessment and therapeutic interventions for ADHD derives currently from extrapolation from studies with children (and more recently adults) without ID. Further outcome research is required for a range of therapeutic interventions, including pharmacotherapy and behavioural and other nondrug treatments specifically in people with ID across the lifespan.

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Autism Spectrum Disorder

16

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Introduction

Autism is a neurodevelopmental condition defined clinically by impairments in social interaction and communication, accompanied by restricted, repetitive and stereotyped patterns of behaviour, interests and activities (American Psychiatric Association, 2000). It is a syndrome diagnosis underscored by different aetiologies, the latter contributing to multifaceted clinical presentations throughout the life span. The proposed fifth edition of the American Psychiatric Association's diagnostic criteria (DSM-5; scheduled for 2013) combines the social and communication criteria into a single criterion along with restricted behaviour (American Psychiatric & DSM-5 Development, 2011). The current category of pervasive developmental disorders (PDD) that includes autism, Asperger syndrome, atypical autism and PDD-NOS is to be replaced by a single category: autism spectrum disorder (ASD).

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Clinical manifestations of ASD (hereafter autism) overlap with those of other conditions such as attention deficit hyperactivity disorder (ADHD), intellectual disabilities (ID) and fragile X syndrome (the most common identifiable inherited cause of ID and autism; Turk, 2011). Autism is associated with a high prevalence of coexisting medical conditions such as gastrointestinal problems (Buie, Campbell, et al., 2010; Buie, Fuchs, et al., 2010; Ibrahim, Voigt, Katusic, Weaver, & Barbaresi, 2009), sleep disturbances (Malow & McGrew, 2008) and epilepsy (Turk et al., 2009). Recent evidence has raised the possibility that autism and epilepsy may even represent a different syndrome from autism alone (Turk et al., 2009). These medical comorbidities contribute further to different developmental challenges and adult outcomes. Autism is more common in males but this may in part be related to females showing a different autism phenotype (Kopp, Kelly, & Gillberg, 2010).

Overlapping clinical presentations, difficulties in reliably identifying different autism subgroups and other evidence have led to the proposal that there may be different autism spectra (Wing, Gould, & Gillberg, 2011) with the most appropriate term for the whole group of disorders inferred here being the 'autisms' (Coleman & Gillberg, 2011; Wing et al., 2011).

Autism is not a psychiatric disorder. However biological, psychological, social, educational, work and other circumstances associated with autism predispose those with autism to greater

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risk for mental health problems (National Institute for Health and Clinical Excellence, 2011, 2012). In this chapter we focus on psychopathology (psychiatric symptomatology as described in DSM/ICD; World Health Organization, 1993) that arises when autism and ID are coexisting conditions. Noting a discrepancy between clinical descriptions of psychopathology in autism and the experience of people on the spectrum, we also compare these clinical descriptions with the (often very moving) accounts of individuals with autism. We have adopted the terminology proposed by Donna Williams, herself with autism, 'Outside-In' and 'Inside-Out'(Williams, 1996). We explore whether an Inside-Out perspective may contribute to our understanding of psychopathology observed in those with autism. An improved understanding can be anticipated to have implications for treatment and prevention of such psychopathology that may arise.

Determining whether the addition of autism adds to the risk of mental ill health in those with ID has implications for care and service provision. In the UK, with the Autism Act (2009), and subsequent government initiatives (Department of Health, 2010a, 2010b; National Audit Office, 2009, 2010), there is increasing support for those with autism in the general population but there remain concerns that the needs of those within ID services are still not being recognised.

Finally, drawing on evidence from clinical research and narratives from people with autism, we make suggestions for future research to shed light on the complex interplay of circumstances that contribute to mental health and psychopathology in those with both autism and ID.

Prevalence of Autism in the General Population

Prevalence estimates for autism range from 0.72 to 1.57 % for child populations (less than 16 years old; Baird et al., 2006; Barbaresi, Colligan, Weaver, & Katusic, 2009; Baron-Cohen et al., 2009; Centers for Disease Control and Prevention, 2009; Posserud, Lundervold, Lie, & Gillberg, 2010) and 0.55 to 1.4 % for adult populations (16 years plus; Brugha et al., 2011; Chang et al., 2003; Cimera & Cowan, 2009; Nylander & Gillberg, 2001). The most commonly quoted rate is 1 %. The rate of unrecognised autism has been found to be as high as 40 % for children (Baron-Cohen et al., 2009) and 91 % for adults (Nylander & Gillberg, 2001).

Rates of ID in autism have been reported in two of these studies of children: estimated prevalences were 55 % (Baird et al., 2006) and 41 % (Centers for Disease Control and Prevention, 2009), respectively. Earlier reviews of ID in children with autism reported estimates of up to 88 % (Table 1 in Bryson, Bradley, Thompson, & Wainwright, 2008). More recent rates of ID are lower likely because they include a wider range of individuals on the autism spectrum and because of improved recognition and therefore higher rates of high-functioning autism.

Prevalence of Autism in ID

Diagnosing autism can be particularly uncertain where there are:

- A lack of information about early life
- Complex coexisting mental health disorders (e.g. ADHD, conduct disorder or attachment disorder)
- Sensory impairment (e.g. blindness or deafness) or motor disorder such as cerebral palsy
- Mental age less than 18 months equivalent (National Institute for Health and Clinical Excellence, 2011)

Autism screening (Charman et al., 2007) and assessment tools (National Institute for Health and Clinical Excellence, 2011) generally have greater efficacy when used with individuals who do not have significant other disabilities. The Diagnostic Interview for Social and Communication Disorders (DISCO; Leekam, Libby, Wing, Gould, & Taylor, 2002; Wing, Leekam, Libby, Gould, & Larcombe, 2002) assessment of autism is unique in that it identifies autism across the range of functioning including mild to profound ID. The ability of other gold standard assessment tools such as the Autism Diagnostic Interview (ADI; Lord, Rutter, & Le Couteur, 1994) and the Autism Diagnostic Observation Schedule (ADOS; Lord et al., 2000) to differentiate autism among individuals with

Table 16.1 Studies of psychopathology comparing individuals who have intellectual disabilities (ID) with and without autism (ASD)

Study Rating on criteria A B C D	Population and identification/ assessment of ASD	N ASD +ID	N ID	Measure of psychopathology	Results for ASD + ID vs. ID
I: Population-based studies					
Bradley, Summers, Wood, and Bryson (2004) 1 3 1 3	Adolescents with severe ID (mean age 16, IQ < 40). Matched on age, gender and non-verbal IQ. ASD identified using ADI-R	12	12	Diagnostic assessment for the severely handicapped (DASH-II)	Reaching 'clinical significance': anxiety (42 % vs. 0 %), mania (67 % vs. 8 %), depression (50 % vs. 8 %; all $p < 0.05$). Mean of 5.3 'clinically significant disorders' vs. 1.3 for those with no ASD. Higher mean total score (44.8 vs. 16.2) and higher scores on anxiety, mania and depression scales (all $p < 0.05$)
Bradley and Bolton (2006) 1 3 2 3	Adolescents with ID (aged 14–20, IQ < 75). Matched on gender, age and non-verbal IQ. ASD identified using ADI-R	36	36	Clinical diagnosis using SAPPA, ICD-10, research diagnostic criteria (RDC)	Greater history of ≥1 episode of psychiatric disorder, 47 % vs. 17 %; higher mean total number of episodes, 0.89 vs. 0.23 (both $p < 0.05$). Most frequent diagnoses: depression (22 % vs. 8 %), adjustment (22 % vs. 5 %), mixed (11 % vs. 1 %) and bipolar disorder (5 % vs. 0 %) (these differences were not significant)
Bradley and Isaacs (2006) 1 3 2 3	See Bradley and Bolton (2006)	31	31	See Bradley and Bolton (2006). ICD-10 symptoms of inattention, hyperactivity and impulsivity (IHI)	More likely to have ≥1 symptom of I, H and I or any one of IHI (55 % vs. 23 %); more likely to meet criteria for hyperactivity, impulsivity or any one of IHI (52 % vs. 19 %) (all $p < 0.05$). Not significantly more likely to reach full criteria for ADHD (23 % vs. 7 %, $p = 0.07$) or hyperkinetic syndrome (13 % vs. 3 %, $p = 0.2$)
Bradley et al. (2011) 1 3 2 3	See Bradley and Bolton (2006)	36	36	See Bradley and Bolton (2006). Compulsive behaviour checklist, tic checklist, stereotypy checklist, self-injury and self-restraint checklist	Higher mean number of conditions (3.4 vs. 1.8; $p < 0.001$). Higher number of fears and phobias, compulsions, stereotypies and observed anxiety symptoms ($p < 0.05$). No difference in number of tics, self-injurious or self-restraining behaviours. No difference in number meeting criteria for anxiety (25 % vs. 17 %), phobic disorder (19 % vs. 11 %) or Tourette syndrome (6 % vs. 3 %)

(continued)

Table 16.1 (continued)

Study Rating on criteria A B C D	Population and identification/ assessment of ASD	N ASD +ID	N ID	Measure of psychopathology	Results for ASD + ID vs. ID
II: ID register-based studies					
Melville et al. (2008) 0 1 2 3	Adults. Matched on gender, age and severity of ID. ASD identified using C21st Health Check and assessed by clinical diagnosis	77	154	Clinical diagnosis (DC-LD, ICD-10 and DSM criteria)	No significant differences on the number with mental ill health: excluding problem behaviours (21 % vs. 23 %) and including problem behaviours (48 % vs. 40 %); incidence of mental ill health over 2 years (16 % vs. 19 %) or recovery from mental ill health over 2 years (36 % vs. 48 %) (all $p > 0.05$). Those with ASD and problem behaviours less likely to recover over a 2-year period (6 % vs. 38 %)
Morgan et al. (2003) 1 1 2 0	Adults. ASD identified using PDD-MRS and assessed by clinical diagnosis using ICD-10	164	400	Clinical diagnosis based on ICD-10	41 % had a psychiatric disorder (65 % of those with mild ID, 35 % of mod ID and 40 % of severe ID). Higher rates of depression (20 % vs. 3 %) and bipolar disorder (11 % vs. 2 %), both $p < 0.05$, no difference in prevalence of schizophrenia
III: ID service-based studies					
Hill and Furniss (2006) 1 0 1 0	Children and young adults (aged 8–29) with severe ID and challenging behaviour. Sample split into ASD/no ASD using cut-off on DASH-II ASD subscale	69	13	Aberrant behaviour checklist, DASH-II	Higher number met criteria on DASH-II for anxiety (77 % vs. 39 %), mania (77 % vs. 39 %), mood disorder (64 % vs.46 %) and schizophrenia (30 % vs. 8 %) (all $p < 0.05$). Higher scores on organic, anxiety, stereotypies and mania subscales ($p < 0.05$). No significant differences on ABC irritability, lethargy, stereotypy or hyperactivity scores
Totsika et al. (2010) 0 0 1 1	Older adults (aged 50+). Subsample matched on functioning. ASD identified and assessed using 3 DAS items	87	195	PIMRA or PAS-ADD, aberrant behaviour checklist	No difference in number meeting criteria for psychiatric caseness in unmatched sample (32 % vs.23 %, $p = 0.09$) or matched sample (37 % ($N = 65$) vs. 27 %, $p = 0.11$), higher ABC score for unmatched group only (38 vs. 19, $p < 0.01$)

LoVullo and Matson (2009) 0 1 0 1	Adults with severe ID. Compared: ASD+ID vs. ID vs. ASD+ID+psychopathology. Matched on severity of ID. Included those with an existing clinical diagnosis of ASD according to DSM-IV/ICD-10	84	42	Autism spectrum disorders-comorbidity for adults (ASD-CA)	Higher scores for ASD+ID vs. ID on total (10 vs. 4), anxiety (0.8 vs. 0.05), irritability (3 vs. 1.4), hyperactivity (2.4 vs. 1.2) and depression (0.7 vs. 0.3) scores (all $p<0.05$). Significantly higher scores for ASD+ID+psychopathology vs. ID on total (8), anxiety (0.4), irritability (2.6) and hyperactivity (2.6) (all $p<0.05$). No significant differences for ASD+ID vs. ASD+ID+psychopathology groups
Matson, Rivet, Fodstad, Dempsey, and Boisjoli (2009) 0 1 2 1	Adults with severe ID. Matched on gender. Included those with an existing diagnosis of ASD according to DSM-IV	90	287	Clinical diagnosis (DSM-IV-TR), Vineland ABS	Higher number with an additional axis I diagnosis (33 % vs. 13 %). Significantly lower adaptive skills for those with ASD and axis I diagnosis compared to those with ASD and no-axis I diagnosis and those with ID alone
Smith and Matson (2010) 0 1 0 3	Adults with severe ID. Matched on age and gender. Compared: ASD+ID vs. ASD+ID+epilepsy vs. ID vs. ID+epilepsy. Included those with an existing diagnosis of ASD using DSM-IV/ICD-10	50	50	ASD-CA	Higher scores on anxiety and hyperactivity subscales for ASD+ID+epilepsy vs. ID+epilepsy. Higher scores on irritability, hyperactivity and depressive subscales for ASD+ID+epilepsy vs. ID (both $p<0.05$). Higher scores on irritability subscale for ASD+ID vs. ID
IV: ASD service-based studies					
Bakken et al. (2010) 0 1 1 0	Adults and adolescents (aged 14+). ASD confirmed by clinical diagnosis using ICD-10	62	132	Psychopathology in autism checklist (PAC)	Overall higher rates of any psychiatric disorder (53 % vs.17 %), psychosis (25 % vs. 9 %), depression (37 % vs. 15 %), anxiety (34 % vs. 9 %) and OCD (13 % vs. 3 %), all ($p<0.05$). No differences among those with a psychiatric disorder for depression (70 % vs. 87 %), OCD (24 % vs. 17 %) or anxiety (64 % vs. 52 %). Mean no. of disorders in those with a psychiatric diagnosis=2.1 for each group

(continued)

Table 16.1 (continued)

Study Rating on criteria A B C D	Population and identification/ assessment of ASD	N ASD +ID	N ID	Measure of psychopathology	Results for ASD+ID vs. ID
Brereton, Tonge, and Einfeld (2006) 0 1 0 1	Children and adolescents (aged 4–18). 86 % of ASD group had ID (ASD confirmed using ABC, CARS and DSM-IV structured clinical interview)	367	550	Developmental behaviour checklist-parent version (DBC-P)	Higher mean total (61 vs. 43), disruptive (13 vs. 11), self-absorbed (15 vs. 9), communication disturbance (5 vs. 4), anxiety (9 vs. 7), ADHD (7 vs. 5) and depression (5 vs. 4) scores; all $p<0.001$ for MANCOVAs controlling for age, gender and ID
V: Mental health service-based studies					
Charlot et al. (2008) 0 1 1 2	Adult inpatients with depression. Matched on age/gender/ID severity. Included those with an existing clinical diagnosis of ASD according to DSM-IV	13	40	Mood and anxiety semi-structured interview for patients with ID (MASS)	Prevalence of comorbid anxiety: 62 % vs. 38 %. Higher mean number of symptoms: 12 vs. 10, $p=0.05$. No differences on individual symptoms or clusters, except for decreased sleep ($p=0.05$)
Lunsky, Gracey, and Bradley (2009) 0 1 2 1	Adult mainstream mental health patients. Matched on gender and inpatient/outpatient status. Included those with an existing clinical diagnosis of ASD	23	23	Clinical diagnosis using ICD-9	Lower rate of psychosis (26 % vs. 78 %, $p<0.01$). No significant difference on other disorders. Lower rate of psychosis compared with 23 mental health patients with no ASD or ID (83 %, $p<0.01$)
McCarthy et al. (2010) 0 1 2 1	Adult from a specialist mental health in intellectual disability service. Included those with an existing clinical diagnosis of ASD according to ICD-10 criteria	124	562	Clinical diagnosis using ICD-10, challenging behaviour (CB, according to DAS-B)	More likely to have CB (88 % vs. 62 %, $p<0.01$). For those with CB, lower rates of schizophrenia (10 % vs. 19 %) or any psychiatric disorder (29 % vs. 52 %) for those with ASD ($p<0.05$). For those without CB higher rates of schizophrenia, 40 % vs. 16 %, $p<0.05$. After controlling for the level of ID, gender and age, there was no association between comorbid psychopathology and presence of CB

Tsakanikos et al. (2006) 0 1 2 1	See McCarthy et al. (2010)	147	605	Clinical diagnosis using ICD-10, DAS-B	Lower rates of psychiatric disorder (36 % vs. 55 %) and personality disorder (PD; 3 % vs. 9 %, both $p<0.01$). No significant differences for anxiety (4 % vs. 8 %), depression (6 % vs. 9 %) or psychosis (16 % vs. 19 %). Significant association between ASD and PD when age, gender and severity of ID were accounted for
Underwood et al. (2012) 0 3 2 1	See McCarthy et al. (2010). Diagnosis of ASD/no ASD confirmed using ADOS	53	48	Clinical diagnosis using ICD-10, DBC-adult version (DBC-A)	Lower rates of psychiatric disorder: 46 % vs. 85 %, more likely to reach cut-off for psychiatric caseness on DBC-A (total score >50): 60 % vs. 11 %. Presence of ASD significantly predicted a higher total DBC-A score and greater likelihood of score >50 when age, gender and severity of ID were accounted for

Studies are grouped together according to the way in which they identified their samples:
I: Population-based studies that identified all (or all in a specific age range) with ID in a geographical area (including those with previously undiagnosed ID)
II: Studies that identified all with an existing diagnosis of ID in a geographical area (e.g. sourced from an ID register)
III: Studies that sourced participants from an ID service (e.g. a residential placement for individuals with ID)
IV: Studies that sourced participants with ASD from an ASD service
V: Studies that sourced participants from a mental health service
Criterion A: *Screening for ASD* 0 = Method with a high threshold/lower level of potential identification (e.g. existing clinical diagnoses). 1 = Method with a low threshold/higher level of potential identification (e.g. standardised ASD screening tool)
Criterion B: *Confirmation/diagnosis of ASD* 0 = Did not use any standardised *diagnostic* assessment for ASD. 1 = Used a clinical diagnostic assessment for ASD according to standardised clinical criteria (DSM/ICD/RDC). 2 = Used a gold standard ASD diagnostic assessment (e.g. ADI) but there was no face-to-face contact with participant. 3 = Used a gold standard ASD diagnostic assessment (e.g. ADI, ADOS, DISCO) that included face-to-face contact with participants
Criterion C: *Assessment of psychopathology* 0 = Compared scores on a psychiatric symptom tool rather than presence/absence of a psychiatric disorder/diagnosis. 1 = Used a psychiatric symptom tool with cut-off scores for psychiatric caseness or specific diagnoses. 2 = Carried out a comprehensive psychiatric assessment using standardised clinical diagnostic criteria (DSM/ICD/RDC)
Criterion D: *Matching of participants with and without ASD 0* = No matching or attempt to account for (a) severity of ID/IQ/standardised measure of functioning or (b) age or (c) gender. 1 = Data analysis took into account all three of a, b and c or participants were matched on at least one of a, b or c. 2 = Group matching on all three of a, b and c or groups were compared and no differences found for a, b or c. 3 = Case-by-case matching on all three of a, b and c
ABC autism behaviour checklist, *ADI* autism diagnostic interview, *ADOS* autism diagnostic observation schedule, *CARS* childhood autism rating scale, *DAS* disability assessment scale, *MANCOVA* multivariate analysis of covariance, *PAS-ADD* psychiatric assessment schedule for adults with developmental disorders, *PDD-MRS* pervasive developmental disorder in mental retardation scale, *PIMRA* psychopathology instrument for mental retarded adults, *SAPPA* schedule for the assessment of psychiatric problems associated with autism (and other developmental disorders)

the most severe intellectual disability remains challenging. Care has to be taken that for each individual, the items considered are developmentally or otherwise appropriate (Bryson et al., 2008). Increasing severity of ID is associated with other disabilities such as sensory and motor impairments that will further complicate the developmental history and manifestations of possible autism. Autism assessments conducted by clinicians and researchers experienced in autism and ID, together with the opportunity for discussion to develop consensus around complex cases, seem likely to offer the most accurate assessment available at the present time.

A review of earlier studies spanning from 1978 to 2005 (Table 2 in Bryson et al., 2008) showed prevalence rates of autistic disorder in ID populations of between 19.8 and 40 % when the broader spectrum of the triad of autistic impairments were included. Of the further eight studies subsequently available for review, six reported on adults (Bouras, Cowley, Holt, Newton, & Sturmey, 2003; Cooper, Smiley, Morrison, Williamson, & Allan, 2007; La Malfa, Lassi, Bertelli, Salvini, & Placidi, 2004; Morgan et al., 2002; Saemundsen et al., 2010; Totsika, Felce, Kerr, & Hastings, 2010), one on adolescents only (14–20 years; Bryson et al., 2008) and one reported on children and adolescents (4–18 years; de Bildt, Sytema, Kraijer, & Minderaa, 2005). Prevalence estimates for autism ranged from 7.5 to 39.2 % for adults, 28.2 % for adolescents and 17.7 % for children. Diagnosis of autism prior to the study identification tended to be low: less than half of the adolescents had been previously diagnosed. Pre- and post rates reported in two of the adult studies (La Malfa et al., 2004; Saemundsen et al., 2010) were 7.8 and 9 % (pre) to 39.2 % and 21 % (post), respectively. Two studies reported greater rates of autism with greater severity of ID (de Bildt et al., 2005; Morgan et al., 2002) and one reported that autism prevalence decreased with age (Morgan et al., 2002). Recent evidence (Underwood, 2011) indicates there may be significant numbers of people with autism in adult clinical populations with ID who have remained undiagnosed; these individuals tend to be in older age groups for whom it can be anticipated getting an early history may be problematic. Such findings may account in part for the lower estimated rate of autism found in one study (Melville et al., 2008).

These research studies support the view that unless specifically looked for and systematically assessed, autism may not be identified in the general population or in populations with ID (Emerson & Baines, 2010; National Institute for Health and Clinical Excellence, 2012). Where such robust identification occurs, autism can be calculated to be up to 24 (children) and 28 (adults) times greater in the population with ID than in the general population.

Psychopathology in ID and in Autism

There is good evidence for greater mental ill health in populations with ID compared to general populations (children: Emerson & Hatton, 2007); (adults: Cooper et al., 2007; Cooper & van der Speck, 2009). Multicentre studies of children with autism indicate that psychiatric disorders are common and frequently multiple; high levels of anxiety, mood, disruptive disorders and ADHD are reported (Leyfer et al., 2006; Simonoff et al., 2008).

While there is a lack of epidemiological research on adults with autism (Brugha et al., 2009, 2011), two robust clinical longitudinal studies of mental health in adults with autism are reported: (1) 122 adults with 'possible childhood onset neuropsychiatric disabilities' and normal IQ were found to have high rates of mood (53 %), anxiety (50 %), especially obsessive-compulsive disorder (OCD; 20 %), chronic tic disorder (20 %), psychotic disorders (12 %) and ADHD (43 %; Hofvander et al., 2009); (2) a follow-up study of 135 adults referred for evaluation of autism as children showed that 16 % had developed new onset disorders (and a further 8 % uncertain new onset disorder), with these disorders falling into two main groups: OCD (some with catatonia) and affective disorders (Hutton, Goode, Murphy, Le Couteur, & Rutter, 2008). An increase in rates of anxiety and depression is also described in adults with Asperger's syndrome (see review by Woodbury-Smith & Volkmar, 2009).

Table 16.2 Different perspectives in understanding distress behaviours and psychopathology in autism

Outside-In perspective: DSM, ICD and other clinical diagnoses based on behaviours of concern	Examples of behaviours of concern	Inside-out perspective: people with autism: first person autobiographical accounts when engaging in these behaviours of concern	Comments on the behaviours of concern by those with autism and those supporting people with autism
Obsessive-compulsive disorder (OCD) (for similarities and differences of repetitive behaviour between OCD and autism, see Zandt, Prior, & Kyrios, 2007)	Repeating the same behaviours over and over (such as opening or closing doors, putting items back in place, arranging things in certain order) may or may not be upset if interrupted but will seek to return to this behaviour	'Reality to an autistic person is a confusing, interacting mass of events, people, places, sounds and sights. There seem to be no clear boundaries, order or meaning to anything. A large part of my life is spent just trying to work out the pattern behind everything. Set routines, times, particular routes, and rituals all help to get order into an unbearable chaotic life' (Jolliffe, Lansdown, & Robinson, 1992)	Focusing on something in order to avoid overload (and the 'autonomic storm'; see text page 8)
	Obsessional rituals—rocking, rhythmic head banging, spinning objects, perimeter hugging, needing to touch everything in the room before settling down	'if you start feeling reality slipping away you want to focus on something – pain or flicking a light switch on or off' …. I like sameness, need to know what's coming next, things are more coherent, under pressure things tend to fragment ….' (Weeks, Undated)	Obsessional rituals may reflect (1) the autistic individual's construction of the world by means of a very different set of sensory information (as seen in visually impaired children) 'using her eyes as much as her ears, using her hands as much as her eyes' and (2) checking up on a distorted sensory perception with an alternative sense, e.g. touch on distorted vision (Albano, 2003; Bogdashina, Date unknown)
	Obsessive and unusual interests (such as in parts of objects)	'my attention was firmly set on my desire to lose myself in spots and I'd ignore the garble'; 'I learned eventually to lose myself in anything I desired, the patterns on the wallpaper or the carpet, the sound of something over and over again, the repetitive hollow sound I would get from tapping my chin' (Williams, 1992)	'The tendency of some autistic people to constantly touch themselves and objects around them may be an attempt to stabilize body and environmental boundaries' (Grandin, 2000a)
	Repetitive speech or echolalia	'I used to repeat the same words over again as this made me feel safer'	Nazeer, a person with autism, writes about his school friends with autism and their search for coherence (Nazeer, 2006)

(continued)

Table 16.2 (continued)

Outside-In perspective: DSM, ICD and other clinical diagnoses based on behaviours of concern	Examples of behaviours of concern	Inside-out perspective: people with autism: first person autobiographical accounts when engaging in these behaviours of concern	Comments on the behaviours of concern by those with autism and those supporting people with autism
		'Trying to keep everything the same reduces some of the terrible fear' (Jolliffe et al., 1992)	'Echolalia or the constant disconnected use of a particular word or phrase … described as the desire for local coherence, the preference that autistic people frequently demonstrate for a limited though immediate form of order' as a protection against complexity or confusion …'(Nazeer, 2006) '… Earlier I mentioned the way that autistic people regularly seek local coherence…. I rarely go out without a crocodile clip in my pocket although recently my cell phone has begun to substitute for it' (Nazeer, 2006) 'The rule probably tended to work. It achieved local coherence. We could make sense of it' (Nazeer, 2006) 'Andre tries to reach local coherence to protect himself by putting more complex things in the background and focusing instead on tapping a pen or lining up empty glasses' (Nazeer, 2006)
Stereotypies	Repetitive movements or vocal sequences that appear to be invariant in form without any obvious eliciting stimulus or adaptive function, e.g. body rocking, mouthing and complex hand and finger movements (Baron et al., 2006)	'rocking and spinning were other ways to shut out the world when I became overloaded with too much noise' (Grandin, 2006)	Coping mechanism to attain homeostasis and calm (functional relationship between stereotypies and arousal (HR) has been found) (Baron et al., 2006)

		'what other people call odd hand movements and what people refer to as grimaces are not meant to be annoying, they too give a sense of control, safety and perhaps pleasure' (Jolliffe et al., 1992)	'Many autistic adults deal with discomforting events by returning to a regime – it may be as simple as rocking … or involve multiplying three digit prime numbers' …. and … 'The search for coherence: other coherentizing behaviours include 'whirling on own initiative, running to and fro, bouncing, walking on toes, mouthing and licking things' (Nazeer, 2006)
		'I [Tito Mukhopadhyay] am calming myself. My senses are so disconnected, I lose my body. So I flap (my hands). If I don't do this, I feel scattered and anxious … I hardly realized I had a body … I needed constant movement, which made me get the feeling of my body' (Blakeslee, 2002) 'many of my fixations had a sensory basis' (Grandin, 1992a, 1992b). 'fixations can be tremendous motivators' (Grandin, 1992a, 1992b) 'Autistics are captivated by movements and inner rhythms … these provide a stabilizing action that grounds the child … they focus upon some point of themselves to help calm them especially when they are threatened' (O'Neill, 1998)	
Anxiety disorders including panic, fears and phobias	Low threshold startle response, constant high levels of anxiety, agitation, hyper vigilance, and avoidance of objects and situations; may (or may not) show physiological signs of anxiety and may have rapid pulse in the absence of other physiological signs of anxiety	'from the moment I get in (to the supermarket) there's all the sounds, movements, smells, shapes, colours, shadow and shine from all directions all hitting me at once … my mind get flooded with this array of contrasts at a pace that's much too fast for my conscious mind to process' (Williams, 1995)	Responses to sensory hypersensitivities and distortions

(continued)

Table 16.2 (continued)

Outside-In perspective: DSM, ICD and other clinical diagnoses based on behaviours of concern	Examples of behaviours of concern	Inside-out perspective: people with autism: first person autobiographical accounts when engaging in these behaviours of concern	Comments on the behaviours of concern by those with autism and those supporting people with autism
		'Life is bewildering, a confusing, interacting mass of people, events, places and things with no boundaries' (Jolliffe et al., 1992)	One adult with autism interviewed by Cesaroni and Garber (1991) theorises that certain frightening sounds can act as a trigger for disorganisation of processing similar to epileptic seizures triggered by a flashing light
		'I always hated to be touched. I wanted to experience the feelings of being hugged but it was just too overwhelming … like a great overwhelming tidal wave of stimulation I reacted like a wild animal … loud noises were also a problem, often feeling like a dentist's drill hitting a nerve' (Grandin, 2006)	'A 'side effect' of [visual] perception in bits, is a sense of fear that is not specifically related to objects but is originated in the fact that the first encounter with a physical object is a particle one' (Van Dalen, 1995) VanDalen compares this with a confrontation with a silhouette in the dark: 'one knows that something is there but it is not altogether immediately clear what it is' (Bogdashina, Date unknown)
		'All things coming from the outside must be gentle, sometimes devoid of emotion, so as not to overwhelm.' (O'Neill, 1998)	
As overload intensifies, increasing difficulty filtering and prioritising information occurs and the body's self-defences system (ANS) is triggered to 'flight', 'fight' and 'freeze' (reactive behaviours) and eventually to an autonomic storm (a massive increase in sympathetic nervous activity—a hyperstress response; Parady, 2011)			
'**Behaviour problems**'—with behavioural judgments (such as 'refusal', 'manipulative', 'attention seeking', 'deliberate') based on the clinicians' sensory experience of reality, and not on that of the person with autism	Withdrawal into own activities	'I would sit for hours watching and dribbling sand between my fingers … I went into a trance which cut me off from the sights and sounds round me' (Grandin, 2006)	'Flight' responses

Oppositional defiant disorder (ODD)	Refusing to engage or doing the opposite of what was requested	'Auditory and tactile input often overwhelmed me. Loud noise hurt my ears. When noise and sensory stimulation became too intense I was able to shut off my hearing and retreat into my own world' (see Grandin, 2000a, 2000b)	Looking for coherence and a point of reference when in overload
	Avoidance of places and situations	Grabbing and Clinging: 'The din made the ground under my feet disappear and I could neither see nor feel the world round me. Up and down were suddenly in the same place and I had no idea where my feet were. So as not to fall over or explode from inside, I had to grab the fence where I was standing, pressing myself against it and holding on hard. I had to feel something that stood still, something anchored in a world that had become totally unpredictable.' (Gerland, 2003; page 130)	
	Grabs or clings onto others; 'No' may intensify this unwanted behaviour	'If you were being FOREVER forced (at times none too patiently) to do upsetting functions or at times acutely painful ones, just because everyone else does it with no discomfort, AND expects you to be the same; would that make you outgoing, and a party personality? Or would you turn away from your tormentors, acting as if you were uncomfortable or afraid or possibly frustrated with them?' (Bogdashina, 2010; page 61)	
A criterion for autism	Avoidance of eye contact	'someone looking directly into my eyes felt like an attack' (Nony, 1993) 'People have no idea how unbearably painful it is for me to have to make eye contact.' (Jolliffe et al., 1992) 'Agony' (Grandin, 2006)	Avoidance of direct perception helps to avoid sensory overload and pain
Conduct disorder including aggression and property destruction	Suddenly lashes out (aggression); this may appear unprovoked and out of the blue or in response to certain triggers	'Tantrums occurred when I became tired or stressed out by too much noise … my behaviour was like a tripping a circuit breaker. One minute I was fine, the next minute I was on the floor kicking and screaming like a crazed wildcat.' (Grandin, 2006)	'Fight' responses and behaviours of an autonomic storm

(continued)

Table 16.2 (continued)

Outside-In perspective: DSM, ICD and other clinical diagnoses based on behaviours of concern	Examples of behaviours of concern	Inside-out perspective: people with autism: first person autobiographical accounts when engaging in these behaviours of concern	Comments on the behaviours of concern by those with autism and those supporting people with autism
Intermittent explosive disorder			Some of these behaviours may represent flashbacks to previous trauma and or abuse
Self injurious behaviour	More severe and sustained aggression directed towards self, others or the environment; hits, pinches and bits self	'There was a rip through the centre of my soul. Self-abuse was the outward sign of an earthquake nobody saw. I was like an appliance during a power surge. As I blew fuses my hands pulled out my hair and slapped my face. My teeth bit my flesh like an animal bites the bars of its cage, not realising the cage was my own body. My legs ran round in manic circles, as though they could outrun the body they were attached to. My head hit whatever was next to it, like someone trying to crack open a nut that had grown too large for its shell. There was an overwhelming feeling of inner deafness – deafness to self that would consume all that was left in a fever pitch of silent screaming.' (Williams, 1995)	'Fight' responses and behaviours associated with an autonomic storm
Disruptive behaviour and impulse control disorders		'… you're going to run out into the street where the traffic is, or run full tilt into a brick wall, anything to stop it [the panic]. I'd much rather have pain – pain is one over-riding sensation rather than getting a whole lot of jumbles' (Weeks, Undated)	
'Challenging Behaviours'			
Autism-related catatonia	1: New onset posturing (freezing), stereotypies, alteration in level of activity	'if I get a lot of sensory overload, I just shut down, become catatonic. I get what's known as fragmentation … like being turned into forty channels at once … if you get this when you are four, the result is total panic' (Weeks, Undated)	ANS response of 'Freeze'—resulting in catatonic-like states (Dhossche, Shah, & Wing, 2006; Fink, Taylor, & Ghaziuddin, 2006; Loos Miller & Loos, 2004; Loos & Loos Miller, 2004; Wing & Shah, 2000) sometimes referred to as 'meltdown' or 'shutdown' states

Catatonia associated with schizophrenia or mood disorder	2: Slowed down, unusual motor patterns and gait, especially in response to requests; may show normal motor movements in self initiated activities and in some environments compared to others	'like having 4 drill sergeants screaming conflicting orders at you at once and if you don't do everything right away you will be in big trouble and you don't know what to do first so you stand there being yelled at'. (see list by Loos) (Loos Miller & Loos, 2004)	Shyness and being on the more passive end of interaction on the ASD spectrum may increase vulnerability to these shutdown and catatonic (Loos Miller & Loos, 2004; Wing & Shah, 2000) withdrawing being characteristic rather than acting aggressively in response to distress
	3: Increased slowness also affecting speech	What makes the shut down better? '… time, sleep, rhythmic rocking, spinning, "stimming", working puzzles and spending quiet time alone …' (Loos Miller & Loos, 2004)	'In academic settings when pressured by an adult to perform tasks that were difficult, she became unresponsive, sleepy, immobile, and limp to the touch for several minutes and then fell asleep in a chair for as briefly as 10 min and up to 2 h. These "shutdown" states were always triggered by social stress of a certain kind and they became more severe and frequent over a period of about a year' (Loos & Loos Miller, 2004)
	4: Difficulties initiating and completing actions with increased reliance of physical and verbal prompts by others	Williams recalls as a child she was afraid of 'the big black nothingness coming to get me'. As an adult she recognised the syndrome as 'sensory flooding triggering such a degree of overload as to cause an epilepsy-like total shut down on the processing of incoming information' (Williams, 2003)	
	5: Increased passivity and lacking in motivation (Wing & Shah, 2000)	'I can't take the fire alarm. I go completely stiff. If it goes off and the house was burning round me, I should have to be physically removed from the room.' (speaker with autism—National Autistic Society Conference Cardiff. 2011)	

Psychopathology When Autism and ID Coexist

Given the evidence cited it might be hypothesised that the presence of autism would add to the risk for psychopathology in those with ID. Current evidence indicates that the interplay of autism and ID on psychopathology is complex. In a review of 24 studies published during 2006 and 2007, McCarthy (2007) concluded that children with ID and autism have a higher prevalence of ADHD, mood disorders, catatonia and repetitive behaviours compared to children without autism. However, in a subsequent review of the literature spanning from 2003 to 2010 concerning adults with ID with and without autism, methodological challenges were more apparent and conclusions less certain (Underwood, McCarthy, & Tsakanikos, 2010). The authors commented on (1) the difficulties of assessing psychopathology in adults; (2) the need to ensure that autism is attributed accurately; (3) the importance that studies match groups on age, gender and severity of ID; and (4) the need to collect data on a broader range of measures including health, social funcv-tioning and service users experiences of mental health services. In one population-based study, apparent differences in mental ill health between ID groups with and without autism were accounted for by factors other than autism, including ability level and Down syndrome (Melville et al., 2008). A more recent series of studies including mental health clinic-based matched groups found participants with ID and autism were less likely to have a psychiatric disorder but had significantly lower health and social functioning than those without no autism (Underwood et al., 2012; Underwood, 2012). The group with autism and ID were also more likely to exceed the cut-off score for 'psychiatric caseness' on the Developmental Behaviour Checklist for Adults (DBC-A; Mohr, Tonge, Einfeld, & Gray, 2004). These latter studies taken together point to the important difference between (a) clinical evaluation of psychiatric disorder and (b) psychopathology as measured by screening tools and checklists; the former embeds symptoms and behaviours in a psychiatric diagnostic formulation while the latter do not differentiate between behaviours that may underpin psychiatric disorder from those related to other aetiologies (such as autism, ID or unrecognised medical conditions).

Two studies, subsequent to the literature reviews above, reported further on psychopathology in individuals who have ID, with and without autism. Using a Psychopathology in Autism Checklist (PAC), greater rates were reported of 'any psychiatric disorder', psychosis, depression, anxiety and OCD in an autism-ID group ($N=62$) compared to an ID only ($N=132$) group of adolescents and adults (all $p<0.05$; Bakken et al., 2010). In a population-based sample of 36 individually matched adolescent ID and autism-ID groups, using a standardised semi-structured clinical evaluation of psychiatric disorder, significant differences were reported in compulsive behaviours ($p<0.001$), stereotypies ($p<0.001$), anxiety ($p<0.001$) and fears and phobias ($p<0.05$) (Bradley, Ames, & Bolton, 2011). These background (non-episodic) behaviours were independent from significant differences in episodic disorders (e.g. depressive, adjustment; Bradley & Bolton, 2006) and clinically significant inattentive, hyperactive, impulsive behaviours and ADHD (Bradley & Isaacs, 2006) in the same matched groups. This population- based study specifically distinguished relapsing and remitting psychiatric illness (e.g. adjustment disorder, depression, bipolar affective disorder) that tended to occur for the first time in adolescence, from disorders that may appear more insidiously during childhood and continue into adolescence but are not specifically episodic although may escalate during episodic illness (e.g. ADHD). This study and others (Cooper et al., 2007; Howlin & Moss, 2012; Hutton et al., 2008; Mouridsen, Rich, Isager, & Nedergaard, 2008) demonstrate the complexity of presenting psychopathology, coexisting diagnoses and the need to take into account individual historical patterns of mental distress.

Additionally there is emerging evidence that circumstances external to the individual such as staff attitudes (Rose, 2011), life events (Hulbert-Williams & Hastings, 2008), traumatic experiences

(Martorell et al., 2009) and other supports (Allen, 2008; Cooper et al., 2007; Smiley et al., 2007) need to be taken into account when studying psychopathology in ID.

As briefly noted above, the concept of psychopathology covers a broad range of mental distress—from clearly identifiable psychiatric disorders to behaviours and symptoms that are inferred to be psychiatric in origin but may be difficult to confirm definitively because of the limited cognitive and communication capacities of the individual with ID and autism. Some behaviours that appear to be mental distress have been shown to be a medical condition, e.g. GI related (Buie, Campbell, et al., 2010; Buie, Fuchs, et al., 2010). Alternatively psychiatric distress may be correctly identified but its origins not fully recognised, e.g. early trauma and other life events (Bradley, Sinclair, & Greenbaum, 2012). These diagnostic issues become more problematic with increasing severity of ID (and autism) and require comprehensive biopsychosocial longitudinal evaluation to unravel the aetiological complexities of the presenting symptoms (Bradley & Hollins, 2010).

From the evidence reviewed above, it is apparent that robust determination as to whether autism, when coexisting with ID, contributes further mental distress, requires, at a minimum, the following methodological criteria to be met:

(a) All in the autism group have been correctly identified with autism and none in the ID group have autism (Criteria A and B: see Table 16.1).
(b) Psychiatric disorder is correctly identified (Criteria C, Table 16.1) as well as aetiological factors wherever possible.
(c) Groups with and without autism are carefully matched on gender, age and level of functioning (Criteria D, Table 16.1).

Details of studies designed specifically to compare psychopathology in individuals who have ID with and without autism are shown in Table 16.1. Each study has been evaluated according to the criteria above. As can be seen there is great variation in the extent to which these criteria are met for each study adding uncertainty to the interpretation of the findings.

However, apart from these methodological challenges there is another circumstance that is becoming more apparent when comparing autism and non-autism groups: that is, the continued practice of considering psychopathology in autism only through the lens crafted for understanding psychopathology in the general population. This limited perspective and its implications are explored in the next section.

Mental Distress Associated with the Condition of Autism

People with autism describe a different experience of the world we share, some referring to themselves as neuro-atypicals (Autism Network International, Williams and Jim Sinclair (http://www.autreat.com/History_of_ANI.html; http://www.autreat.com/)) compared to those without autism (neurotypicals). Significant differences in brain structures and neurobiological functioning in autism have been found (Baron, Groden, Groden, & Lipsitt, 2006; Courchesne, Webb, & Schuman, 2011; Minshew, Scherf, Behrmann, & Humphreys, 2011) that underpin these different perceptual and psychological experiences (Baron et al., 2006; Bogdashina, 2003, 2010). It can be anticipated that mental health disturbances in those with atypical neuro-functioning may be both different as well as similar to those with typical neuro-functioning. Understanding how people with autism experience the world differently from the general population may illuminate more accurately the nature of the mental health disturbances they experience, the extent to which these overlap and are different from the general population, as well as provide opportunities for more targeted interventions and treatment.

Processing differences and difficulties in social and communication domains, particularly where verbally mediated content is concerned (e.g. speech), are well documented (Groen & Buitelaar, 2011; Tager-Flusberg, Edelson, & Luyster, 2011). Not so well documented in mental health research in autism, are the specific hyper- and hyposensitivities to sensory input (e.g., touch, taste, smell, hearing and vision;

Bogdashina, 2003; Caldwell & Horwood, 2008; Grandin, 2006; O'Neill, 1998; Williams, 1999a). These different sensory sensitivities have been found to be pervasive, multimodal and persistent across age and ability in children and adults with autism (Billstedt, Carina Gillberg, & Gillberg, 2007; Leekam, Nieto, Libby, Wing, & Gould, 2007). Included in these sensory differences are hyposensitivities to proprioception, making it difficult for the individual to know what he/she is doing and hence to atypical motor patterns such as rocking, climbing, swinging, banging and walking on toes, to help restore a sense of body boundaries (Ayres, 1972; Ayres & Robbins, 2005; Caldwell & Horwood, 2007). Some individuals with autism meet criteria for sensory processing disorder (SPD), a proposed new disorder in DSM-5, characterised by persistent atypical over- or under-responsivity to neutral sensation (Sensory Processing Disorder (SPD) Scientific Work Group (SWG), 2007, Appendix A; 2008, Appendix B). Added to these different sensory experiences, there are autism-specific difficulties with regard to regulating behavioural responses to contingencies in the outside world: change, transition and choice being especially problematic in these regards (Caldwell, 2006; Caldwell & Horwood, 2007; Grandin, 1992a, 1992b; O'Neill, 1998; Robinson, 2011; Williams, 1996), as well as difficulties in regulating emotional responses to internal contingencies such as embarrassment and any form of emotional warmth (Caldwell, 2006; Caldwell & Horwood, 2007; O'Neill, 1998).

Inside-Out Experience in Autism

The inability of the brain to keep up with filtering and processing incoming sensory stimuli (including proprioceptive, emotional and cognitive perceptions) is described by those on the spectrum as 'overload'. When the resulting confusion becomes too overwhelming, the brain 'crashes', in what is termed the 'autonomic storm'. This experience and resultant behaviours are vividly described in a video simulation created by a woman with autism to help 'neurotypicals' understand how powerful, dramatic and debilitating this experience is [http://www.youtube.com/watch?v=BPDTEuotHe0]. As such, it draws attention to 'ordinary' environments and circumstances that are intolerable for many with autism; without this understanding their behaviour may be misinterpreted as to its aetiology and intent:

> …I process information at a slower pace than average. My brain is like a dial-up modem versus a cable modem. Non-autistic brains are like cable modems that take in several data packets at a time. My brain doesn't work that way. If you try to send it more data than it can handle, it crashes…… I hear everything four times louder than a non-autistic person … if I don't remove myself from the situation I immediately go into sensory overload.[1] To someone watching, I appear to cop an attitude or fly into a rage …. When I'm in overload I'm in a fight or flight response. Acting angry is a defense mechanism …once I am in full overload there is nothing I can do but ride the meltdown until I've expelled all my emotional energy. I just kinda lose my head and explode and then I'm fine …

Others with autism have also written about their experiences of the sometimes sliding scale between overload and the autonomic storm. Williams (1995), for example, describes floating images in her brain and 'running, running, running' to try and keep up. Gerland (2003) describes how as a child she experienced 'a constant shudder down my spine ……. It was like the sound of screeching chalk on a blackboard turned into a silent concentration of feeling, then placed in the back of my neck. From there, so metallic, the feeling radiated out into my arms, clipped itself into my elbows but never came to an end, never ever came to an end'. For Williams (1999a), sensations that began in the spine radiated even further, to her feet, 'like cracks in an earthquake'. She describes being in terror, 'it felt like death coming to get me… surprising how many times a

[1] There is some confusion in the exact use of the term 'overload'. Weird Girl Cindi is using it to refer to the condition that other authors refer to as 'Fragmentation', 'Meltdown', and more recently, in the light of increasing understanding of what is happening in the brain, 'The Autonomic Storm' (see Table 16.2 for further descriptions of this condition and experience of those with autism, pp. 245–251).

day I could be dying and still alive'. Descriptions by children include reference to 'my head running away', 'being in a car crusher' (see Table 16.2, for further descriptions of the experience of overload, pp. 245–251).

These experiences of fragmentation and sensory chaos accord well with the proposal by Ramachandran (2011) of a distorted salience landscape, involving overreaction by the autonomic nervous system (ANS) to (what may appear to a non-autistic person as) apparently 'trivial' incoming stimuli, triggering an autonomic storm accompanied by an increase in sweating and other responses of the body's self-defence system. The salience theory has been tested by measuring the Galvanic Skin Response (GSR) which showed a heightened response to a wide range of stimuli in children with autism compared with children not on the spectrum (Ramachandran, 2011).

Autism-specific differences in autonomic functioning and anxiety have been extensively investigated by Groden and colleagues (finding differences in attention, habituation, general arousal and cardiovascular response; Baron et al., 2006; Goodwin et al., 2006; Goodwin, Groden, Velicer, & Diller, 2007) and inferred in clinical descriptions of individuals who 'shut down' (Loos Miller & Loos, 2004; Loos & Loos Miller, 2004) or become catatonic (Dhossche et al., 2006; Fink et al., 2006; Wing & Shah, 2000) seemingly in response to life events they experience as overwhelming.

Some people on the autism spectrum describe their autonomic storm as initiated by an unusual or fizzy sensation in the back of the neck (Gerland, 2003; Williams, 1999b). They use words such as 'confusing', 'painful' and 'agonising' to describe its onset (further descriptions by authors with autism in Table 16.2, pp. 245–251). One man described the pain triggered by his hypersensitivity to light as 8–9 out of 10 compared to 4–5 caused by kidney stones (Caldwell; personal communication). Weeks (Undated) says of the autonomic storm, 'I would do anything to stop it, crash my head against the wall, run in front of a car'. Such symptoms, not infrequently described in metaphors that focus on immediate sensory experiences and visual imagery, run the risk of being misinterpreted as psychotic if these unique ways of communicating distress are not recognised (Howlin, 2004, Chap. 10; Palucka, Bradley, & Lunsky, 2008).

Barron and Barron (1992); Gerland (2003); Grandin and Scariano (1986); Jolliffe et al. (1992); Nazeer (2006); and Williams (1999a) also provide accounts of their response to these painful effects of sensory overload. Grandin (1992a, 1992b) reports 'By focusing on the spinning coin I could cut out other sounds'. When Gerland (2003) heard the sound of a scooter revving up she would lose all sense of where up and down were and had no idea where her feet were. She would seek proprioceptive reassurance, clinging to the railings in order to have a physical point of reference.

Nazeer (2006) describes how, when it became noisy in the pub, his friend Andre would spill some beer on the bar and doodle in it. Focusing on this activity enabled his friend to shut out the overloading chatter. Barron describes how, when he switched the lights on and off, it gave him a wonderful sense of security (Barron & Barron, 1992). He knew what he was doing and it was exactly the same each time. These repetitive patterns and clinging grabbing behaviours in those with autism should be seen as coping strategies against the potential threat of being overwhelmed. In the absence of being able to cut out the overloading sensory clutter, they essentially attempt to find coherence in a world marked by altered and distorted sensory and proprioceptive inputs (Nazeer, 2006; see also Table 16.2 for further descriptions of repetitive behaviours and stereotypies functioning as coping strategies, pp. 245–251).

The instinct of the non-autistic world is to try to limit these repetitive behaviours. However, if the individual is stopped from engaging with their point of focus, they run the risk of becoming overloaded by incoming sensory stimuli (Gillingham, 1998). Levels of agitation and distress rise. Misattribution of intentional states may occur if the non-autistic perspective fails to recognise that these are attempts by the individual to cope with their sensory distress in whatever way he/she can; behaviours otherwise considered as

inappropriate or even provocative, deliberate, aggressive and 'challenging' (e.g. increased grabbing and squeezing directed towards others in a desperate attempt to restablise perception of body boundaries) can result. As the perceived threat increases, sensory distortions become more urgent (see the video simulation previously mentioned) and the repetitive behaviours become more agitated. Other defensive strategies employed either contingently or in sequence, if previous attempts to reduce overload have failed, are those of the body's self-defence ANS response appearing as:

- Flight: avoidance such as shutting eyes, pulling clothes over head and turning or running away
- Fight: aggression towards the perceived offending circumstance such as others or objects in the environment or against self, the individual being unable to distinguish between what is or is not their own body
- Freeze: shutdown of everyday functions—either partial or verging towards catatonic-like states

These coping strategies overlap and the triggers are not always simple. For example, in terms of eye contact, eyes may be screwed up (1) to avoid light that is too intense and therefore painful; (2) to shut out patterns or colours that set off the zooming, swirls and breakup of visual intake; or (3) when the individual is with another person, where eye avoidance occurs associated with the severe pain that can be caused by hypersensitivity to the body's emotional responses ('emotional overload'). Jolliffe et al. (1992) write, 'people have absolutely no idea how painful it is for me to look at them'. Williams (1999a) simply calls it 'agony'.

As the brain shuts down on some of its functions, Williams (1996) describes how at times she could either see or feel or vice versa but not both at once. So she could either see her hand but not feel it as part of her or feel it but not see it. As far as she was concerned, it was just a thing floating in front of her and she kept trying to push it away. The experience of the brain shutting down completely is described by one child as how sometimes he can hear his teacher but sometimes everything disappears and he has no idea what is happening. He says his teacher does not believe him (Caldwell, personal communication). Finally, the whole body may simply refuse to move resulting in 'shutdown' (Loos Miller & Loos, 2004; Loos & Loos Miller, 2004) and catatonic-like states (Wing & Shah, 2000).

In people with autism these coping strategies in response to environments that are experienced as overwhelming, result in clinical presentations (e.g. repetitive behaviours) that overlap with behaviours (e.g. OCD) characteristic of psychiatric disorders seen in the general population (Table 16.2). Evidence from electrodermal studies of children with autism has highlighted the use of self-stimulation activities in order to calm hyperresponsive activity of the sympathetic ('fight or flight') branch of the ANS (Hirstein, Iversen, & Ramachandran, 2001). In order to ensure accurate psychiatric diagnosis (and hence appropriate targeted treatment), it is important to differentiate between symptoms that herald psychiatric disorder and autism-specific distortions resulting from sensory sensitivities and differences in brain processing and ANS system functioning. These two perspectives focusing on repetitive behaviours, stereotypies, anxiety disorders and behaviours associated with the flight, fight and freeze responses of the ANS are further elucidated in Table 16.2.

While these personal accounts are from more articulate people on the autism spectrum, similar repetitive compulsive behaviours and anxiety behaviours are common in those less articulate and lower functioning. Additionally, clinical interventions aimed at reducing overstimulating environments, attending to sensory issues and increasing coherence (e.g. familiar, predictable environments, planning around transitions, care providers using Intensive Interaction and Sensory Integration approaches), result in reduction of these behaviours in lower functioning individuals as they do for the more able on the spectrum (Caldwell & Horwood, 2008).

Summary

In this chapter our focus has been on (1) understanding psychopathology in autism and (2) determining whether the prevalence of

psychopathology in those with autism and ID is greater than those with ID alone.

1. Psychopathology in Autism

We have identified two different perspectives in understanding mental distress in autism—perspectives that touch directly on our understanding of clinical presentations, aetiology and consequently on treatment and prevention of mental health disturbances that arise. We refer to these two perspectives as (a) the 'Outside-In' approach, essentially applying DSM/ICD criteria to presenting symptoms and behaviours to make psychiatric diagnoses, and (b) the 'Inside-Out' approach, poignant descriptions by people who describe what it is like to live with sensory hypersensitivities, sensory integration deficits, brain processing problems and ANS dysregulation.

Importantly the Outside-In perspective infers psychopathology for some behaviours that people with autism actually describe as helping them to cope with sensory perceptual distortions and overload consequent to these autism-specific neurobiological differences in brain function. The Outside-In perspective derives from an evidence base of psychiatric disorder as this presents in the general population. Guidelines (also evidence based) as to how to treat these disorders have been developed from this perspective. However, if applied inappropriately to people with autism, treatments will be offered that may be inappropriate and indeed have serious outcomes (Esbensen, Greenberg, Seltzer, & Aman, 2009; Propper & Orlik, 2009).

The alternative Inside-Out perspective from people with autism is that the world picture (associated with neurobiological differences in autism) is totally chaotic. In an attempt to cut down on unfiltered sensory overload—and so make sense of their situation—the individual may (1) focus on a particular object or theme, idea or script (repetitive behaviour), (2) hide their eyes or run away (avoidance), (3) shut down the processing of one or more of the senses or freeze (catatonia) or (4) attack self or others (aggression). Underlying these behaviours is a desperate search for coherence (Table 16.2).

This Inside-Out perspective also has implications for the identification of those with autism who may (or may not) have ID. Many of these self-advocate authors describe difficulties at school and mistaken functioning and academic abilities which come to light only when their sensory needs are accommodated.

It is hard for those without autism and without the physical and psychological impacts of these hypersensitivities, sensory integration deficits and ANS dysregulation to understand the profound impact these have on all aspects of daily life; difficulties in perspective taking between those with and without autism may be more mutual than currently acknowledged. In relation to catatonia seen in autism, Hare and Malone (2004) report that existing labels may obscure an expression of the underlying autism condition, one which warrants a more in depth understanding of the individual's sensory, perceptual and neurocognitive functioning.

2. Psychopathology and the Impact of Autism Coexisting with ID

While separate studies of children and adult populations with ID (of which up to a third may have autism) and populations with autism (across functioning levels but mostly reported for those without ID) have shown increased psychopathology compared to the general population without these disabilities, it remains unclear whether autism coexisting with ID gives rise to greater psychopathology than ID alone. We have identified the need for (1) robust identification of autism and matching of autism and non-autism groups on variables known to impact on psychopathology and (2) careful identification of mental health disturbances that considers longitudinal understanding of patterns, as well as aetiology, of these disturbances. When these criteria are met, the evidence is pointing to greater psychopathology in children and adolescents where ID and autism are coexisting.

Meeting these research criteria for adults is more difficult and the picture is less clear. This may relate in part to less robust identification of autism in adult populations and therefore less accuracy in identifying ID groups with and without autism for comparison.

However, it is noteworthy that to date comparison studies of psychopathology in ID, with and without autism, have not considered the autism-specific differences we have described in this chapter, differences that would be anticipated to impact significantly on behaviour in everyday settings: for example, when reporting anxiety behaviours noting whether there are any autism-specific hypersensitivities and whether accommodations have been made to reduce exposure to distressing and sometimes painful 'everyday' sensory events.

Implications for Future Research on Psychopathology in ID with and Without Autism

Population-based studies, controlling for relevant variables and using robust screening and assessment tools, will determine whether there are differences in behaviours related to mental health between those who have ID with and without autism. Relevant variables already acknowledged in current matching studies (see Table 16.1 Criteria) include chronological age, gender and level of functioning (acknowledging that those with autism may function differently depending on whether verbal or non-verbal skills are being tested). Syndrome diagnosis is increasingly being recognised as relevant to specific types of mental ill health (Society for the Study of Behavioural Phenotypes (SSBP), 2011) and matching on these is recommended, though difficult in practice given the relatively low prevalence of these syndromes.

As noted, understanding the aetiology of any differences found between autism and non-autism groups is likely to be restricted if these behaviours are viewed only through the lens of our current understanding of mental health disorders (DSM criteria) in the general population. Assessment tools developed to measure autism-specific behaviours across the life span and across levels of functioning will more accurately identify those behaviours that are associated with autism and those that represent the onset of psychiatric disorder. The DISCO is one such tool; the use of this diagnostic approach resulted in the first systematic identification of (a) atypical sensory responses and found these to be present in over 90 % of those with autism (Billstedt et al., 2007; Leekam et al., 2007) and (b) autism-associated catatonia present in 17 % individuals (Wing & Shah, 2000). Neither of these conditions had been previously identified using tools developed from DSM criteria. We recommend that these neurobiological differences in autism (e.g. hypersensitivities) and consequent autism-specific vulnerabilities in 'ordinary' environments are taken into account when studying psychopathology in autism.

Clinical practice attests to effective outcomes in reducing anxiety, repetitive behaviours and catatonia when environments and supports that are offered acknowledge these autism-specific vulnerabilities and needs; in our clinical practice we refer to these environments as 'autism friendly'. Future research might evaluate the impact on mental health of implementing such autism-friendly environments and supports. Using an analogy to reading ability, the latter can only be accurately evaluated when the person who requires glasses to see the script is provided with glasses; likewise differences in 'psychopathology' (DSM, ICD criteria) may only be evident when the individual with autism is provided with appropriate supports and does not have to struggle so hard to make sense of their world and retreat into repetitive behaviours and fight, flight and freeze responses when these are not offered.

Finally, more formal involvement of self-advocates with autism in future mental health-related research might ensure that conceptualisation of mental health disorders in autism is not biased in favour of the non-autism perspective. This should include more efforts to understand the experiences of individuals with ID, not just those with high-functioning autism.

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Behavior Problems and Psychopathology

17

Robert D. Rieske and Johnny L. Matson

Introduction

It was once believed that individuals with intellectual disabilities (ID) were not capable of developing the same emotional disorders as the normal population. The term "dual diagnosis" is a relatively new term in the field of intellectual and developmental disabilities. Today we are well aware that those with ID can suffer from both psychiatric and behavioral disorders and that these individuals can suffer from the full range of current psychiatric disorders (Matson & Barrett, 1982). However, the prevalence rate of these disorders is not well understood and is clouded by differences and problems in sampling, assessment, and methodology. Regardless, studies have shown that those with ID suffer from a wide range of challenging behaviors including aggression, property destruction, and self-injurious behavior (Matson, Boisjoli, Rojahn, & Hess, 2009; Poppes, van der Putten, & Vlaskamp, 2010; Tsakanikos, Underwood, Sturmey, Bouras, & McCarthy, 2011). The presence of comorbid psychiatric disorders has also been found to occur at a higher rate in individuals with ID as well as those with challenging behaviors (Gustafsson & Sonnander, 2004; Iverson & Fox, 1989; Rojahn & Meier, 2009; Rutter, Tizard, Yule, Graham, & Whitmore, 1976).

Prevalence Rates of Challenging Behaviors

The presence of challenging behaviors in individuals with ID is not a new concept. However, the way that we view and approach treatment of challenging behaviors, and their relation to comorbid psychopathology, is a relatively novel idea in the body of research. Determining the prevalence rates of challenging behaviors is important to understand how they develop and function in this particular population. In one study of individuals with profound ID, the prevalence rate of challenging behaviors was as high as 82 % displaying at least one challenging behavior (Poppes et al., 2010). Self-injurious behavior, defined in this study as refusing food, hitting head, biting self, and grinding teeth, was found to occur in 82 % of the sample. The same percentage was also found to display stereotypical behavior defined in this sample as screaming and shouting as well as repetitive hand movements. Aggressive and destructive behavior, such as pulling and grabbing others or pinching others, was found to occur in 45 % of the sample. It was reported that all of these behaviors occurred on a frequency ranging from hourly to monthly but that aggressive behaviors occurred more on a

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weekly basis, whereas stereotypical and self-injurious behaviors occurred more on a daily basis. However, within this population of individuals with profound ID, it was also found that caregivers rated these challenging behaviors as being low in severity with very few individuals engaging in severe challenging behaviors (Poppes et al., 2010).

The prevalence rate of challenging behaviors has generally been researched in populations of individuals with ID that are institutionalized, living in a residential care center, or attending a special school. Few research studies have viewed challenging behaviors according to the total population. One such study was conducted by Qureshi and Alborz (1992) in England among a population of approximately 4,200 adults receiving services for ID. Qureshi and Alborz reported that about 16.7 % of the population showed some type of problem behavior and that 7 % exhibited more severe problems. They also mentioned that the prevalence varied according to socioeconomic status and other demographic characteristics of the seven different districts included in the study.

Emerson et al. (2001) looked to extend these results using much of the data from the Qureshi and Alborz (1992) and other related studies and collecting additional data from some of the original districts. They reported similar prevalence rates with 10–15 % of the population exhibiting some type of challenging behavior. They examined challenging behaviors according to four categories, including aggression, destructive behavior, self-injurious behavior, and other behaviors. The majority of those with challenging behaviors exhibited two or more of the general categories of challenging behaviors, and these individuals were more likely to be males and adolescents or young adults.

Prevalence Rates of Psychopathology

Early studies of comorbid psychopathology have attempted to accurately assess rates of psychiatric disorders in individuals with ID. Rutter and colleagues reported psychopathology rates at 4–5 times that of children with normal intelligence (Rutter et al., 1976). The identification of psychopathology in individuals with ID has proven to be more difficult than in the general population. Prevalence rates of psychiatric disorders in individuals with ID have ranged anywhere from 15.7 % (Cooper et al., 2007) to 54 % (Gustafsson & Sonnander, 2004) depending on differences in sampling, definition and classification of psychiatric disorders in the population, and use of screening and assessment tools (Rojahn & Meier, 2009). There is still much work to be completed in the assessment of the prevalence rate of specific psychiatric disorders in individuals with ID. New tools and assessments are being created and researched to help with this endeavor, and as research progresses, the field of intellectual and developmental disabilities should strive to reach a consensus on the definition and classification of such disorders including appropriate diagnostic criteria.

Interaction of Challenging Behavior and Psychopathology

It is important to understand that challenging behaviors are not simply symptoms of psychopathology or ID but are distinct disorders (Sturmey, Laud, Cooper, Matson, & Fodstad, 2010a, 2010b; Tsiouris, Mann, Patti, & Sturmey, 2003). Past literature has described challenging behaviors as "behavioral equivalents" of psychiatric disorders which are described as atypical manifestations of the disorder in individuals with ID (Sovner & Hurley, 1982a, 1982b). While it is generally recognized that the manifestation of psychiatric disorders is different than in the general population, it has not been shown that challenging behaviors are symptoms of specific psychiatric disorders except in the case in which behaviors are included in the criteria of the disorder (e.g., repetitive and stereotyped behaviors in autism).

In one study, Tsiouris et al. (2003) found that challenging behaviors such as self-injury and aggression were not predictors of, nor associated with, symptoms of depression and therefore

should not be used in the diagnosis or treatment of depression in those with ID. These findings were supported by a larger study assessing behavioral equivalents of depression in a sample of 693 adults with ID. They found no behavioral equivalents for symptoms of depression and therefore advise against the use of maladaptive behavior rating scales in the diagnosis of depression in those with ID (Sturmey et al., 2010a). A factor analysis completed in the same study showed that symptoms of depression and challenging behaviors were loaded onto separate factors of the *Diagnostic Assessment for the Severely Handicapped-II* (*DASH-II*). Although some researchers have found associations between challenging behavior and psychopathology, there is little empirical evidence supporting the claim that challenging behaviors are behaviorally equivalent to psychiatric symptoms (Rojahn & Meier, 2009). Rojahn and Meier (2009) suggest that although challenging behaviors may be nonspecific indicators associated with psychopathology, they are not symptoms of a specific psychiatric disorder.

McCarthy et al. (2010) found that of those with an ID (with or without comorbid autism spectrum disorders [ASD]), presence of challenging behaviors was not significantly different between males and females, the young and old, or between those with or without a psychiatric diagnosis. Felce, Kerr, and Hastings (2009) reported that studies have generally shown that the presence of challenging behaviors in adults with an ID is associated with higher rates of psychiatric disorders. To determine if the association found in previous studies was artificial or not, they conducted a study controlling for presence of an ASD and adaptive behavior functioning in adults with ID. They reported that challenging behaviors were significantly higher among participants in the psychiatric group. Further, the level of adaptive functioning acted as a moderator in such a way that those with lower adaptive abilities displayed greater frequency and severity of challenging behaviors.

In a recent study completed by Kearney and Healy (2011), they reported several findings regarding the relationship between challenging behaviors and psychopathology. They reported that those with severe challenging behaviors displayed greater psychiatric symptoms than those without challenging behaviors on the following subscales of the *DASH-II* (Matson, 1995): anxiety, mood, mania, impulse, organic, PDD/autism, schizophrenia, stereotypies, SIB, and eating. However, this study did not examine specific challenging behaviors and how they relate to specific disorders but rather simply observed the relationship between more severe challenging behaviors and greater psychiatric symptoms.

In a study by McCarthy et al. (2010) of adults with an ID with or without a comorbid ASD diagnosis, they found that of those with challenging behaviors, individuals with an ASD were significantly different than those without an ASD in relation to psychiatric symptoms. Interestingly, those with an ASD diagnosis had lower rates of psychiatric disorders. They found, as has been shown by previous research, that those with an ASD diagnosis tended to be male, younger, and have lower intellectual functioning. Due to this finding, the authors examined their results while controlling for ID, gender, and age. The presence of challenging behaviors was independent of psychiatric symptoms. Only ASD and severe ID were shown to be reliable predictors of challenging behaviors.

Much of the research examining the relationship of challenging behaviors and psychopathology has not looked at specific behaviors or disorders. In a study conducted by Jenkins, Rose, and Jones (1998), they examined differences between aggressive and non-aggressive challenging behaviors. Comparisons between measures of challenging behavior and measures of psychopathology in those with ID suggest more of an overlap of non-aggressive challenging behaviors and comorbid psychopathology than aggressive behaviors. In other words, even though aggressive behaviors have been linked to psychopathology, these behaviors also occur concurrently with a host of other problems related to their ID (e.g., lack of social skills, adaptive skills, and coping skills). In one recent study, researchers found that children diagnosed with an ID and/or ASD suffered from significantly more sleep problems and

anxiety and displayed more challenging behaviors than typically developing children (Rzepecka, McKenzie, McClure, & Murphy, 2011). In this study, they also found that a large percentage of the variance (42 %) in challenging behaviors could be accounted for based on medication, sleep problems, and anxiety. Jenkins et al. (1998) have even reported that non-aggressive challenging behaviors were more predictive and characteristic of a comorbid mental disorder. Felce et al. (2009) also reported that challenging behaviors were more common in those with psychiatric problems and that this relationship was more prominent in those with more severe ID.

The Role of Social Skills

Social skills play an important part in the relationship of challenging behaviors and psychopathology in individuals with ID. In one recent study, Kearney and Healy (2011) reported that those with severe challenging behaviors displayed greater deficits in social skills than those without challenging behaviors as measured by the *Matson Evaluation of Social Skills for Individuals with Severe Retardation* (*MESSIER*). Kearney and Healy (2011) also reported that individuals who presented with comorbid psychiatric problems (as measured by the *DASH-II*) also had greater deficits in social skills than those without. This finding may suggest that deficits in social skills, which are common components of developmental and intellectual disabilities, increase the likelihood of challenging behaviors, which may serve as a maladaptive way of communicating.

In adults with ID, Matson, Fodstad, and Rivet (2009) found that social behaviors, as measured by the *MESSIER*, were predictive of some types of challenging behaviors including aggression/destruction and total behavior problems. It was found that positive nonverbal social behaviors, as well as general positive and negative social behaviors, were more predictive of these challenging behaviors. The authors also reported that in adults with a comorbid ASD, the presence of negative behaviors was more predictive of challenging behaviors (specifically aggression, property destruction, and stereotypies) than the absence of positive social skills.

Autism Spectrum Disorders

ASDs are a common comorbid disorder among persons with ID. People with an ASD also exhibit higher rates of challenging behaviors. Due to this fact, it is important to discuss ASDs and the role they play in the ID population. In a recent study by McCarthy et al. (2010) of adults with ID with or without a comorbid ASD diagnosis, they found that those with a comorbid ASD diagnosis were approximately 4 times more likely to display challenging behaviors as compared to those without an ASD. This finding was also supported by Matson, Fodstad, et al. (2009) who reported that in a study of adults with severe ID, those with comorbid ASD displayed more challenging behaviors than those with an ID alone. Additionally, Matson and Rivet (2008) reported that those diagnosed with autistic disorder displayed more challenging behaviors than people with ID alone but that individuals with pervasive developmental disability not otherwise specified (PDD-NOS) played an intermediary role between the two in terms of severity of challenging behaviors as well as the number of concomitant types of challenging behavior.

For children, researchers found that those with an ASD evince greater levels of challenging behaviors than both typically developing children and atypically developing children with a current Axis I disorder (Matson, Wilkins, & Macken, 2009). They also reported that more than 94 % of their sample of children with ASD displayed some type of challenging behavior and that challenging behaviors were positively correlated with severity of autism symptomology; in other words, those with more severe presentations of autism displayed higher rates of challenging behaviors.

It has been commonly believed that individuals with ASD and comorbid ID may have higher rates of psychopathology than individuals with ID alone. Some studies have shown higher rates of depression in individuals with comorbid

autism as compared to those without (Morgan, Roy, & Chance, 2003). However, in a study of 752 adults with or without comorbid autism, it was reported that those with autism were no more likely to receive a psychiatric diagnosis than those without autism and, they were in fact, less likely to receive a personality disorder diagnosis (Tsakanikos et al., 2006). Even though the research regarding psychopathology in this population with comorbid autism has shown no differences, these individuals still tend to receive more psychiatric diagnoses and psychotropic medications for the treatment of challenging behaviors (Sturmey, Seiverling, & Ward-Horner, 2008).

Physical Disabilities and Challenging Behaviors

It is also important to consider other contributing factors to challenging behaviors in addition to psychopathology. Medical and physical variables are important to consider and should be accounted for in an assessment of challenging behaviors. Recently, a group of researchers conducted a systematic review of the literature to determine possible links between specific physical conditions and challenging behaviors. The researchers reported that challenging behaviors were directly related to enuresis, pain related to cerebral palsy, and chronic sleep problems (de Winter, Jansen, & Evenhuis, 2011). The authors also found that the specific challenging behavior of self-injury was associated with visual impairments. This was also supported by other researchers in a study examining rates of challenging behaviors in individuals with profound ID. They reported that individuals with visual, tactile, and psychiatric problems exhibited higher rates of challenging behaviors (Poppes et al., 2010). de Winter et al. (2011) reported that variables not found to be a factor included hearing impairments, bowel incontinence, mobility impairments, and epilepsy. The authors also noted that research regarding the association between challenging behavior and physical conditions is lacking and many of the existing studies lack methodological rigor (de Winter et al., 2011).

Assessment of Challenging Behaviors and Psychopathology

The assessment of challenging behaviors and comorbid psychopathology is an important component of psychological evaluations and treatment development. Although research regarding comorbid psychiatric disorders and challenging behaviors is in its infancy for this population, several tools exist that have extensive research supporting their validity and utility. The following are several assessment tools that have been developed for use with this population.

Psychopathology Instrument for Adults with Mental Retardation

The first measure designed to assess for comorbid psychopathology in those with ID, the *Psychopathology Instrument for Adults with Mental Retardation (PIMRA)*, was introduced in 1983 (Kazdin, Matson, & Senatore, 1983). The *PIMRA* is a brief interview which also included a caregiver report to address the issue of inaccurate or difficult self-reporting common in those with ID. The *PIMRA* is a 56-item test based on the DSM-III which measures symptoms of schizophrenia, depression, psychosexual disorders, adjustment disorders, anxiety, somatoform disorders, and personality disorders. The *PIMRA* is a measure that has been highly studied (Matson, Belva, Hattier, & Matson, 2012) and has proven psychometric properties (Matson, Kazdin, & Senatore, 1984). Several studies have also been published examining the validity of the *PIMRA* (Masi, Brovedani, Mucci, & Favilla, 2002; Sturmey & Bertman, 1994; Swiezy, Matson, Kirkpatrick-Sanchez, & Williams, 1995). The *PIMRA* has been widely used across the world and has been translated into several languages including Swedish (Gustafsson & Sonnander, 2005), Dutch (van Minnen, Savelsberg, & Hoogduin, 1994), as well as Italian, Spanish, and Norwegian (Matson, Belva, et al., 2012).

Developmental Behavior Checklist

The developmental behavior checklist (*DBC*) (Einfield & Tonge, 1995) is a measure designed specifically for the assessment of behavioral and emotional problems of children ages 4–18 with developmental and intellectual disabilities. The *DBC* now has several different versions including a parent, teacher, and adult version as well as a short form and daily monitoring form. The *DBC* has a wide research base supporting its use and psychometric properties (Dekker, Nunn, & Koot, 2002; Mohr et al., 2011). It has also been translated into several languages around the world including German (Steinhausen & Metzke, 2011), Finnish (Koskentausta & Almqvist, 2004), as well as Chinese, Spanish, Italian, and multiple other languages. The *DBC* parent and teacher versions include five subscales including disruptive/antisocial, self-absorbed, communication disturbance, anxiety, and social relating. The adult version has a different factor structure including subscales of disruptive, self-absorbed, communication disturbance, anxiety/antisocial, social relating, and depressive scales. One of the advantages of the *DBC* is that it can be used to continually track behaviors to observe changes and effectiveness of behavior treatment plans and can be used throughout the life span (Mohr et al., 2011).

Assessment for Dual Diagnosis

The assessment for dual diagnosis (*ADD*) (Matson & Bamburg, 1998) is a measure which assesses for symptoms of comorbid psychopathology in individuals with mild and moderate ID. The measure consists of 79 items rated by the frequency of the behavior in the previous 2 weeks. Ratings for duration and severity are also collected for any items endorsed as occurring on the frequency scale. The measure includes 13 subscales based on DSM-IV criteria of mental disorders including symptoms of mania, depression, anxiety, post-traumatic stress disorder, substance abuse, somatoform disorders, dementia, conduct disorder, pervasive developmental disorders, schizophrenia, personality disorders, eating disorders, and sexual disorders. The measure is a parent/caregiver report that can be conveniently administered during an interview. The *ADD* was designed as a screening instrument and should be used by clinicians in conjunction with behavioral observations, additional assessments, and clinical judgment. The *ADD* can assist a clinician in identifying clusters of symptoms of psychiatric disorders to aid in differential diagnosis and alert the clinician to areas which may require further assessment. The *ADD* has been shown to have good psychometric properties (Matson & Bamburg, 1998) including good concurrent validity with the *Social Performance Survey Schedule* (Matson, Anderson, & Bamburg, 2000) and validation of the depression subscale against the *ADAMS* Depressed Mood subscale (Esbensen & Benson, 2006).

Diagnostic Assessment for the Severely Handicapped-II

Much like the *ADD*, the *DASH-II* (Matson, 1995) is a measure which assesses for symptoms of comorbid psychopathology in individuals with ID. Unlike the *ADD*, the *DASH-II* has been designed for use with individuals with severe and profound ID. The *DASH-II* is an 84-item measure that is administered to a parent or caregiver during an interview. The *DASH-II* rates behaviors occurring over the last 2 weeks and includes eight separate subscales including impulse disorders, organic, anxiety, mood, mania, pervasive developmental disorder/autism, schizophrenia, and stereotypies. The measure also includes five additional subscales for self-injurious behavior, elimination disorders, eating problems, sleep problems, and sexual disorders. The *DASH-II* is designed to be used in conjunction with direct observation and additional assessments to help clinicians in the differential diagnosis of comorbid psychiatric disorders.

The *DASH-II* has been reported to have strong psychometric properties (Matson, 1995). The reliability and validity of individual subscales were completed over several different studies.

Matson and Smiroldo (1997) examined the psychometric properties of the mania subscales of the *DASH-II* showing good convergent validity with the DSM-IV criteria, correctly classifying 90.9 % of manic individuals and 100 % of controls. Other subscales were also assessed by examining characteristics of individuals with autism (Matson et al., 1996), stereotypies and self-injurious behavior (Matson et al., 1997), depression (Matson et al., 1999), and other subscales. The *DASH-II* has also been validated against the *Aberrant Behavior Checklist* (Paclawskyj, Matson, Bamburg, & Baglio, 1997), the *Young Mania Rating* scale (Laud & Matson, 2006), and several other assessments. Overall, the *DASH-II* is a widely used and extensively studied assessment to assist in the identification and differential diagnosis of comorbid psychiatric disorders in those with severe and profound ID.

Nisonger Child Behavior Rating Form

The Nisonger child behavior rating form (*NCBRF*) (Aman, Tassé, Rojahn, & Hammer, 1996) is another measure which includes 66 items on challenging behaviors and 10 items on social competence. The measure is a parent/teacher report that can be completed in a short amount of time and requires little training to administer. The social competence scale measures social behaviors that have occurred in the last month and are broken into two subscales including compliance/self-control and positive/adaptive. The problem behavior scale measures behaviors that have occurred in the last month and include five factors: antisocial behavior/defiance, hyperactivity/inattention, withdrawal/depression, negative self-image/self-injury, and anxiety.

The psychometric properties of the *NCBRF* were examined by Aman et al. (1996) in a study of 369 children with ID and reported high internal and inter-rater reliability. Additionally, the measure has been translated into French for use in a Canadian population (Tassé, Morin, & Girouard, 2000) and has been used in antipsychotic drug treatment outcome studies (Aman, De Smedt, Derivan, Lyons, & Findling, 2002; Snyder et al., 2002) and multiple other studies. However, much of its available research has been used for similar drug treatment studies and lacks research supporting its clinical utility.

Reiss Screen for Maladaptive Behavior and the Reiss Scales for Children's Dual Diagnosis

The *Reiss Screen for Maladaptive Behavior* (Havercamp & Reiss, 1997; Reiss, 1988) was the second instrument created for assessing comorbid psychiatric disorders in adolescents and adults with ID. The measure is a 36-item informant-based rating scale that is administered to parents, caregivers, teachers, etc. The measure includes eight subscales including items measuring aggressive behavior, autism, psychosis, paranoia, behavioral signs of depression, physical signs of depression, dependent personality disorder, and avoidant personality disorder. The measure includes national norms for differing levels of ID from mild to severe.

The *Reiss Scales for Children's Dual Diagnosis* (Reiss & Valenti-Hein, 1994) is another measure created for the assessment of comorbid psychopathology in children and adolescents with ID. It is a 60-item child and adolescent version of the *Reiss Screen* and was created to fill the need of measures designed for use with children. The measure consists of ten subscales including anger/self-control, anxiety disorder, attention deficit, autism, conduct disorder, depression, poor self-esteem, psychosis, somatoform behavior, and withdrawn/isolated.

The *Reiss Screen for Maladaptive Behavior* and *Reiss Scales for Children's Dual Diagnosis* are both widely used and well-established measures of dual diagnosis. Both measures are meant to be used as screening tools and were introduced in a time that lacked appropriate scales for challenging behaviors and comorbid psychopathology in those with ID. The measures have since been translated into several languages, and additional research regarding psychometric properties has

been conducted (Havercamp & Reiss, 1997; Prout, 1993; Reiss, 1988, 1990; Reiss & Valenti-Hein, 1994) including comparisons with the *Aberrant Behavior Checklist* and other related measures (Matson, Belva, et al., 2012).

The Psychiatric Assessment Schedule for Adults with a Developmental Disability

The psychiatric assessment schedule for adults with a developmental disability (*PAS-ADD*) (Moss et al., 1993) was designed for use with adults with an ID to assess for comorbid psychopathology based on criteria from the ICD-10. The *PAS-ADD* is comprised of a three-tiered interview consisting of the *PAS- ADD Checklist*, the *Mini PAS-ADD*, and the *PAS-ADD 10 Clinical Interview*. The revised version of the *PAS-ADD Checklist* (Moss, 2002) is composed of 25 items which are used to produce three subscale scores measuring affective or neurotic disorder, possible organic condition, and psychotic disorder. Multiple psychometric studies have been completed (Moss et al., 1998; Sturmey, Newton, Cowley, Bouras, & Holt, 2005) including psychometrics of a German translation of the *PAS-ADD Checklist* (Zeilinger, Weber, & Haveman, 2011). The checklist is meant to be used as a screener with elevations warranting further assessment with its counterparts.

The *Mini PAS-ADD* is a shortened version of the *PAS-ADD 10 Clinical Interview*. Consisting of 86 items, the Mini PAS-ADD assesses for psychiatric symptoms of depression, anxiety, expansive mood, obsessive-compulsive disorder, psychosis, autism spectrum disorder, and other unspecified disorders (e.g., dementia and organic problems). The most recent version of the *PAS-ADD Clinical Interview* (*PAS-ADD 10*; Costello, Moss, Prosser, & Hatton, 1997) is based on criteria of the ICD-10 and has made necessary improvements over earlier versions including better rating of sleep problems and an additional module to measure psychotic symptoms. It is a semi-structured interview which assesses for a broad range of psychiatric disorders and is the most comprehensive of the three assessments.

The *PAS-ADD* is a highly researched battery of assessments that have been used in multiple studies of psychopathology in individuals with ID. It has been translated and studied in several languages including German (Zeilinger et al., 2011) and Norwegian (Borge & Gitlesen, 2003) and has proven adequate psychometric properties (Matson, Belva, et al., 2012).

Other Measures

Other measures have been used to assess challenging behaviors in children and adults with ID. One example is Achenbach's *Child Behavior Checklist* (CBCL; Achenbach & Rescrola, 2000). This is a commonly used measure in the field of child psychology in the assessment of challenging behavior and psychopathology. However, it was not designed for use with individuals with ID and has only recently been researched for such a use. Studies using the CBCL in this population have not validated its use in assessment of psychopathology and have shown poor psychometric properties (Embregts, 2000) and a different factor structure than use with the typical population (Borthwick-Duffy, Lane, & Widaman, 1997). The same could also be said for the *Adult Behavior Checklist* (*ABCL*; Achenbach & Rescrola, 2003). The measure was not created for use with individuals with ID. However, in a recent study investigating the utility of the *ABCL* in the assessment of psychopathology in those with ID, Tenneij and Koot (2007) found that the *ABCL* was a reliable and possibly valid measure in the assessment of psychopathology in those with mild ID. Nevertheless, research regarding the use of this measure in such a population is lacking (Matson, Belva, et al., 2012) and requires increased empirical support before use in such a population could be justified.

Autism-Specific Measures

It is also important to consider other measures created for specific comorbid diagnoses. One of the most common and debilitating disorders among those with ID is ASD. As mentioned

earlier, these individuals are also at increased risk for challenging behavior and possibly certain types of psychiatric disorders. The dual nature of autism and ID further clouds the diagnosis of additional psychiatric disorders and requires measures designed specifically for use with those diagnosed with an ASD. Two such measures that assess for comorbid psychopathology and challenging behaviors in those with an ASD are the *Baby and Infant Screen for Children with aUtIsm Traits* (*BISCUIT*; Matson, Boisjoli, & Wilkins, 2007) and the *Autism Spectrum Disorder Battery* for both children (*ASD-Child*; Matson & Gonzalez, 2008) and adults (*ASD-Adult*; Matson, Terlonge, & Gonzalez, 2006). Each of these batteries includes three separate measures for the assessment of autism symptomology, comorbid psychopathology, and challenging behaviors.

BISCUIT

The *BISCUIT* (Matson, Boisjoli, et al., 2007) is an assessment battery designed for use with infants between the ages of 17 and 37 months of age. The first section of the *BISCUIT* (Part 1) is a diagnostic measure, which will not be discussed in this chapter (see Matson, Boisjoli, Hess, & Wilkins, 2010; Matson, Wilkins, & Fodstad, 2011; Matson, Wilkins, Sevin, et al., 2009 for further discussion).

Part 2 of the *BISCUIT* is a measure of comorbid problems common in those with an ASD. The measure consists of 57 items comprising five subscales including tantrum/conduct behavior, inattention/impulsivity, avoidance behavior, anxiety/repetitive behavior, and eating/sleep problems. Part 3 of the *BISCUIT* is a measure of behavior problems that are common in those with an ASD. The measure consists of 15 items comprising 3 subscales including aggressive/destructive behavior, stereotypies, and self-injurious behavior. Both measures have strong reported psychometric properties (Matson, Wilkins, Sevin, et al., 2009), and over 80 studies have been published on the measure at this point. These measures are meant to be used as screeners in an interview setting with parents or caregivers. If an individual has significant elevations on any of the given scales, then further assessment may be necessary to examine specific deficits and comorbid problems.

ASD-Child

The *ASD-Child* battery (Matson & Gonzalez, 2008) is a comprehensive assessment which includes three separate measures including a measure of autism symptomology (*ASD-Diagnostic-Child*), comorbid psychopathology (*ASD-Comorbidity-Child*), and problem behavior (*ASD-Problem Behavior-Child*). The battery was designed to be used with children between the ages of 2–18 years of age and is completed independently by parents or caregivers. The first section of the *ASD-Child* battery is the diagnostic measure (*ASD-DC*) and will not be discussed in this chapter (see Matson, Mahan, Hess, Fodstad, & Neal, 2010 for further discussion).

The comorbidity measure of the *ASD* (*ASD-CC*) is a 39-item measure that is comprised of seven subscales which measure tantrum behavior, repetitive behavior, worry/depressed behavior, avoidant behavior, under-eating, conduct problem, and overeating. The *ASD-CC* is one of the first measures designed specifically for assessing comorbid psychopathology in children with an ASD (Matson & Nebel-Schwalm, 2007) and therefore helps clinicians in determining if psychiatric symptoms are better accounted for by an ASD or comorbid psychopathology. The challenging behavior measure (*ASD-PBC*) has two subscales measuring externalizing and internalizing behaviors consisting of 18 items. The measure includes items of aggression, disruptive behavior, self-injurious behavior, and stereotypic behavior. This measure was also designed to be used as a screener specifically for children with an ASD diagnosis and should be used in conjunction with other assessments and clinical observations.

ASD-Adult

The *ASD-Adult* battery (Matson et al., 2006) also includes three distinct measures including diagnostic (*ASD-DA*), comorbidity (*ASD-CA*),

and problem behavior (*ASD-PBA*) scales. Much like the name suggests, the *ASD-Adult* battery is designed to be used with individuals 18 years of age and older. The *ASD-Adult* battery is a parent or caregiver informant-based measure and, in the case of residential or community placement, should only be administered to caregivers that have experience working with the individual for a minimum of 6 months. The diagnostic scale (*ADS-DA*) measures symptoms of autism and will not be discussed in this chapter (see Matson, Wilkins, & Gonzalez, 2007 for further discussion).

The comorbidity scale (*ASD-CA*) is a 37-item measure that includes five subscales measuring anxiety/repetitive behaviors, conduct problems, irritability/behavioral excesses, attention/hyperactivity/impulsivity, and depressive symptoms. The problem behavior scale (*ASD-PBA*) is a 19-item measure that includes four subscales including measures of self-injurious, aggressive/destructive, disruptive, and stereotypic behaviors.

Functional Assessment of Challenging Behaviors

Once a target challenging behavior has been identified, either through checklists, caregiver interviews, or direct observations, it is important to determine the antecedents and maintaining factors of the behavior. A functional assessment is necessary to determine why a behavior occurs and what is reinforcing the maladaptive behavior. Several options are available for completing a functional assessment. A functional assessment may include a brief functional analysis through checklists or caregiver interviews, direct observation and tracking of challenging behaviors, and possibly an experimental functional analysis (EFA). Each of these assessment methods has its own pros and cons and should be used with careful consideration by the clinician depending on the situation and presentation of challenging behaviors.

Brief Functional Analysis

A brief functional analysis generally includes the use of some type of checklist or caregiver interview based on direct observations to help determine the function of a behavior. Several tools exist to help in this endeavor including the *Motivational Assessment Scale* (Durand & Crimmins, 1989), the *Behavior Problems Inventory* (Rojahn, Aman, Matson, & Mayville, 2003), and the *Questions About Behavioral Function* (*QABF*; Matson & Vollmer, 1995). These types of scales are based on research from EFA discussed in the following section. Brief functional analysis assessments generally require less time and training than EFA and are usually more cost-effective. The most widely studied of these instruments at this time is the *QABF* (Matson, Tureck, & Rieske, 2012).

Questions About Behavioral Function

The *QABF* was developed by Matson and Vollmer (1995). Since then there have been two other forms that have been developed and published including a short form (*QABF-SF*; Singh et al., 2009) and a form to be used with severe mental illness (*QABF-MI*; Singh et al., 2006). The *QABF* was developed as a way to overcome the time-intensive method for identifying behavioral function of EFA. The *QABF* consists of 25 items that measure five factors based on behavioral functions including attention, escape, nonsocial, physical, and tangible. The *QABF* is a parent/caregiver report conducted during an interview based on direct observations of the informant. This requires that the informant has known the individual and is familiar with their challenging behaviors for a minimum of 6 months. The target behavior must be operationally defined according to the method set out by Kazdin (1985) and should contain inclusion and exclusion criteria for the behavior.

The *QABF* has been shown to have strong psychometric properties (Paclawskyj, Matson,

Rush, Smalls, & Vollmer, 2000). In a study of behavior functions, the results of the *QABF* were found to be more analogous to EFA results than the results of the *Motivation Assessment Scale* (*MAS*; Paclawskyj et al., 2000). Additionally, Hall (2005) found the *QABF* and EFA data to be more similar than descriptive methods (observational data) in a study of pica, self-injury, and aggression.

Experimental Functional Analysis

EFA is a clinician-intensive process in which mediating variables of target challenging behaviors are manipulated to determine which factors are maintaining the behavior. This process is broken down into multiple sessions involving variable manipulations and reversals in the natural environment and could take several weeks to complete (Iwata, Vollmer, & Zarcone, 1990). EFA involves the experimental manipulation of attention, access to tangibles, opportunities to escape unwanted tasks, etc. EFA can be useful in many settings to determine a causal relationship that can be experimentally tested between different functions and challenging behaviors. However, there are several things to consider before conducting an EFA with an individual exhibiting challenging behavior.

There are several instances in which conducting an EFA might not be appropriate or safe. First, an EFA should not be used for challenging behaviors that have a low frequency of occurrence (Matson & Minshawi, 2007). Due to the nature of EFA, it would be improbable to determine the function of a behavior which only occurs once per day or even a few times per day. EFA is really only useful for behaviors with a high frequency that can be tested and manipulated several times in a session. Second, the risks involved in the manipulations of behavioral functions should be considered. For example, if an individual displays a certain behavior as an escape from demanding or unwanted tasks but is given a desired tangible or attention, it is possible that the individual may quickly make that connection and attempt to use the challenging behavior in the future to gain access to desired tangibles or attention. Therefore, the behavior now has multiple functions which can complicate behavioral treatment and may cause undue stress to the individual as well. Third, the safety of both the individual and the assessor should also be considered. EFA may not be appropriate for aggressive behaviors and self-injurious behaviors. Due to the nature of EFA, it is required that the behavior occurs without interruption in order to manipulate the functions and therefore may exclude attention given during an intervention. In certain situations, it would not be safe, and possibly unethical, to allow an individual to engage in a challenging behavior that causes harm to self or others. Finally, due to the large amount of resources required for completing an EFA (training, time, reinforcers, etc.), it may not be an economically viable option for many programs and clinicians.

Economic Concerns (EFA vs. BFA)

The training involved in completing an EFA is also very time intense and costly. Because of the time and cost constraints on many clinicians, this method has been proven to be inefficient for determining behavioral functions for the majority of clients in developmental centers and similar settings. Brief functional assessments have been shown to be more time and cost-effective, require less training, and are as accurate (if not more accurate) in determining functions of behavior than EFA (Hall, 2005).

Tarbox et al. (2009) found that brief functional analysis methods (e.g., *QABF*) and EFA both produced relatively conclusive functions of behaviors for all of their children exhibiting challenging behaviors as compared to descriptive assessment methods (e.g., tracking of frequency, intensity, and duration as well as perceived behaviors, antecedents, and consequences). Although the two methods did not always agree, there was more agreement between the *QABF* and EFA than either had with descriptive assessment methods (Tarbox et al., 2009). Therefore, due to the resources

necessary for completing an EFA and research showing the effectiveness of brief functional analysis methods such as the *QABF*, it may be more intuitive to use a brief functional analysis method for most cases of functional analysis and turning to EFA for more complex cases in which the *QABF* and similar assessment tools cannot determine the function of a behavior. EFA may also be warranted if behavioral treatments based on functions of behavior have continued to be unsuccessful.

Behavior Support Plans

A Behavior Support Plan (BSP) is an important part of treatment and ongoing assessment of challenging behaviors in individuals with ID. BSPs have been utilized in several different settings including developmental institutions, group homes, and even home-based supports. In order to have a successful and appropriate BSP, it is imperative that a proper assessment of challenging behaviors has been completed which identifies the function of the challenging behaviors. Once the function of the behaviors has been identified, appropriate prevention and intervention strategies can be created and implemented as well as functionally equivalent replacement behaviors. Although prevention and intervention strategies are an important and critical part of a BSP, appropriate and functionally equivalent replacement behaviors are the key to a successful treatment plan. This feature is also one of the most misunderstood and most difficult components of a successful BSP. In order to determine a functionally equivalent replacement behavior, it is important to conduct a proper assessment of comorbid psychopathology and challenging behaviors through the assessment methods discussed in this chapter. After determining the challenging behaviors to be targeted in a BSP, it is critical to determine functions of the target behavior(s) through a functional assessment through the methods discussed previously. Replacement behaviors should meet the needs fulfilled by the challenging behavior by addressing the function of the behavior.

Psychotropic Drug Treatment of Challenging Behaviors

Before concluding this chapter, it is important to discuss the treatment of challenging behaviors via antipsychotics and other psychotropic medications. It has become commonplace for individuals with ID and/or ASD to be prescribed psychotropic medications for the treatment of challenging behaviors. Such medications include typical and atypical antipsychotics, multiple antipsychotics, anxiolytics, antidepressants, mood stabilizers, and antiepileptic medications (Matson & Neal, 2009). Researchers have questioned the use of such medication in the treatment of challenging behavior for several decades. Baumeister and Sevin (1990) discussed the lack of evidence and success in using medications to control challenging behaviors. They also stated that few individuals with ID receive medications for psychiatric disorders, likely due to the problems in diagnosing discussed above, but that treatment of challenging behaviors such as aggression, self-injurious behavior, and stereotyped behavior with psychotropic medication was commonplace in this population. They warned against the continued use of pharmacologic control of such behaviors without more methodologically rigorous studies guided by a theoretical framework.

Almost a decade later, Kennedy and Meyer (1998) provided a similar argument in which they state that the empirical evidence supporting the use of psychotropic medications for challenging behaviors is lacking. They question not only the methodological rigor of previous research but also the effectiveness and adverse effects of the medications being used. They provided several suggestions to improve research in this area as well as appropriate use and monitoring of medication side effects.

More recently, Matson and Neal (2009) discussed the same topic examining advances made in the literature and our current state of knowledge on the topic. They call attention to the tendency in the literature to consider all maladaptive problem behaviors ranging from different types

of aggression and self-injurious behavior to noncompliance and disruptive behavior as challenging behavior and note that researchers do not distinguish these behaviors one from another in the body of research. Researchers also fail to take into account the function of the behavior in the psychotropic treatment of the individuals' challenging behaviors. The authors warn against the adoption of psychotropic medications for the treatment of challenging behaviors citing the lack of empirically supported evidence and the apparent disconnect between practitioners and researchers in adhering to rules for evidence-based practice. This factor is especially important when the consequences of newer generation drugs are still unknown and the side effects of more traditional medications are very serious.

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Forensic and Offending Behaviours

18

Glynis Murphy and Jon Mason

Introduction

During the eugenics era, people with intellectual disabilities (ID) were thought to be especially likely to break the law (e.g. Goddard, 1912; Clarke, 1894, quoted in Brown & Courtless, 1971). As Perske reminded us recently (Perske, 2007), in the infamous Buck v. Bell case in the USA in 1927, 'Chief Justice Oliver Wendell Holmes said that people with ID were "a sap on the strength of the state"'. Careful research since those days has suggested that this is not so. Nevertheless, people with intellectual disabilities are particularly vulnerable in the Criminal Justice System (CJS), to not understanding their rights, not understanding the process and not being offered appropriate services. This chapter will review what is known about the prevalence of people with intellectual disabilities in the CJS, their characteristics and their vulnerabilities, as well as considering services to support people with intellectual disabilities when they break the law.

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Prevalence

A number of studies of crime and delinquency in the general population have suggested that boys/young men with lower IQs are more likely to be convicted of crimes than those who are more able (Farrington, 1995; Farrington & West, 1993; Herrnstein & Murray, 1994). Of course, this may be at least partly because they are less good at evading arrest than more able boys/young men, and recent evidence using self-report of criminal behaviour in adolescents suggests that criminal behaviour is not a function of IQ, when social deprivation is taken into account (Dickson, Emerson, & Hatton, 2005).In any case, such studies treat IQ as a continuous variable, and it is usually not clear in these studies how many people actually have an intellectual disability (i.e. IQ <70, with significant deficits in adaptive behaviour, present in the developmental period). Instead, in order to consider the prevalence of people with intellectual disabilities, we need to turn to other research, which takes one of two forms, neither of them perfect. Some studies take total populations of people with ID, and they examine what proportion of them are involved in the CJS. The others take the approach of examining parts of the CJS, such as prison, and asking what proportion of people there have intellectual disabilities.

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Total Populations and Those with ID Who Offend

In these studies, it is asserted that the total number of people with intellectual disabilities is known, and the percentage of them in the CJS is the subject of the study. For example, Hodgins (1992) examined convictions for a total Swedish birth cohort of 15,117 people born in Stockholm in 1953, who were followed up for 30 years. Evidence of ID was taken from registers of the children who were placed in special classes at school as a result of ID (this included 1.5 % of the men and 1.1 % of the women). Hodgins reported that the likelihood of conviction for a man with ID was 3 times as high as for those without disabilities. For women with ID the likelihood was nearly 4 times as high as for women without disabilities. The odds ratios were even more extreme for violent offences (5 times higher for men with ID and 25 times higher for women).

A further study by Hodgins, Mednick, Brennan, Schulsinger, and Engberg (1996) of a total population of over 300,000 people, in Denmark, born between 1944 and 1947, followed up at age 43 years, gave similar results: people with ID (excluding those with serious mental illness), who had had admissions to psychiatric wards, had an increased risk of committing offences of various kinds compared to people who had never been admitted (risk ratios were 5.5 and 6.9 for women and men respectively, for crimes entered onto the computerised criminal record system, which came into operation in 1978 in Denmark).

The problem with these two studies, though, is that we cannot be certain that all those identified as having ID definitely did have, since identification is by a proxy measure (e.g. attendance at special school or psychiatric hospital), nor can we be sure that all those with ID were included, for similar reasons. Moreover, people with ID may not commit more crimes: they may simply be easier to convict (see below under "Vulnerabilities of People with ID in the CJS").

In the UK, some total population studies have been undertaken focusing on people receiving ID services in particular geographical areas. Lyall, Holland, & Collins (1995), in a small study in Cambridge, found that 2 % of the 385 people, known to services for people with ID, had been in contact with the police as potential suspects over the previous year. McNulty, Kissi-Deborah, and Newsom-Davies (1995), in another small study of two residential providers for people with ID (serving 180 residents), in London, found 9 % of people had contact with the police over the previous year, as suspects. In the Cambridge study, none of the individuals was prosecuted and only one was formally cautioned, whereas in the London study, most were cautioned and about one third were charged (however, the numbers in both studies were very small). These differences may partly have resulted from the fact that the London sample was entirely recruited from residential services. Arguably, people with challenging behaviour (including that of the offender type) were more likely to be in such services than living with families, probably resulting in a higher figure for the people living in residential services.

In a third and larger study, McBrien, Hodgetts, and Gregory (2003), based in a city with a general population of almost 200,000, examined the numbers of convictions and contacts (as suspects), with the Criminal Justice Service, of all 1,326 adults known to ID services. It transpired that:

- 0.8 % of the 1,326 were serving a current sentence
- 3 % had a conviction of some kind (current or past)
- A further 7 % had had contact with the CJS as a suspect but had no conviction
- An additional 17 % had challenging behaviour that was 'risky', in the sense that it could have been construed as offending

This study suggests that about 10 % of the people in touch with ID services will have had contact with the CJS at some time in their lives, as suspects. Vaughan, Pullen, and Kelly (2000), in a study of mentally disordered offenders and community teams in Wessex (total population 1.8 million), found a slightly higher figure: 13 % of the people known to ID Teams fitted a definition of 'mentally disordered offender' (a larger percentage showed challenging behaviour).

In all probability, these kinds of studies overestimate the proportion of all the people with ID at risk of offending, since it is thought that around 1/3 to 2/3s of adults with mild disabilities lose touch with services when they leave school (Richardson & Koller, 1985), and the studies only count those in touch with services (they miss people with mild ID who are not known to disability services and who may or may not become involved in the CJS).

Studies of People in the CJS and the Proportion with ID

An alternative way of examining prevalence is to look at Criminal Justice Services and to ask how many people there have ID. A number of studies have examined the proportions of people with ID in the police station, in courts, in prisons, on probation and in hospitals following convictions for offences. There are two problems with such studies: first, the measurement of intellectual disability is often poor, rarely uses gold standard individualised IQ testing and almost never includes measures of adaptive functioning; secondly, such prevalence figures can only be properly understood in the context of the filters and thresholds that operate to keep people out of the CJS, which differ from country to country and even state to state within the same country (see Fig. 18.1 for a diagram of these filters in England). For example, very few jurisdictions convict people with severe ID through the CJS, preferring to divert them from custody (though in some jurisdictions such individuals can receive 'mandatory care' if they commit serious crimes, Sondenaa, Rasmussent, Palmstierna, & Nottesstad, 2008). Moreover, it is known that where offending behaviour takes place in disability services, one of the filters operating is whether staff decide to report the person's behaviour to the police. Several studies have shown that staff may well be reluctant to report possible crimes, so that those reported to the police must be only a proportion of the total number committed (Lyall, Holland, & Collins, 1995; McBrien & Murphy, 2006). In addition, many countries have 'diversion from custody' schemes, so that assessments of people referred to such schemes will of course yield very high rates of ID (e.g. Mannynsalo, Putkonen, Lindberg, & Kotilainen, 2009), compared to those not so referred.

Nevertheless, two UK studies have examined the numbers of people with ID appearing at police stations. For example, Gudjonsson, Clare, Rutter, and Pearse (1993) assessed the ID of 156 people arrested for questioning at two London police stations, using a short form of the WAIS-R. They found that 9 % of the total sample had an IQ score below 70 and 34 % had a score below 75 (representing the bottom 5 % of the general population), suggesting that a significant proportion of those detained by the police had an intellectual impairment. Similar findings were reported by Lyall, Holland, & Collins (1995), who screened 251 people appearing at a Cambridge police station for questioning. Participants were screened using a brief questionnaire (adapted from Clare & Gudjonsson, 1993), consisting of questions about the individual's reading and writing skills, whether they had received extra help at school and/or if they had attended a special needs school. They reported that 5 % of the 251 people had attended a school for children with mild or severe ID, and a further 10 % had attended schools for children with emotional/behavioural difficulties or a learning support unit within a mainstream school. Again, the results suggested that substantial numbers of those appearing at police stations have ID. As a result of such studies, many countries now include training for the police in ID, and a recent project in the state of Victoria in Australia, interviewing 229 policemen, estimated that most policemen encounter a person with an ID about 3 times a week and the police reported their training as useful in helping identify people with ID, though many requested more such training (Henshaw & Thomas, 2012).

In most jurisdictions, following questioning in the police station, if a person is charged with an offence, a court appearance will follow. Very few studies have been conducted which have assessed all those appearing before a court but, in two cohort studies in Australia, Hayes looked at the prevalence of people with ID appearing before

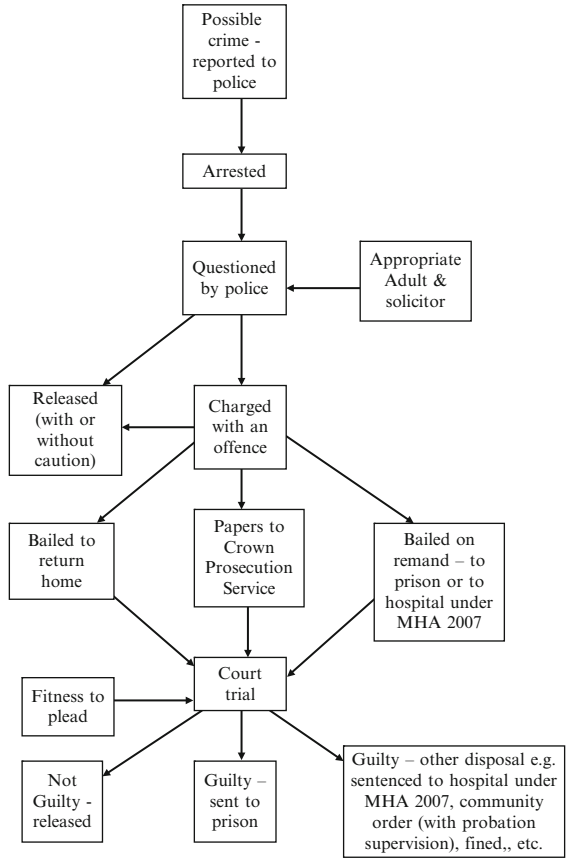

Fig. 18.1 Filters in the criminal justice system in England

Magistrate's courts in New South Wales. In the first, she assessed 113 people appearing before 4 local (2 urban and 2 rural) NSW Magistrate's courts. Fourteen per cent of the suspects were found to score below IQ 70; a further 9 % scored between IQ 70 and 79 (Hayes, 1993). In the second study, Hayes (1996a) concentrated solely on two rural courts (incorporating a large aboriginal population) and demonstrated that 21 % of those tested scored below IQ 70, and a further 36 % scored between IQ 70 and IQ 79. In both studies, however, Hayes used the Matrices section of the Kaufman Brief Intelligence Test (K-BIT) on the grounds that it was a culture-fair test (Hayes, 1996b) and it seems possible that this is not entirely culture-fair. A later study in four new South Wales Magistrates courts, using the K-BIT and the Vineland Adaptive Behaviour Scales, reported 10 % of participants scored <70 on the K-BIT, and 12 % scored below 70 on the VABS-2, but only 3.5 % scored below 70 on both (Vanny, Levy, Greenberg, & Hayes, 2009).

Following appearance in court, people with ID may be released unconvicted, or they may be convicted and subject to a variety of consequences, including imprisonment, hospitalisation, probation or more minor consequences (see Fig. 18.1). Research that has examined the proportions of people in these parts of the CJS is discussed below.

Studies of people with ID in the prison system have a long history (e.g. Woodward, 1955 quoted an analysis by Sutherland of over 300 studies conducted between 1910 and 1928 in the USA).

Table 18.1 Prevalence of offenders with intellectual disabilities in prisons

Author and year of study	Location of study	Number of participants	Test(s) used	% of prisoners with ID
Brown and Courtless (1971)	Inmates in USA prisons	90,000 (80 % of prison pop)	Large variety	9.5 %
MacEachron (1979)	Inmates in 2 USA prisons	436 of the 3,938 total pop.	Variety	1.5–5.6 % (depending on how measured)
Denkowski and Denkowski (1985)	20 prisons in USA[a]	191,133	WAIS-R	0.2–5.3 % (state to state variations)
Coid (1988)	1 prison in England	Retrospective study, 10,000	None specified	0.34 %
Gunn, Maden, and Swinton (1991)	UK, 16 prisons, 9 young offender (institutions)	404 youths and 1,365 men	None specified	0.4 %
Murphy, Harnett, and Holland (1995)	1 London prison (remand)	157 men	WAIS-R	0 % <IQ70
				5.7 % <IQ75
Birmingham, Mason, and Grubin (1996)	1 prison, northern UK (remand)	569 men	None specified	1 %
Brooke, Taylor, Gunn, and Maden (1996)	13 Prisons and 3 YOIs in UK	750 youths and men	Quick test	1 %
Murphy, Harrold, Carey, and Mulrooney (2000)	Prisons in Ireland	CHECK	K-BIT 2	29 %
Hayes, Shackell, Mottram, and Lancaster (2007)	One NW prison in UK (male)	140 randomly selected men (10 % of prison's population)	WAIS-III and VABS	WAIS: 7 % <70
				VABS: 10 % <70
				Both <70: 3 %
Crocker, Cote, Toupin, and St-Onge (2007)	Pretrial holding centres for men in Montreal, Canada	281 men assessed from a pool of 895	EIHM (French Canadian IQ test)	19 % in the 'probable ID' range, i.e. IQ <70
Sondenaa et al. (2008)	6 prisons in the north of Norway	143 prisoners (136 men and 7 women), randomly selected	WASI and HASI	WASI: 11 % <70
				HASI: 10 % <70
				Both: 7 % <70
Herrington (2009)	One young offender prison (18–21-year-old males) in London, UK	185 randomly selected prisoners	K-BIT 2 and VABS	K-BIT: 10 % <70
				VABS: 0 % <70
				Both: 0 % <70
Holland and Persson (2011)	All sentenced male prisoners released from prison in a 3-year period in Victoria, Australia	7,805 prisoners	WAIS-III	1.3 % with IQ <70

[a]Denkowski and Denkowski also look at IQ scores on group tests in 16 other institutions. These have been omitted

The numerous investigations have led to divergent opinions as to the approximate number of people with ID in the prison system, some asserting that in the USA, for example, 10 % is the correct prevalence figure (e.g. Brown & Courtless, 1971) while others argue that this simply reflects poor methodology (MacEachron, 1979). Figures from most recent studies are summarised in Table 18.1. Generally, it seems that studies from American, Irish and Australian prison populations produce higher prevalence rates than those from prisons in the UK, possibly reflecting the increased diversion from custody in the UK. Those studies that use a measure of adaptive behaviour, as well as IQ, tend to find the lowest rates. It also seems that geographical differences exist within some countries, such that some states in the USA record far higher prevalence rates than others

(Noble & Conley, 1992). However, much of the variation in prevalence rates results from the use of different tests (e.g. full individual IQ tests vs. quick screening tests), the cut-offs employed (those that include 'borderline ID' always get much higher figures), the administration of the tests by personnel with varying degrees of training, the time at which the tests are administered (e.g. whether these are done at times when individuals may be highly stressed), the type of institution (it may be that some prisons are more prone to receive offenders with ID than others) and the methodology employed in the study (McBrien, 2003; Murphy & Mason, 2007; Noble & Conley, 1992; Uzieblo, Winter, Vanderfaeillie, Rossi, & Magez, 2012). A large meta-analysis (Fazel, Xenitidis, & Powell, 2008) concluded that people with ID are represented but not overrepresented in prisons (0.5–1.5 %). Interestingly, at least until recently, despite the considerable research in this area, few prisons seem to use routine screening for ID within the UK (Talbot, 2008) or USA (Scheyett, Vaughn, Taloy, & Parish, 2009).

Probably one of the least restrictive consequences following conviction is a sentence requiring the individual to report to a probation officer. The probation service, both in the UK and the USA, provides for the supervision of offenders within the community. Little research has been done on the prevalence of mental disorder amongst those on probation but, where government policy emphasises the least restrictive alternative and community-based treatment wherever possible, the probation service may well provide supervision after conviction. Consequently, it may be the case that people with ID are increasingly appearing before the probation service, rather than being sent to hospital or prison. In the UK, Mason and Murphy (2002a, 2002c) found prevalence rates of 6 and 7 % of probationers with an IQ under 70 and a deficit in social functioning, in two separate studies in Kent. Interestingly, an earlier US study reported that a similar rate of 6–7 % of people on probation and parole in Missouri had developmental disabilities (Wood, 1976, quoted in Noble & Conley, 1992).

Characteristics of People with Developmental Disabilities Who Offend

Very few people with *severe* ID are to be found in the CJS, which may be surprising, given that challenging behaviour is known to become more common in people with more severe disabilities. However, it results at least in part from the requirement in law, in many jurisdictions, that the court must show not just *actus reus* but also *mens rea* on the part of an individual before conviction. Moreover, in the UK and the USA, for state/federal prosecutions, decisions are made by the Crown Prosecution Service (CPS) or the District Attorney (DA) about whether cases should go to court on a number of criteria, including the likelihood of conviction and public interest. It is probable that where staff or carers do call the police when someone with severe ID engages in potentially criminal behaviour, either the police themselves or the CPS/DA, in these countries, judge it not to be in the public interest to proceed. In some countries there are special provisions for people with severe intellectual disabilities who have committed serious crimes, so that in Norway, Finland and New Zealand, for example, they can be sentenced to mandatory or involuntary or secure care (Mannynsalo et al., 2009; Nottestad & Linaker, 2005; Sondenaa, Linaker, & Notestad, 2009), while in the UK, they may be sentenced to hospital under the Mental Health Act 2007.

People with ID who are convicted of offences are mostly young and mostly male, though no more so than might be expected when compared to other offender populations without disabilities (Barron, Hassiotis, & Banes, 2004; Cockram, 2005a; Noble & Conley, 1992). Most studies, however, have shown an overrepresentation of people from ethnic minorities with ID in courts/prisons, especially in the USA and Australia, often more than would be predicted from other suspects or offenders without disabilities (Cockram, 2005a; Hayes, 1993, 1996a; Holland & Persson, 2011; Noble & Conley, 1992).

It seems likely that this may reflect a variety of social disadvantages, including disproportionate levels of social deprivation and poverty, as well as poorer access to defence lawyers for people from ethnic minorities.

The kinds of crimes which people with ID commit have been a matter of dispute for years. Early assertions, drawn from Walker and McCabe's (1973) data on people detained under the Mental Health Act 1959 in England, in a particular year, claimed that sexual offences and arson were particularly common amongst people with ID (e.g. Prins, 1980, p. 92; Robertson, 1981). In fact, this did not follow from Walker and McCabe's data: the figures simply showed that a disproportionate number of the sexual and arson offences *of their sample* were committed by the proportion of people detained under the Mental Health Act on the grounds of 'mental subnormality' and 'severe mental subnormality' (as they were then termed). Nevertheless researchers continue to assert that people with ID are particularly likely to commit arson (Devapriam, Raju, Singh, Collacott, & Bhaumik, 2007) or to be sexual offenders (Lindsay et al., 2002), almost always on the basis of biased samples. These kinds of data should not be interpreted as having any implications for the proportion of *all* sexual and arson offences committed, since, firstly, not all such crimes are reported to the police (especially in the case of sexual offences), and, secondly, it is known that only a small minority of such offenders are diverted into hospitals under the Mental Health Act in the UK (so that a hospitalised group is a biased sample of all such offenders with mental health needs or ID). Similar arguments have been put by Noble and Conley (1992) in relation to data from various studies in the USA. Assertions of the same kind, drawn from assessments of people referred for forensic psychiatric evaluations (Hawk, Rosenfield, & Warren, 1993; Mannynsalo et al., 2009; Rasanen, Hakko, & Vaisanen, 1995) can be subject to the same criticism.

The only way to derive a true picture of the types of crimes committed by people with ID would be through the offence records for a total population sample (and even then, only *documented* offences would be counted). In fact, this is a difficult task: following school-leaving age, the numbers of people with mild degrees of ID, in touch with services, drop by three quarters to two thirds, as people 'blend' into the 'ordinary' community, apparently needing no further services (Richardson & Koller, 1985). In the absence of a total population sample, the most that can be said is that, where the types of offences of people with ID have been analysed, the range seems similar to those of other offenders (Barron et al., 2004; Cockram, 2005a; Glaser & Deane, 1999; Hodgins et al., 1996).

The best studies, which are still not perfect, involve comparison of large unbiased populations of offenders with and without ID, and there seem to be only small differences in types of offences reported. MacEachron (1979) compared offenders with ID to those with borderline abilities in two US prisons and found no significant differences in the types or severity of the most recent offence, the length of current sentence, the degree of recidivism, participation in rehabilitation programmes, recommendations for parole, degree to which parole had ever been revoked and the use of probation as a juvenile. Crocker et al. (2007) in their French Canadian study of 281 men in pretrial holding centres found no differences in the history of types of offences between those with ID and those without. Cockram, in her longitudinal study of offenders with and without ID charged with offences in Western Australia, examined the *most serious offences* in all of her sample and found that while people with ID were slightly more likely to be sent to prison for offences against good order and offences against property than people without ID, they were slightly less likely to be sent to prison for offences against the person (sexual offences, assault, murder, etc.) and much less likely to be sent to prison for drug or drunk driving offences (Cockram, 2005a, Table 2). Holland and Persson (2011) in their large study of people leaving prisons in Victoria, Australia, also found prisoners with ID were more likely to have had a property offence and less likely to have had a drug offence

than non-ID prisoners. They were not significantly more likely to have committed sexual offences or violent offences than non-ID prisoners. Meanwhile, Lindsay et al. (2010) in their study of patterns of offending and pathways into services for 477 people with ID in the UK reported that verbal and physical aggression were the most common offences and sexual offences were rarer (but perpetrators of these tended to be more frequently sent to high secure settings).

Some characteristics of offenders with ID have been confirmed in almost all studies, however. It appears that the social background of people with ID who have offended is very often characterised by social deprivation and many investigations have reported a high incidence of social deprivation and family breakdown/disorder in childhood (e.g. Barron et al., 2004; Day, 1988; Lindsay et al., 2010; Wheeler et al., 2009; Winter, Holland, & Collins, 1997), long histories of antisocial or 'challenging behaviour' (e.g. Day, 1988; Lindsay et al., 2010; Wheeler et al., 2009; Winter et al., 1997), high rates of adult unemployment with low rates of higher qualifications (Barron et al., 2004; Cockram, 2005a; Murphy et al., 1995) and a raised incidence of abuse in their own histories (Barron et al., 2004; Lindsay, Law, Quinn, Smart, & Smith, 2001; SOTSEC-ID, 2010). Nevertheless, relatively few of the studies have included comparison groups of non-disabled offenders, so that findings on social background are sometimes difficult to interpret. One exception, MacEachron (1979), in her study of people in prison in Maine and Massachusetts, found that, compared to people in the borderline range for IQ, those with ID were on average slightly older (in their 30s), less well educated, less likely to be abusing drugs and were from larger families (the average number of children was seven). Otherwise, the groups were very similar (e.g. both groups were highly likely to be unemployed, equally likely to have other disabilities and equally unlikely to be married). Moreover, in attempts to predict offence severity and length of sentence from intellectual, social and legal variables, MacEachron found few differences between the disabled and non-disabled groups, and she concluded that the intellectual differences between the two groups were fairly immaterial, with the social and legal variables 'more germane to the problem of being an offender than … intelligence'. Crocker et al. (2007) in Canada found a similar lack of demographic differences between those in pretrial holding centres with and without ID, the main difference being that fewer of the ID group had completed high school. Cockram's longitudinal study in Western Australia, of people with and without ID charged with offences, reported similar results, with few demographic differences between those with and without ID, though people with ID had somewhat higher rates of unemployment and lower rates of qualifications than those without ID. She also found people with ID were significantly more likely to get a custodial sentence than the non-disabled comparison group (Cockram, 2005a).

It seems likely that there is a high prevalence of mental health needs amongst people with ID who have committed offences (McGee & Menolascino, 1992; Noble & Conley, 1992). Studies from specialist treatment units and/or forensic services in the UK and the USA have reported very high rates of mental health problems and histories of abuse in those with ID, both men and women (Day, 1988; Lindsay et al., 2002; Lindsay, Smith, et al., 2004; Murphy & Clare, 1991; O'Brien, 2002; White & Wood, 1988). This is not surprising of course in health service facilities. However, in Crocker et al.'s (2007) study of 281 men held in pretrial holding centres in Canada, rates of mental health needs were the same in ID and non-ID groups, showing high rates of mental health needs in both.

Vulnerabilities of People with ID in the CJS

People with ID suffer a number of disadvantages in the CJS and these are of three kinds:

- Not understanding information given to them, such as about their rights
- Being suggestible and acquiescent on interview
- Not making wise decisions at crucial points in the CJS

Some of these difficulties are exacerbated by inequities in the legal process: for instance, Brown and Courtless (1971) found 8 % of defendants with ID in the USA were not represented by a lawyer. However, other disadvantages arise when people with ID are treated just like any other suspects in the CJS, and these difficulties basically stem from suspects with ID not fully understanding their rights, nor understanding the legal process and other matters thereafter, as their relatives often recognise (Cockram, Jackson, & Underwood, 1998).

Vulnerabilities in Police Stations and Courts

In England, on arrest, the police are required to 'caution' individuals. The exact words of the caution change from time to time, and in England they were altered in 1994, when the right to silence was modified. The current caution is as follows:

> You do not have to say anything. But it may harm your defence if you do not mention when questioned something which you later rely on in court. Anything you do say may be given in evidence.

Studies in England have demonstrated that many people with ID did not fully understand the older (simpler) caution (Clare & Gudjonsson, 1991), and several studies have demonstrated that even the general population (Clare, Gudjonsson, & Harari, 1998) and non-disabled suspects frequently struggle to understand the new caution (Fenner, Gudjonsson, & Clare, 2002). Indeed the middle sentence of the current English caution is so complex that some of the police were unable to give a full account of its meaning (Clare et al., 1998).

In addition, in England and Wales, when suspects arrive at the police station, they are given a written 'Notice to Detained Persons', which reiterates the caution and also tells them that they have a right to have someone informed of their arrest, to have a legal representative and to consult the Codes of Practice (Home Office, 1995; for revised Codes, see Home Office, 2012). However, analysis of the written 'Notice' has shown that it requires a reading age which people with ID are very unlikely to attain (Gudjonsson, 1991), and it contains such complex wording that many people with ID cannot understand it, even if they have it read to them, which the police are not obliged to do (Clare & Gudjonsson, 1991). As a result, Clare and Gudjonsson (1992) developed an experimental version of the 'Notice', with simplified wording and demonstrated that it was far easier to understand than the version in use in police stations (the Home Office declined to adopt the new version however).

Similarly, in the USA, since the Miranda v. Arizona case of 1966, suspects have to be warned before interrogation that they have a right to remain silent, that what they do say may be used in court and that they have a right to a lawyer (the so-called Miranda rights). Suspects are allowed to waive these rights if the waiver is made 'voluntarily, knowingly and intelligently' (Fulero & Everington, 1995). Much as in the case of the English 'Notice to Detained Persons', the written form of the Miranda warning is too complex for people with ID to be able to comprehend it (Fulero & Everington, 1995; O'Connell, Garmoe, & Goldstein, 2005).

At some point during suspects' stay at police stations, they are likely to be interviewed by the police. At such times, while people with ID may be able to accurately recount an event that has occurred (Kebbell & Hatton, 1999; Perlman, Ericson, Esses, & Isaacs, 1994), they tend to be much more suggestible and acquiescent, on average, than people without such disabilities, under questioning (Finlay & Lyons, 2002; Heal & Sigelman, 1995). Beail (2002) has argued that the usual tests employed to assess acquiescence and suggestibility need to be interpreted with care but, nevertheless, in the police station, such vulnerabilities would make it more likely that people would acquiesce to suggestions made to them by the police and be led by leading questions into self-incrimination (Cardone & Dent, 1996; Clare & Gudjonsson, 1993; Everington & Fulero, 1999). This tendency may be exacerbated by the fact that many people with ID misunderstand legal terms which are basic to the legal process:

Smith (1993), for example, found that about 20 % of the people referred for pretrial competency assessments in South Carolina did not understand the terms 'guilty' and 'not guilty', such that some actually had the meanings of the words reversed. Talbot (2008), in her study of people with intellectual disabilities in prisons in the UK, found that people with ID had often been confused and unable to understand what was happening in the police station, and in court, as the quotations below demonstrate:

> There was a solicitor, one police lady and two other people. I don't know why they were there, police talk maybe. It was somebody I didn't know before I got in trouble with the police. I didn't know if it was someone who could have helped me'. (Prisoner with ID talking about his time being interviewed in the police station, p. 18)
>
> To be truthful, I couldn't understand them. They talk so fast, they were jumping up and down saying things. I gave up listening. (Young offender with ID talking about his time in court, p. 21)
>
> I just felt out of place, being in court, that's the only way I can explain it. Everyone was talking. I didn't know what was going on'. (Prisoner with ID talking about his time in court, p. 21)

Most people with ID in Talbot's study found their own lawyers helpful, though they did not all have lawyers in the police station. Some people, however, found they could not even understand their lawyers, though they recognised the lawyers were trying to help:

> The solicitor tried to talk to me but used big words and I found it difficult to understand. The solicitor came and spoke to me in the cell and when she left I thought 'What was all that about?' (Prisoner talking about when his lawyer was talking to him, p. 23).

People with ID may also fail to predict the likely events in the criminal justice process and, perhaps because they are not familiar with the process, they may make very unwise decisions. For example, Clare and Gudjonsson (1995) showed people with ID and people without ID a film of a man making a confession to a burglary, which he had done, and then confessing to a murder, which he had not done. People without ID were aware of the seriousness of this and did not think the man would be able to get bail, nor correct his false confession in court. A large proportion of the people with ID, though, thought that despite making a false confession to a very serious crime in the police station, the man would be allowed to go home and would be able to correct the error later in court (Clare & Gudjonsson, 1995).

False Confessions

No doubt because of all these difficulties, it appears that false confessions by people with ID are not uncommon in a number of countries and are especially likely in juveniles with ID (Drizin & Leo, 2004; Gudjonsson, 1992; Kassin et al., 2010; Leo & Offshe, 1998; Perske, 1991, 2005, 2008, 2011). Perske, in the USA, has collected 75 cases so far of people with ID who have made false confessions. The cases have arisen mostly in the southern states; they mostly involved men, who were often poor and/or homeless, and who were frequently black (Perske, 2011). Commonly, the cases entailed very lengthy questioning by the police, in the absence of lawyers (Perske, 2005). All of these individuals in Perske's list were convicted, and some were executed, as the USA uses the death sentence. Of those not executed, some have since received a pardon (e.g. as a result of DNA evidence); others have been exonerated and released; others remain in prison.

In England and Wales, in recognition of some of these vulnerabilities of people with ID, special provisions were brought in, under the *Police and Criminal Evidence Act 1984*, in particular the audiotaping of police interviews (so that the manner of police questioning can be analysed) and the provision of an 'Appropriate Adult' (AA) for 'vulnerable' suspects (including those with developmental disabilities). Somewhat similar provisions were also made in Australia (Baroff, Gunn, & Hayes, 2004), and many states in the USA also now record custody statements (Sullivan, 2004, quoted in Perske, 2007).

In England and Wales, the Appropriate Adult's (AA) role in the police station is to protect vulnerable suspects from their tendency to 'provide information which is unreliable, misleading or self-incriminating' (Home Office, 1995, 2012).

However, there appear to have been two main problems with the Appropriate Adult scheme:

- Firstly, it is difficult for the police to evaluate when someone has a developmental disability (see below) so that many people are not provided with an Appropriate Adult, even though they are entitled to one (Bean & Nemitz, 1994; Medford, Gudjonsson, & Pearse, 2000).
- Secondly, Appropriate Adults, who may be parents, carers or social workers who have never met the individual in question, often do not speak during the police interview and seem unclear about their role (Pearse & Gudjonsson, 1996).

Many areas in the UK have since brought in Appropriate Adult training schemes, in order to improve the effectiveness of this important provision.

Vulnerabilities in Prison

Where people with ID are sentenced to prison, the vulnerabilities they experienced in the police station and in court, of course, follow them into prison (Talbot, 2008). As a result, they find understanding information difficult in prison, as it is usually in written form (which typically they cannot read). They also struggle with filling in forms, which most prisons require when prisoners wish to order a meal, request a visitor or book an appointment with a doctor when unwell. In Talbot's study in the UK prisons, many prisoners with ID struggled with written information, and 78 % of them said they had trouble with filling in forms (Talbot, 2008), as these quotations show:

> Its been a nightmare. Basically I don't know what the rules and regulations are. When you come in they give you a huge induction pack and tell you to look at it, you don't get any help. I have told them I need help but they don't pay any interest to me (p. 28).
>
> I couldn't fill in the visiting forms when I first came so I missed visits, then I was told what to do and someone filled it in for me (p. 33)
>
> I can't understand some of the forms, there are words that I don't know and I just get mad again (p. 34)
>
> I know you have to fill in a form but I wouldn't know what to put on it (female prisoner with ID talking about what to do when unwell, p. 42)

It was not unusual for prisoners with ID in Talbot's study to say they found it very difficult to ask for help, due to lack of confidence, shame, embarrassment or fear of ridicule. Moreover, Talbot found people with ID in prison were significantly less likely to know their release date and to have a job in the prison, and they were more likely to have clinically significantly depression and anxiety, than people without ID in prison (Talbot, 2008, 2009).

In many countries people with ID in prisons are excluded from work training and from the treatment programmes designed for mainstream offenders (Hayes, 2004; Sondenaa et al., 2008; Talbot, 2008). In the UK a prisoner called Dennis Gill took the government to court (via judicial review) for breach of the Disability Discrimination Act, on the grounds that he had been told he had to do a treatment programme in order to earn parole, but at the same time he was excluded from the programme on the grounds of his ID, so that he had actually served twice his tariff. Gill won his case (Straw & Lomri, 2010).

Diversion Out of the CJS

In most countries, diversion out of the CJS can occur at a number of points. For example, the police may decide not to proceed with a case involving a person with ID, or following an appearance in court, the person might be referred to community-based services for people with ID or sent to hospital, either on remand or once convicted. In some countries, hospital provision following offending is not available, and some of these countries have alternative provision, such as 'secure care' or 'mandatory care' (Norway, Finland, New Zealand) for some offenders with ID.

In England and Wales, courts can only divert people to hospital if it has been established in court that the individual is unfit to plead or if she/he falls within the broad category of 'mental disorder' of the Mental Health Act (MHA) 2007. While the numbers of those with mental illness diverted from courts and detained under the MHA are increasing, the numbers with ID are

decreasing: for example, on a census day at the end of March 2004, out of a total of 14,000 people detained under the MHA 1983, only 1,092 were detained under mental impairment/severe mental impairment (ID).

In the UK, Canada, the USA, Australia and elsewhere, people with ID can be found 'unfit to plead' (England and Wales) or 'not competent to stand trial' (USA) or there can be a finding of 'insanity in bar of trial' (Scotland) and they can be diverted out of the CJS (Baroff et al., 2004; Bonnie, 1992a, b; Brewster, Willox, & Haut, 2008; Rasch, 1990; Vanny, Levy, & Hayes, 2008). In England and Wales, this procedure was based on criteria set out in *R v. Pritchard 1836*, where it was established that the crucial issues were whether the accused could understand the proceedings so as to make a defence, challenge a juror and comprehend the evidence (MacKay, 1990). Nowadays fitness to plead is usually said to be based on five criteria: ability to plead, ability to understand the evidence, ability to understand the court proceedings, ability to instruct a lawyer and knowing that a juror can be challenged (Grubin, 1991a; MacKay & Kearns, 2000). The criteria in Scotland include being unable to understand the charge, or instruct a solicitor, or follow proceedings in court; those in the USA and Australia are very similar (Baroff et al., 2004).

Under early legislation on fitness to plead in both the UK and the USA, there was no trial of the facts, and people with ID who were found unfit to plead were detained in hospital and were supposed to return to court once they became fit to plead. In fact, people with ID rarely returned to court, disproportionate numbers of them remaining in hospital without ever returning to court for a trial (see Grubin, 1991a, 1991b for England; Chiswick, 1978, for Scotland). This of course had major civil rights implications, and there was subsequently a change in the law in Scotland, and in England and Wales, such that the facts of the case had to be tried and a broader set of disposal options was available (in Scotland the changes followed the Second Thomson Report (Scottish Home and Health Department Crown Office, 1975 and in England the relevant changes were first contained in the *Criminal Procedure* (*Insanity and Fitness to Plead*) *Act 1991*)). In both Scotland and England and Wales however (Brewster et al., 2008), relatively few people have been found unfit to plead (Brewster et al., 2008; MacKay & Kearns, 2000), perhaps because diversion from the CJS, through the use of the *Mental Health Act*, often allowed more flexibility. For example, MacKay and Kearns (2000) found that the number of people found unfit to plead only rose from an average of 12 per year across England and Wales, to 33 per year, after the changes to the legislation, only a quarter of these being people with ID (the remainder had mental health problems).

In the USA and Canada, in contrast, 'competence' hearings were very much more common, arising in between 2 and 8 % of felony cases in the USA, for example (Hoge, Bonnie, Poythress, & Monahan, 1992), meaning that there were thousands of competency hearings per year (Steadman, Monahan, & Hartson, 1982). Trial judges were required to order competence evaluations whenever significant doubts were raised as to the defendant's mental competence, and failure of defence lawyers to consider such an evaluation could invalidate subsequent conviction, if competence was later established as an issue (Bonnie, 1992a). Commonly, competence was judged on the 'Dusky' criterion: 'whether the defendant has sufficient present ability to consult with his lawyer with a reasonable degree of rational understanding and whether he has a rational as well as factual understanding of the proceedings against him' (*Dusky v. United States*, *1960*). More recently the criteria have been considered to be threefold: understanding the nature and seriousness of the charge, understanding the nature and purpose of the court proceedings and being able to assist one's lawyer in providing a defence (Baroff et al., 2004).

In the early years of competence hearings, difficulties arose in the USA that mirrored some of those in the UK: for example, one study found that 50 % of those found 'incompetent' and sent to hospital in Michigan were never released (Hess & Thomas, 1963), and McGarry (1971) reported that, after being found 'incompetent', more people left hospital, in Massachusetts, by

dying than by any other route. Subsequently, following the case of *Jackson v. Indiana*, in 1972, the US Supreme Court ruled that those held in hospital following an incompetency hearing could not be kept there for an unreasonable length of time. Thereafter, the average length of time in hospital following incompetency hearings fell to around 6 months to a year, and people with mental health needs were frequently treated in hospital and returned to court rapidly, as had been the original intention (Steadman et al., 1982). Where someone's competence to stand trial was considered 'untreatable', as presumably it might be for people with intellectual disabilities, the state was required to proceed with a civil commitment or drop the charges.

Some studies in the USA have shown that as few as 2 % of defendants with ID have pretrial evaluations of competence to stand trial (Brown & Courtless, 1971), though more recent figures suggested this had increased somewhat (Smith & Broughton, 1994). In practice, competence to stand trial has often been assessed by informal interview (not always competently conducted according to Brewster et al., 2008). In the USA, though not in the UK, a number of formal test protocols have been developed for the purpose of assessing fitness to plead (Grisso, 1986), including at least one which is highly sophisticated, the McArthur Adjudicative Competence Assessment or McCAT-CA (Hoge et al., 1997). Only two measures have been designed specifically for people with ID, however, one being a brief screening test (Smith & Hudson, 1995) and the other a more thorough assessment, the CAST-MR (Everington, 1990; Everington & Luckasson, 1987).

When competence is assessed in the USA, in people with ID referred for evaluation, it appears that about 35 % are judged not competent to stand trial (Petrella, 1992; Smith & Broughton, 1994). For example, in a 5 year cohort of people with ID assessed for competence to stand trial in South Carolina, approximately 2/3s were judged competent and 1/3 not competent to stand trial (this latter group had a lower mean IQ of 58, compared to those judged competent, who had a mean IQ of 64). Those judged not competent were less often sent to jail (0 %, as opposed to 55 % of those judged competent), less often put on probation (13 %, compared to 31 %) and mostly were dismissed back home (47 %, compared to 7 %) or referred to the department of 'Mental Retardation' (40 %, as compared to 7 %). In theory, those found incompetent to stand trial should be returned to court for trial, if and when they become competent. A recent review found only two training programmes in the literature that aimed to restore competence (Salekin, Olley, & Hedge, 2010), and it is difficult to be sure how effective these programmes are; one finding only 18 % of their sample of people with ID were returned to competence after receiving training (Anderson & Hewitt, 2002), while the other reported 75 % had become competent within a year (Morris & Parker, 2008).

In addition to 'competence' in the USA, 'culpability' or criminal responsibility may be assessed. Essentially, the former refers to whether the defendant understands the charges and court proceedings and can instruct his/her lawyer, while the latter refers to whether the defendant knew right from wrong at the time of the offence (Smith & Broughton, 1994). Criminal responsibility is usually judged by the McNaughten rule, the so-called right-wrong test (which states that a person is not legally responsible if she/he was 'labouring under such a defect of reason from disease of the mind, as not to know the nature and quality of the act he was doing; or, if he did know it, that he did not know what he was doing was wrong' (Grisso, 1986)). Mental health professionals have often been accused of confusing competence to stand trial and culpability (Johnson, Nicholson, & Service, 1990), and a number of investigations have addressed the extent to which the two characteristics co-occur in particular individuals (Johnson et al., 1990; Petrella, 1992; Smith & Broughton, 1994). In general, for people with ID, it appears that it is rarer to be judged not criminally responsible than to be judged not competent to stand trial and, amongst those judged not responsible, most will have also been judged not competent (though these relationships are different for people with mental health problems—see Johnson et al., 1990).

Improvements in Practice in the CJS

At present, people with ID who break the law generally enter the CJS much as other people would, though perhaps with more confusion and less appreciation of their circumstances than most. Essentially, their treatment within the CJS depends on the extent to which their disability is recognised by those coming into contact with them and that often determines their course through the system. Most research, though, has demonstrated that the identification of people with ID in the CJS is poor in the pretrial phase in the USA, Australia and the UK (Scheyett et al., 2009; Talbot, 2008). In England where some special provisions (such as an *Appropriate Adult*) exist for people with ID and/or mental health needs in the police station, Gudjonsson et al. (1993) found that only about one fifth of the people who needed this special help were identified, and others have suggested even lower rates of identification nationally (Bean & Nemitz, 1994). Likewise, reports from court diversion schemes in the UK (James, 1996; Pakes & Winstone, 2010) have shown that they are very variable, but on the whole, few people assessed in these projects have ID, implying that many cases may simply be missed. Cooke (1991), for example, in Scotland, reported that in a consecutive series of 150 offenders referred for psychological/psychiatric treatment before prosecution (so-called primary diversion), none appeared to have ID, and Joseph and Potter (1993) in a London diversion scheme, operating at two magistrates' courts, found only four people (2 %) had ID (formal assessment was not employed). Moreover, *after* conviction, Talbot (2008) reported that there was no widely accepted routine screening for ID in prisons in the UK, though prisons did often test for literacy skills if and when prisoners went to their education and skills departments.

Nevertheless, special protections before and during trials do exist, and there is some evidence that they are being increasingly recognised and more often used, though not yet to anything like their full extent. Thus, for example, in some police stations in the UK, people brought in for questioning are screened by the custody sergeant in order to try to ensure that the protections available at the police station are made available (Clare, 2003). Likewise, a brief screening test for the presence of ID has been developed in Australia (Hayes, 2002) and in the UK (Mason & Murphy, 2002b), and a brief screening test to assess competence to stand trial has been developed in the USA (Smith & Hudson, 1995) in order to allow rapid screening of all those who will appear in court and may need a full competency assessment. In addition, there is beginning to be an increase in the willingness of agencies in the CJS and health services to work together, to provide support for those proceeding through the CJS (e.g. easy read information—see Hollins, Clare, Murphy, & Webb, 1997; Hollins, Murphy, Clare, & Webb, 1997) and to ensure treatment for those people with ID who may otherwise be likely to remain at risk of offending (Murphy & Clare, 2012).

In the UK, following the Talbot (2008) research and the Bradley report (Bradley, 2009), the Department of Health instigated a trial of screening for ID in three prisons, using the LDSQ (McKenzie & Paxton, 2006). The results showed that about 7 % of prisoners screened positive for ID (almost certainly an overestimate), and two of the prisons were imaginative in providing assistance to those identified, one inviting in the relevant local community team to come and provide a full assessment and another referring the prisoners to a service providing support inside the prison which would also follow up those who left prison and provide support in the community. Despite this successful screening programme, the Ministry of Justice is still currently debating whether to roll out such screening, partly on the grounds of the resources it would require in prison services. In fact, of course, while screening would be very welcome in prisons, it would be better done at the police station stage, provided the information could then follow suspects through the CJS (allowing a variety of kinds of help at many stages in the system). The government in the UK seems reluctant to enable this to happen. Similar debates are happening elsewhere (e.g. parts of Canada are considering screening for ID in prisons, Crocker, personal communication).

Another attempt to reduce the disadvantages suffered by people with ID in courts is that of so-called 'mental health courts', which began in the USA in 1997, spreading to 100 courts in 2012 in the USA (Burke, Griggs, Dykens, & Hodnapp, 2012), also used in parts of Australia (Vanny et al., 2008), and which are currently being trialled in some places in the UK. The courts recognise that people with mental health needs are not appropriately placed in the CJS and the courts prioritise treatment over punishment. Initial studies of reconviction rates have suggested that mental health court defendants go a longer period of time without reoffending than do defendants in ordinary courts. Burke et al. (2012) showed that about 11 % of defendants in mental health courts in one county in Tennessee had ID and mental health needs (dual diagnosis) and that those with dual diagnosis were younger, more often male and African-American, had lower levels of social support, less education and less employment than non-ID defendants. Burke et al. have presented no data as yet on reconviction rates.

Another important development in the USA, given the existence of the death penalty there, together with the likelihood of false confession and wrongful imprisonment, is the decision in *Atkins v. Virginia* in the US Supreme Court in 2002. In what has become known as the Atkins decision, Justice John Paul Stevens in the Supreme Court declared that executions of people with ID violated the eighth amendment that forbids cruel and unusual punishment. Before this, although 18 states had already banned the death sentence for people with ID, a survey showed that 44 people with ID had been executed in the USA between 1976 and 2002 (Perske, 2007). Moreover, there had been heated argument about the definitions and measurement of 'mental retardation'. The definition of ID proposed by Justice Stevens included significant deficits in general intelligence, together with significant deficits in adaptive behaviour, present since the developmental period (i.e. what used to be called mental retardation in the USA). A debate followed about exactly how to measure adaptive behaviour (Dwyer & Frierson, 2006), with some states asserting that a person had to have significant deficits in all ten areas of adaptive behaviour listed by the AAIDD manual (Luckasson et al., 1992), others asserting people only had to have significant deficits in the three dimensions introduced in the revised AAIDD manual (Luckasson et al., 2002) and other states making no assertions. In some states in the USA, a measure called the Street Survival Skills Questionnaire was used to establish levels of adaptive behaviour, while others argued that the inadequate standardisation of that test disadvantaged people with ID who were Atkins claimants, in overestimating their adaptive behaviours (Denkowski & Denkowski, 2008). Nevertheless, Denkowski himself has been banned from practising forensic psychology in Texas for arguing that some Atkins claimants deserved to be executed on the grounds that their socially deprived backgrounds depressed their IQs so that they did not 'really' have an ID (http://standdown.typepad.com/weblog/2011/04/texas-defender-service-on-the-denkowski-sanction.html).

Assessment, Treatment and Risk Management

Early attempts to provide assessment and treatment for people with ID who were at risk of offending concentrated on behavioural and medical methods, with limited success. Increasingly, however, specific measures have been developed for the psychological assessment of people at risk of aggression (Benson & Ivins, 1992; Walker & Cheseldine, 1997; Taylor, 2002), people who set fires (Murphy & Clare, 1995) and men who engage in sexually abusive behaviour (Lindsay, Whitefield, & Carson, 2007). Recently, cognitive-behavioural treatment has been seen as the method of choice (Murphy & Clare, 2012), and a number of studies have now demonstrated that such techniques as anger management training are effective in reducing self-rated anger (Allen, Lindsay, MacLeod, & Smith, 2001; Benson, Johnson Rice, & Miranti, 1986; Rose, West, & Clifford, 2000; Taylor, Novaco, Gillmer, & Thorne, 2002; Whitaker, 2001) and that cognitive-behavioural treatment for sexual offenders can

reduce recidivism (Lindsay & Smith, 1998) and cognitive distortions (Heaton & Murphy, in press; Lindsay, Marshall, Neilson, Quinn, & Smith, 1998; Lindsay, Neilson, Morrison, & Smith, 1998; Lindsay, Olley, Jack, Morrison, & Smith, 1998; SOTSEC-ID, 2010; Williams, Wakeling, & Webster, 2007) and improve empathy (Heaton & Murphy, in press; Rose, Jenkins, O'Connor, Jones, & Felce, 2002; SOTSEC-ID, 2010). Nevertheless, few of the studies investigating the effectiveness of CBT in reducing offending-type behaviour have involved control or comparison groups of any kind (Benson et al., 1986; Lindsay & Smith, 1998; Rose et al., 2000; Taylor et al., 2002). Even fewer have involved randomised control trials, as noted by Barron, Hassiotis, and Banes (2002), Lindsay (2002) and Taylor (2002), although one small RCT has appeared that examined the effectiveness of anger management training (Willner, Jones, Tams, & Green, 2002) and a larger trial of this kind is underway (Willner, personal communication). Despite this burgeoning of treatment available in some settings, it is often the case that people with ID are excluded from mainstream services because of their ID but excluded from ID services because they are too able (Hayes, 2004; Murphy & Clare, 2012). Moreover, in many countries treatment programmes are not available and/or not suitable in prison settings for people with ID (Holland & Persson, 2011; Talbot, 2008), though in the UK, in the wake of the Gill case, some programmes such as the basic Thinking Skills programme are being adapted for people with ID in prison (the sex offender treatment programme, SOTP, had already been adapted—see Williams et al., 2007; Williams & Mann, 2010).

Whether or not treatment can be provided for people with ID at risk of offending, services frequently employ risk assessment and risk management methods, so as to ensure public safety and to reduce the risk of reoffending. According to Cockram's longitudinal study (Cockram, 2005b), and to Holland and Persson (2011), people with ID are more likely to be rearrested and reconvicted than people without ID, though Gray, Fitzgerald, Taylor, MacCulloch, and Snowden (2007) found them less likely than non-disabled offenders to be reconvicted. As Halstead (1997) and Johnston (2002) have commented, many services for people with ID simply employ structured clinical judgments of risk for this purpose, and, while this method of risk assessment may have some face validity, there have been criticisms that such methods are not as good as actuarial methods in predicting risk at least in the non-disabled population (Grove, 2000; Monahan, 2002).

For non-disabled offenders, risk is predicted using measures that combine actuarial historical variables (such as age, gender, numbers of previous offences), with clinical variables (such as diagnoses, PCL-R scores). The result is that a variety of measures abound, such as the Violence Risk Appraisal Guide (VRAG) (Quinsey, Harris, Rice, & Cromier, 1998) and the Sex Offender Risk Appraisal Guide (SORAG) (Quinsey, Rice, & Harris, 1995), the Historical Clinical Risk-20 (HCR-20) (Webster, Eaves, Douglas, & Wintrup, 1995), the Sexual Violence Risk-20 (SVR-20) (Boer, Hart, Kropp, & Webster, 1997), the SONAR, the Static-99 (Hanson & Thornton, 1999) and the Rapid Risk Assessment of Sexual Offence Recidivism (RRASOR) (Hanson, 1997). A number of large-scale studies have found that these instruments do predict risk well and that the instruments are often equally good as each other in their predictive ability for non-disabled offenders (Barbaree, Seto, Langton, & Peacock, 2001; Harris et al., 2003; Sjostedt & Langstrom, 2002). Nevertheless, research into the use of these instruments is only just beginning in relation to people with ID at risk of offending (Lindsay & Beail, 2004), and it is likely that parts of the instruments will need adapting (e.g. some risk measures for sexual offending consider that being single and being unemployed are risk factors for reoffending, but actually just about all men with ID who have at least one sex offence are likely to be single and unemployed). Recent attempts to validate use of risk assessments instruments with people with ID include, for example, the following: Harris and Tough (2004) have found that the RRASOR did predict sexual offending amongst men with ID and sexually abusive behaviour, Quinsey, Book, and Skilling (2004) and Gray et al. (2007) reported

that the VRAG was a good predictor of violent behaviours in people with ID, and Gray et al. (2007) and Lindsay et al. (2008) found the HCR-20 was predictive of reoffending for people with ID. However, McMillan, Hastings, and Coldwell (2004) noted that clinical prediction was as good as actuarial prediction of violent incidents within a forensic institution. One of the problems with these instruments though is that researchers and clinicians sometimes adapt the instruments for people with ID without making it clear exactly how they were adapted. However, Boer, Frize, Pappas, Morrissey, and Lindsay (2010a, 2010b) have now provided details of how the HCR-20 and SVR-20 should be adapted.

Increasingly, there have been attempts to improve prediction of reoffending by non-disabled offenders, by including dynamic factors (such as mood) in the risk measure, to supplement the normally static actuarial and clinical factors. Attempts to do this for people with ID are just beginning (Boer, Tough, & Haaven, 2004) and some measures have shown some initial success: Lindsay, Murphy, et al. (2004), for example, have developed a measure called Dynamic Risk Assessment and Management System (DRAMS) which is showing promise as a predictor of aggressive incidents in residential settings, and Boer et al.'s ARMIDILO-S, a risk measure for predicting reoffending in sex offenders, is also showing promise (Blacker et al., 2011; Sindall & Murphy, submitted).

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Part V

Interventions and Services

Psychopharmacology

19

Shoumitro Deb

Abbreviations

ABC	Aberrant Behaviour Checklist
ADHD	Attention Deficit Hyperactivity Disorder
ASD	Autistic Spectrum Disorder
BPI	Behavior Problems Inventory
CGI	Clinical Global Impressions Scale
DASH	Diagnostic Assessment for the Severely Handicapped
DISCUS	Dyskinesia Identification System
DOTES	Dosage Record and Treatment Emergent Symptom Scale
ECG	Electroencephalograph
ESRS	Extrapyramidal Symptoms Rating Scale
ID	Intellectual disabilities
ITT	Intention to treat
MOAS	Modified Overt Aggression Scale
NCBR-F	Nisonger Child Behavior Rating Form
NICE	National Institute for Health and Clinical Excellence
NNT	Number needed to treat
OCD	Obsessive Compulsive Disorder
ONE	Objective Neurological Examination
PIMRA	Psychopathology Instrument for Mentally Retarded Adults
RCT	Randomised controlled trial
RUPP	Research Unit of Pediatric Psychopharmacology
SIB	Self-injurious behaviour
SOME	Summation of Maladaptive Expression
SSRIs	Selective serotonin reuptake inhibitors
VAS	Visual Analogue Scale

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Introduction

It has been reported that 20–50 % of people with intellectual disabilities (ID) receive psychotropic medications (Deb & Unwin, 2007a). It has been reported that 36 % of those who receive psychotropic medications do not have a psychiatric diagnosis (Clarke, Kelley, Thinn, & Corbett, 1990). People with ID often receive multiple medications and often at a high dose (Deb & Fraser, 1994). In a recent prospective 12-month follow-up study of 100 adults who have been seen by psychiatrists in the UK in their outpatient clinics for the management of aggressive behaviour, Unwin, Rashid, and Deb (2011) found more than 90 % of the participants received psychotropic medications. Of them 66 % received antipsychotics, 42 % antiepileptic, 35 % antidepressants, 14 % antianxiety/beta blockers, and

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43 % as required medications, and 23 % received medications to counteract adverse effects of other psychotropic medications. In a consensus study among psychiatrists in the UK on the use of medication for the management of aggression and self-injurious behaviour (SIB), Unwin and Deb (2008) found that most psychiatrists preferred to use a non-medication-based management first. However, if they had to use a psychotropic medication, the order of preference was usually an antipsychotic followed by an antidepressant followed by a mood stabiliser. Among the antipsychotics, the newer antipsychotics were preferred in the order of risperidone followed by olanzapine, quetiapine, amisulpride, aripiprazole and clozapine. Among the antidepressants, the order of preference was citalopram followed by fluoxetine, sertraline, escitalopram, mirtazapine, paroxetine, venlafaxine and fluvoxamine. Psychiatrists usually considered psychotropic medications under the following circumstances: (a) failure of non-medication-based interventions; (b) risk/evidence of harm to others, property and self; (c) high frequency and severity of problem behaviours; (d) to treat an underlying psychiatric disorder or anxiety; (e) to help with the implementation of non-medication-based interventions; (f) risk of breakdown of the person's community placement; (g) lack of adequate or available non-medication-based interventions (although this should not be used as a rationale for using medication); (h) good response to medication in the past; and (i) patient/carer choice.

The main concerns for using psychotropic medications in the absence of a diagnosed psychiatric disorder are that these medications in general are not licensed for the management of problem behaviour without a psychiatric diagnosis; there is potential for adverse effects particularly if these medications are used for a long time, yet it is difficult to withdraw these medications once started because of the potential withdrawal symptoms and resurgence of problem behaviour and the lack of evidence for the effectiveness of psychotropic medications in the absence of a psychiatric diagnosis. Therefore, systematic reviews have been carried out on the effectiveness of different types of psychotropic medications which have been summarised in this chapter. The evidence base for the effectiveness of different psychotropic medications for different types of psychiatric disorders such as schizophrenia, depressive disorders and anxiety disorders is summarised in the respective National Institute for Health and Clinical Excellence (NICE) guidelines in the UK (www.nice.org.uk) and in similar guidelines in other countries. These guidelines should be used for people with ID with different psychiatric disorders in the absence of any specific guidelines for these people. Therefore, in this chapter the evidence for the use of different psychotropic medications for people with ID only for the management of problem behaviour in the absence of any psychiatric disorder is summarised. This chapter concentrates primarily on studies on adults, although for antipsychotics data from children's studies as well have also been presented.

These systematic reviews were carried out in order to develop a national and an international guide for the management of problem behaviour in people with ID (Deb et al., 2009; Unwin & Deb, 2010). As advised by the Guideline Development Group (GDG), any study that included less than 10 participants in their study was excluded from the systematic reviews. In this chapter, Table 19.1 summarises the overall findings in terms of total number of papers included in the systematic reviews on different psychotropic medications. In this chapter findings from the randomised controlled trials (RCTs) of antipsychotics have been presented in some details and the rest of the studies are summarised in the respective tables.

Antipsychotic Medications

As expected the highest number of studies found in the systematic reviews was on antipsychotics. Here only the RCTs based on the new generation of antipsychotics are summarised. Among the new generation of antipsychotics, risperidone is studied most frequently as this is the most commonly used antipsychotic in the UK for the management of problem behaviour in ID. Further information on the effectiveness of antipsychotic medications for the management of problem behaviours in adults with ID is provided in Deb,

Table 19.1 Summary findings of systematic reviews of different psychotropic medications

Drug	*N*	RCT (number of participants included in different studies)	Prospective (number of participants included in different studies)	Retrospective (number of participants included in different studies)
Antipsychotics	12	3 (39 vs. 38; 30; 28 vs. 29 vs. 29)	6 (15, 15, 18, 20, 33, 34)	3 (17, 20, 24)
Antidepressants	10	1 (10)	7 (10, 14, 15, 16, 19, 20, 60)	2 (14, 33)
Antiepileptics	4	1 (10)	1 (28)	2 (22, 28)
Lithium	4	3 (20 vs. 22; 52; 26)	0	1 (74)
Naltrexone	4	2 (33, 24)	1 (15)	1 (56)
Psychostimulants	0	0	0	0
Antianxiety/buspirone	1	0	1 (26)	0
Diet/vitamins	1	1 (Pica: 128, control: 30)	0	0

Sohanpal, Soni, Unwin, and Lenôtre's (2007) systematic review. Further information on the effectiveness of antipsychotic medication in children with ID is presented in Unwin and Deb's (2011) recent systematic review.

There are three RCTs among adults with ID (Gagiano, Read, Thorpe, Eerdekens, & Van Hove, 2005; Tyrer et al., 2008; van Den Borre et al., 1993). There are six RCTs among children with ID with or without Autistic Spectrum Disorder (ASD) (Aman, De Smedt, Derivan, Lyons, & Findling, 2002; Buitelaar, van der Gaag, Cohen-Kettenis, & Melman, 2001; Research Unit of Pediatric Psychopharmacology-RUPP, 2002; Shea et al., 2004; Snyder et al., 2002; Van Bellinghen & De Troch, 2001). RUPP (2002) and Shea et al. (2004) primarily included children with ASD, some of whom also had ID, whereas Aman et al. (2002) and Snyder et al. (2002) primarily included children with ID but excluded those who had ASD. Of these four studies, only the RUPP study (2002) was not sponsored by a pharmaceutical company. Three of the RCTs involving children were continued for many weeks using open-label designs (Findling, Aman, Eerdekens, Derivan, & Lyons, 2004; RUPP Continuation Study, 2005; Turgay, Binder, Snyder, & Fisman, 2002).

Adult Studies

van Den Borre et al. (1993) included 37 adults (15–58 years) with ID in their study who showed aggression, SIB, agitation, hyperactivity and irritability. It is not clear whether or not the authors excluded participants who had a diagnosis of psychiatric disorder. Risperidone ($N=30$ after seven drop outs) 4–12 mg/day was used as an add-on to the existing medications. A crossover RCT design was used, which included 1 week wash out followed by 3 weeks RCT followed by 1 week wash out followed by 3 weeks crossover RCT. Primary outcome measure was Aberrant Behaviour Checklist (ABC) total score. Secondary outcome measures included Clinical Global Impression (CGI), Visual Analogue Scale (VAS) (target behaviours), extrapyramidal symptoms (Extrapyramidal Symptoms Rating Scale: ESRS), blood tests, electroencephalograph (ECG) and the participants' weight.

In the first phase there was 16 % drop in the total ABC score in the risperidone group and 15 % in the placebo group. In the second phase there was 27 % drop in the total ABC score in the risperidone group and 0 % in the placebo group. The difference in phase one was not statistically significant but the difference in phase two was. There was statistically significant improvement in the risperidone group according to CGI ($p<0.01$) (both phases). However, there was no statistically significant change according to VAS. There was also no change between the two groups in the ECG or the ESRS score. However, the participants in the risperidone group showed sedation 10 times more commonly than the placebo group. Blood tests did not detect any statistically significant change in the two groups.

Risperidone was found to be more efficacious in this study. However, conflicting results were found in two phases of the study in that two groups did equally well in phase one and the risperidone group did better only in phase two. It is therefore possible that the same group of participants continued to show improvement irrespective of the intervention used. There was also conflicting results found according to different outcome measures. For example, the risperidone group did better according to the total ABC score and CGI, but not according to VAS.

The other problems with the study included a very short wash out period, which also increased the chance of contamination from potentially withdrawal symptoms as being rated as problem behaviour, and short follow-up period. Authors did not clarify how many participants were included in each group. The method of randomisation and blinding were not described, and the IQ level or gender ratio was not specified in the paper. The total score of ABC is not valid; hence, most studies now use the Irritability (ABC-I) subscale. As authors did not exclude underlying psychiatric disorders, it is possible that in some cases, risperidone may have improved behaviour by treating the underlying psychiatric disorder. The dose of risperidone is higher than what is usually used for problem behaviour now.

Gagiano et al.'s (2005) study included 77 adults (18–57 years) with ID who did not have a diagnosis of psychiatric disorder. The first phase of the study was a parallel design RCT in which 39 participants were randomly allocated to the risperidone group and 38 into the placebo group. The RCT lasted for 4 weeks after which 58 participants continued to receive risperidone in an open-label design for another 48 weeks. Participants received risperidone as an add-on to other medications at a dose of 1–4 mg/day (mean dose: 1.8 mg/day) both in the RCT and in the open-label study.

The primary outcome measure was the ABC total score, and secondary outcome measures included Behavior Problems Inventory (BPI), CGI-S and VAS (target behaviours). According to the authors, 52 % in the risperidone group improved as opposed to 31 % in the placebo group (number needed to treat; NNT = 5). There was a statistically significant improvement in the ABC total score in the risperidone group compared with the placebo group ($p = 0.036$) and also according to CGI ($p < 0.05$). In the risperidone group, 23–41 % complained of somnolence and mean weight gain was 3.8 ± 0.6 kg. There was no difference between the groups in the QTc interval according to ECG and extrapyramidal symptoms according to the ESRS.

Overall this is a good quality study and supports the use of risperidone among adults with ID, included a reasonable number of participants (although the study could still be underpowered!), the overall design was good. However, the ABC total score lacks validity, and ABC-I score instead should have been used as the primary outcome measure. The follow-up period in the RCT of 4 weeks is short. The pharmaceutical company sponsored the study.

Tyrer et al. (2008) in a multicentre parallel design RCT randomly allocated 86 adults with ID and aggressive challenging behaviours into three groups, namely, risperidone (mean dose of 1.07–1.78 mg/day), haloperidol (mean dose of 2.5–2.94 mg/day) and placebo. Clinical assessments of aggression, aberrant behaviour, quality of life, adverse drug effects and carer burden, together with measurement of total costs, were recorded at 4, 12 and 26 weeks. The primary outcome was change in aggression after 4 weeks treatment according to the Modified Overt Aggression Scale (MOAS).

Aggression declined dramatically with all three treatments by 4 weeks, with placebo showing the greatest reduction according to MOAS median score (79 % as opposed to 57 % for combined medication groups) ($p = 0.06$). Placebo treatment was also cheaper than the other two treatments over a 6-month period in terms of total costs (Tyrer et al., 2009).

However, the risperidone group showed a higher level of aggression at the baseline compared with the placebo group and had the highest level of improvement according to the ABC-I subscale. The period of follow-up of 4 weeks when the data were analysed was short and the participant number is small, which may not have provided adequate power to the study.

Children Studies

Aman et al.'s (2002) study included 115 children (87 included) (5–12 years) with ID. The authors have excluded children with ASD. A multicentre parallel design RCT was used in which 43 children were randomly allocated to risperidone group in order to receive 1.2 mg/day mean dose and 44 allocated to the placebo group. Children were followed up for 6 weeks at the end of which Nisonger Child Behavior Rating Form (conduct problem subscale) (NCBR-F) was used as the primary outcome measure along with ABC-I, BPI, VAS and CGI as secondary outcome measures. According to the authors, 15.2 % of children in the risperidone group as opposed to 6.2 % in the placebo group showed significant improvement. Adverse effects in the risperidone group included headache and somnolence, but not extrapyramidal symptoms. Mean weight gain in the risperidone group was 2.2 kg as opposed to 0.9 kg in the placebo group.

Overall this seems to be a good quality study and supports the use of risperidone among children. However, the study could still be underpowered and the follow-up period was short. The improvement was not defined.

Findling et al. (2004) followed up 107 children from Aman et al.'s (2002) study in an open-label study for 48 weeks' extension. The same outcome measures as in Aman et al.'s (2002) study were included such as NCBR-F, ABC-I, CGI-I, BPI and VAS. Fifty (47 %) children completed the trial. Improvement with risperidone at 1.51 mg/day mean dose was maintained for 48 weeks. Although the dropout rate was high, they are not always necessarily due to the adverse effects of risperidone.

RUPP (2002) study included 101 children (5–17 years) with ASD, 74 of whom had ID and 12 borderline intelligence. The authors used multicentre parallel design RCT for 8 weeks in which 49 children were randomised to receive 0.5–3.5 mg/day mean dose of risperidone and 52 to receive placebo. The primary outcome measure was ABC-I and the secondary measure was CGI-I.

In the risperidone group, there was 57 % mean reduction in the ABC-I score at follow-up as opposed to 14 % in the placebo group ($p<0.001$). Similarly 69 % in the risperidone group and 12 % in the placebo group, respectively, showed much or very much improvement according to CGI ($p<0.001$). Average weight gain for the risperidone group was 2.7 ± 2.9 kg as opposed to 0.8 ± 2.2 kg in the placebo group ($p<0.001$). A higher proportion of children in the risperidone group reported increased appetite, fatigue, drowsiness, dizziness and drooling ($p<0.05$). In the subsequent open-label study, two-thirds of subjects with a positive response to risperidone at 8 weeks maintained the improvement at 6 months.

Overall this is a good quality study and supports the use of risperidone among children. Cohort size is still relatively small and the follow-up period is relatively short.

RUPP Continuation (2005) study was conducted in two phases. In phase one, 63 children (5–17 years) with ASD (53 with ID and seven with borderline intelligence) continued to receive risperidone at a mean dose of 1.96 mg/day in an open-label trial for 4 months. In phase two, 38 children with ASD (31 with ID and five with borderline intelligence) were allocated randomly in a double blind study either to continue to receive risperidone or being replaced by placebo for 8 weeks. The ABC-I subscale was used as the main outcome measure.

At the end of phase one, the change in ABC-I score was small and nonsignificant and the average weight gain was 5.1 kg ($p<0.001$). In phase two, 63 % of the children showed relapse in problem behaviour in the gradual placebo substitution group as opposed to the 13 % that continued to receive risperidone.

Risperidone showed persistent efficacy and good tolerability for intermediate length of treatment for children with ASD and ID. It seems that the adverse effect such as somnolence disappeared after a few weeks, but the problem with weight gain persisted. It is not clear whether or not the authors took into account the behavioural adverse effect of withdrawal, which may disappear after a few weeks.

Shea et al.'s (2004) study included 79 children (5–12 years) with ASD of whom 42 had ID and ten with borderline intelligence. The authors used a multicentre parallel design RCT in which 40 children were randomly allocated to receive 1.17 mg/day mean dose of risperidone and 39 to receive placebo for 8 weeks. ABC, NCBR-F, VAS, CGI-C and safety measures were used as outcome measures.

The children in the risperidone group showed 64 % improvement in the ABC-I score as opposed to 31 % in the placebo group ($p<0.01$). The authors also reported significant improvement in the risperidone group according to all ABC subscales, NCBR subscales and VAS. There was CGI global improvement in 87 % of the risperidone group as opposed to 40 % in the placebo group ($p<0.001$). Adverse effects, particularly the extrapyramidal symptoms, were comparable between the two groups. However, mean weight gain in the risperidone group was 2.7 kg as opposed to 1 kg in the placebo group, and somnolence was reported by 78 % of the risperidone group as opposed to 8 % in the placebo group.

Overall this is a good quality study and supports the use of risperidone among children. However, the study sample was relatively small and the follow-up period relatively short. One major criticism of the study is that the children were excluded if they did not respond to risperidone previously. This is likely to produce a major bias in the study. Also there was no correction for multiple testing (Type I error).

Snyder et al. (2002) included in their study 110 children (5–12 years) with ID (52 %) and borderline intelligence (48 %). In a 6-week parallel design RCT, the authors randomised 53 children to receive risperidone at a mean daily dose of 0.98 mg (range 0.4–3.8 mg/day) and 57 children to receive placebo.

NCBR-F-conduct behaviour subscale, ABC, BPI, VAS, CGI and cognitive assessments were used as outcome measures. There was 47 % reduction in the NCBR-F subscale score in the risperidone group as opposed to 21 % in the placebo group ($p<0.001$). The authors also reported a significant improvement in the risperidone group according to all ABC subscales, BPI ($p<0.01$), VAS ($p<0.001$) and CGI ($p=0.001$). The common adverse effects in the risperidone group included weight gain of 2 kg ($p<0.001$), somnolence, headache, appetite increase and dyspepsia. Extrapyramidal symptoms were also more common (13 %) in the risperidone group as opposed to placebo group (5 %) ($p=0.25$).

Overall this is a good quality study and supports the use of risperidone among children. However, the cohort size was relatively small and the follow-up period was short.

Turgay et al.'s (2002) study is the continuation of Snyder et al.'s study (2002). The authors continued to prescribe risperidone on an average dose of 1.38 mg/day to 77 children (5–12 years) with ID and borderline intelligence for 48 weeks in an open-label design. The authors were particularly interested to assess the long-term adverse effects of risperidone among children with ID.

Over the study period, 52 % complained of somnolence, 38 % headache, 36 % weight gain (mean gain was 7.1 kg) and 27 % increased appetite. Prolactin level peaked at 4 weeks and then came down to normal. Extrapyramidal symptoms affected 26 % of the children (mild/moderate). No change was observed in cognitive measures, haematology, vital signs and ECG. Improvement in behaviour was maintained over the 48 weeks of the study.

According to this study, risperidone showed persistent efficacy and good tolerability for intermediate length of treatment for children with ID. Somnolence and weight gain were the common adverse effects. Authors did not check for lipid profile and glucose intolerance.

Two smaller RCTs that included 38 children and adolescents (Buitelaar et al., 2001) and 13 children with ID and ASD (Van Bellinghen & De Troch, 2001) also showed significant improvement in problem behaviour in the risperidone group when compared with the placebo group.

McDougle et al. (1995) in a placebo-controlled RCT included 31 children with ASD, many of whom also had ID. Risperidone (1–6 mg/day) was compared with placebo for the management of repetitive behaviour, SIB, aggression and autism symptoms. Nine out of the 11 participants with ID in the risperidone group improved compared with two out of 13 in the placebo group. Overall, risperidone was found to be superior to placebo on all measures. Mild sedation was reported with risperidone.

Aripiprazole

So far only a handful of papers have been published on the efficacy of aripiprazole in the management of problem behaviour in people with ASD, some of whom also have ID. All these papers are published from the USA and included only children with ASD and no adults. Of these studies, only two are RCTs, both of which are conducted by the pharmaceutical company that produces aripiprazole.

Owen et al. (2009) studied the effect of aripiprazole on the irritability and challenging behaviour in 98 children with ASD in a placebo-controlled RCT over 8 weeks. Aripiprazole showed significant decrease in ABC-I score and significantly greater improvement in CGI-I compared with the placebo group.

Marcus et al. (2009) studied 218 children aged 6–17 years with ASD in a placebo-controlled RCT for the management of irritability. The aripiprazole group showed significant improvement according to the irritability, stereotypy and hyperactivity subscale of ABC. The aripiprazole group also showed significantly greater improvement in CGI and the quality-of-life measures.

Summary

Most evidence for the new antipsychotic medications was based on RCTs on risperidone apart from two RCTs on aripiprazole. There are also some RCTs conducted on the older antipsychotics such as chlorpromazine and haloperidol (see Table 19.2). It appears from the RCTs available so far that there is at present equivocal evidence for the efficacy of risperidone among adults with ID with problem behaviours, two studies showing positive and one showing negative findings. According to the evidence based on studies on children with ID (with or without ASD), risperidone seems to be effective in the management of problem behaviours. However, the main concern about using risperidone is its adverse effects such as somnolence and weight gain (not much evidence is available from the RCTs on other adverse effects such as metabolic and cardiac). Long-term follow-up studies among children are reassuring, showing that initial improvement continues over many weeks and overall, the adverse effects are tolerable.

Table 19.2 Number of studies using older antipsychotic medications

Type of problem behaviour studied (number of studies)	Range of number of participants included in different studies	Randomised
SIB (>21)	1–141	2
Stereotypy (14)	1–100	11
Aggression (22)	3–316	6
Hyperactivity (26)	6–396	10

Antidepressants

On the whole, ten studies were found in the systematic review (see Table 19.3 for the characteristics of these studies). Further information on the antidepressants is provided in the systematic review by Sohanpal et al. (2007).

Of these studies, there was one RCT (Lewis et al., 1995), which investigated the effectiveness of the tricyclic antidepressant clomipramine. The remaining studies explored the effectiveness of selective serotonin reuptake inhibitors (SSRIs). One cohort study (Troisi et al., 1995) and two open trials (Cook et al., 1992; Markowitz, 1992) looked at the efficacy of fluoxetine. Of the prospective case-series studies, there was one regarding fluoxetine (Bodfish & Madison, 1993), two on fluvoxamine (La Malfa et al., 1997, 2001) and one on paroxetine (Davanzo et al., 1998). In addition, there was one retrospective, uncontrolled study on paroxetine (Janowsky et al., 2005) and one on both paroxetine and fluoxetine (Branford et al., 1998).

Summary

The existing evidence on the use of antidepressants for the management of problem behaviour in adults with ID is scant. The study on clomip-

Table 19.3 Summary of systematic review on antidepressants

Author/evidence category (EC)	Medication/average daily dose	Target behaviour	Type of study	*N*/duration of follow-up (FU)	Outcome measures	Results
Troisi, Vicario, Nuccetelli, Ciani, and Pasini (1995), EC III	Fluoxetine 20 mg (as add-on)	Aggression (all had co-morbid epilepsy)	Prospective uncontrolled	*N* = 19/FU: overall median of 36 weeks	Modified Overt Aggression Scale (MOAS)	In 47 % there was deterioration, 42 % showed no appreciable change and 11 % had some improvement
Bodfish and Madison (1993), EC III	Fluoxetine dose range 20–80 mg (as add-on)	Compulsive behaviour, SIB, aggression (5 had co-morbid epilepsy)	Prospective uncontrolled	*N* = 16/FU: 4 months	Unspecified outcome measure; documentation of discrete episodes of target behaviour	44 % were classified as responders. 67 % of non-responders had increased mean level of target behaviour
Cook, Rowlett, Jaselskis, and Leventhal (1992), EC III	Fluoxetine dose range 20–80 mg (as add-on)	Perseverative behaviours including SIB to complex rituals (2 had co-morbid psychiatric illness)	Prospective uncontrolled	*N* = 23 but 10 applicable adults/ FU: variable—7–467 days	CGI	60 % improved, 40 % showed no improvement
Markowitz (1992), EC III	Fluoxetine dose range 20–40 mg (as add-on)	Aggression, SIB, obsessive-compulsive behaviours, social relatedness (most had co-morbid psychiatric illness)	Prospective uncontrolled	*N* = 20/FU: 3 months	Direct caretaker observation	60 % markedly improved, 20 % moderately, 10 % mildly, 10 % had no improvement and 5 % discontinued due to adverse effects
La Malfa, Bertelli, and Conte (2001), EC III	Fluvoxamine 250 mg	Aggression, aversive behaviour	Prospective uncontrolled	*N* = 60/FU: 6 weeks	Handicaps, behaviour, and skills schedule (HBSs), DOTES	Severity of aggression decreased with medication
La Malfa, Bertelli, Ricca, Mannucci, and Cabras (1997), EC III	Fluvoxamine 300 mg	Aggression, SIB	Prospective uncontrolled	*N* = 14/FU: 6 weeks	3-hourly functional analyses, CGI, PIMRA, DASH	Functional analysis showed decrease in aggression and DASH SIB score showed reduction. CGI showed improvement after 4 weeks of treatment

Lewis, Bodfish, Powell, and Golden (1995), EC I	Clomipramine titrated up to 225 mg (add-on in *N*=4) vs. placebo	Stereotypy, repetitive SIB and compulsive behaviour	RCT crossover	*N*=10/FU: 19 weeks	ABC, 5-point Likert scale for intensity of repetitive behaviour, treatment emergent side effects scale	Improvement was seen in body and object stereotype behaviours. Six improved in 1 or more repetitive behaviours
Janowsky, Shetty, Barnhill, Elamir, and Davis (2005), EC III	Paroxetine dose range 10–40 mg (as add-on)	Aggression towards others, SIB, destructive behaviours	Retrospective uncontrolled	*N*=38 but 14/relevant FU: 6 months	Psychologists' target behaviour ratings, 7-point rating scale for global and specific maladaptive behaviours	SIB and destruction/ disruptive behaviour ratings significantly decreased, aggression ratings did not significantly decrease
Branford, Bhaumik, and Naik (1998), EC III	Paroxetine dose range 20–40 mg and fluoxetine dose range 20–80 mg (add-on for both)	Perseveration of rituals, maladaptive behaviour such as aggression and SIB (10 had co-morbid epilepsy)	Retrospective Uncontrolled	*N*=33/FU: –	CGI	Overall SSRIs showed in 40 % of cases no benefit, 24 % deteriorated and 36 % had a reduction in perseverative and maladaptive behaviour
Davanzo, Belin, Widawski, and King (1998), III	Paroxetine 35 mg (add-on in *N*=8)	Aggression, SIB	Prospective uncontrolled	*N*=15/FU: 4 months	Observations of severity and frequency of target behaviours	Only severity, not frequency of aggression reduced over the whole treatment period

EC—I, randomised controlled trial (RCT); II, controlled study without randomisation; III, other nonexperimental studies such as case series, *SIB* self-injurious behaviour, *ABC* Aberrant Behaviour Checklist, *CGI* Clinical Global Impressions Scale, *DOTES* Dosage Record and Treatment Emergent Symptom Scale, *PIMRA* Psychopathology Instrument for Mentally Retarded Adults, *DASH* Diagnostic Assessment for the Severely Handicapped

ramine showed beneficial effects (Lewis et al., 1995), but the cohort size was very small ($N = 10$). However, responses to the SSRIs were varied; whereby some studies reported clear favourable results (Janowsky et al., 2005; La Malfa et al., 1997, 2001; Markowitz, 1992), some showed negative effects (Bodfish & Madison, 1993; Branford et al., 1998; Troisi et al., 1995) and other studies demonstrated both positive and negative outcomes (Cook et al., 1992; Davanzo et al., 1998). This discrepancy in findings, therefore, makes it difficult to come to a definite conclusion regarding the effectiveness of antidepressants in this context.

Improvements were largely reported in SIB and perseverative/compulsive behaviours. It may, therefore, be the case that medications were in actual fact treating underlying behaviours that are part of the Obsessive Compulsive Disorder (OCD) spectrum for which SSRIs are indicated anyway. Not surprisingly the antidepressants were most effective in the management of problem behaviour when depression or anxiety was present in the background. In a number of cases, deterioration in behaviour is reported which may have been caused by the adverse effects of some of the antidepressants.

In general, the majority of the evidence based on open trials and case-series studies was fraught with methodological concerns. The small sample sizes meant that the studies were statistically underpowered and often control groups were not recruited. There was a dearth of validated outcome measures utilised and where more than one assessor conducted the outcome measurements, inter-rater reliability was not contemplated.

The efficacy of antidepressants certainly deserves more attention in research, as there is evidence to suggest (Unwin et al., 2011) that these medications are used commonly in the management of problem behaviours in people with ID. This review does not suggest that they are ineffective but that there is not enough good quality evidence for their usefulness at present.

Mood Stabilisers (Lithium and Antiepileptic Medication)

Summary of the findings from the mood stabiliser systematic review is presented in Table 19.4. Further information on the effectiveness of mood stabilisers is presented in Deb et al.'s systematic review (2008). Eight studies on mood stabilisers were extracted through the systematic reviews, one of which on lithium (Tyrer et al., 1993) was published in a book, which was not peer reviewed. The other four studies included one retrospective case-series study on lithium (Langee, 1990). There were one prospective (Verhoeven & Tuinier, 2001) and another retrospective (Ruedrich et al., 1999) case series, both on the effectiveness of sodium valproate. The fourth was a retrospective study of effectiveness of topiramate in the management of problem behaviours in adults with ID (Janowsky et al., 2003). Two further studies explored the effects of lithium, one of which consisted of adults and children with ID (Tyrer et al., 1984) and the other adults only (Craft et al., 1987). The third relevant study was on carbamazepine (Reid et al., 1981).

Summary

There are only a small number of RCTs on mood stabilisers primarily on lithium. However, the RCTs on lithium are dated and of poor quality as they included primarily inpatients, included small number of patients and used questionable outcome measures that are not validated. Some studies showed effectiveness of lithium on particular problem behaviours, but not on others. There is also a major concern for using lithium on patients with severe ID who cannot consent to treatment because once started it is difficult to withdraw lithium. Therefore, it may not be ethical to prescribe lithium to someone who cannot consent to a treatment which has potential long-term major adverse effects and narrow window between therapeutic serum level and toxic level.

Table 19.4 Summary findings of systematic review of mood stabilisers (lithium and antiepileptic medications)

Author/evidence category (EC)	Medication/average daily dose	Target behaviour	Type of study	*N*	Outcome measures	Results
Tyrer, Aronson, and Lauder (1993), EC II	Lithium 500 mg adjusted to achieve 0.5–0.8 mmol/L plasma levels	Aggression, SIB, destructive behaviour, tantrums, hyperactivity	Controlled, crossover	52	VAS	54 % improved on lithium, 44 % unchanged and 2 participants dropped out
Langee (1990), EC III	Lithium dosage adjusted to achieve 0.7–1.2 mmol/L plasma levels	Aggression, SIB, hyperactivity	Retrospective uncontrolled	66	Behaviour disturbance index (severity × frequency)	47 % improved of whom 77 % required additional medication and 53 % remained unchanged
Craft et al. (1987), EC I	Lithium dosage adjusted to achieve 0.7–1.0 mmol/L plasma levels	Aggression	Controlled, crossover	Study group: 22, controls: 20	Scores of counts of frequency and severity of target behaviour	73 % improved, 9 % got worse and 18 % remained unchanged. 30 % improved on placebo
Verhoeven and Tuinier (2001), EC III	Sodium valproate 1,345 mg	SIB, aggression, hyperactivity, disorganised behaviour, Stereotypies, impulsivity	Prospective uncontrolled	28	VAS, CGI	68 % showed some degree of improvement and 32 % minimally improved or remained unchanged
Ruedrich, Swales, Fossaceca, Toliver, and Rutkowski (1999), EC III	Sodium valproate 920 mg	Various but primarily SIB and aggression	Retrospective uncontrolled	28	Monthly behaviour counts, CGI	71 % markedly improved, 21 % mildly improved, 1 remained unchanged and 1 got worse
Janowsky, Kraus, Barnhill, Elamir, and Davis (2003), EC III	Topiramate 202 mg	Aggression, SIB, destructive/disruptive behaviour	Retrospective uncontrolled	22	Cumulative frequency recordings, global severity ratings	74 % improved, 1 remained unchanged and 4 got worse
Reid, Naylor, and Kay (1981), EC II	Carbamazepine 25–26 µg/L	Overactivity	Controlled, crossover	10	Nurse's behaviour ratings	Overall, 40 % improved on carbamazepine and 40 % on placebo
Tyrer, Walsh, Edwards, Berney, and Stephens (1984), EC II	Lithium 500 mg adjusted to achieve 0.5–0.8 mmol/L plasma levels	Aggression	Controlled, crossover	26	VAS, nurse behaviour ratings	68 % improved on lithium

Evidence categories—I, randomised controlled trial (RCT); II, controlled study without randomisation; III, other nonexperimental studies such as case series, *SIB* self-injurious behaviour, *VAS* Visual Analogue Scale; *CGI* Clinical Global Impressions Scale

In some people with severe and profound ID, it may not be possible to carry out blood tests that are mandatory. There are also potentially less toxic alternatives to lithium available which may not require regular blood tests. Within this context it is difficult to recommend lithium for use in people with severe and profound ID unless absolutely necessary. In Unwin et al.'s (2011) prospective 12-month follow-up study, there is little evidence of the use of lithium by the UK psychiatrists. Unfortunately currently there is not much evidence for the effectiveness of other mood stabilisers such as sodium valproate, carbamazepine and lamotrigine, which may provide a better alternative to lithium. However, lack of evidence does not mean that there is evidence that these antiepileptic mood stabilisers are not effective in the management of problem behaviour in people with ID.

Antianxiety Medications/ Beta-Blockers

King and Davanzo (1996) reported in a prospective uncontrolled study of 26 adults with ID (age range 25–63 years) (46 % male) of the effect of buspirone 25–60 mg/day (average 52 mg/day) on aggression and/or SIB. This study did not show any improvement from buspirone.

Summary

There is little evidence currently to recommend any antianxiety medication for the long-term management of problem behaviours in people with ID. The benzodiazepine group of medications carries the risk of tolerance and dependence in the long run. The evidence for the effectiveness of buspirone is currently poor, therefore, cannot be recommended. However, for the general population, some SSRIs, SNRI, pregabalin and quetiapine are now recommended treatment for anxiety-related disorders (Bandelow et al., 2008; NICE guide on the management of anxiety disorders; www.nice.org.uk). In the field of ID, some antipsychotics are prescribed in a smaller than antipsychotic dose to manage problem behaviours with the assumption that at a lower dose, antipsychotics work as an antianxiety medication, although the evidence to support this assumption currently is not available from the literature.

Opioid Antagonists

On the whole, four studies were found in the systematic review on the opioid antagonists that included adults (see Deb & Unwin, 2007b). Three of the studies were prospective trials (Sandman et al., 1993, 2000; Willemsen-Swinkells, Buitelaar, Nijhof, & Van Engeland, 1995) and one was a retrospective case-series study (Casner, Weinheimer, & Gualtieri, 1996). Only one study on children (Campbell et al., 1993) is described in this chapter. The characteristics of these studies are summarised in Table 19.5.

Summary

There are only a handful of RCTs on naltrexone that included a small number of participants, different doses and crossover design, which has its drawbacks. The findings are equivocal in that some showed beneficial effect from naltrexone and others did not. One study showed differential effect depending on the dose, particularly the higher dose being effective and lower doses being noneffective.

Psychostimulants

Most studies of psychostimulants have been used on people with a diagnosis of Attention Deficit Hyperactivity Disorder (ADHD). Therefore, almost all the studies used ADHD symptoms as outcome measures than problem behaviour per se, although problem behaviours are often included in the outcome measures as part of the ADHD symptoms. Therefore, it is difficult to find any evidence to prove effectiveness of psychostimulants specifically for the management of problem behaviour per

Table 19.5 Summary of findings from systematic review on the opioid antagonists

Author/evidence category (EC)	Medication/average daily dose	Target behaviour	Type of study	*N*	Outcome measures	Results
Sandman et al. (2000), EC III	Naltrexone single dose 0.5, 1.0 or 2.0 mg/kg	SIB	Double blind, prospective (follow-up of Sandman et al., 1993)[a]	15 (including 1 under 18 years of age)	Direct observations	Subgroup of participants showed a decrease in SIB for 1 year after acute exposure to naltrexone. Five showed an increase in SIB after long-term treatment
Casner et al. (1996), EC III	Naltrexone 96.8 mg	SIB	Retrospective uncontrolled	56	Retrospective review of behavioural data	57 % responded to naltrexone. Blind review showed a 50 % reduction in SIB in 25 % of participants
Willemsen-Swinkells et al. (1995), EC I	Naltrexone *N* = 19: 50 mg, *N* = 14: 150 mg	SIB	Controlled, crossover	33	ABC, CGI, target symptom checklist, some had direct observations	No therapeutic effect with naltrexone
Sandman et al. (1993), EC III	Naltrexone doses of 0.5, 1.0 and 2.0 mg/kg	SIB	Prospective trial[a]	24 (including 1 under 18 years of age)	Direct observations, ONE, adapted version of Conners' Parent-Teacher Questionnaire, SOME, DISCUS, stereotypies checklist	Naltrexone decreased SIB by 50 % at a dose of 2 mg/kg in 52 % of participants. Single-dose effect of at least a 25 % reduction was apparent in 86 %
Campbell et al. (1993), EC I	Naltrexone 1 mg/kg	Aggression	RCT	Study group: 23, controls: 18	CGI and NGI combined to give Global Clinical Consensus (GCC), aggression rating scale, naltrexone side effects checklist	No therapeutic effect with naltrexone

Evidence categories—I, randomised controlled trial (RCT); II, controlled study without randomisation; III, other nonexperimental studies such as case series, *SIB* self-injurious behaviour, *ABC* Aberrant Behaviour Checklist, *CGI* Clinical Global Impressions Scale, *ONE* Objective Neurological Examination, *SOME* Summation of Maladaptive Expression, *DISCUS* Dyskinesia Identification System—Condensed User Scale

[a]No comparison was made between treatment vs. placebo and, hence, these trials cannot be allocated an evidence category rating of II or I

se in people with ID without a diagnosis of ADHD. One study by Aman and Singh (1982) used an RCT design to compare methylphenidate with placebo for the management of different problem behaviours among 28 participants (age 13.6–26.4 years) with ID. Overall no significant effect was found from the medication.

Vitamins and Others

The only study available on the effectiveness of diet (zinc supplement) on the management of problem behaviour (pica) did not include a proper placebo control group (Lofts, Schroeder, & Maier, 1990). Therefore, it is difficult to draw any conclusion from this study on the effectiveness of diet.

Conclusion

The evidence presented in this chapter on the effectiveness of psychotropic medications has to be interpreted with caution. Most studies in this field are case reports on a small number of participants. It is known that studies with positive findings are more likely to be published than studies with negative findings. This is likely to create a reporting bias for the published case reports. There are only a few RCTs, but they often used a small cohort size, resulting in insufficient statistical power to draw firm conclusions. The outcome measures used are often not appropriate or validated. The method of selection of the control and the experimental group is not always clear or appropriate, and outcome data are often not presented in an appropriate manner. For example, most studies neither quote the 'number needed to treat' (NNT) nor use analysis based on the 'intention to treat' (ITT) model. Most studies do not distinguish symptoms of psychiatric illness from those of problem behaviours, and often researchers do not take into account the existence of autistic and ADHD symptoms in the context of problem behaviour. Also in many studies, participants with comorbid psychiatric disorders were not excluded. It, therefore, remains unclear whether the psychotropic medications used in these studies treated the underlying psychiatric condition or the problem behaviour per se. It is important, however, to recognise the difficulty in carrying out RCTs involving people with ID (Oliver-Africano et al., 2010), particularly because of securing consent in adults who lack capacity. Subsequently these people are deprived of the opportunity to have treatments that are based on strong evidence.

Problem behaviours are usually long-standing; therefore, short follow-up periods used in most studies meant that it is not possible to know whether patients would derive any benefit in the long term. Only a long-term follow-up will determine the effect of many confounding factors such as environmental changes that are concomitant with the use of psychotropic medications. Most studies do not take into account the confounding effect of concomitant non-medication-based management of behaviour, which may have a profound effect on the behaviour. Similarly in most studies the antipsychotics were used as an add-on therapy, which made it difficult to tease apart the confounding effects of the other medications that have been used simultaneously. For example, the use of antiepileptic medications is common among adults with ID (Deb, 2007) and these medications may have an effect on the behaviour. However, an RCT design should take care of some of these confounding factors.

Another problem of interpreting the case report-based data is that many patients who showed improvement on a particular medication may have had an unsuccessful trial of other medications that have been shown to be effective in other case studies. Therefore, the individualised response to specific medication is always going to be difficult to determine. There may be many causes for problem behaviours among people with ID and many factors including medical, psychological and social may influence behaviour. It is, therefore, imperative to carry out a detailed assessment of the causes and consequences of problem behaviours before an intervention is implemented. However, none of the studies provide any detail of behaviour analysis. This sort of issue could be addressed by including an overall

quality of life measure. Future studies should also assess the effect of interventions on family carers' burden and cost-effectiveness.

On the basis of the evidence available, it is difficult either to recommend or to refute the use of psychotropic medications for the management of problem behaviours in people with ID. Furthermore, there is no evidence to show effectiveness of particular psychotropic medication for particular problem behaviours. In the absence of this evidence, guidelines have been developed in order to provide advice to clinicians when using psychotropic medications for the management of problem behaviours in people with ID (Banks et al., 2007; Deb, Clarke, & Unwin, 2006; Deb et al., 2009; Einfeld, 2004; Reiss & Aman, 1998; Unwin & Deb, 2010). These guides advise that a thorough assessment of the causes and effects of the problem behaviours including organic, psychiatric, psychological and social factors should be carried out before a medication is prescribed. Before initiating medication, a formulation should be documented including the assessment and a rationale for the use of medication. Non-medication-based management of problem behaviours should always be considered and be used either instead of or along with medication when necessary. People with ID and their carers as well as the multidisciplinary team should be fully involved in the decision-making process from the outset (Hall & Deb, 2008). There are accessible versions of information leaflets (with audio versions) on psychotropic medications (Unwin & Deb, 2007) freely available for downloading from the web (www.ld-medication.bham.ac.uk). These should be handed over to patients and their carers where appropriate. The time, methods and personnel to conduct the follow-up assessment should be recorded at the outset. Both the impact of the intervention on the behaviour as well as the adverse events should be assessed as objectively as possible, if necessary using validated instruments. At each follow-up, the original formulation should be reassessed; non-medication-based interventions should be considered along with the possibility of withdrawing medication. The psychotropic medication, if needed, should be used with as small a dose as possible for as short a period of time as necessary. If medication is withdrawn, a relapse plan should be in place and the possibility of withdrawal symptoms in the form of problem behaviours should be considered before taking a decision to reinstate any psychotropic medication. The ultimate aim of the management should be symptom reduction as well as to improve the quality of life of the individual with intellectual disability.

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Psychological Treatments

20

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Evidence-Based Practice and Intellectual Disabilities

Psychological and behavioral disorders have significant impacts on the daily lives of millions of people (Kessler & Wang, 2008; Merkiangas et al., 2010) with depression alone accounting for 12.1 % of years lived with disability (Ustun, Ayuso-Mateos, Chatterji, Mathers, & Murray, 2004). Approximately half the population meets the criteria for one DSM-IV mental disorder within their lifetime (Kessler & Wang, 2008). Many of the most impairing mental disorders have an onset during childhood or adolescence and can persist throughout an individual's lifetime (Kessler & Wang, 2008), and nearly one in six children experience some form of developmental disability (Boyle et al., 2011). The health care costs are also very large (Croen, Najjar, Ray, Lotspeich, & Bernal, 2006; Simon, Ormel, von Korff, & Barlow, 1995).

Rationale for an Evidence-Based Approach

Advances in psychological treatment have often been dogged by treatment fads and the vociferous arguments of some clinicians as to the importance of intuition and clinical judgment in research and practice (Chambless & Hollon, 1998; Dunnette, 1966; Lilienfeld, 2010). This is somewhat understandable. Many scientific theories and research findings conflict with personal experience, common sense, or standard practice (Lilienfeld, 2010). Highly publicized treatment fads publicized with a string of anecdotes provoke demand for new and apparently promising interventions for people desperate for help, even from treatments that lack scientific evidence such as specialized diets, chelation, and hyperbaric oxygen (Metz, Mulick, & Butter, 2005). Unfortunately, intuition and passion—both lay and professional—are often inaccurate guides to treatment evaluation and selection. Thus, fad treatments, including harmful treatment fads, continue after their ineffectiveness has been empirically supported (Lilienfeld, 2010). For these reasons, there has been a tremendous push in recent years to determine which interventions are evidence-based so that practitioners can be informed about and use effective treatments (Levant & Hasan, 2008; Sturmey & Hersen, 2012a, 2012b).

The American Psychological Association defined evidence-based practice in psychology as "the integration of the best available research

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with clinical expertise in the context of patient characteristics, culture and preferences" (American Psychological Association, 2006, p. 273). For a treatment to be considered evidence-based, it must meet certain operationalized criteria. For example, Chambless and Hollon (1998) defined "efficacious interventions" as those that are "demonstrated in controlled research in which it is reasonable to conclude that benefits observed are due to the effects of treatment and not to chance or confounding factors such as the passage of time, the effects of psychological assessment, or the presence of different types of clients in the various treatment conditions" (p. 8). Most authorities define EBTs only in terms of a certain number of well-conducted randomized controlled trials (RCTs); some also include a specified number of well-conducted small *N* experiments (Chambless & Hollon, 1998; Horner et al., 2005).

A well-designed RCT can be defined in terms of a well-conceptualized research question; a design that answers that question; a clearly defined and relevant population; an appropriately described and conducted randomization procedure; complete and clear reporting of participants recruited and retained at all stages of the study; the use of manualized interventions; treatment integrity data; evaluation on appropriate, valid, and reliable dependent measures; appropriate data analysis procedures; and some evaluation of the social significance of behavior change (Chambless & Hollon, 1998; Mohrer et al., 2010). Well-conducted small *N* experiments can be characterized as having adequate baseline, clear experimental control, treatment integrity data, a reliably measured dependent variable, and social validity (Chambless & Hollon, 1998; Horner et al., 2005). It is also desirable for both RCTs and small *N* experiments to also have generalization and maintenance data. EBP should also disseminate evidence-based practices that address common, treatment-responsive problems that effectively and efficiently treat the person's problems and also result in wise investment of limited public resources that result in economic benefit to the person or society who pays for treatment. EBP should also be responsive to the informational and training needs of practitioners by making information readily available in an economic and appropriately summarized and synthesized form that easily answers the practitioner's clinical question and provides ready access to treatment manuals and supporting training (Addis, Wade, & Hatgis, 1999; Strauss, Glasziou, Richardson, & Hayes, 2011; Torrey et al., 2001).

EBP remains controversial to some. Many clinicians perceive that evidence and practice may not match (Addis et al., 1999) and that EBP fails to take into account their apparently unique cases and environments that they work in. There are a number of misconceptions about EBPs that should be clarified. First, many clinicians object to EBP because they believe that manualized treatments may disrupt the therapeutic relationship (Addis et al., 1999; Lilienfeld, 2010), limit therapist creativity, and may not be applicable to complex treated outside research settings. The threat to the therapeutic alliance may be overstated (Carroll, Nich, & Rounsaville, 1997) since manualized treatments may result in therapeutic alliance that is stronger than those in treatment-as-usual (Addis et al., 1999). Similarly, some studies have found that manualized treatments developed to treat a narrowly defined population may be quite effective in more heterogeneous populations (Lilienfeld, 2010).

The failure to adhere to EBP is personally and financially costly. Individuals with depression, panic attacks, and eating disorders often fail to receive empirically supported psychotherapies and behavioral interventions (Lilienfeld, 2010). Similarly, one-third or more of individuals with autism spectrum disorders receive non-EBPs, such as sensorimotor integration therapy and facilitated communication, which have been assessed in multiple studies as being ineffective (Lilienfeld, 2007, 2010) and perhaps even harmful. Many people with IDs continue to receive unevaluated treatments, such as TEACCH, counseling, and psychotherapy. The use of ineffective and unevaluated interventions not only potentially wastes client and staff time and money but also deprives clients, their family, and society of the benefit of available EBPs. Ineffective and unevaluated treatments also do harm to families when they falsely raise their hopes and deceive

them before dashing these hopes when these treatments fail (Lilienfeld, 2010). Such treatments fail to reduce family members' distress by leaving family members to face untreated problems when solutions are available.

Unevaluated and ineffective treatments also sometimes do positive harm. For example, false abuse allegations stemming from facilitated communication (Lilienfeld, 2007, 2010) pose significant harm to family members. There is also the potential for client deterioration following different forms of ineffective psychotherapy, such as relaxation for individuals with panic disorder, and substance abuse treatment programs (Lilienfeld, 2007). A second example comes from a recent report from the BBC which showed that only approximately 20 % of children with ASD in France are placed in educational settings and many are institutionalized where they received psychoanalysis and a psychoanalytic treatment known as "packing" in which they are restrained in frozen wet blankets for approximately 45 min (Schofield, 2012). Thus, we should critically assess which psychological treatments are EBPs and develop and implement ways to disseminate such practices in typical service settings. The following section examined the current state of the evidence base for people with IDs for common psychological treatments, including behavioral interventions, sensory treatments, and other psychological treatments.

When planning this chapter the authors optimistically hoped to review 10 or 12 meta-analyses and systematic reviews of EBP and ID and ASD. We searched PubMed using the terms "(meta-analysis) OR (systematic review) AND ("ment* AND retard*)" on March 21, 2012, and found 3,590 abstracts. We retained only papers that included a systematic search of the literature and that may or may not have used a quantitative analysis and/or synthesis of effect sizes of psychological treatments. We excluded papers on medical treatments, such as psychotropic medications (see Sturmey & Maffei-Almodovar, 2012), diets, and acupuncture. Thus, we retained reports from expert panels, systematic narrative reviews, and quantitative meta-analyses of both RCTs and small *N* experiments. After some preliminary online searches and reference checking, the authors stopped searching systematically any further once more than 90 relevant systematic reviews and meta-analyses were located. When target behaviors or treatments were identified, additional searches were conducted. Thus, this chapter does not present a complete overview of the existing evidence base. Rather, we present a selection of this literature to illustrate the status of evidence for different treatments and some of the issues this evidence raises.

Presenting this literature is challenging, not only because of the large amount of evidence but also because it can be organized in numerous ways depending upon the question being asked. Papers differ with respect to at least three important features: The treatment(s), target behavior(s), and population(s) are reviewed. This presents a complication. A review can be organized by any other of these three features, but these features may not correspond to the question the reader has in mind. For example, this chapter is organized by treatment type, for example, "What is the evidence that music therapy is effective?" However, a reader might have questions about a specific target behavior or population, such as "What are the effective treatments for depression in adults with mild ID?" Due to the large amount of research on behavioral interventions, this section is further divided into general reviews; reviews of some illustrative behavioral interventions, such as differential reinforcement; and treatments of some illustrative target behaviors, such as SIB.

General Reviews

Consensus Panels

One method to identify evidence-based practices is to assemble a panel of experts, ask them to identify and evaluate evidence and/or make ratings to evaluate the effectiveness of different treatments, and to synthesize and disseminate these data in a consensus statement. One of the earliest such consensus statements came from the National Institutes of Health (NIH, 1989) consensus development conference (CDC) statement on treatment of destructive behaviors. The CDC was asked to answer six questions including

"What are the approaches to prevent, treat, and manage these [destructive] behaviors? … What is the evidence that these approaches, alone or in combination, eliminate or reduce destructive behaviors? … What are the risks and benefits associated with these approaches? … What recommendations can be made at present regarding the use of different approaches? … What research is needed on approaches for preventing, treating, and managing destructive behaviors?" (NIH, 1989, p. 3). The CDC panel complied and extended an extensive bibliography; produced, presented, discussed, and revised background papers; drafted and represented a preliminary report for public comment and review; and revised and disseminated the report. The panel identified five treatment approaches: (1) Behavior reduction approaches included aversive conditioning, such as shock therapy, restraint, overcorrection, facial screening, time out, response cost, verbal reprimand, and extinction; (2) behavior enhancement approaches included DRO and DRI; (3) environmental/skills acquisition approaches include compliance training, self-management, communication, and functional independence training approaches; (4) ecological approaches and stimulus-based treatments included scheduling situations and settings associated with low rates of problem behavior and modifying stimuli that trigger destructive behavior; and (5) pharmacological methods included a wide array of psychotropic medications.

The panel reviewed treatment studies that focused on single treatments where possible and used a criterion of 90 % reduction in destructive behavior over baseline and 90 % reduction over baseline in 1–10 days for treatments for rapidity of effect. The CDC concluded that (1) treatment should include multiple elements; (2) treatment may require behavior enhancement methods; (3) treatment should be based on an assessment of medical and psychiatric conditions and a functional analysis of behavior; (4) psychotropic medication was used at disturbingly high levels without robust scientific validation; and (5) behavior reduction procedures should be used "…only if the exigencies of the clinical situation require such restrictive interventions and only after appropriate review. These interventions should only be used in the context of a comprehensive and individualized behavior enhancement treatment package" (p. 15). The panel also called for more research on skills acquisition, new pharmacological approaches, and research on the origins and natural history of destructive behavior. Although based on a careful analysis of the evidence, the last recommendations concerning the possibility of behavior reduction procedures caused an uproar of protest that resulted in the suppression of the report for 2 years (Foxx, 2005).

A later consensus statement was reported by Rush and Frances (2000) and outlined expert consensus guidelines related to people with ID for psychosocial treatment of psychiatric and behavior problems. Eighty-six percent of 48 experts rated the effectiveness of applied behavior analysis, managing the environment, client and/or family education, cognitive behavior therapy (CBT), supportive counseling, and psychotherapy. They also rated 13 specific ABA interventions including behavioral parent and teacher/staff training and social skills and communication training. There was almost uniform agreement on the ratings of the appropriateness of the seven treatments: ABA, managing the environment, and client/family education were rated as first-line treatments ("usually" and "extremely appropriate treatments"). CBT, classical behavior therapy, supportive counseling, and psychotherapy were ranked lowest as second- or third-line treatments ("usually/extremely inappropriate treatments"). Behavioral parent and staff training, social skills and communication training, and accelerating differential reinforcement procedures were uniformly rated as first-choice treatments. In contrast, punishment-based interventions were rated consistently as "least appropriate." Rush and Frances' panel-rated behavioral interventions as more appropriate than other interventions and positive behavioral interventions as more acceptable than punishment-based interventions.

There have been numerous consensus panels relating to early intervention and ASD promulgated by numerous state, national, insurance, and professional organizations which have sometimes resulted in some states mandating insurance

coverage for early behavioral intervention (National Autism Center, 2009; New York State Department of Health, 1999; Scottish Intercollegiate Guidelines Network, 2007; United Healthcare, 2011). These are now so numerous that a comprehensive listing is beyond the scope of this chapter. These panels often ask two related questions: (1) Does intensive early intervention produce any benefits to young children with ASD? (2) Does intensive early intervention result in recovery from ASD? On the first question many panels agree that there is some or good evidence of some effects for some children and perhaps large effects for some subset of children. On the issue of recovery, there is much less agreement across panels, with a few supporting the conclusion that some children may recover, whereas other panels conclude that this is not the case due to the methodological limitations of the studies and the lack of long-term follow-up to support that conclusion. These conclusions are often supported by numerous meta-analyses of RCTs published after 2008 which address a gradually expanding literature on this topic (Eldevik et al., 2009, 2010; Howlin, Magiati, & Charman, 2009; Ospina et al., 2008; Reichow & Wolery, 2009; Spreckley & Boyd, 2009; Virues-Ortega, 2010; Warren et al., 2011). Again, these meta-analyses reach different conclusions perhaps reflecting the fact that they are based on different literatures and use different inclusion criteria and different criteria for "effective."

A consensus panel is a social and political process in which scientific results are evaluated by the panel members who attempt to reach consensus and then by members of the public and government who may or may not agree with the conclusions of the consensus panel. The reasons for agreement and disagreement among scientists and between scientists and the public and government are numerous. Not everyone believes in a spherical earth or biological evolution; there are flat-earthers; some governments fund teaching of creation science and busily suppress inconvenient scientific evidence. As both the history of NIH consensus statement on destructive behavior (Foxx, 2005) and the diverse conclusions reached by different consensus panels on early intervention for ASD show, the consensus panel method is a flawed social process; it is perhaps remarkable that sometimes consensus panels do reach correct conclusions.

General Systematic Reviews and Meta-analyses

Table 20.1 summarizes some illustrative general systematic reviews and meta-analyses. Here we will consider three illustrative. One early important meta-analysis of psychosocial and other interventions comes from Didden et al. (1997). This meta-analysis was notable because it expanded the database of previous meta-analyses (Lennox et al., 1988; Lundervold & Bourland, 1988; Scotti, Ujcich, Weigle, Holland, & Kirk, 1996) by reviewing data from a 30-year period, increasing the number of journals included, and using hand searches of those journals to identify potential papers. The authors conducted a meta-analysis of 482 single-subject treatment studies where intervention was directed toward reducing problem behaviors. The populations, target behaviors, and treatment settings varied widely. There was often evidence of efficacy. For example, based on PND, Didden et al. found that 20.3 % of the treatment procedures were "highly effective," 37.5 % were "fairly effective," 21.9 % "questionably effective," and 20.3 % were found to be "unreliable," suggesting that psychosocial interventions were generally effective.

A more recent systematic review confirmed and expanded these findings. The National Autism Center (2009) building upon earlier expert panels (e.g., New York State Department of Health, 1999) assembled a pilot team and multiple expert panels which identified 7,038 abstracts, of which 5,978 were retained for further review. They included both RCTs and small *N* experiments that included participants with ASD and were peer-reviewed empirical articles reporting change in the behavior of children or adolescents 21 years of age or under. Many of these participants must also have had intellectual disabilities and so there may be relevance to people with ID generally.

Table 20.1 General systematic reviews and meta-analyses and psychosocial treatments of developmental disabilities

References	Description	Main findings and conclusions
Lennox, Miltenberger, Spengler, and Erfanian (1988)	Systematically reviewed 7 journals from 1981 to 1985 and identified 162 studies of decelerative procedures including both psychosocial and medication interventions which they classified into three levels of intrusiveness. They calculated percentage reduction from baseline to intervention	Differential reinforcement and overcorrection accounted for 33 % of interventions. All behavioral procedures resulted an average of more than 50 % reduction in the target behavior. Psychotropic medication resulted in less than 50 % reduction in the target behavior. Mean effect sizes were 62 %, 54 %, and 63 % reduction in target behavior for levels 1, 2, and 3 interventions, respectively. Psychotropic medication had an unusually small effect size (12 %). Some treatments, such as overcorrection for socially inappropriate behavior, were counter-therapeutic
Lundervold and Bourland (1988)	Reviewed 62 experimental studies of treatment of aggression, self-injury, and property destruction. Rated effect size on a three-point scale (0 ≤ 50 % reduction), 1 = 51–74 % reduction, 3 = 75–100 % reduction	The most commonly used interventions were differential reinforcement with or without punishment (57 % of studies). The most effective treatments were response interruption + DRO/DRI and facial screening (mean effect size = 1.8); DRI was second (mean effect size = 1.6). The mean effect sizes for antecedent, reinforcement, and punishment; for antecedent and reinforcement control; and for DRO were 1.2, 1.0, and 0.0, respectively
		Ninety-two percent of studies did not report collateral effects and only 2 % reported negative side effects
Matson and Taras (1989)	Hand searched 23 journals from 1967 to 1987 and found 382 studies on punishment and alternative methods to reduce problem behavior. No effect sizes were calculated	The most common procedures were combined aversives and reinforcement (52 %); non-painful stimuli, such as overcorrection, restraint, and visual screening (46 %); painful stimuli, such as ammonia and sock (43 %); extinction (36 %); and positive procedures, such as reinforcement, increased activities, and self-monitoring (26 %)
Didden, Duker, and Korzilius (1997)	Hand searched 30 journals focused on intellectual and developmental disabilities, treatment procedures for problem behaviors from 1968 to 1994 and found 482 articles. Conducted a meta-analysis using percentage of non-overlapping data	The overall effect size was a mean PND of 73.41 % (SD = 33.42 %). The mean PND for the combined treatment procedure was 80.92 % (SD = 29.64 %) and the mean PND for primary treatments was 71.27 % (SD = 34.13 %). There was a higher mean PND for interventions based on experimental analysis (M = 82.60 %) than ABC analysis (M = 71.54 %), scatterplot and Motivation Assessment Scale (M = 66.02 %), or informal assessment (M = 65.83 %). The authors concluded that 26.5 % of behaviors can be treated quite effectively and 20.3 % of the primary treatment procedures are highly effective
Scotti, Ujcich, Weigle, and Holland (1996)	Reviewed 22 journals over a 5-year period (1988–1994) on intervention research for challenging behavior for persons with developmental disabilities. They found 179 studies	The interventions were most commonly carried out in community settings and the most frequently used interventions included differential reinforcement and skills training. Over 100 different combinations of two or more interventions were used across the studies. Forty-eight percent of studies used some form of functional assessment. Few studies reported data on the majority of standards of practice, but there were increases in the reported use of functional assessment, monitoring of collateral behaviors, follow-up data, generalization, and active programming. The results largely support meaningful improvements in the state of the intervention literature since the time of several previous reviews

(continued)

Table 20.1 (continued)

References	Description	Main findings and conclusions
Prout and Nowak-Drabik (2003)	Searched PsycINFO and MEDLINE from 1968 to 1998 and found 92 studies of psychotherapy. Experts rated outcome on two, 5-point Likert scales of outcomes and effectiveness. Reliability on coding study features and outcomes were 98 % and 87 %, respectively. Nine high-quality studies were then meta-analyzed using Cohen's *d*	Thirty-three percent of studies were behavioral, 13 % CBT/cognitive, 15 % analytic/dynamic, 2 % humanistic, and 37 % others. The mean ratings for outcomes and effectiveness were 3.15 and 2.72 based on 83 studies. The mean effect size was 1.01
Heyvaert et al. (2012)	Searched 3 databases, hand searched reference sections of reviews and 31 journals resulting in 285 studies with 598 individuals. This was used to generate effect sizes and hypotheses concerning treatment efficacy and mediating variables	Interventions were highly effective effect size (=2.96, $p<0.0001$). Age, ASD, type of challenging behavior (aggression, destructive), and interventions type (antecedent, caregiver education) all mediated effect sizes

Entries are ordered chronologically

There were 724 retained papers with 775 total studies which the team rated according to quality using a six-point rating scale. Treatments were rated as "beneficial," "ineffective," "adverse," or "unknown." The team then grouped treatments into 38 types and rated the evidence for each treatment as "established," "emerging," "unestablished," or "ineffective/harmful."

There were 11 established treatments. Five were established treatments for problem behaviors: antecedent package, behavioral package, comprehensive behavioral treatment for young children, modeling, and self-management. There were also 22 emerging treatments, of which only two (imitation training and multicomponent package) were rated as emerging treatments for problem behaviors. Of the 11 established treatments, most were effective for increasing skills such as communication skills, self-regulation, and other alternate behaviors. Thus, the authors concluded that "approximately two-thirds of Established Treatments were developed exclusively from the behavioral literature ….. [and] of the remaining one-third, 75 % represent treatments for which research support comes predominantly from the behavioral literature…" (p. 52). Absent from both established and emerging treatments were many commonly used treatments such as cognitive, sensory, dietary, counseling, and psychotherapeutic interventions.

Didden et al. (2012) provides the most contemporary systematic review of treatments for ID and focused on aggressive behavior, SIB, stereotypic behavior, rumination, food refusal, sleep problems, anxiety disorders, mood disorders, and offending. Here we will illustrate their conclusions by considering aggressive behavior and their overall conclusions. For aggression, Didden et al. concluded that several ABA interventions were effective including DR, noncontingent reinforcement, extinction, and FCT. They also concluded that CBT was effective for individuals with borderline or mild ID, perhaps because of the inclusion of behavioral interventions. They also concluded that social skills training and mindfulness procedures were possibly effective

for people with mild or moderate ID, although it is possible that the effect of mindfulness training is mediated by changing the behavior of caregivers, rather than any hypothesized cognitive mechanism. In summarizing the literature, Didden et al. concluded that a variety of ABA treatments were effective for a wide range of problems and that CBT was effective for aggression, mood disorder, and offending. A range of treatments were classified as possibly effective, including chronotherapy and DR for sleep problems; cognitive therapy, psychotherapy, and light therapy for mood disorder problems; and psychotherapy for offending. Finally, the following therapies were identified as ineffective: SIT and gentle teaching for aggression, SIT for stereotypy, and DR for pica. These conclusions are notable because they begin to illustrate effects of the growing evidence base to answer more nuanced questions, such as which therapies are effective for which target behavior and which therapies have been evaluated and shown to be ineffective.

There have also been several systematic reviews and meta-analyses related to specific populations (see Table 20.2) and specific target behaviors (see Table 20.3). Tables 20.1, 20.2, and 20.3 illustrate the large and growing evidence-based literature. Thus, there are now many systematic reviews and meta-analyses of psychosocial and related treatments. A partial listing of these include (a) general systematic reviews and meta-analyses of a wide array of behaviors (see Table 20.1); (b) systematic reviews and meta-analyses of specific populations such as ASD (Case-Smith & Arbesman, 2008; Ma, 2009; Matson et al., 1996; National Autism Center, 2009; Seida et al., 2009; Sinha et al., 2006), especially related to early intervention (Dawson & Burner, 2011; Diggle & McConachie, 2009; Doughty, 2004; Eldevik et al., 2010; Granpeesheh et al., 2009; Howlin et al., 2009; Kuppens & Onghena, 2012; Levy, Kim, & Olive, 2006; Makrygianni & Reed, 2010; McConachie & Diggle, 2006; McGahan, 2001; Odom et al., 2003; Ospina et al., 2008; Reichow & Wolery, 2009; Rogers, 1998; Rogers & Vismara, 2008; Spreckley & Boyd, 2009), mild ID (Didden et al., 2006), and adults with ASD (Bishop-Fitzpatrick, Minshew, & Eack, 2013); (c) specific problem behaviors, such as aggression (Brosnan & Healy, 2011), pica (Bell & Stein, 1992; Hagopian, Rooker, & Rolider, 2011; McAdam et al., 2004, 2012), stereotypic behavior (May, Kennedy, & Bruzek, 2012), fears and phobias (Jennett & Hagopian, 2008; White, Oswald, Ollendick, & Scahill, 2009), self-injury (Campbell, 2003; Christiansen, 2009; Horner, Carr, Strain, Todd, & Reed, 2002; Kahng, Iwata, & Lewin, 2002; Sturmey, Maffei-Almadovar, Madzharova, & Cooper, 2012), sleep problems (Schreck, 2001), rumination (Lang et al., 2011), pediatric feeding disorders (Sharp, Jaquess, Morton, & Herzinger, 2010), and externalizing behavior disorders (Lundervold & Bourland, 1988); (d) teaching a wide range of adaptive behavior (Banda & Grimmet, 2008; Bellini & Akullian, 2007; Flynn & Healy, 2012; Goldstein, 2002; Luckett, Bundy, & Roberts, 2007; McConnell, 2002; Palmen et al., 2012; Schwartz & Nye, 2006; Snell et al., 2010) including social skills training (Bellini, Peters, Benner, & Hopf, 2007; Burgess & Turkstra, 2006; Gillis & Butler, 2007; Rao, Beidel, & Murrary, 2008; Reichow & Volkmar, 2010; Vaughn et al., 2003; Williams-White, Keonig, & Scahill, 2007), augmentative and alternative communication skills (Ganz, Earles-Vollrath, et al., 2012; Ganz et al., 2011; Millar, Light, & Schlosser, 2006; Schlosser & Lee, 2000; Schlosser & Sigafoos, 2006; Schlosser & Wendt, 2008), self-management (Lee, Simpson, & Shogren, 2007), vocational interventions (Taylor et al., 2012), and purchasing skills (Xin, Grasso, Dipipi-Hoy, & Jitendra, 2005); (e) specific behavioral interventions that are part of PBS (Carr et al., 1999; Marquis et al., 2000) and school-wide components of PBS (Solomon, Klein, Hintze, Cressey, & Peller, 2012); (f) functional analysis of behavior problems (Hanley, Iwata, & McCord, 2003); (g) FCT (Mancil, 2006); (h) differential reinforcement (Chowdhury & Benson, 2010; Petscher, Rey, & Bailey, 2008); (i) choice interventions (Kern et al., 1998; Shogren, Faggella-Luby, Bae, & Wehmeyer, 2004); (j) PECS (Flippin, Reszka, & Watson, 2010; Ganz, Davis, Lund, Goodwyn, & Simpson, 2012; Hart & Banda, 2010; Preston & Carter, 2009; Tien, 2008; Tincani & Devis,

Table 20.2 Systematic reviews and meta-analyses of treatments for specific populations

Population (references)	Description	Main findings and conclusions
Autism (Matson, Benavidez, Compton, Paclawskyj, & Baglio, 1996)	Reviewed 251 studies from hand searches of unspecified journals and psychological abstracts. Articles were then classified on their use of positive, aversive, and combined treatments with people with autism	The most commonly addressed problems were stereotypy (22 %), aggression (17 %), and SIB (14 %). The following procedures were reported: positive (53 %), aversive (20 %), extinction (1 %), and combined (26 %). Positive interventions were used in almost all interventions to increase skills such as social skills, language, daily living skills, and academic skills
Children with autism (Granpeesheh, Tarbox, & Dixon, 2009)	This general review article describes components of comprehensive ABA treatment programs, reviews research on effectiveness, and discusses issues related to collaboration between ABA and psychiatry	The authors concluded that ABA treatment programs for individuals with autism are supported by a significant amount of scientific evidence and are therefore recommended for use and that individual care would likely benefit from a greater degree of collaboration between practitioners in the fields of ABA and psychiatry
Autism (Krebs et al., 2009)	Conducted an "umbrella review" of 30 systematic reviews of treatment up to May 2007	Most systematic reviews were behavioral ($N=9$) or communication therapy ($N=7$). Most interventions reviewed were effective compared to no treatment, but there was little evidence of the relative effectiveness of one therapy over another. Most systematic reviews were rated as being of poor quality and vulnerable to bias
Mild ID (Didden, Korzilius, van Oorsouw, & Sturmey, 2006)	Eighty articles published between 1980 and 2005 were examined on the effectiveness of behavioral and psychotherapeutic treatments for challenging behaviors in individuals with mild mental retardation. Percentage of non-overlapping data (PND and PZD) was calculated	Mean PND and PZD across studies were 75 % ($SD=30$ %) and 35 % ($SD=32$ %), respectively. Mean PND was 80 % in studies using a functional analysis. Relatively high mean ES was found for FCT, antecedent control, and positive practice (but low ES for self-management etc.). Behavioral interventions for challenging behaviors are effective with people with mild mental retardation
Sallows and Graupner (2012)	Reviewed consensus panels and meta-analyses of treatment of comprehensive treatment programs, including ABA, TEACCH, developmental interventions, such as SCERTS, social skills interventions, increasing social skills, communication interventions, and inappropriate behavior	Concluded that: 1. The only effective, efficacious, and specific intervention was comprehensive ABA programs 2. Naturalistic methods were efficacious, but not comprehensive 3. Efficacious methods included video modeling, self-management, peer-mediated interventions, learning scripts, increasing toy play for self-stimulation, DRO for problem behavior, punishment for problem behavior (which consensus panels recommend combining with reinforcement), and PBS 4. Possibly efficacious interventions included priming, social stories, FCT, and behavioral momentum 5. Interventions that are not evidence-based due to the lack of evidence include TEACCH and social skills groups due to the lack of evidence and perhaps poor design of social skills groups
Harvey et al. (2009)	Updated Scottie et al. by searching ERIC and Psych Lit and 22 journals from 1988 to 2006. A pool of 1,086 articles was reduced to 72 Three hundred and five participants with high reliability	Seventy-six percent of articles were AB designs and 23 % were true experiments. The authors calculated multiple effect size measures. Results are complex due to use of more than one effect size measures. They concluded that "psychological (behavioral) treatments … can clearly reduce even the most severe challenging behavior. However, there is no one intervention used alone or in

Note that papers are presented chronologically and those that dealt with specific behaviors in specific populations were placed in Table 20.3

Table 20.3 Systematic reviews and meta-analyses of specific target behaviors

Target behavior	Description	Main findings and conclusions
Augmentative and alternative communication (Schlosser & Lee, 2000)	Searched 46 journals from 1976 to 1995 using searches of databases, hand searches, and footnote chasing. Reviewed 50 small *N* experiments with 232 comparisons of treatment phases. Calculated PND using PND and percentage of overlapping data (POD). Focused on generalization and maintenance	Participants were children with multiple disabilities, intellectual disabilities, autism, and physical disabilities. Forty-five percent of interventions were "highly effective" (PND > 90 %) and 43 % were "fairly effective" (PND = 70–90 %). Train and hope and multiple exemplar training produced the largest effect sizes for generalization
Children with autism (Goldstein, 2002)	Sixty studies published between 1978 and 2000 on speech and language interventions for children with autism were reviewed	The most commonly used interventions included signing, total language training, discrete trial teaching, natural language milieu training, communication training to replace challenging behavior, social interactions, and group interventions
Pica (McAdam, Sherman, Sheldon, & Napolitano, 2004)	A systematic review of small *N* experiments	The most commonly evaluated interventions were noncontingent food (*N* = 5), overcorrection (*N* = %), and physical restraint (*N* = %)
FCT in children with autism (Mancil, 2006)	Conducted a systematic review without a meta-analysis. Searched 4 behavioral journals by hand and found 8 small *N* experiments on FCT for children with autism	When implementing the procedures during initial development stages, under controlled conditions, research teams produced positive behavioral and communication results. Some research studies reported increases in communication and decreases in challenging behaviors. Limitations still exist with respect to maintenance and generalization and the lack of natural change agents
AIT in children and adults with autism (Sinha, Silove, Wheeler, & Williams, 2006)	The authors systematically reviewed six randomized controlled trials of auditory integration training for adults and children with autism published between 1993 and 2000	Of the six RCTs, three showed no benefit from AIT over control conditions and the remaining three found AIT to be effective at 3-month follow-up with measures of questionable validity. Based on the inconsistent results, the authors concluded that the use of AIT is not supported by the literature
Hand mouthing (Cannella, O'Reilly, & Lancioni, 2006)	A systematic review which identified 23 studies from 1995 to 2004 from ERIC, PsycINFO, and MEDLINE and coded treatment into 7 categories	18/20 intervention studies produced positive results and 2 produced mixed results. There were a wide range of interventions, including environmental enrichment and choice, multicomponent interventions, naltrexone, and response blocking (see McAdam, Breitman, Levine, & Williams, 2012, for an updated meta-analysis)
Video modeling and video self-modeling in children and adolescents with autism (Bellini & Akullian, 2007)	Twenty-nine studies were identified between the years of 1980 and 2005 on video modeling and video self-modeling for individuals with autism spectrum disorders. Effect sizes were calculated using percentage of non-overlapping data points	Interventions (antecedent, response contingent, and noncontingent procedures) were moderately effective with mean PND = 80 %. Similar effects were found for maintenance and generalization (mean PND = 83 % and 74 %, respectively). Video modeling was somewhat more effective (*M* PND = 81 %) than video self-modeling (*M* PND = 77 %). These interventions were most effective in targeting functional skills and response contingent procedures were more effective than either antecedent or noncontingent procedures

Activity schedules in children and adults with autism (Banda & Grimmet, 2008)	A systematic review of 13 studies on activity schedules for individuals with autism (children to adults) was conducted. Studies were published between the years of 1993 and 2004	Multiple baseline and reversal designs were the most frequently used design in studies. Activity schedules were shown to be effective for all 31 participants across the 13 studies. Social interactions, coping with transitions, and on-task behaviors were all enhanced with the use of activity schedules. There were also decreases shown in disruptive behaviors (e.g., tantrums, screaming, and aggression toward others)
Behavioral interventions for cognitive and language skills and adaptive behavior in preschoolers with autism (Spreckley & Boyd, 2009)	Six randomized controlled trials of applied behavioral interventions for preschool children with autism were reviewed. Four of these studies, published between 2000 and 2007, had standardized mean difference scores calculated	Based on the results of the standardized mean difference calculations, applied behavioral interventions (e.g., parent training) were not significantly better than standard care for cognitive outcomes (SMD = 0.38), expressive language (SMD = 0.37), receptive language (SMD = 29), or adaptive behavior (SMD = 0.30)
Elopement (Lang et al., 2009)	Identified 10 studies with 53 participants	Positive outcomes identified in 80 % of papers. Concluded that function-based treatments may be most effective. Five experiments were rated as providing "conclusive" evidence of treatment effectiveness
Skin picking Lang, Didden, et al. (2010)	Searched several databases and conducted hand searches of journals to identify 223 studies of which identified 16 studies with 19 participants were retained	The most common intervention was DR followed by antecedent- and punishment-based interventions. The mean PND was 97 % (75–100 %) and maintenance was demonstrated from 2 to 35 months
Social skills and autism (Wang & Spillane, 2011)	Searched multiple databases from 1994 to 2008 and identified 7,070 papers of which 13 were retained with 43 participants	Using HLM meta-analysis, the mean effect size was 1.27 (range 0.65–2.31). Both peer-mediated and video-modeling interventions were effective
Rumination and operant vomiting (Lang et al., 2011)	Searched several databases and conducted hand searches of journals to identify 114 papers of which 21 studies 32 participants were retained	Interventions used included dietary manipulation, interventions based on automatic reinforcement, and interventions based on attention or escape functions. Only 3 studies were classified as providing "certain" evidence of treatment effectives and 10 "at the preponderance" level of certainty
Social and self-help skills in individuals with ASD (Flynn & Healy, 2012)	Twenty-two studies were collected between the years 1987 and 2010 on the topic of treatments for social and self-help skills for individuals with ASD. A descriptive review was conducted as categorized by treatment type	Peer-mediated instruction (PMI) was determined to be a very naturalistic teaching method which permitted flexibility in procedure (e.g., incorporating modeling, direct instruction, prompting). Pivotal-response training shared many of the advantages of PMI. Social skills groups were not as advantageous; however, they were shown to be effective in some studies. Video modeling and script fading procedure were promising. More research is needed on all these intervention techniques
Adaptive behavior in higher functioning (IQ > 70) adults with ASD (Palmen, Didden, & Lang, 2012)	Searched 4 databases from 1990 to 2010 and conducted hand searches to identify 106 abstracts of which 20 studies with 116 participants were retained	Interventions addressed social, academic, vocational, and domestic skills of which 65 % had positive outcomes. Five studies were rated as providing "certain" evidence of effectiveness

2011); (k) video and video self-modeling (Bellini & Akullian, 2007; Delano, 2007; Mason, Ganz, Parker, Burke, & Camargo, 2012; Shukla-Mehta, Miller, & Callahan, 2010); (l) behavioral momentum for compliance (Lee, 2005); (m) antecedent exercise (Allison, Faith, & Franklin, 1995; Lang, Keogel, et al., 2010; Sowa & Meulenbroek, 2012); (n) activity schedules (Banda & Grimmet, 2008; Stromer, Kimball, Kinney, & Taylor, 2006); (o) self-management for children with autism (Lee, Simpson, & Shogren, 2007); (p) training caregivers to implement ABA (Brookman-Frazee, Stahmer, Baker-Ericzen, & Tsai, 2006; Rispoli, Neely, Lang, & Ganz, 2011; van Oorsouw, Embregts, Bosman, & Jahoda, 2009) and improve interaction skills (Hamelin & Sturmey, 2011); (q) CBT for children with ASD (White, 2003); (r) facilitative communication (Mostert, 2001); (s) SIT (Lang et al., 2012; May-Benson & Koomar, 2010; Ottenbacher, 1982; Vargas & Camilli, 1998); (t) academic skills such as reading skills in children with disabilities (Browder, Wakeman, Spooner, Ahlgrim-Delzell, & Algozinne, 2006) and children with Down syndrome (Naess, Melby-Lervag, Hulme, & Lyster, 2012) and mathematical problem solving (Montague & Dietz, 2009); and (u) social stories (Reynhout & Carter, 2006, 2011) and peer tutoring (Stenhoff & Lignugaris-Kraft, 2007).

Specific Behavioral Treatments

The previous sections reviewed general treatment systematic reviews and meta-analyses but did not specifically search for behavioral treatments; however, these general reviews identified a very large amount of evidence on behavioral interventions. Thus, this section will illustrate the evidence for the effectiveness of specific behavioral intervention procedures (Table 20.4).

Activity Schedules

Activity schedules are written or pictorial prompts which people with disabilities learn to use to prompt appropriate behavior. The term "activity schedule" is used in the literature in two ways. The first way the term is used is to refer to

Table 20.4 Systematic reviews and meta-analyses of specific behavioral intervention methods

Treatment	References	Description	Main findings and conclusions
Activity schedules	Banda and Grimmet (2008)	A systematic review of 13 studies on activity schedules for individuals with autism (children to adults) was conducted. Studies were published between the years of 1993 and 2004	Multiple baseline and reversal designs were the most frequently used design in studies. Activity schedules were shown to be effective for all 31 participants across the 13 studies. Social interactions, coping with transitions, and on-task behaviors were all enhanced with the use of activity schedules. There were also decreases shown in disruptive behaviors (e.g., tantrums, screaming, and aggression toward others)
Video modeling and video self-modeling	Bellini and Akullian (2007)	Twenty-nine studies were identified between the years of 1980 and 2005 on video modeling and video self-modeling for individuals with autism spectrum disorders. Effect sizes were calculated using percentage of non-overlapping data points	Interventions (antecedent, response contingent, and noncontingent procedures) were moderately effective with mean PND = 80 %. Similar effects were found for maintenance and generalization (mean PND = 83 % and 74 %, respectively). Video modeling was somewhat more effective (*M* PND = 81 %) than video self-modeling (*M* PND = 77 %). These interventions were most effective in targeting functional skills, and response contingent procedures were more effective than either antecedent or noncontingent procedures

(continued)

Table 20.4 (continued)

Treatment	References	Description	Main findings and conclusions
Positive behavior support	Marquis et al. (2000) and Carr et al. (1999)	Positive behavior support for aggression, SIB, property destructions, tantrums, and combinations of these, Carr et al. (1999) searched for articles that (1) were published between 1985 and 1996; (2) in peer-reviewed journals; (3) in English; (4) used DSM or AAMR criteria for developmental disability and other diagnoses; (5) must address aggression, SIB, property destruction, tantrums, and combinations; and (6) used antecedent, reinforcement, or both strategies. Hand searched all journals mentioned in 4 previous meta-analyses; searched reference; and searched topography × disability searches in 9 online databases. This yielded 216 articles from 36 journals of which 109 were retained	PBS was widely applicable to serious problem behavior and could be used in typical settings. PBS is evolving, for example, through increased use of pre-intervention assessments and short-term maintenance (1–5 months) There were modest to substantial increases in adaptive behavior and about half of PBS interventions resulted in >90 % reduction in challenging behavior, of which two-thirds are maintained. Few studies addressed broad lifestyle changes. Studies with pre-intervention assessment produced twice as large an effect as those without
Communication interventions	Goldstein (2002)	Sixty studies published between 1978 and 2000 on speech and language interventions for children with autism were reviewed	The literature provides little direction in terms of service delivery models or the intensity of services that are more likely to maximize communication intervention efforts
Time delay	Browder et al. (2006)	Reviewed 30 experiments on time delay published from 1975 to 2007	Concluded that time delay was an evidence-based practice for teaching picture and sight word recognition
Active support	Hamelin and Sturmey (2011)	A systematic review was conducted on two studies that used experimental designs to evaluate active support as used in agencies for individuals with developmental disabilities. One study was published in 1999 while the other was published in 2007. Percentage of non-overlapping data (PND) and percentage of all non-overlapping data (PAND) were calculated	Only one experiment, out of three identified in the two studies, showed a clear functional relationship between active support with "ineffective" to "questionable" percentage of non-overlapping data points (intervention = 54–66 %, follow-up = 30–50 %) effect sizes and acceptable percentage of all non-overlapping data points effect sizes (intervention = 78–85 %, follow-up = 90–93.3 %). Based on these data, active support only meets Chambless and Hollon's (1998) criterion for a "promising treatment" but not an evidence-based practice
Safety skills	Dixon et al. (2010)	Searched PubMed from 1970 to 2009. Of 509 studies, 13 were retained. Fourteen additional studies were identified from references	Prompting, reinforcement, and role-play appeared to be effective interventions to teach safety skills

a very particular technology developed by McClannahan and Krantz (1999) in which non-social prompts from behind are used to teach the person with a disability to obtain their schedule book, follow their picture schedule, and put materials away; prompts are then faded by shadowing and fading so that eventually the individual behaves independently with no or minimal social prompts. The term is also used more broadly to refer to any form of activity, picture or photographic schedule, or other visual supports to prompt appropriate behavior but which may be taught by a variety of methods; for example, TEACCH includes extensive use of visual supports, but their use is taught by a variety of informal methods. Recently, this technology has been extended to teach children to follow activity schedules on computers (Stromer et al., 2006).

Banda and Grimmet (2008) searched ERIC and PsycINFO and identified 13 data-based experimental studies of individuals with autism published in peer-reviewed journals. There were 28 children in the studies, all of whom were 14 years old or younger and three adults aged 22–40. Target behaviors included increasing on-task behavior, play, and independent work and decreasing problem behaviors. Change agents included psychologists, teachers, and graduate students. Seven studies took place in classroom settings. The authors concluded that activity schedules were effective for all participants. Generalization was assessed in six studies, and social validity was measured in five with positive results in all cases. Banda and Grimmett concluded that multiple features of activity schedules may have promoted their effectiveness such as having predictable, discrete units where the completion of one behavior could serve as a discriminative stimulus for the next and the incorporation of modeling in their use. Future research could extend this study by calculating effect sizes and conducting a meta-analysis of the results.

Video and Video Self-Modeling

Video modeling refers to presenting a videotaped model of appropriate behavior, and video self-modeling refers to presentation of a video of the person behaving appropriately. As video technology has become cheaper, more portable, and more widely available, including through phone technology, video and video self-modeling has become a more readily available intervention.

We identified two meta-analyses of video and video self-modeling. The first comes from Bellini and Akullian (2007) who searched ERIC and PsycINFO from 1980 to 2005, conducted an ancestral search, and hand searched two journals. There were 29 small *N* experiments in peer-reviewed journals which examined of video and video self-modeling in children aged 3–21 with ASD. The mean reliability for coding article features was 98 % and for PND calculations was 100 %. There were 73 participants in the 23 studies aged 3–20 years. The authors found a moderately high PND both for video modeling (15 studies, mean PND = 81 %) and for video self-modeling (7 studies, mean PND = 77 %) and moderate to low generalization effect sizes for maintenance and generalization based on smaller numbers of studies. These modeling procedures were effective for increasing self-help, communication, and adaptive behavior skills. The authors concluded that video modeling and self-modeling meet the criteria for an effective treatment. They also noted that video-modeling interventions are difficult to fully assess since they are most often one component of a treatment package and cannot be evaluated in isolation. Future studies are needed where social validity and treatment fidelity are assessed; however, the authors still suggested that there was sufficient support for video modeling as an evidence-based practice.

A more recent meta-analysis comes from Mason, Ganz, Parker, Burke, and Camargo (2012) who meta-analyzed 42 studies with 126 participants. The number of experiments is larger than the previous meta-analysis perhaps because it was conducted later and perhaps because it included both participants with ASD and with other developmental disabilities. The authors used IRD as their effect size measure. The mean IRD was 0.82 (0.81, 0.83) (“large”) and was larger for participants with ASD (mean = 0.83) than for participants with developmental disabilities

(0.68, $p=0.05$). Effect sizes were significantly larger when video modeling was part of a multicomponent treatment package (mean IRD = 0.88) than when used alone (mean IRD = 0.73, $p<0.05$), and when video modeling was used alone with people with ID, the effect size was "small" (mean IRD = 0.40). A second meta-analysis from Mason, Ganz, Parker, Burke, and Camargo (2012) extended the previous database to 56 small *N* experiments with 177 participants and 233 IRDs. Again, the mean effect size was IRD = 0.82 (0.80, 0.82). A refinement to the earlier meta-analysis was a disaggregation analysis which showed that effect sizes were significantly largest for video modeling with reinforcement (mean IRD = 0.86 [0.84, 0.88]) than for either video modeling alone (mean IRD = 0.81 [0.79; 82]) or packages containing video modeling (mean IRD = 0.75 [0.73, 0.77]), although the magnitude of these differences was quite small.

These three meta-analyses nicely illustrate the developing literature in this area: Bellini and Akullian (2007) made the simple conclusion that video modeling meets the criteria for evidence-based practice. Some 5 years later, using a larger literature and a more sophisticated statistical analysis, Mason, Ganz, Parker, Boles, et al. (2012) and Mason, Ganz, Parker, Burke, and Camargo (2012) can make more nuanced conclusions about the relative effect sizes of procedural variations and different populations.

PECS

PECS is a behavior analytic manualized pictorial communication system to teach children to exchange icons and strips to communicate to teach children to request and comment. The PECS system includes six phases which progress from exchanging an icon to request a preferred item with no distractors present (phase 1) to commenting in response to a partner's questions such as "What do you see?" PECS has become highly popular and widely disseminated. Recently there have been an increasing number of empirical articles on PECS, and this is reflected in four meta-analyses (Flippin et al., 2010; Ganz, Davis, et al., 2012; Hart & Banda, 2010; Preston & Carter, 2009); here we will only discuss the most recent meta-analysis as it is the most comprehensive and offers the most nuanced analysis of the literature.

Ganz et al. (2012) searched three databases using terms related to autism and PECS and found 168 articles and chapters of which 13 were retained for meta-analysis. The mean effect size was IRD = 0.56 (0.49, 0.62, range −0.51 to 0.95, 94 effect sizes, 13 studies), indicating a large positive, but varied outcome. Effect sizes for target variables, such as acquisition of specific PECS communication skills, were larger (IRD = 0.65 [0.59, 0.73] 52 effect sizes, 7 studies) than for nontarget behaviors, such as challenging behavior, social behavior, and acquisition of speech (IRD = 0.45 [0.35, 0.56] 42 effect sizes, 6 studies). Although there was no evidence that age mediated effectiveness, students with autism alone had larger effect sizes (mean IRD = 0.69 [0.61, 0.76], 43 effect sizes, 9 studies) than those with autism and ID (IRD = 0.59 [0.42, 0.78], 2 studies 6 effect sizes). They also found that studies that only implemented the first four phases of PECS had smaller effect sizes (IRD +0.33 [−0.18, 0.83] 2 effect sizes, 2 studies) than those that implemented all six phases (IRD = 0.84 [0.72, 0.96] 6 effect sizes, 2 studies). The authors concluded that PECS "has potential to positively impact student outcomes, albeit additional factors may account for effects" (p. 411). Using Parker et al.'s (2009) criteria, the omnibus, target, and nontarget behavior effect sizes would all be judged "small or questionable." As Ganz et al. noted, the range of outcomes is very wide, suggesting that sometimes PECS results in very large effect sizes and sometimes no effects. Although client characteristics such as ID were associated with smaller effect sizes, of more importance to practitioners is the larger effect sizes when all six phases were implemented, suggesting that clients do best when they complete the entire program, rather than only the first four phases. Again, as with the literature on video modeling, these four meta-analyses illustrate a progression from answering a relatively simple question ("Is this an evidence-based practice?")

to more nuanced and helpful questions ("What aspects of the treatment, that we can manipulate, result in larger effect sizes for specific clients?").

Choice

Choice interventions refer to several procedures in which an individual is given a choice of items, activities or people, or the sequencing in which activities take place; it can also refer to a procedure that caregivers implement or a skill that a client uses independently. Choice also has two different connotations: choice may be seen as a philosophically desirable component of services that are respectful of the person's autonomy or a behavioral phenomenon in which an organism selects one reinforcer from one or more concurrently available schedules of reinforcement. Choice-based procedures have often been used to increase desirable behavior, such as client engagement, and to decrease challenging behavior. There has been one systematic review (Kern et al., 1998) and one meta-analysis of choice interventions (Shogren et al., 2004). In this section we will review the meta-analysis.

Shogren et al. (2004) searched PsycINFO and ERIC to identify articles on choice, problem behavior, and developmental disabilities published before March 2003. They retained 13 studies on choice making as an intervention for problem behavior with 30 participants, all but one of which included children, with developmental disabilities resulting in 56 unique PND and 59 unique PZD scores. The overall mean PND was 65.7 % and the overall mean PZD score was 42.3 %. These effect sizes were of "questionable" magnitude (Scotti et al., 1996). The standard deviations of effect sizes were quite large, suggesting that sometimes effect sizes were very large and sometimes negligible. A disaggregation showed no effects of choice training, type of choice offered, type of activity, type of behavior or functional assessment/analysis, etc.; other than that the PZD was much large for boys than girls (53.7 % vs. 15.6 %). These findings provide preliminary support for the benefit of providing choice-making opportunities as an intervention for problem behavior. This meta-analysis did not report data on adaptive behavior and is based on literature that is nearly 10 years old, so it is possible that a more recent meta-analysis might reach different and more qualified conclusions.

Differential Reinforcement

Differential reinforcement refers to reinforcing some responses and withholding reinforcement from other responses in order to increase some and decrease other behavior. There are at least three procedural variations. In DRO any behavior other than the target behavior is reinforced; in DRA some specified alternate behavior is reinforced; and in DRI a behavior that is physically incompatible with the target behavior is reinforced. DR has been extensively used since the 1960s and is a commonly used behavioral intervention method but depends upon accurate identification and control of reinforcers, correct calculation, modification of the reinforcement interval, and schedule thinning.

There have been several systematic reviews and meta-analyses of DR with individuals with ID and/or ASD (Chowdhury & Benson, 2010; Lennox et al., 1988; Petscher et al., 2008; Prochnow-LaGrow, 1984; Vladescu & Kodak, 2010). We will illustrate this literature with one recent study that explicitly applied Chambless and Hollon's (1998) criteria for evidence-based practice. Petscher et al. (2008) searched ERIC and abstracts from JABA and JEAB from 1977 using terms for DRA including FCT. The initial search identified 538 articles of which 116 articles with 277 participants were retained. Eighty-two percent of participants were children and 18 % were adults, and the most common target behavior was combinations of aggression and disruption. The authors concluded that three forms of DRA were well established (DRA with and without extinction for destructive behavior and DRA with extinction for food refusal) and that DRA plus NCR was an "experimental treatment", since there was only one multiple baseline experiment with only two participants in the only study that the authors found. Chowdhury and

Benson (2011) made more qualified conclusions. Their systematic review of various DR procedures was based on 31 studies with 48 participants published between 1980 and 2009. These authors distinguished three variations in DR: DR alone, DR as part of a package, and DR combined with punishment. They found evidence that DR alone and packages with DR were effective. They also noted, however, several examples where DR alone was ineffective and was only effective when combined with punishment. Such negative effects, echoing the earlier NIH consensus panel (National Institutes of Health, 1989), raise the question of whether DR had been conducted adequately before punishment was added.

Positive Behavior Support

Positive behavior support (PBS) aims to improve the opportunities, lifestyle, and competence of people with disabilities in integrated community settings that result in durable change and a rich lifestyle. PBS uses both traditional behavioral interventions, such as positive reinforcement and choice that appear to be nonpunitive interventions (Carr et al., 1999, 2002), and system-side interventions, such as school-wide positive discipline (Solomon et al., 2012). Marquis et al. (2000) and Carr et al. (1999) conducted a systematic review and meta-analysis of PBS in studies published between 1985 and 1996 which focused on interventions for individuals with developmental disabilities, such as ID, ASD, PDD-NOS, and associated dual diagnoses and self-injury, aggression, property destruction, and tantrums. The authors reviewed 109 studies and extracted effect size data using percentage change over baseline on reduction of problem behaviors, increasing adaptive behaviors, stimulus generalization, response generalization, and maintenance using both antecedent- and consequence-based interventions. Most studies applied PBS to elementary school-age children, and the majority of subjects had severe to profound intellectual disability and engaged in self-injury or aggression (Carr et al., 1999). The number of studies using PBS, the use of functional analyses, and use of stimulus-based interventions rather than reinforcement-based interventions increased over time. Over two-thirds of studies demonstrated 80 % or more reductions of problem behaviors and over 94 % showed increases in positive behaviors. While there were consistently strong maintenance effects shown, only 6.6 and 7.1 % of studies reported data on stimulus and response generalization.

Carr et al. (1999) concluded that PBS for children with developmental disabilities was generally effective but noted a number of limitations with their study including (a) the use of non-PBS interventions used in conjunction with PBS; (b) relatively few studies conducted in community settings by typical change agents such as parents and teachers in school and home settings; and (c) limited data on stimulus generalization, response generalization, social validity, and lifestyle changes. Other limitations in this meta-analysis included the use of percentage change as the effect size metric and the fact that it is now based on literature that is over 15 years old and is thus incomplete.

School-wide PBS has become an increasingly popular approach that has already been widely disseminated at organization- and statewide levels. Solomon et al. (2012) recently conducted a meta-analysis of 20 articles published over a 16-year period. Based on 21 effect sizes, they reported "low to medium effect sizes" (p. 115) and noted that there were "promising early trends in the data across dependent variables"; however, this meta-analysis illustrates the challenges of conducting good experimental work in large organizations over extended periods of time with many participants. For example, 8 of the 20 studies used nonexperimental AB designs (Table 20.2, pp. 113–115). Further, only one study included data on treatment integrity but only measured treatment integrity by reporting the number of hours and sessions of consultation rather than actual implementation of PBS strategies by teachers.

The evidence presented by Carr et al. and Marquis et al. combined with other more recent meta-analyses of specific behavioral interventions such as differential reinforcement and choice

(see above) demonstrates that these intervention methods do indeed meet and exceed criteria for evidence-based practices. Data on school-wide PBS interventions is currently lacking and of poor quality; hence, we cannot yet conclude that school-wide PBS is an evidence-based practice.

Behavioral Interventions for Specific Challenging Behavior

There have been several systematic reviews and meta-analyses of behavioral treatment for specific problems. This section will illustrate two applications: anxiety disorders and self-injury.

Anxiety Disorders

Behavioral treatment of anxiety disorders has received considerable attention in individuals with ID for many years (Jennett & Hagopian, 2008). More recently, several researchers have also attempted to treat anxiety disorders in higher functioning teenagers and adults with Asperger syndrome resulting in at least one systematic review (White et al., 2009) and two meta-analyses (Lang, Koegel, et al., 2010; White et al., 2009) as well as two more recent RCTs (Reaven, Blakely-Smith, Culhane-Shelburne, & Jepburn, 2012; Sung et al., 2011).

Jennett and Hagopian (2008) searched PsycINFO and PubMed from 1970 to February 2007 crossing disability with fear terms to yield 38 candidate articles, of which 13 small *N* experiments with 28 children and adults were retained (four group designs were rejected). All experiments used behavioral packages and all packages included exposure and reinforcement. Thus, Jennet and Hagopian concluded that behavioral packages were "an established treatment" and perhaps exposure and reinforcement were the effective components of the package.

Lang, Regester, Lauderdale, Ashbaugh, and Haring (2010) searched PsycINFO, ERIC, and MEDLINE, crossing disability terms with anxiety or CBT, and searched reference sections of candidate articles, relevant author names, and journals from January to July 2009. This yielded 164 potential studies of which nine experiments with 111 participants were retained. Participants were mostly individuals with Asperger and "higher functioning autism" treated mostly with manualized CBT modified for this population. The most common dependent variables were psychometric self-report measures. Only one of nine studies (Wood, Drahota, et al., 2009) was considered high quality, limiting the possibility of firm conclusions. Wood, Drahota, et al. (2009) compared CBT with treatment-as-usual in a group of 40 children aged 7–11 years with ASDs, anxiety, and IQs over 70. They found large reductions in proportion of children who met diagnostic criteria for an anxiety disorder (64 % vs. 9 %) and parent ratings of anxiety symptoms (Wood, McLeod, Piacentini, & Sigman, 2009), although no effects were found on child-reported anxiety symptoms. There were also increases in child's adaptive behavior and reductions in parental intrusive involvement in child's daily routines (Drahota, Wood, Sze, & Van Dyke, 2011).

Two additional RCTs of CBT for anxiety in individuals have evaluated the effectiveness of CBT for higher functioning children with anxiety. Sung et al. (2011) found *no* differences between CBT and a social recreational group since both groups showed improvement over baseline. The authors described the social recreational group as a second, effective therapy; however, it could equally be described as an active placebo condition suggesting that CBT was ineffective. In contrast, both Wood's papers and Reaven et al. (2012) found robust evidence that a CBT program, which both included many behavioral components such as exposure, relaxation, and parent training, produced large effects on reductions in anxiety diagnoses and global rating of symptoms. In addition, a recent small-scale RCT for combined anxiety and social skills training for children aged 12–17 years and anxiety disorders also reported significant effects (White et al., 2013). The positive results from two well-conducted RCTs (Reaven et al., 2012; Wood, Drahota et al., 2009) and White et al. (2013) suggest that CBT is an evidence-based practice for individuals with Asperger syndrome

or ASD in older children and adolescents with IQs over 80; as elsewhere, the issue of whether the cognitive therapy components contribute anything to the treatment effects remains to be evaluated. Thus, practitioners should include exposure and reinforcement (Jennett & Hagopian, 2008) and consider training parents and other caregivers to deliver treatment and promote generalization of treatment sessions.

Self-Injurious Behavior

Expert panels, such as Rush and Frances (2000) and the National Autism Center (2009), have all concurred that behavioral interventions should be first-line treatment for SIB and there is little evidence for alternative effective treatments. There is a very extensive literature on behavioral interventions for SIB dating back to the mid-1960s which is reflected in several systematic reviews and meta-analyses of SIB generally (Christiansen, 2005, 2009; Denis, Van den Noortgate, & Maes, 2011; Kahng et al., 2002; Sternberg, Taylor, & Babkie, 1994; Sturmey et al., 2012) as well as specific forms of SIB, such as hand mouthing (Cannella et al., 2006), skin picking (Lang, Didden, et al., 2010), rumination (Lang et al., 2011), and pica (Bell & Stein, 1992; Hagopian et al., 2011; McAdam et al., 2004, 2012). This literature is predominantly behavioral and behavior analytic and reaches broadly similar conclusions to the consensus panels. The recent meta-analysis by Christiansen (2009) will illustrate this literature.

Expanding upon her earlier meta-analysis of treatment of SIB in individuals with autism (Christiansen, 2005), Christiansen (2009) searched electronic databases, conducted an ancestral search, and hand searched relevant journals for research published between 1964 and 2008. She identified 224 experiments with 343 participants and analyzed the data using HLM. The overall effect size was large and significantly different from zero, indicating that treatment was very effective. She also ranked treatments by effect size and found that combined aversive and non-aversive treatments were most effective, followed by aversive interventions alone, combined aversive and communication interventions, and least effective were non-aversive interventions alone. Finally, she found that sensory interventions for SIB were ineffective.

Sensory Interventions

Sensory Integration Therapy

Individuals with DD have greater risk of sensory impairments than others (Baranek, 2002), and some believe they also have increased sensitivities or unusual reactions to sensory stimuli (Baranek, 2002; Sinha et al., 2006). Based on the hypothesis that abnormalities in processing sensory information, sometimes referred to as "sensory processing disorder," result in a wide range of problems in adaptive and maladaptive behaviors, SIT procedures, such as sensory integration and sensory diets, and procedures such as brushing, swings, and balls to increase and/or reorganize sensory functioning have been developed. Although popular and subject to quite extensive research (although often of mediocre quality), the evidence for effectiveness is negative.

The American Academy of Pediatrics reviewed three meta-analyses (Baranek, 2002; May-Benson & Koomar, 2010; Vargas & Camilli, 1999) and one recent study. They concluded that "pediatricians should not use sensory processing disorder as a diagnosis" and "should recognize and communicate with families about the limited data on the use of sensory-based therapies for childhood developmental and behavior problems" (p. 1188). They also concluded that "One recent small study cautions … about the possible negative behavioral effects of sensory integration therapy" (p. 1188). The three meta-analyses they cite (Baranek, 2002; May-Benson & Koomar, 2010; Vargas & Camilli, 1999) and one additional meta-analysis (Lang et al., 2012) all reached the same conclusion. For example, Lang et al. (2012) identified 25 studies of SIT with individuals with ASD. Only three studies reported positive outcomes, but these studies contain many serious methodological flaws

Table 20.5 General challenging behavior

Target behaviors and/or methods (authors)	Search methods	Main findings
Problem behavior in children and adults with ASD (Campbell, 2003)	Searched 15 journals from 1966 to 1998 and identified 117 articles with 181 participants. Calculated effect size using mean percentage reduction over baseline, PND, and PZD	The most commonly used treatments were DRO (13 %) and punishment (11 %); most articles used a functional assessment
		The mean percentage reduction over baseline was 76 %, mean PND = 85 %, and mean PZD = 43 %. Effect size was not a function of target behavior type. Effect sizes were larger for interventions that used functional assessments than interventions that did not. Studies that used a functional analysis had larger effect sizes than those that used functional assessments
Aggression (Brosnan & Healy, 2011)	Conducted search of 3 databases and hand searched references for studies of children aged 3–18 years with ASD alone, ID, or both published from 1980 to 2009 that treated aggression. Identified 18 studies with 31 participants. Treatment classified into antecedent manipulations, reinforcement-based strategies, and consequential control	Most common topographies were hitting ($N = 18$), biting ($N = 16$), kicking ($N = 16$), pinching ($N = 10$), and scratching ($N = 9$). Reported that all interventions decreased the target behavior and that interventions using functional behavioral analyses resulted in consistently at or near zero levels of target behavior, sometimes with rapid response suppression
	Did not calculate effect sizes	
Challenging behavior (Heyvaert et al., 2012)	Searched the literature from 2000 to 2011 a search of 2 online databases, 31 journals, and a reference search from candidate articles from the previous 2 types of searches for small *N* experiments to reduce challenging behavior. This yielded a database of 285 studies with 598 participants	Treatment of challenging behavior is highly effective but varied between studies and participants. Mediating treatment variables that resulted in larger treatment effects included manipulating antecedent factors, and informing education training the environment. A subsequent analysis only found manipulating antecedents to produce larger treatment effects

precluding a firm conclusion that SIT was effective. They concluded that practitioners should not use SIT with children with ASD. A systematic review of seven studies of weighted vests (Stephenson & Carter, 2009) and a more recent empirical study (Leew, Stein, & Gibbard, 2010) also reached similar conclusions. At least two small *N* experiments with seven participants have compared SIT with ABA and found that ABA was more effective in reducing SIB (Mason & Iwata, 1990) and a range of challenging behaviors (Devlin, Healy, Leader, & Hughes, 2011). For a critique of this literature and defense of SIT, the reader is referred to Clark (2012) and Schaaf and Blanche (2011) and for a reply to the critiques to Healy, Hughes, Leader, and Devlin (2011). The present authors concur with the majority and agree that there is no well-conducted research supporting the effectiveness of SIT or weighted jackets at this time and two small *N* experiments supporting the superiority of behavior analysis. Although it is possible that SIT was not implemented adequately in the evaluations, the onus remains on advocates of SIT to operationally define and reliably measure both eligibility criteria and treatment characteristics and to conduct methodologically sound experiments to demonstrate if SIT is an effective treatment (Table 20.5).

Music Therapy

There have been at least two meta-analyses of music therapy. Whipple (2004) conducted a

meta-analysis of nine studies and reported mean $d = 0.83$. Only three of the studies, however, were published and at least one included behavioral interventions, such as contingent music, and other studies included procedures such as background music, raising the issue of whether the treatments that were included in this meta-analysis were actually music therapy. Similar concerns over evidence for the effectiveness of music therapy are found in Gold, Wigram, and Elefant's (2006) Cochrane review which identified only three small-scale studies (total $N = 24$) of music therapy over a period of a few days to a week. The mean SMD was 0.36 and 0.50 for verbal and gestural communication skills, respectively, and there were no effects on challenging behavior. Further, the effects on communication skills were most likely merely eliciting existing communication skills, rather than teaching new behavior. Thus, there is little evidence that music therapy is an evidence-based practice, and for those behaviors where there is marginal evidence for effectiveness, there are alternate methodologies with much greater evidence to support their use. A third meta-analysis by Gold, Voracek, and Wigram (2004) of music therapy for child and adolescent psychopathology also did not conclude, due to lack of evidence, that music therapy was an evidence-based practice for autism or other developmental disabilities (Table 20.6).

Snoezelen

Snoezelen is a Dutch intervention in which a facilitator offers multimodal sensory stimuli in a specially designed room in order to balance relaxation and activity. There have been two systematic reviews of Snoezelen (Lancioni, Cuvo, & O'Reilly, 2002; Lotan & Gold, 2009). Both reached similar conclusions however, and since Lotan and Gold (2009) also conducted a meta-analysis, we will discuss their results. Lotan and Gold searched PubMed and ERIC and Lancioni et al.'s references and identified 28 studies, of which 13 met inclusion criteria. Effect sizes were medium to large (Hedges $g = 0.63$–2.63). The authors, however, also described that none of the studies were RCTs and "showed weak methodological structure" (p. 211) and that "The results give some initial support to the assumption that Snoezelen has value as a therapeutic approach; yet sufficient rigorous research… is lacking." Thus, at this time there is not any good quality evidence that Snoezelen is an evidence-based practice.

Auditory Integration Therapy

AIT involves having the subject listen to specially modulated music that is designed to match with the sensitivities often found in individuals with autism for certain frequencies. There are several different versions of AIT. In a systematic review of six RCTs on AIT, Sinha et al. (2006 and Sinha, Silove, Hayen, and Williams (2011) found mostly inconsistent results and emphasized that there was insufficient evidence to justify its continued use.

Cognitive Behavior Therapy and Cognitive Therapy

CBT is "treatment that seeks overt behavioral change by teaching [individuals] to change thoughts and thought processes in an overt, active manner" (Durlak, Fuhrman, & Lampman, 1991, p. 205) which attempts to correct cognitive distortions and deficiencies by teaching emotional recognition, stress and anxiety management, cognitive restructuring, and self-reflection (White, 2003), as well as homework assignments to practice relaxation, coping, and social skills outside the therapy session. Thus, CBT uses both cognitive and behavioral interventions to correct problematic underlying cognitive processes and structures to produce behavior change. CBT has been extensively studied across all age ranges and for a variety of problems (Ozabaci, 2011), such as depression (Cuijpers, van Straten, Andersson, & van Oppen, 2008; Cuijpers, van Straten, Warmerdam, & Andersson, 2008), personality disorders (Leichsenring & Leibing, 2003), anxiety disorders (Townsend et al., 2010),

Table 20.6 Systematic reviews and meta-analyses of specific behavioral intervention methods

Treatment	Description	Main findings and conclusions
Activity schedules (Banda & Grimmet, 2008)	A systematic review of 13 studies on activity schedules for individuals with autism (children to adults) was conducted. Studies were published between the years of 1993 and 2004	Multiple baseline and reversal designs were the most frequently used design in studies. Activity schedules were shown to be effective for all 31 participants across the 13 studies. Social interactions, coping with transitions, and on-task behaviors were all enhanced with the use of activity schedules. There were also decreases shown in disruptive behaviors (e.g., tantrums, screaming, and aggression toward others)
Video modeling and video self-modeling (Bellini & Akullian, 2007)	Twenty-nine studies were identified between the years of 1980 and 2005 on video modeling and video self-modeling for individuals with autism spectrum disorders. Effect sizes were calculated using percentage of non-overlapping data points	Interventions (antecedent, response contingent, and noncontingent procedures) were moderately effective with mean PND = 80 %. Similar effects were found for maintenance and generalization (mean PND = 83 % and 74 %, respectively). Video modeling was somewhat more effective (*M* PND = 81 %) than video self-modeling (*M* PND = 77 %). These interventions were most effective in targeting functional skills and response contingent procedures were more effective than either antecedent or noncontingent procedures
Communication interventions (Goldstein, 2002)	Sixty studies published between 1978 and 2000 on speech and language interventions for children with autism were reviewed	The literature provides little direction in terms of service delivery models or the intensity of services that are more likely to maximize communication intervention efforts. The literature provides little direction in terms of service delivery models or the intensity of services that are more likely to maximize communication intervention efforts
Noncontingent reinforcement (Carr et al., 2000)	Searched three databases and the *Journal of Applied Behavior Analysis* and identified 59 studies of which 24 were retained	The most commonly used procedure was fixed time schedules combined with extinction and schedule thinning (11 studies) followed by fixed time schedules with extinction (7 studies). Eighty-five percent of participants were successfully treated. Fixed time schedule plus extinction and schedule thinning was a *well-established* treatment (mean PND = 71 %, 11 studies). Fixed time schedule plus extinction was a *probably efficacious* treatment (mean PND = 67 %, 8 studies). Variable time schedule plus extinction was classified as probably efficacious (mean PND = 81 %, 3 studies). Fixed time schedule alone was classified as experimental (mean PND = 84 %, 2 studies). Fixed time schedule with thinning was also classified as experimental (mean PND = 93 % 1 study)
Active support (Hamelin & Sturmey, 2011)	A systematic review was conducted on two studies that used experimental designs to evaluate active support as used in agencies for individuals with developmental disabilities. One study was published in 1999 while the other was published in 2007. Percentage of non-overlapping data (PND) and percentage of all non-overlapping data (PAND) were calculated	Only one experiment, out of three identified in the two studies, showed a clear functional relationship between active support with "ineffective" to "questionable" percentage of non-overlapping data points (intervention = 54–66 %, follow-up = 30–50 %) effect sizes and acceptable percentage of all non-overlapping data points effect sizes (intervention = 78–85 %, follow-up = 90–93.3 %). Based on these data, active support only meets Chambless and Hollon's (1998) criterion for a "promising treatment" but not an evidence-based practice

Differential reinforcement (Chowdhury & Benson, 2010)	Thirty-one studies published between 1980 and 2009 were reviewed on differential reinforcement to reduce maladaptive behaviors in adults with developmental disabilities	Of the 31 studies, 15 reported DR to be an effective intervention when used independently. Of the remaining, 10 studies found DR to be useful as part of a treatment package, and six found a DR contingency used independently to be ineffective and only observed treatment effects when an aversive component was added. Very few studies assessed the use of DR in older adults and in individuals with mild ID; these reflect areas of future research. A methodological analysis found that several studies did not report information on key methodological variables (e.g., conducting a functional analysis, stimulus preference assessment)
Differential reinforcement of alternative behavior (Petscher et al., 2008)	One hundred and sixteen studies were reviewed on the use of DRA for the challenging behaviors of individuals with developmental disabilities. The reviewed studies were published between the years of 1977 and 2007	The authors found that DRA has been successful at reducing behaviors on a continuum from relatively minor problems like prelinguistic communication to life-threatening failure to thrive. DRA with and without extinction was found to be well established for treating destructive behavior of those with developmental disabilities and to combat food refusal. More research needs to be done using research designs that meet the criteria of the APA Division 12 Task Force
Applied behavioral intervention (Spreckley & Boyd, 2009)	Six randomized controlled trials of applied behavioral interventions for preschool children with autism were reviewed. Four of these studies, published between 2000 and 2007, had standardized mean difference scores calculated	Based on the results of the standardized mean difference, calculations applied behavioral interventions (e.g., parent training) were not significantly better than standard care for cognitive outcomes (SMD = 0.38), expressive language (SMD = 0.37), receptive language (SMD = 29), or adaptive behavior (SMD = 0.30)
Compliance (Lee, 2005)	Used online searches, hand searches of reference sections, and journals to identify experiments evaluating hi-p sequences to teach compliance published between 1987 and 2001. This resulted in 28 studies. Effect sizes were calculated using PND	The mean PND was 77 % ("effective"). Effect sizes were larger in preschool (PND = 83 %) and middle/high school settings (mean PND = 83 %) than adult settings (PND = 48 %). Effect sizes were larger when initial compliance was greatest, when there was praise for compliance, and when the time between the hi- and low-p instruction was shortest
Training paraprofessionals (Rispoli et al., 2011)	Searched ERIC, MEDLINE, and PsycINFO crossing disability terms with paraeducator, paraprofessional, and assistant; searched a list of journals from 2010 to 2011 and relevant author names. This yielded 361 abstracts, of which 12 small *N* experiments with 39 paraprofessionals were retained	7/12 studies reported positive and 5/12 produced mixed effects in paraprofessional skills. Of 6 studies reporting effects on child behavior, all reported positive effects

behavioral maladjustment (Durlak et al., 1991), anger (Hamelin, Travis, & Sturmey, in press; Saini, 2009), and aggression (Ozabaci, 2011). Thus, for many adult disorders, CBT is commonly an evidence-based practice (Sturmey & Hersen, 2012b) although it is associated with a wide range of effect sizes (Cuijpers, van Straten, Andersson, & van Oppen, 2008; Cuijpers, van Straten, Warmerdam, & Andersson, 2008; Durlak et al., 1991; Leichsenring & Leibing, 2003; Ozabaci, 2011; Townsend et al., 2010).

There have been reports of CBT for a range of problematic behavior in people with disabilities (Hamelin et al., in press). Most well-conducted studies have been in the areas of anger and anxiety disorders in higher functioning people with ASD (see section on "Anxiety Disorders"), but not in areas such as anxiety or depression in adults with disabilities (although there has been at least one RCT or CBT for depression in adults with ID, the study was non-blind and had many other methodological problems).

There is an extensive literature on CBT and maladaptive anger which provided strong support for its use in the general population (Saini, 2009), although there are very few well-controlled studies of CBT for anger in people with disabilities. Hamelin et al. (in press) found a mean effect size of 1.05, which is somewhat larger than the means of 0.60 and 0.83 that Saini (2009) reported in the general population for CBT and cognitive therapy, respectively. Although Hamelin et al.'s results are promising, they were based on only two well-controlled studies. Other limits to this literature include the need for treatment fidelity checks, active control groups, measures of clinical significance, information that could be used to rule out threats to internal validity (Hamelin et al., in press), and reliable and valid outcome measures beyond staff and self-report.

The number of RCTs of CBT with people with disabilities is increasing slowly (Hassiotis & Sturmey, 2010), although the quality of the studies is often poor. An important question that remains to be answered is whether the changes in behavior that may occur in these studies are due to the cognitive or behavioral components of the treatment package. To date, dismantling studies that have addressed this in the literature on CBT and depression uniformly support that it is the behavioral rather than cognitive components of CBT that are the active components of the package (Cuijpers et al., 2012; Ozabaci, 2011; Sturmey, 2009). There is little evidence that CT alone is effective with individuals with disabilities at this time, and the effectiveness of behavioral components used alone, for example, in the treatment of fears and phobias (Jennett & Hagopian, 2008), suggests that, as in other populations, behavioral interventions may be the effective components of this package.

Psychotherapy and Counseling

Psychotherapy and counseling have only been evaluated with typical adults with psychiatric disorders using true experiments. These treatments often require relatively well-developed verbal and cognitive skills, which is why they are often not used with children, adolescents, or individuals with ID. Meta-analyses have revealed that these interventions are limited to small effect sizes over conditions such as care-as-usual (Bower, Rowland, & Hardy, 2003; Svartberg & Stiles, 1991), and psychotherapy was shown to be inferior to alternative treatments (e.g., CBT, behavioral, experiential) used in some studies (Svartberg & Stiles, 1991). Also, the components of these treatments are heterogeneous and variables such as treatment length and population have varied considerably (Bower et al., 2003; Svartberg & Stiles, 1991). The limited effectiveness and lack of consistency in defining "psychotherapy" indicate the increased need for additional research on these interventions.

Conclusion

The evidence base for ID and related developmental disabilities is now very large: There are over 100 meta-analyses! Meta-analyses of psychosocial interventions uniformly agree that psychosocial interventions are effective for many socially significant behaviors, including increasing

skills and independence and decreasing challenging behavior. Almost all expert panels, with the exception of some on intensive early intervention, have also reached this conclusion. Thus, we no longer have to proceed "with compassion while awaiting the evidence" (King, 2005); we have a mountain of evidence to guide treatment that is both compassionate and effective. A notable trend in some areas is that we can move beyond the simple question of whether a treatment is evidence-based or whether we have a treatment that is better than nothing to beginning to answer more interesting and clinically meaningful questions such as the following: Which is the *most* effective treatment for this clinical problem? What target behavior(s) is this treatment most effective for?

Despite this large quantity of evidence, there are many important gaps in this literature. Some of these gaps include an absence of evidence for common practices, such as person-centered planning, TEACCH, cognitive therapy, many aspects of PBS, and some forms of ABA, such as fast-paced teaching and stimulus pairing to establish secondary reinforcers. Thus, future research could address these gaps by conducting systematic reviews and meta-analyses to identify the present status of the evidence and to stimulate future research in these areas. The majority—but not all—of the literature relates to children and adolescents. The majority of evidence relates to behavior modification and behavior analysis: Several meta-analyses have reported hundreds of small *N* experiments with hundreds of participants. Given the weight of the evidence and the robustness of the effects across populations, settings, and target behaviors and the almost uniform agreement from many different consensus panels, behavioral treatments are clearly the first-choice treatments in many situations.

It is useful to distinguish three kinds of evidence that are absent from the literature. First, there are those treatments that have not been evaluated with well-designed studies where there is evidence, but it is of poor quality and cannot yet support the conclusion that a treatment is evidence-based, such as psychotherapy, cognitive therapy, counseling, SIT, and system-wide PBS. Future research might demonstrate that these interventions may be effective, but the evidence to support that conclusion is not yet available. There are also some treatments which have no evidence yet available. In both of these situations, when there are alternate effective treatments, practitioners should use existing evidence-based practices. If practitioners deviate from evidence-based practices, then practitioners have a heavy burden to positively show that their choice of an unevaluated treatment is beneficial to the client. Minimally, this can be done by collecting pre- and post-data using meaningful, reliable, valid, and socially significant outcome measures; collecting data more frequently using AB designs is preferable as it is both a more sensitive measure of outcome. Additionally, it gives useful feedback to both practitioner and client as to treatment effectiveness and whether an ineffective treatment should be abandoned and replaced with an evidence-based practice. There are also treatments that have been evaluated in well-designed studies and found to be ineffective (AIT, weighted jackets) or sometimes positively harmful (FC). In these cases, the continued use of ineffective or potentially harmful treatments places an especially heavy burden on practitioners to demonstrate treatment effectiveness and lack of harm.

There are numerous significant gaps in the literature, the most prominent of which is the absence of a meta-analysis of psychosocial interventions for adults with ID, including common psychiatric problems, such as anxiety, depression, and schizophrenia, especially in community settings with typical change agents, and for some common target behaviors such as toilet training. Second, there are very few economic analyses of psychosocial interventions outside of early intervention and psychotropic medication, although some evidence suggests that community-based ABA for adults results in reductions in problem behavior without increasing costs (Hassiotis et al., 2011).

Perhaps the most significant gap is the absence of dissemination of EBP. There may be two reasons for this. First, until recently it was assumed that there was insufficient evidence to identify EBPs and hence nothing could be done:

This chapter shows this is not the case. We can now identify EBPs for common problems, such as choice, differential reinforcement, and behavioral interventions for problem behavior based on functional assessments. Even though this literature now exists, it is not available in any useful or readily accessible format for service providers and planners to access. Researchers should devise methods to make this information more readily available in formats that are easily accessible, easy to understand, and readily searchable. A more significant gap is the absence of readily available manualized treatments and service models that can be used to train and support local practitioners to implement EBPs accurately. This remains a major hurdle in implementing EBPs.

We end on a note of optimism: Research has now identified a number of EBPs for people with ID and ASD for common, socially significant problems that may lead to an improvement in the quality of life and potentially result in economic benefits to society, such as avoidance of resources wasted on ineffective or harmful therapies. Doubtless, the literature on identifying EBPs and making more refined conclusions related to EBPs will continue to grow, but we can now begin to implement and disseminate EBPs with people with ID and ASDs.

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Community Mental Health Services

21

Jean O'Hara, Eddie Chaplin, Jill Lockett, and Nick Bouras

Introduction

This chapter outlines developments in mental health community services and future directions. An international perspective is given although most of the research and current developments come from the UK. In the last 50 years there has been a common trend internationally towards community integration for people with intellectual disabilities. In many areas this has coincided with the development of specialist mental health care (Davidson & O'Hara, 2007). However in spite of these developments, there is still a wide disparity as to how care is delivered or provided for people with intellectual disabilities and mental health needs. This disparity exists not only between but also within countries, both in terms of consistency and the philosophy that underpins it (Holt et al., 2000). It is the case even in more advanced areas of the world; people with intellectual disabilities are still more likely to suffer from poor access to mental health services (Holt et al., 2000; Vanstraelen, Holt, & Bouras, 2003). Advances in health-care science have served to highlight the gaps in service provision, e.g. for those with additional needs such as autism spectrum disorders (ASD) and attention deficit hyperactivity disorder (ADHD), whilst genetic findings suggest that more attention should be given to the relationship between psychosis and neurodevelopmental disorders, including autism (Craddock & Owen, 2010; Owen, O'Donovan, Thapar, & Craddock, 2011). In comparison to the general population, people with intellectual disabilities have lower life expectancy and higher rates of unmet health needs (Balogh, Ouellette-Kuntz, Bourne, Lunsky, & Colantonio, 2008; Janicki, Dalton, Henderson, & Davidson, 1999), with different patterns of illness and complex interactions between co-morbidities (Williams, 2011). In spite of this, access to health care is more difficult for people with intellectual disabilities (Disability Rights Commission, 2006; MENCAP, 2007; Michaels, 2008) both in primary care and secondary care, which includes mental health services. In spite of having an increased understanding of the mental health needs of people with intellectual disabilities, e.g. clinical presentation, assessment and diagnostic techniques, this knowledge and awareness have largely remained within specialist services. The changing commissioning landscape, the move to personalised health budgets, greater patient empowerment in health-care delivery in

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general and the increased morbidity of this population have brought new challenges and a greater urgency to share these advances across the range of mental health care (Cain et al., 2003; Davidson et al., 2003; Her Majesty's Government, 2011; Janicki et al., 2002; Stawksi, Davidson, & Merrick, 2006).

History and Policy

Following deinstitutionalisation the emphasis of policy has been towards inclusion and independence within local communities. In the USA there was 'Combating Mental Retardation' (Kennedy, 1962), and later the 'A Charge We Have to Keep: A Road Map to Personal and Economic Freedom for Persons with Intellectual Disabilities' (President's Committee for People with Intellectual Disabilities, 2004). In the UK, Enoch Powell, the Minister of Health infamous 'Water Tower Speech' as part of the Address to the National Association of Mental Health Annual Conference, 9 March 1961, advocated the reduction of the number of hospital beds and a move towards a local authority community infrastructure for people with mental health problems. For people with intellectual disabilities, the future direction was not clear, but there was still a belief that there was a need for alternatives and more information to understand the predicament of this group.

This was followed a decade later by the UK government White paper and subsequent report, 'Better services for the mentally handicapped', which proposed a move away from hospital care (Department of Health and Social Security, 1971, 1980). In terms of mental health provision, 'Better services for the mentally ill' (Department of Health and Social Security, 1975) was published with similar aspirations, but without indication to the process.

The development of services and health-care systems has differed across countries, and the quest towards specialist mental health services has taken a number of different paths and priorities, which has led to different interfaces and models. In Ireland a 'bottom-up' approach in some areas has existed where 'parents and friends' groups' set up community-based residential services as a template for the deinstitutionalisation and normalisation in the community of the more traditional campus-based services (Cain et al., 2010). In Australia the Second National Mental Health Plan (Australian Health Ministers, 1998) identified the need for improved access, treatment and care, although its subsequent evaluation found little evidence of progress. A call for better co-ordination of existing services followed although there was no mention of the need for specialist expertise or clinical services (Australian Health Ministers, 2003). In Spain in 2006, a specialist service model has been developed, inspired by the Mental Health in Learning Disabilities (MHiLD) model in South East London, UK (see Chaplin & O'Hara, 2008) and has been adopted as a part of the Catalan Mental Health Plan (2006–2010) (Cain et al., 2010; Departamento de Salut, 2006). In Holland the delivery of mental health care has moved towards community care and regional centres, resulting in a spectacular decline in the number of beds in mental hospitals, increased admissions, decreased length of stay and closure of the large asylums (Ravelli, 2006), with evidence to suggest that outreach represented an effective and efficient alternative to hospital treatment for people with intellectual disabilities (Van Minnen, Hoogduin, & Broekman, 1997).

In Canada, the 2006 Canadian guidelines for primary care of adults with developmental disabilities (Williams, 2011) were updated with the remit to make evidence-based recommendations to address the particular health issues of adults with developmental disabilities. This included a recommendation to avoid long-term use of antipsychotic medications for behavioural issues in the absence of an underlying mental illness, which is echoed in published good practice guidelines in the management of challenging behaviours (Deb & Unwin, 2007; Royal College of Psychiatrists, British Psychological Society, & Royal College of Speech and Language Therapists, 2007).

In spite of the international impetus for change brought about by a philosophical shift in how we view people with intellectual disabilities, the reality is that service development, provision and

delivery are still reliant on a number of factors. These include the interpretation of what is required by those providing and commissioning services, informed legislative and policy frameworks that offer clear direction on how to achieve change, the resources that are available to develop services, the varying attitudes and culture of those currently running services and the absence of a single care model. The lack of a consistent approach has in many areas affected performance in terms of outcomes and commitment to client-centred care. Mansell (2005) offers two main explanations to explain the variability of services:

1. The ideology of institutions persists so only an increase in supported living will address it.
2. The problem lies with weak implementation of the model rather than something intrinsic to the model, with some community services providing no more in terms of meaningful activity that the institutions.

The competing paradigms and perceptions of how intellectual disability affects the individual and how society sees and accommodates them has acted in some cases as a barrier to joined up services. History has created a blame culture that has often sought to simplify complex issues where things have gone wrong, e.g. safeguarding concerns (abuse) within institutions to group homes. Too often blame has been contributed to single professional groups rather than examining the culture and context in which adverse events occur. Ideological approaches, often advocated by prominent individuals, have shaped the history of societal attitudes, care and service delivery and organisational design. In operational terms such fixed beliefs can be as harmful as any prejudice, as they may not only ignore or minimise the highly complex needs of a minority of people with intellectual disability (perhaps for the benefit of the majority) but also compromise the working relationships and at worst prevent delivery of multiagency services. In the UK, for example, a number of scandals have provoked a knee-jerk reaction when a few voices dominate the scene and result in a reactive response of rapid closure, e.g. Cornwall (Commission for Healthcare Audit and Inspection, 2006) resulted in rapid closure of institutions and NHS long-stay beds (only for this need to be picked up by the independent sector), rather than taking the opportunity to look towards community provision. These issues are not just a problem for the NHS as the recent Winterbourne View closure has demonstrated and where a serious case review has been announced (Care Quality Commission, 2011). Why such events occur, where and when they do, is not fully understood, but from the largest institution to supported community living arrangements, there will always be the need to safeguard against the occurrence of such events given the vulnerability of the people we serve. We should not lose sight of community services as the preferred model for those with complex needs or who challenge services. Often outdated systems that continue regardless of what good commissioning guidance tells us (Department of Health, 1999a, 1999c; Department of Health & Office of the National Director of Learning Disabilities, 2007) often means there is a lack of preparation or proper acknowledgement of complexity of need to offer the levels of support required. The issue of financial support for this type of provision will mean different countries have as a result dealt with this shift in different ways (Balogh et al., 2008). A lack of investment in local services particularly in some areas of the UK is often due to high use of out-of-area services. In some areas reactive approaches have replaced an overall strategy to develop local community infrastructures and pathways.

The concept of 'normalisation' (Wolfensberger, 1972, 1980) has pushed the community agendas and we have seen the development and introduction of new roles, although not without some misinterpretation of what this meant, causing differences in implementation. Psychiatry in the UK soon found its niche within the new emerging social models by specialising in the assessment and treatment of mental health needs of people with intellectual disabilities; however, this has not been a uniform development as in other counties such as the Netherlands, psychiatrists distanced themselves from the care of this group. However both approaches saw the need for greater input from other disciplines to meet the needs of this group, e.g. clinical psychology, teaching, speech

and language therapy and social work. In this era of change, new roles such as behavioural support workers came into existence. In the UK, nursing has come full circle from its role and legitimacy being questioned to the current development of the profession (Royal College of Nursing, 2010). At the point of deinstitutionalisation, social care service models were developed that had no place for nurses who in many areas were 'demonised' and were forced to relinquish their roles to take up social care or behavioural support roles and were no longer accountable to their professional body to uphold standards expected of nurses. In mental health care, nurses from both intellectual (learning) disability and mental health branches have been embraced by the new specialist mental health services. Intellectual disability nursing will to some extent always be vulnerable to political, theoretical and philosophical influences. Currently there is a shortage of intellectual disability nurses, and numbers in the UK continue to fall despite increasing numbers of people with intellectual disabilities who would benefit from their support (Gates, 2011).

Community Mental Health Services

The delivery of mental health care as we have described is both variable and at times haphazard for people with intellectual disabilities. A lack of vision and development of services has meant for long periods the mental health care for this group has not been considered.

In the UK there are variations in how mental health services for people with intellectual disabilities in the community are delivered and by whom, e.g. Community Intellectual/Learning Disabilities Teams (CLDT), mainstream Community Mental Health Teams (CMHT) and specialist mental health teams for people with intellectual disabilities (Bouras, 2004). From a policy perspective (Department of Health, 2001) people are expected to access mainstream mental health services with specialist mental health services available to meet those with the most complex mental health needs that are unable to be managed within mainstream services. In the UK there are three main approaches to mental health care for people with intellectual disabilities:

1. Within a Community Intellectual Disabilities Team (Hassiotis, Barron, & O'Hara, 2000). These are primary care based with interfaces with general practitioners and are usually in or connected to primary health care teams (O'Hara, 2000; Slevin, Truesdale-Kennedy, McConkey, Barr, & Taggart, 2008).
2. Mainstream health services. This includes primary care (accessed within the communities, e.g. GPs (family doctors)) and secondary care mental health services.
3. Specialist mental health services specifically for people with intellectual disabilities:
   (a) The Mental Health in Learning Disability (MHiLD) Service—(Bouras, Cowley, Holt, Newton, & Sturmey, 2003; Bouras & Holt, 2001; Chaplin & O'Hara, 2008)
   (b) Virtual Mental Health (LD) Team: (Islington model) (Hall, Higgins, Parkes, Hassiotis, & Samuels, 2006)
   (c) Managed functional needs-led teams (Tower Hamlets model) (O'Hara, 1998)

The three models outlined above often provide a secondary or tertiary care function, and some pathways include access to specialist inpatient mental health services. The Virtual Model is delivered within existing mental health services whereas the MHiLD service is a stand-alone service that has clear care pathways to mainstream adult mental health services. The needs-led teams are made up of integrated multidisciplinary and multi-agency (health and social care) teams specialising in different aspects of care, with one team dedicated to mental health and behavioural problems.

In spite of specialist models, mental health provision is still seen as a key component within Community Intellectual Disabilities Teams, and this traditional approach is still the preferred model in many areas, especially where there is a lack of specialist mental health clinicians. In some areas the approach has become outdated because of a failure to develop interfaces and care pathways to primary and secondary care mental health services.

Specialist mental health community services are able to address mental health issues that can

have potential negative consequences if left untreated or unidentified. This may occur more in areas where eligibility criteria to mainstream services only reflect the needs of emergency cases or for people with chronic and severe enduring mental health problems and not take into account how existing cognitive and social impairments can impact on the onset or maintenance of mental illness. As a result individuals who do not receive early intervention may go on to require more restrictive approaches than necessary. The need for specialist mental health services for people with intellectual disabilities is now widely recognised in policy and professional bodies (Department of Health, 2001; Royal College of Psychiatrists, 1996, 2003, 2011). The first specialist services were developed in the 1980s both to assist the hospital closure programmes and to provide a service in their absence. In areas that were slow to respond, it was soon clear that the expected level of improvement in behavioural and mental health issues was not realistic, and in some cases the opposite was true with increased rates of depression, isolation from former communities, loss of friends and increase in behavioural problems. This 'relocation syndrome' (Bouras, Kon, & Drummond, 1993) appeared to be more apparent in people whose move was not accompanied by adequate planning. In the absence of a uniform approach as to a preferred model of specialist mental health care, arguments persist as to the best way to provide services; this lack of consensus has widened inequalities (Pritchard & Roy, 2006).

There is a plethora of guidance on joint working to provide local person-centred services with local 'Learning Disability' Partnership Boards overseeing strategies for effective commissioning, engagement and delivery of services. The NHS Commissioning Frameworks advocate a need for plurality of provision outlined within its operating frameworks (Department of Health, 2011) and the push in local authorities for the tendering and contracting out of placement support has had an adverse effect on the provision of highly specialised placements for those with co-morbid conditions, closer to home. The rise of independent (private or third) sector provision for this cohort has maintained a strong out-of-area focus for provision and fuelled an artificial demand that could have been responded to by local decision making and care planning closer to home and the developmental of community infrastructures and pathways (Chaplin, Paschos, O'Hara, McCarthy, et al., 2010). The lack of local investment has created an artificial demand and has been associated with a rise in inpatient services particularly in the independent sector and out-of-area placements (Allen, 2008; Beadle-Brown, Mansell, Whelton, Hutchinson, & Skidmore, 2006; Chaplin, Kelesidi, et al., 2010; Chaplin & Xenitidis, 2010; Mansell, Beadle-Brown, Skidmore, Whelton, & Hutchinson, 2006; Whelton, 2009). This type of reactive policy has demonstrated a lack of attention to detail and failure to properly consider and to learn lessons from the past and to provide robust integrated community structures for both commissioning and provision of high quality care for all local needs.

A lack of direction and investment in providing local services for those with mental disorders and intellectual disabilities has had unacceptable consequences for a minority and this lack of planning for this group is now impacting on those with complex needs transitioning from children to adult services. It means moving to a placement out of the local area, usually far away from family and friends, and often into a more restrictive environment to get treatment (Chaplin & Xenitidis, 2010). The national average is 34 %; however, in inner London, up to 63 % of people with ID requiring specialist services have been placed out of area (Emerson & Robertson, 2008). In spite of lobbying and policy documents, this situation persists for many (Department of Health, 1999c) and illustrates the shortfalls in many local services with regard to joined up working (Department of Health, 1999c; Department of Health & Office of the National Director of Learning Disabilities, 2007; Royal College of Psychiatrists, 1996, 2003). The lack of local provision impacts on local commissioning practices that is less likely to be focussed on individuals, contrary to accepted guidance; see the National Service Frameworks (Department of Health, 1999b; Foundation for People with Learning Disabilities, Valuing People Support Team, & National Institute for Mental Health in

England, 2004), the Reed Report (1992) Green Light Tool Kit (Foundation for People with Learning Disabilities et al., 2004) and the revised Mansell Report (2007).

Service Models

The provision of community mental health services has developed at different rates across England and Wales in the UK. In terms of the development of mental health services for people with intellectual disabilities, much of the research available has occurred in the UK where the development of specialist mental health services for people with intellectual disabilities has influenced a number of areas, leading to a better understanding of the diagnosis and treatment of mental disorder as part of an increasing evidence base. Distinguishing what level of service is offered to people is difficult as many of the names that once described services, e.g. assertive outreach and intensive home treatment, have become interchangeable and bear no or little resemblance to the original philosophies they were based upon (Marshall & Lockwood, 1998). In Chaplin's (2004) review there was 'no clear evidence to support either model with research being often of a poor quality, lacking replication, and outcome measures were often inappropriate or varied between studies'.

The UK700, the largest UK study of assertive outreach for service users with psychosis, was criticised for not using traditional definitions in defining assertive outreach. The UK700 also examined a group of people with borderline intellectual functioning (IQ 71–85) within adult mental health services. This group had better results than other groups in terms of reduction in hospital admissions, days spent in hospital and increased user satisfaction compared to other groups (Hassiotis et al., 2001). A later study that compared intensive case management vs. standard community treatment for people with intellectual disabilities did not find any significant differences (Martin et al., 2005). There are a number of reasons for this which the authors elude to which include small sample size; the two treatment arms may have been too similar and the applied methodology of randomised controlled trials (RCTs) for this type of research (see Medical Research Council, 2004). In the absence of any new RCTs, research suggests that mainstream services alone cannot meet the mental health needs of people with intellectual disabilities (Chaplin, 2009). Another study comparing assertive outreach for people with ID with Assertive Outreach reported recruitment difficulties, only getting a sample of 30 and a blurring of what the two approaches meant in terms of care in everyday practice (Oliver et al., 2005).

The continuing debate over service models and service configurations over the two last decades has led to attempts to provide a framework to examine services using set criteria, e.g. 'the matrix model' (Moss, Bouras, & Holt, 2000). This model attempted to provide an international framework and considered inputs, processes and outcomes and influenced the comparison of services across five countries, (BIOMED-MEROPE project, Holt et al., 2000). The measurement of the effectiveness and service outcomes even with a framework is difficult as services often change over time in terms of function, who they cater for, the needs people present with, the level of risk, what areas people want to study, etc. This is due to a number of factors including deinstitutionalisation, change in communities (regeneration, urban, rural, deprived, affluent, etc.), the introduction of market forces and the independent sector, different commissioning systems, blurred boundaries in terms of health-care structures, responsibilities and organisation, a more risk averse culture evidenced by more restriction orders for offenders or those at risk of offending and the return to locked doors (UK), the greater awareness of mental health problems and 'new' specialist groups such as those with autism spectrum conditions. These, and other variables, will dictate changes to the core population for many services. This makes comparison difficult not only between services but also within the same service. The potential confounding variables invariably will leave room for questioning. For example, in terms of demographics, specialist units often provide for people who are less able than those who access acute mental health care (Lunsky, Bradley, Durbin, & Koegl, 2008).

Table 21.1 Outcomes examined within the eight studies (from Balogh et al., 2008)

Outcomes measured	Selected headlines	Study
Behaviour-related outcome		Coelho (1993), Dowling et al. (2006), Lowe, Felce, and Blackman (1996), Martin et al. (2005) and Van Minnen et al. (1997)
Evaluated overall psychological and psychiatric function		Martin et al. (2005) and Oliver et al. (2005)
Length of stay and annual admissions	Reported that ICT services with small caseloads significantly decreased the number of days in hospital (Hassiotis et al., 2001)	Hassiotis et al. (2001) and Allen (1998)
Carer burden as an outcome		Lowe et al. (1996), Martin et al. (2005) and Oliver et al. (2005)
Studies identified beneficial interventions	Significant increase in adaptive behaviour and a decrease in maladaptive behaviour in the intervention group (Coelho, 1993). Consistent improvements in behaviour among control group participants who received bereavement counselling working with people with an intellectual disability (Dowling, 2006)	Coelho (1993), Dowling (2006) and Hassiotis et al. (2001)
Quality of life	Quality of life outcomes improved in the control group (Martin et al., 2005). Oliver et al. (2005) found no significant differences	Hassiotis et al. (2001), Lowe et al. (1996), Martin et al. (2005) and Oliver et al. (2005)

However, in reality, is this always the case? Tajuddin, Nadkarni, Biswas, and Watson (2004) reported that within the two outcome periods studied, the demographics of those admitted had changed. These changes are more evident when services are examined longitudinally so we can examine changes in the core client group and advances in assessment and treatment. All of which make comparison difficult. It is therefore understandable that there is no quantifiable evidence to suggest one approach over another is better in terms of outcomes. What may be more important to consider is how the model views the person, their health, support networks and their role within society. With ever changing populations and the changing roles of services over time, older studies are still of value in that they inform practice and highlight trends. Allen (1998) examined services in Wales, over 20 years, capturing the trend towards community provision. The reality is that whatever the model, many local areas lack specialist services necessitating the need for out-of-area placements, causing a knock-on-effect to families (Emerson & Robertson, 2008); the need expressed 20 years ago in many areas is yet to be fulfilled.

Balogh et al. (2008) as part of a Cochrane review (Table 21.1) aimed to assess the effects of organisational interventions for the mental and physical health problems of persons with an intellectual disability. They considered 114 studies of which eight met the eligibility criteria for inclusion. Six were RCTs, one was a controlled before and after study and one was an interrupted time series.

Many of the studies reviewed found no differences between control and experimental groups (Allen, 1998; Lowe et al., 1996; Martin et al., 2005; Oliver et al., 2005; Van Minnen et al., 1997) consistent with the view that current evidence was not adequate to come to any conclusion over best service design (Chaplin, 2004).

In developing modern services, the need for robust interfaces that join up are now more transparent with the advent of care pathways. This has, for example, helped to go some way to addressing the needs of people with neurodevelopmental disorders who traditionally fell between services because they met neither eligibility criteria (Chaplin, O'Hara, Holt, & Bouras, 2009). In one area this has been addressed by providing a single point of access to a newly launched

Neurodevelopmental Disorders Pathway in South East London (UK). This single point of entry now ensures all referrals are discussed and considered and allocated to the appropriate service area, e.g. community mental health in intellectual disability, community or outpatient developmental disorders services (for assessment of ADHD or autism for those without clear evidence of intellectual disability), community intellectual disability services, neurodevelopmental disorders inpatient services or signposted to more appropriate services. Within this umbrella of services, all managed within 'service lines', there is a regular audit to address clinical outcomes, user satisfaction and efficiency. The bringing together of these services has broken down traditional referral barriers and has begun to allow for sharing of expertise and cross-fertilisation of ideas and practices between clinicians and service areas, including assessment models, management of risk, offending behaviours, adult safeguarding issues and working with service users and carers as 'consultants'. This new Neurodevelopmental Disorders pathway, officially launched in 2012, is an innovative model yet to be evaluated in terms of how it addresses the needs of referrers and commissioners, as well as issues of eligibility, managed pathways of care, the extent to which it is outcomes and recovery focused and if it is sensitive to service user experience and feedback.

New Challenges for Providing Local Services and Care Pathways

Chaplin, Paschos, and O'Hara (2010) put forward 'six steps to better services':

1. A sign up for local services that afford people with ID appropriate access to both mainstream and specialist mental health services
2. Service interfaces designed to complement the role of specialist mental health services
3. Local proactive commissioning strategies that are clear about service delivery for this group
4. The use of academic centres to develop the evidence base for this service user group
5. Further research into effectiveness of outcomes related to clinical models
6. Joined up working to provide partnerships towards joint initiatives such as training to both ID and mental health services

In an ever changing environment of new initiatives such as 'direct payments', 'personalised health budgets', 'payment by results' and 'clinical commissioning groups' to empower individuals and focus on service user and clinical outcomes, there is pressure to redesign and to overhaul current systems. However this is in the context of a dearth of specialist commissioning and expertise in complex groups that is highlighted by current and historical gaps (Chaplin, Paschos, & O'Hara, 2010). The changing demographic of people with ID to increasingly diverse populations with more complex co-morbidity and increased age expectancy has increased local expectations; Mansell (2007) provides a sharp message for commissioners and care providers about the determinants of health outcomes for the intellectual disabilities community and the experiences of many people with ID regarding their overall mental and physical well-being, early diagnosis and disease manage programmes. There is an expectation of 'world-class commissioning', which states, 'It is critical that commissioners are able to distinguish and make appropriate investment in services to meet this genuine need whilst preventing inappropriate admissions to isolated and outdated models of service provision or purchasing services commissioned by other PCTs a long distance from peoples' home community' (Department of Health & Office of the National Director of Learning Disabilities, 2007). Inequalities have been highlighted by both Death by Indifference (MENCAP, 2007) and Healthcare for All (Michaels, 2008), which reported evidence of inadequacy in both commissioning and provider structures and the failings to service users and their families. The challenge for services is to develop local integrated managed care pathways, to strengthen the role of community teams (whether MHLD, CMHT or CLDT) and interfaces with inpatient services (and primary care), to support an early discharge plan when admission is necessary, to come up with creative ways of supporting individuals at home to avoid admission where appropriate and to develop a tiered

service response at times of crises (Carnaby, Roberts, Lang, & Nielsen, 2011; Royal College of Psychiatrists, 2011).

The Green Light Toolkit (Foundation for People with Learning Disabilities et al., 2004) was to enable a shared action plan for integrated services and integrated commissioning to tackle the themes and demonstrate progress on improved service outcomes, and in 2008, the NHS and Local Authority Commissioning were underpinned by key guidance, most notably 'No Secrets—Safeguarding Adults, a Consultation Review of the No Secrets Guidance' (Department of Health, 2008) and 'Valuing People Now' (Department of Health, 2009).

Since the UK General Election of May 2010, the Coalition Government has stepped away from the 'Valuing People Now' framework and the Green Light toolkit does not feature as an assessed performance indicator for health-care delivery in the 2011/2012 or 2012/2013 NHS Operating Frameworks (Department of Health, 2011). In parallel there has been a seismic shift away from grant support for residential, supported living and complex placements through social care budgets. The Coalition Government has introduced NHS structural changes through the 2010 Health and Social Bill (currently at the House of Lords report stage at the time of writing) that would hold the majority of the health budget, some £60 billion of public funds. The emerging shakeup of commissioning agencies and strategic health authorities has allowed, even in these early stages, for a distraction away from key performance indicators for service users with intellectual disabilities and refocused energy on acute hospital relationships and disease programmes (The King's Fund, 2011). The impact of the political environment, the reduction of performance frameworks and the shift of staffing structures in commissioning agencies have left a gap in knowledge and pace in many local integrated pathways. Moreover, where strong integrated commissioning and providing relationships existed, they are being stretched because of the retreat of grant support, pre-existing financial flexibility and the loss of organisational memory.

The King's Fund in its assessment of the Health and Social Care Bill and its likely impact outlined a 10-point plan for commissioning priorities (Imison et al., 2011).

Of the ten priorities, points *One*, *Five* and *Six* provide useful support for the enabling of better local community commissioning and provision of services for people with intellectual disabilities in that:

Priority One—active support for self-management

Priority Five—Improving the management of patients with both mental and physical needs

Priority Six—Co-ordination through integrated health and social care teams

Advocate a model in keeping with evidence and learning from research regarding the achievement of local health outcomes.

The Intellectual Disability Partnership Boards focussing on their integrated commissioning of effective services might also have added two more priorities:

New Priority Eleven—the drive for education and employment opportunities for all (Wanless, Appleby, Harrison, & Patel, 2007)

New Priority Twelve—patients as commissioners, having a local voice to shape priorities and test benefits realisation

In short the commissioning of ID services in the UK is often at odds with best practice guidance and a rapid revisiting of 'Green Light' (Foundation for People with Learning Disabilities et al., 2004), and 'Healthcare for All' (Michaels, 2008) within the more modern parlance of the King's Funds Priorities for Commissioners Programme (Imison et al., 2011) is required to ensure that levels of unmet need, out-of-area placements and poor community infrastructures in a range of service provision are remedied.

Central to service development is the need for a strategic approach to see how issues such as demographic changes, e.g. ageing population, increasing complexity of need, changing service user and family expectations, geographical disparity, types of cases referred and large out-of-area populations, will be catered for. It is in this context more resources need to be placed to find and evidence effective models of care for people with intellectual disabilities and mental health problems. The disparity of service provision is highlighted operationally by service design, care

pathways, funding streams, commissioning patterns, staffing patterns, resources and philosophy.

Service Users

In terms of partnerships in developing services and listening to service users, the majority of guidance advocates the meaningful involvement of service users (Department of Health, 2001, 2009; Foundation for People with Learning Disabilities et al., 2004). Mutual governance designed to strengthen partnerships arrangements is central to that agenda (Monitor, 2006) and integral to NHS Foundation Trusts in the UK. In spite of this there is little evidence on the impact of user involvement and the process has been difficult in some areas (Joseph Rowntree Foundation, 1999) perhaps due to poor preparation and lack of negotiation (Branfield & Beresford, 2006).

Often in the absence of an organisational direction, the reliance is on individuals or teams to ensure involvement (Chaplin, Halls, Carlile, Hardy, & Joyce, 2009). A recent survey from East London sought the opinion and experiences of family carers from ethnic minority populations, where a family member had been placed in an out-of-area placement far away from home (Bonell, Ali, Hall, Chinn, & Patkas, 2011). Service user involvement needs to be the central theme running through services. Examples include:

1. NHS Cornwall and Isles of Scilly have appointed eight people with learning disabilities to form an expert learning disabilities user group to help shape the way NHS services are developed in Cornwall.
2. The South London and Maudsley NHS Foundation Trust has developed a two-tier approach within the Behavioural and Developmental Psychiatry Clinical Academic Group (Chaplin & Hardy, 2012; in press). The first tier is designed to give a voice for people currently receiving clinical services. This can be achieved through community groups or satisfaction surveys. This tablet-based instrument features questions designed by service users and includes an opportunity for free text, pictorial representations and voice recording. The second tier is made up of previous service users and carers, which not only takes forward the concerns of current service users but also has a function to audit and advise the senior management team within the organisation. Both these tiers are involved in the wider Trust's initiatives to offer 'ward to Board' coverage. The Trust catchment areas are characterised by its diversity, which brings unique challenges to ensuring that staff engage with service users and their families to develop their voice and input to ensure accessibility and responsiveness of services (Royal College of Psychiatrists, 2011). A recent Delphi consultation project compared the experiences of service users from Black and Ethnic Minority communities with intellectual disabilities and mental health problems accessing services in South East London (Bonell, Underwood, Radhakrishnan, & McCarthy, 2012; in press).

National Voices (2011) emphasises that integrated care takes time to establish, develop and improve and to demonstrate its benefits. As a 'relationship business' this 'relationship capital' should be protected and allowed to grow without continual disruption of policy changes and organisational reform… and that services must be given clear permission to experiment, the aim being to achieve public and social value and not just to save money.

Conclusion

This chapter has provided an international perspective to the development of community mental health services for people with intellectual disability as well as using the current UK situation to demonstrate the issues faced in the development and provision of services. The move towards deinstitutionalisation has occurred at different paces and a number of different models have come about as a result. The comparison of services is made difficult as traditional names that once underpinned philosophical approaches are now used interchangeably across services.

Research has reported benefits of more intensive input, but a lack of studies and difficulty in

defining interventions from community services, changing demographics and eligibility criteria over time has made valid comparisons across, between and within services, just an aspiration. In some cases this might be positive in that as an organic process, we begin to understand what is provided that is more acceptable and meaningful to service users. A strategic direction, informed and driven by service user needs and experiences and based on clinical and research evidence, delivered through integrated but flexible care pathways, is the real challenge for the future. Full implementation of this model, realistically resourced, will bring social and public value to populations and services that are often still marginalised within society and the wider health and social care agenda.

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Inpatient Mental Health Services

22

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Introduction

This chapter provides a review of key issues concerning inpatient psychiatric services for individuals with intellectual disabilities (ID). Such a review is very much impacted by the time in which it is written. A similar chapter written twenty to thirty years prior would focus on long stay hospitals for individuals with ID. However, with the deinstitutionalization movement impacting many parts of the developed world, the purpose of hospitalization has shifted to short term treatment of acute psychiatric issues. This chapter focuses on papers published within the past 10 to 20 years. The first section describes what we know about the profile of individuals with ID that are hospitalized for mental health or behavioural concerns. Next, we review in some detail the two main models of inpatient care: mainstream or general services and specialist units. We will describe what is known about how the two models differ as well as the risks and benefits of each. The third section focuses on clinical considerations and challenges with inpatient care. Finally, the chapter concludes with new directions for models of inpatient care and areas requiring further research.

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Profiles of Inpatients with ID

Inpatients with ID Compared to Other Psychiatric Inpatients

Mental health problems occur in approximately 40 % of adults with intellectual disabilities (ID) living in the community (Cooper, Smiley, Morrison, Williamson, & Allan, 2007), and psychiatric crisis is the primary reason for hospitalisation of such individuals (Balogh, Hunter, & Ouellette-Kuntz, 2005; Lunsky & Balogh, 2010; Morgan & Lowin, 1989). Despite this, deinstitutionalisation has resulted in most individuals with ID receiving outpatient care for their psychiatric issues. As a result, inpatients with ID make up a relatively small but extremely expensive (Polder, Meerding, Bonneux, & van der Maas, 2002) and challenging (Lunsky et al., 2006) group to serve in inpatient psychiatric settings. Further, compared to the general population, individuals with ID visit hospitals much more frequently and have greater needs for specialist services (MENCAP, 1998). Understanding the unique needs and profiles of hospitalisation for inpatients with ID is, therefore, very important.

Several studies have considered differences between individuals with ID and other psychiatric inpatient groups. Generally, findings suggest that

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inpatients with ID tend to be male, younger, living in residential facilities rather than private homes, presenting with comorbid mental health problems and requiring higher levels of recommended care (Burge et al., 2002; Lohrer, Greene, Browning, & Lesser, 2002; Lunsky et al., 2006, 2009). Studies also suggest that inpatients with ID are less likely to be diagnosed with mood or substance abuse disorders compared to inpatients without ID (Burge et al., 2002; Lohrer et al., 2002; Lunsky et al., 2006).

Findings related to discharge patterns between groups have been more mixed. Some studies report that inpatients with ID are more likely to be discharged to an alternative placement from where they came (Burge et al., 2002), whereas others find them *less* likely (Saeed, Ouellette-Kuntz, Stuart, & Burge, 2003). These differences are likely due in part to differences in the social and political climates where studies have been conducted.

Variables Associated with Length of Inpatient Stay

Findings regarding length of stay for inpatients with ID compared to the general population have been varied. Results of a population-based Canadian study found that length of stay did not differ between the two groups, but that individuals with ID were more likely to be rehospitalised within the year (Lunsky & Balogh, 2010). Some studies report similar findings of no difference in length of stay for inpatients with and without ID (Addington, Addington, & Ens, 1993; Burge et al., 2002; Lohrer et al., 2002), whereas others report longer inpatient stays for individuals with ID (Lunsky et al., 2006; Saeed et al., 2003). Despite these inconsistencies, several variables have been reliably linked to longer inpatient stay for individuals with ID including gender (male), mild ID, greater age, independent housing, more mental health issues, discharge to long-term care facilities and admissions which were planned or direct (Addington et al., 1993; Burge et al., 2002; Driessen, DuMoulin, Haveman, & van Os, 1997; Gustafsson, 1997; Huntley, Cho, Christman, & Csemansky, 1998).

Predictors of Hospitalisation

One challenge in identifying consistent predictors across studies is that the reason or need for inpatient services is often based on the local context and is influenced by philosophy, economics and social issues. Inpatient need in a community that has extensive outpatient supports is very different from a community with fewer outpatient supports and low tolerance of risk. Generally, however, several consistent predictors have emerged from the literature. The individuals with ID most likely to be hospitalised are those with severe mental illness, mild intellectual disability, living independently or having severe aggression to self or others (Burge et al., 2002; Cowley, Newton, Sturmey, Bouras, & Holt, 2005). Gender may also be a predictor of admission, with men being more likely to be admitted than women, particularly at younger ages when aggression is more evident (Alexander, Piachaud, & Singh, 2001; Khan et al., 1993; Lunsky et al., 2009).

There are a number of possible reasons why the aforementioned factors appear to predict hospitalisation among inpatients with ID. It may be that inpatient units lack the skills and supports to care for people with more severe disabilities (Burge et al., 2002; Cowley et al., 2005), or that persons with mild ID have lower levels of support in their home environments compared to individuals with severe ID, and are therefore more likely to access hospital care during crisis (Cowley et al., 2005; Gustafsson, 1997). For aggressive and self-injurious behaviour, it is likely that these behaviours become too difficult to manage in the community leading to a greater likelihood of hospitalisation and longer inpatient stays (Burge et al., 2002; Cowley et al., 2005; Lunsky et al., 2006; Morgan & Lowin, 1989; Saeed et al., 2003).

Medical Comorbidities Relevant to Psychiatric Care

Individuals with ID experience poorer health and more poorly managed medical conditions compared to the general population (Beange, McElduff, & Baker, 1995; Cooper, Melville, &

Morrison, 2004; Krahn, Hammond, & Turner, 2006; Lennox, Diggens, & Ugoni, 1997; van Schrojenstein Lantman-De Valk, Metsemakers, Haveman, & Crebolder, 2000). In addition to higher rates of behavioural/mental health problems, individuals with ID experience elevated rates of epilepsy, fractures, skin conditions, poor oral health and respiratory disorders (Davidson, Janicki, Houser, Henderson, & Cain, 2003; Krahn et al., 2006). Individuals with ID often lack access to appropriate primary healthcare (Sullivan et al., 2011) and medical concerns are frequently overlooked because they are poor reporters of their own health issues (Charlot et al., 2011). A number of studies have reported correlations between the health problems of individuals with ID and behavioural or emotional disorders including gastrointestinal disorders, neurological diseases, cardiopulmonary diseases and asthma (Davidson et al., 2003; Kennedy, Juarez, Greenslade, Harvey, & Tally, 2007; Ryan & Sunada, 1997). Fewer studies, however, have examined the prevalence of medical comorbidities among psychiatric inpatients with ID although it has been suggested that many "psychiatric presentations" may be due to unrecognised medical problems (Sullivan et al., 2011). One of these studies, conducted by Charlot et al. (2011), reviewed the medical problems and medications of 198 people with ID admitted to a specialist inpatient psychiatric unit. Results identified 40 % of the sample for which medical problems were identified as the primary contributing factor for inpatient admission. Many individuals had comorbid medical problems, and inpatients with more medical diagnoses had longer inpatients stays. Constipation was the most frequently reported medical comorbidity, followed by gastro-oesophageal reflux disease, seizure disorder and hypothyroidism (Charlot et al., 2011). Older inpatients had more medical problems, as did inpatients with Down syndrome (DS).

Psychotropic Medications

Pharmacotherapy is the most commonly used intervention for individuals with intellectual disabilities and comorbid mental health or behavioural concerns (Matson & Neal, 2009). Despite its widespread use in this population, issues of drug efficacy and safety continue to be widely debated (McGillivray & McCabe, 2006). Furthermore, challenges' differentiating psychiatric disorder from challenging behaviour among people with intellectual disabilities is an area of ongoing concern (Emerson, Moss, & Kiernan, 1999; Holden & Gitlesen, 2003). Unlike the general population, psychotropic medication use in patients with ID is often used to treat non-specific challenging behaviours rather than symptoms of a specific psychiatric disorder (Deb, Sohanapal, Soni, Len, & Unwin, 2007; Tsakanikos, Costello, Holt, Strumey, & Bouras, 2007). Further, medications are often added, but rarely discontinued, even when they are ineffective or causing adverse side effects. This may occur in part because of reluctance from community-based prescribers to make significant medication changes or reduce medication when there are concerns for safety. Although the inpatient environment should be viewed as an opportunity to review and reduce polypharmacy, brief hospitalisations often times have the opposite effect and lead to the prescription of new medications as a short-term solution.

A study of psychotropic drug use among hospitalised individuals with ID in the USA revealed that 86 % of inpatients were prescribed at least two psychotropic medications, 69 % three or more, 44 % four or more and 21 % five or more medications (Charlot et al., 2011). In another study of medication use among inpatients with ID in the Netherlands, antipsychotics were the most commonly prescribed medication, and polypharmacy was linked to psychosis; aggressive, bizarre and attention-seeking behaviour; involuntary measures; and long duration of hospital stay (Stolker, Heerdink, Leufkens, Clerkx, & Nolen, 2001). There was also a tendency to prescribe multiple medications for inpatient stays exceeding one year (Stolker et al., 2001). There is evidence that patterns of medication use may differ from one inpatient programme to another. For example, in a study of inpatients with ID in specialist and general units, inpatients in specialist units experienced overall reductions in their medication regimens from admission to discharge, whereas the number of medications

for inpatients in general programmes increased (White, Lunsky, & Grieve, 2010).

Psychotropic medication use can lead to a number of concerning side effects including constipation, irritability, weight gain/loss, sedation and secondary medical conditions (e.g. metabolic syndrome) (Bradley & Lofchy, 2005; Charlot & Beasley, 2005). As such, the decision to medicate should not be taken lightly and should always be supplemented by non-pharmacological interventions and therapies (Hassiotis et al., 2009). Ideally, the inpatient environment should be viewed as an opportunity to review medications with the goal of reducing polypharmacy when possible, and to implement and monitor multimodal interventions beyond those which are strictly pharmacological.

Summary

Inpatients with ID are a complex and challenging population to serve in the hospital environment. Results suggest that individuals with ID who have mild ID, severe mental illness and aggressive behaviour may be at heightened risk for inpatient admission and longer hospital stay (Burge et al., 2002; Cowley et al., 2005). High rates of medical comorbidities among inpatients with ID and subsequent issues of polypharmacy have also further complicated supporting and treating these individuals. Highly skilled, interdisciplinary teams with specific training and knowledge of ID and mental health are critical to the health and well-being of this population.

Models of Psychiatric Inpatient Services for Individuals with ID

In many countries, the adoption of a more inclusive approach has created a situation in which most people with ID and psychiatric disorders are expected to have their mental health needs met by general psychiatric services rather than specialised psychiatric services for individuals with ID. Such a significant change in service delivery models has raised the question of who benefits from which type of programme. To date, the only studies to compare the two types of programmes come from the UK and Canada. Even though both countries offer both types of services, how services are provided differs. For a full review of specialist and general services, readers are encouraged to review Chaplin (2004, 2009).

Specialist Versus General: A Comparison of Inpatient Profiles

Several studies have examined differences between the profiles of inpatients with ID in specialist and general psychiatric programmes. Findings suggest that patients with ID in specialist programmes are more likely to have a diagnosis of pervasive developmental disorder (PDD) (Alexander et al., 2001; White et al., 2010) and to have higher ratings of challenging behaviour compared to inpatients with ID in general programmes (Lunsky, Bradley, Durbin, & Koegl, 2008). Inpatients in general programmes are more likely to have substance abuse disorders, forensic problems and psychosis or to have attempted suicide (Lunsky et al., 2008; White et al., 2010). Specialist and general programmes report similar levels of recommended care (Lunsky et al., 2008) and risk to self and others are the primary reasons for admission to both units (White et al., 2010).

In terms of inpatient demographics, many similarities have been reported between those using specialist and general psychiatric services (Hemmings et al., 2009; White et al., 2010; Xenitidis et al., 2004). An early UK-based retrospective chart review found that young men with aggressive behaviour were most commonly referred to specialist and general settings (Alexander et al., 2001). However, in the largest comparison to date, inpatients with ID in specialist programmes were more likely to be male and unmarried but less likely to be prescribed psychotropic medication compared to inpatients in general programmes (Lunsky et al., 2008).

Level of Intellectual Disability

Findings of a UK study noted that patients in specialist programmes were more likely to have severe or profound ID and less likely to be living independently, compared to patients of general programmes (Alexander et al., 2001). In a second Ontario study, individuals with ID admitted to specialist programmes had significantly lower Global Assessment of Functioning (GAF) scores at admission than individuals admitted to general psychiatric services (White et al., 2010). Still, some studies report no difference in level of intellectual disability among inpatients with ID in specialist and general programmes (Hemmings et al., 2009; Lunsky et al., 2008).

Clinical Outcomes

There is a considerable dearth of literature comparing the outcomes of patients with ID in specialist and general psychiatric services. However, studies have typically examined outcomes in one setting only, and all studies published until the mid-1990s took a descriptive approach which did not incorporate standardised outcome measures. Moreover, only two studies looked at whether inpatient admission had a differential effect on the clinical outcome for individuals with varying levels of ID. As a result, it is difficult to compare findings between studies since outcome tools vary greatly from one study to another. Most recent studies used scores from the GAF scale (American Psychiatric Association, 2000) as an outcome measure alongside other tools such as the Aberrant Behavior Checklist (Aman & Singh, 1986) and Reiss Screen for Maladaptive Behavior (Reiss, 1986). The appropriateness of the GAF as a measure of patient outcomes in the ID population, especially in cases of severe ID, however, has been questioned (Lunsky et al., 2010; Shedlack, Hennen, Magee, & Cheron, 2005).

Studies conducted in different countries using pre- and post-treatment outcome measures consistently document clinical improvement in specialist inpatient settings (Hall, Parkes, Samuels, & Hassiotis, 2006a; Raitasuo, Taiminen, & Salokangas, 1999; Tajuddin, Nadkarni, Biswas, Watson, & Bhaumik, 2004; Xenitidis et al., 2004). Some studies included ratings completed by caregivers, hospital staff and clients, who also perceived outcomes in specialist units positively (Longo & Scior, 2004; Parkes, Samuels, & Hassiotis, 2007; Samuels, Hall, Parkes, & Hassiotis, 2007). Further, studies conducted in Finland and the UK noted that clinical improvement was maintained 6 months after discharge (Hall et al., 2006a; Raitasuo et al., 1999).

Hall et al. (2006a, b) developed and evaluated an integrated service model of care where adult mental health services work collaboratively with ID services to deliver effective care for all individuals with ID. The mainstay of the model is the provision of specific beds for adults with mild ID and mental disorders within a general psychiatric ward and an associated community "virtual team" comprises professionals in ID and other mental health professionals including ward nursing staff. This structure is supported by an annual training day and twice-a-year review meetings to discuss and resolve organisational and management difficulties (Hall et al., 2006a). In a study of inpatients with ID, patients admitted to an integrated service model reported feeling less worried on admission and settled more easily into the hospital environment compared to patients admitted to general wards (Parkes et al., 2007). Further, inpatients with mild ID reported benefitting from the opportunity to interact with patients without disabilities (Hall et al., 2006a). In another study of integrated service programmes, inpatients with ID receiving services from an integrated programme reported significant improvement in psychiatric symptoms and overall functioning at discharge. Further improvements were noted 6 months after admission (Hall et al., 2006a). It would appear that well-developed hybrid units can lead to positive client outcomes for individuals with ID.

Results of an Ontario-based study examining clinical outcomes of inpatients with mild versus moderate/severe ID in specialist units found significant improvement for both groups, but greater reduction in GAF scores for inpatients with mild ID (Lunsky et al., 2010). No differences were observed between groups in terms of inpatient management, length of stay and likelihood of

readmission (Lunsky et al., 2010). In another Ontario-based study, GAF scores were directly compared among inpatients with ID served in specialist and general units of a teaching hospital (White et al., 2010). Results revealed that patients treated in both programmes showed significant improvement; however, in comparison to patients of the general programme, patients who received specialist care had significantly lower GAF scores during admission and discharge. Interestingly, individuals in general units were prescribed fewer psychotropic medications at admission than those in specialised units, but more at discharge. Findings highlight the trend of individuals with more severe and complex problems receiving care in specialist units (Hemmings et al., 2009; White et al., 2010; Xenitidis et al., 2004).

Satisfaction with Services

Several studies have compared the quality of general and specialist inpatient environments according to caregiver and patient report (Bouras, Holt, & Gravestock, 1995; Longo & Scior, 2004; Parkes et al., 2007; Samuels et al., 2007). Results suggest that caregivers rate specialist units more positively (Bouras et al., 1995; Longo & Scior, 2004; Samuels et al., 2007), whereas findings for general units have been more mixed (Chaplin, 2009; Longo & Scior, 2004; Parkes et al., 2007). Caregivers report that specialist inpatient units provide caring staff, practical help, positive and safe environments, open communication, clear information, good discharge planning and the ability to provide respite care (Longo & Scior, 2004; Samuels et al., 2007). General wards are described as more frightening for inpatients with ID (Parkes et al., 2007), and caregivers suggest that patients are often inappropriately sedated and not provided with the tools necessary to manage their care (Fox & Wilson, 1999).

Length of Inpatient Stay

Studies comparing duration of inpatient admission between specialist and general programmes have consistently reported significantly longer inpatient stays for individuals in specialist programmes (Alexander et al., 2001; Hemmings et al., 2009; White et al., 2010; Xenitidis et al., 2004). For example, in a UK-based study, the median length of inpatient stay was 19.3 weeks for a specialist programme and 5.5 weeks for a general programme (Hemmings et al., 2009). Similarly, findings of a Canadian study found a median duration of 119 days for a specialist group and 26 days for a general group. Moreover, individuals discharged from general units were significantly more likely to stay in the hospital for a period of 3 months or less.

Specialist Versus General: Is There a Winner?

There is a long-running debate over the adequacy of general psychiatric services in meeting the needs of people with ID and mental health issues. Whilst general services have certain advantages, specific concerns have been raised around specialist training, inadequate resources, unsupportive staff attitudes and increased likelihood of exploitation by other patients (Chaplin, 2004; Lunsky et al., 2008). Moreover, there is some evidence that mainstream services can support people with ID, but that general wards are unable to support individuals with ID who have more complex needs (Hemmings et al., 2009).

It is crucial to be cognizant of differences in the health service systems of different countries before making any generalisations. Since there are differences among countries in the way services are funded and organised, as well as the training opportunities offered, access to specialist care may vary widely (Lunsky et al., 2008). For example, there is some evidence that triaging takes place in the UK where health policy and partnerships between health and social services for mental health and ID are in place. Moreover, psychiatric trainees and nurses have access to more training opportunities in the UK where ID is a recognised and accredited subspecialty in psychiatry. Concerns have been raised that in Canada, general psychiatric services may not be serving the needs of people with ID and mental health problems adequately (Lunsky, Garcin, Morin, Cobigo, & Bradley, 2007).

An important point to consider is that research in this field has failed to keep pace with developments in community psychiatric services for patients with ID. Furthermore, research thus far has been largely descriptive, and randomised controlled trials have yet to take place. Studies comparing patient characteristics have produced inconsistent results, and research on patient outcomes is limited. As such, it is difficult to form any robust conclusions (Chaplin, 2011). Finally, although specialist services appear to have certain advantages, it is necessary to accept the fact that these units do not have the capacity to serve all patients with ID (Hall et al., 2006a, b).

Summary

In summary, nonrandomised, largely uncontrolled descriptive studies suggest that general psychiatric services, alone, may not be adequate to manage the complex needs of individuals with ID and mental health problems. Such services may be suitable for short-term stay, but it appears that individuals, especially those with severe disability, might be better served in specialised units or hybrid units which provide specialist input. Directions for future research include the identification of clinical and systemic predictors of specialised care with a focus on clinical and functional outcomes. There is also a need to conduct randomised controlled trials comparing specialist and general programmes to identify key elements of successful programmes for specific patient profiles.

Issues Pertaining to Hospitalisation

Hospitalisation is often viewed as something to be avoided at all costs. As with any intervention, hospitalisation carries a number of potential risks and benefits, and it is important to weigh these issues before proceeding with inpatient admission. As in the general population, a considerable proportion of individuals with ID are admitted because of significant risk of harm to self or others that cannot be managed on an outpatient basis. In some cases, assessing an individual in their community is not possible because their living environment or support systems are too limited or chaotic and there is no caregiver to provide reliable information. In these circumstances, patient risk is deemed so severe that hospitalisation may be the only option, even against the individual or their carer's wishes (Gardner & Hunter, 2003).

Many individuals with ID present with a complex interplay of developmental, psychiatric, behavioural and/or medical issues. For these individuals, hospitalisation may offer the best chance to problem solve issues in an interprofessional, collaborative environment with the opportunity for ongoing observation and assessment of the individual and his or her behaviours. Response to interventions can also be directly assessed without relying exclusively on informant report.

Disruption of Individual's Life and Routines: Access to Alternative Outpatient Services

Even in the best of circumstances, hospitalisation causes considerable disruption to an individual's life and can be very stressful for all individuals involved. Changes to routines and care giving can be particularly difficult for individuals with autism spectrum disorders and individuals with more severe disability who have fewer coping resources. Hospitalisation may also unnecessarily interrupt involvement in school, work or other activities that the individual enjoys and which are vital to their well-being.

In some cases, admission to a particular hospital unit may be inappropriate or preventable, for example, when women with legal involvement or a history of sexual trauma are placed in mixed-gender forensic units for sexual offenders (Berber & Boer, 2004). Similarly, individuals with severe developmental disabilities or autism spectrum disorders should not be placed in general acute units where there is little expertise in the understanding and treatment of individuals with ID. More generally, hospital admission

should be avoided when there is access to adequate and appropriate outpatient clinical services to meet the assessment and treatment needs of the individual.

Environmental and Social Challenges

Adjusting to the physical and social aspects of the hospital environment is difficult for most, and these challenges are magnified for individuals whose cognitive and coping abilities are significantly compromised. The physical environment of hospitals may not be suitable for individuals with sensory, physical or medical conditions including autism, epilepsy and vision impairment. In addition, many individuals with ID require hands-on assistance with activities of daily living (e.g. toileting, bathing), which can be difficult to carry out in the hospital environment. The physical space itself can also be very constraining for individuals whose engagement in activities is limited to motor/physical activities such as running, walking or playing ball. For inpatients with ID who are not allowed to leave the unit for extended periods of time due to safety concerns or risk of elopement, it can be challenging to find meaningful activities within the hospital unit.

The social environment, particularly on an acute unit, can be very overwhelming for inpatients with ID, particularly individuals with an autism spectrum disorder or sensory sensitivities. Noise levels, and proximity and intrusiveness of other patients can add significant distress to what these individuals are already experiencing. In addition, throughout the day individuals must adapt to frequent changes in staff and may also be cared for by unfamiliar persons.

Extended Admission and Discharge

Prolonged inpatient admission for individuals with ID is not uncommon, particularly among specialised psychiatric units (Lunsky et al., 2010; Sovner, Beasley, & Hurley, 1995; Xenitidis et al., 2004). This may occur in part because specialised units provide services to individuals with ID who present with more severe developmental disabilities and challenging behaviours. Unfortunately, however, prolonged admissions are often unrelated to clinical need, but rather due to an inability to discharge the individual to their previous residence, as caregivers no longer feel equipped to support them.

Some inpatients with ID do not understand why or for how long they will stay in hospital. They miss their home and caregivers and can become hopeless and depressed, particularly if they have worked hard to get better, but cannot be discharged because of lack of residential placement. Forensic patients might be more difficult to discharge because of their history and perceived risk of reoffending (Beer et al., 2005; Berber & Boer, 2004). Consequently, they may lose motivation to engage in therapeutic efforts and deteriorate behaviourally and emotionally.

It appears that with the closure of institutions, for some individuals with particularly challenging behaviours, a hospital bed becomes an unfortunate long-term substitution (Lyall & Kelly, 2007). From a service perspective, blocked beds reduce the capacity to provide specialised services and, in turn, create wait lists. More importantly, from the perspective of an inpatient with ID, long hospitalisations, particularly those beyond what is clinically needed, result in loss of skills, reduced quality of life and adjustment issues.

Potential for (Re)traumatisation

It is well documented that individuals with ID experience more trauma and are at a higher risk for traumatisation compared to the general population (Horner-Johnson & Drum, 2006; Peckham, 2007). Events that would not be considered traumatic by individuals of normal intelligence may be very distressing and traumatising for adults with ID whose emotional and cognitive abilities are limited (Martorell & Tsakanikos, 2008; Mitchek, Clegg, & Furniss, 2006). Individuals with ID are also more likely to have a history of physical, sexual or emotional abuse, severe neglect and placement outside of the family home. Hospital admission can trigger these memories, for example, assign-

ing a male staff to a female patient with a history of sexual abuse by a male caregiver.

Individuals with ID are also vulnerable in social interactions and can be at greater risk of assault or exploitation by other patients. Their behaviour can put them at greater risk of assault such as not reading social signals, being intrusive/bothering other patients, being loud or approaching too closely or touching. There is also risk of sexual abuse, which may go undetected if the person acquiesces, but lacks the capacity to make this kind of decision.

Separation from familiar caregivers and supports on which the person is dependent can be very difficult, particularly if the individual does not understand why they are hospitalised and when they will be discharged. In the case of individuals who are not verbal or who have limited language, it may be difficult to explain, prepare for or debrief treatment and management procedures and interventions. It may also be difficult or impossible to ascertain the needs, wishes or reasons for behaviour and fears. Whilst this is also the case in community settings, its impact is amplified in the hospital setting where the individual is exposed to an unfamiliar and changing environment with a large number of staff who do not know the person well.

Use of Intrusive Measures

A lot of progress has been made in recent years towards decreasing the use of mechanical restraints and locked seclusion in hospitals (Fisher, 2003; Gaskin, Stephen, & Happell, 2007). Over the last decade, least restraint policies, staff training and administrative supports aimed at eliminating, or at least reducing, the use of those measures have been introduced (Fisher, 2003). However, in circumstances where an individual is at risk for harming self or others, many of these measures are still in use. A study of 82 inpatients with developmental disabilities in psychiatric hospital wards revealed that women were involved in a disproportionately high number of incidents (e.g. seclusion, restraint, rapid tranquilisation) and were more likely to receive rapid tranquilisation after a violent incident (Sequeria & Halstead, 2001). Men, in contrast, were more likely to be secluded (Sequeria & Halstead, 2001). If aggressive or self-injurious behaviour is the reason for inpatient admission, there is a strong likelihood that the person will be restrained or secluded at some point during their hospital stay (Luiselli, 2009). Witnessing patient restraint or seclusion can also be very distressing for other inpatients with ID (Jones & Kroese, 2007), and they might come to fear that this could happen to them if they become angry or act out. For individuals with a history of sexual or physical abuse, experiencing or witnessing restraining procedures can be extremely re-traumatising.

Patient, Caregiver and Staff Experiences

Many individuals with ID and caregivers of individuals with ID report dissatisfaction with their hospital experience (Fox & Wilson, 1999; Iacono & Davis, 2003; Parkes et al., 2007). Studies suggest that patient needs are generally met in areas of nutrition, medication, mobility and discharge, but that hospital staff are often perceived to have negative attitudes towards people with developmental disabilities and lack skills and knowledge in the area (Iacono & Davis, 2003).

From a service provider perspective, hospital staff report feeling ill equipped to support and care for inpatients with ID, and it can be particularly difficult for hospital staff in general psychiatric units to serve this population when their experience is limited (Lennox, Diggens, & Ugoni, 2000; Lennox & Kerr, 1997). It can also be quite stressful to work in hospital settings when aggression is prominent (Chung & Corbett, 1998; Howard, Rose, & Levenson, 2009; Landon, Yágüez, & Kuipers, 2007). Further, hospital staff experience high levels of aggression and burnout when caring for individuals with ID compared to community or residential staff (Hensel, Lunsky, & Dewa, 2011). Not surprisingly then, several studies document negative attitudes of physicians and nurses towards people with developmental disabilities (Hart, 1998; Lennox et al., 2000; Paris, 1993; Shanley & Guest,

1995). It is likely that inadequate training of healthcare professionals in ID has led to negative attitudes among some healthcare workers and subsequent negative hospital experiences for individuals with ID. Ongoing mandatory training of mental health staff in ID, including specific training on how to prevent or reduce aggression and how to cope when working in an aggressive environment, has the potential to help improve both patient and staff experiences.

Summary

The decision to hospitalise is one which should be considered thoughtfully. It is important to avoid hospitalisation if it is not necessary, but equally important to hospitalise when other options provide less than optimal assessment and treatment. Early hospitalisation may prevent a crisis that could result in criminal charges, loss of housing or caregiver burnt out.

Conclusions and Future Directions

Reviews of existing psychiatric service models are limited, with some studies demonstrating effectiveness for general units (Bouras et al., 1995) and others for specialist units (Raitasuo et al., 1999; Xenitidis et al., 2004). Generally, findings suggest that no single service model has been successful in caring for all inpatients with ID. As such, some researchers have proposed the notion of an integrated service model of care where adult mental health services work collaboratively with ID services to deliver effective care for all individuals with ID (Hall et al., 2006a, b). Further study of these programmes is necessary to facilitate endorsement in other countries and communities. Specifically, studying key programme components (e.g. staff training, agency collaborations, interdisciplinary teams, early intervention, preventative services) and identifying standards of care which are in accordance with the needs of individuals with ID are important. Hospital admission procedures, policies and interventions must be adapted for patients with ID, and services should be monitored regularly to ensure delivery of quality care (Cowley et al., 2005). Further, recognition of how to handle mental health problems among primary care services will help detect and manage psychiatric disorders earlier (McCarthy & Boyd, 2002).

One of the current drivers in mental health is the reduction in inpatient beds (British Medical Journal Group, 2011) and the provision of specialised community support as well as alternatives to admission wards that disrupt as little as possible patients' lives and activities. In this context, there is an imperative that service provision for people with ID should be overhauled with the adoption of a modern, flexible range of services that run the full gamut from inpatient to day patient, to crisis units and to home treatment teams. These services need to be acceptable from a service user and carer perspective. Specialist provision for women only (Kohen, 2001) and step-down facilities for those with severe mental illness and complex comorbidities must also be considered. In a population-based study of inpatients with ID and psychiatric disorder, the majority of inpatients required either intensive outpatient treatment or "step-down facilities" with only 4 % requiring the tertiary level inpatient care they were receiving (Lunsky et al., 2006). Where possible, service users with ID should be supported to access these services, and at other times additional components may have to be delivered directly by professionals specialising in ID. Regardless of the approach chosen to fit local service configurations, we must strive towards evidence-based practice and to evaluate the clinical and cost-effectiveness of the interventions we provide.

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Part VI

Future Directions

23 Psychopathology in Intellectual Disability: Future Directions in Research and Evidence-Based Practice

Jane McCarthy and Elias Tsakanikos

Introduction

All the chapters in the book provide a reflection of current practice and the implications for future clinical practice and service delivery. This evidence must be viewed in the context that advances in the fields of neuroscience and genetic research will change clinical practice and delivery of services for people with intellectual disability in the coming decades. The purpose of this chapter is to highlight key themes for future research and clinical practice.

Future Directions in Research

In the past 25 years, research in the field of psychiatry of intellectual disability has focused on improving recognition, assessment and diagnosis of mental disorder presenting in people with intellectual disability (Moss & Hurley, 2013). In addition there has been an increase in health service-related research such as the development of community-based services particularly around the needs of those with challenging behaviours (O'Hara, Chaplin, Lockett, & Bouras, 2013).

At the same time technology is improving to allow further study of neurodevelopmental disorders such as autism spectrum disorders which will also give a wider understanding into the brain abnormalities of people with intellectual disability (Ecker, 2013).

Over the past decade, progress has been made in the understanding of the genetic and chromosomal causes of intellectual disability particularly the relevance of copy-number variations in the aetiology of intellectual disability (Stankiewicz & Beaudet, 2007). In addition the increased number of genes for X-linked forms of intellectual disability has been recognised as well as the importance of autosomal recessive forms of intellectual disability in which there may be mutations in many different genes (Ropers, 2008). This increased understanding of genetic conditions may lead to a greater understanding of the brain development, so giving rise to potential treatments early in life for those with intellectual disability.

Many countries have also witnessed significant changes in policy to improve the lives of people with intellectual disability with an emphasis on independence, choice and community-based care (Maulik, Harbour, & McCarthy, 2013). The authors in this book have provided a summary of the evidence base with the intention of identifying not only the gaps in knowledge but also the importance of the evidence base in influencing clinical practice today and in the future.

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Part I of the book is an overview of the developments in epidemiology, assessments and diagnosis. The key area identified for future research is the need for an expansion of epidemiological research in low- and middle-income countries on the presentation and prevalence of mental health problems in people with intellectual disability. In order to progress this research, we need to develop assessment tools that can work in countries with limited mental health resources. We also need to evaluate the effectiveness of this improved recognition and early diagnosis on the well-being of people with intellectual disability who may present with mental illness. The development of mental health services in the low- to middle-income countries is therefore an important area for future research including the impact of new policies on the lives of people with intellectual disability in these countries (Maulik et al., 2013). For example, there have been a number of policy initiatives in recent years such as 'Valuing People (DH, 2009)' in the UK and internationally (Cain et al., 2010) but little evidences of the impact of such policies on the lives of people with intellectual disability specifically those with mental health problems.

Parts III and IV of the book look at co-morbid and common clinical conditions.

The evidence base presented for common clinical conditions indicates a greater understanding in how we may recognise and diagnose these conditions through standardised diagnostic assessments used in the wider population but also with tools specifically developed for people with intellectual disability (Hassiotis, Steuber, Thomas, & Charlot, 2013). There is still little reported research on the validation of standardised diagnostic assessments of psychopathology in adults with intellectual disability and co-morbid conditions such as autism spectrum disorder and attention deficit hyperactive disorder (ADHD) (Underwood, McCarthy, & Tsakanikos, 2012; Xenitidis, Maltezos, & Asherson, 2013). Therefore from the perspective of clinical practice, there will be a greater encouragement to use standardised diagnostic tools for assessment and diagnosis to improve standards in clinical practice. This will hopefully allow an increased opportunity to use routinely collected clinical data for research purposes.

Improving the collection of standardised clinical data will allow a teasing out of the risk and protective factors in the causation of mental health problems in people with intellectual disability (Magiati, Tsakanikos, & Howlin, 2013) including those with personality disorders (Flynn, 2013) but also in time will increase our understanding of the wider population suffering from severe mental illness (Hemmings, 2013). It will also allow a more detailed understanding of the specific vulnerabilities, for example, people with autism spectrum disorders who develop mental illness studying specific neurobiological and sensory responses to environmental factors such as noise and light (Bradley, Caldwell, & Underwood, 2013). This will allow clinicians to understand how the environment needs to be modified into more 'autism friendly' and so in the long-term impact on the mental health problems of people with autism spectrum disorders and intellectual disability. It will increase understanding on how the environment impacts on the brain functioning and allow a more personalised approach to interventions and treatments for people with intellectual disability.

Research to date has been focused on the prevalence, presentation and subsequent assessment and diagnosis of psychiatric disorder in people with intellectual disability. This progress has allowed a greater clarity about the classification of psychiatric disorder in the majority of people with intellectual disability and so opened the opportunity for a more informed evaluation of specific interventions. The delineating of diagnostic groups and the overlap of co-morbid clinical conditions such as autism spectrum disorder and ADHD have been aided by the development of diagnostic systems specific to individuals with intellectual disability such as Diagnostic Manual for Intellectual Disability (DM-ID) (Fletcher, Lusches, Stavrakaki, & First, 2007) and Diagnostic Criteria for Learning Disability (DC-LD) (Royal College of Psychiatrist, 2001).

For clinicians working in the field of intellectual disability, a significant amount of our time is seeing people without a defined psychiatric disorder or clinical condition (Alim, Paschos, &

Herne, 2013). Therefore the issue comes in understanding and managing those who present with a spectrum of behavioural problems (Rieske & Matson, 2013). The diagnostic systems designed for people with intellectual disability improve clinical assessment in those presenting with co-morbid conditions and behavioural problems that do not fall under a category of mental disorder and also support a more systematic approach to research (Hamelin, Maum, & Sturmey, 2013).

The past decade has seen an increased understanding in the needs of people with intellectual disability in contact with the Criminal Justice System (Chaplin & McCarthy, 2013; Murphy & Mason, 2013) specifically on improving their recognition of people with intellectual disability and other neurodevelopmental disorders across the criminal justice system. Systematic assessment of risk particularly for people with intellectual disability in community services and effectiveness of adapted interventions for specific offending behaviour, e.g. sex-offending treatment programmes, will be the focus of future research.

Part V of the book reviews the evidence base on interventions in people presenting with a psychiatric disorder or behaviour problems. The evidence base for the effectiveness of psychological interventions has grown in the past 10 years (Sturmey & Hamelin, 2013) but still requires further published work on psychological interventions for common mental disorders such as depression. Deb (2013) in his chapter focuses on the use of psychopharmacology for behavioural problems; however, for specific interventions on psychiatric disorder he refers the reader to the National Guidelines evidence base, used in the wider population. The challenges in undertaking research using psychopharmacological interventions in people with intellectual disability were highlighted through the study undertaken by Tyrer et al. (2009) with the key problem being recruitment of participants. This is why future research in this field must be embedded into day-to-day clinical practice by collecting data on cohorts of individuals across a number of centres not only at national but also at international level. Therefore the research of the future will be about building up databases of routine clinically collected data with standardised assessments. This will progress clinical knowledge alongside the advances in science and technology to increase the biological and genetic understanding of the mental health needs of people with intellectual disability.

The importance of professional networks across countries and continents is critical to the future research into the mental health needs of people with intellectual disability. In order to understand the complex interplay of biological factors such as epilepsy (Ring & Winterhalder, 2013), environmental and social factors in the presentation and outcomes of psychiatric disorder in people with intellectual disability, clinical data is required across a large pool of individuals. Researchers need to support clinicians in developing the new evidence base but just as important is the role of practitioners in the psychiatry of intellectual disability, supporting the research of tomorrow. Therefore research networks such as Challenging Behaviour and Mental Health Specialist Interest Group of the International Association for the Scientific Study of Intellectual and Developmental Disabilities (http://www.iassid.org) and European Association of Mental Health in Intellectual Disability (http://www.MHID.org) are important in supporting such opportunities for future research.

Implications for Practice

There will be an increasing recognition across all countries in the need to support evidence-based care pathways for people with intellectual disability presenting with psychiatric disorder. Care pathways are the key into improving outcomes and ensuring reduced variations in practice by making explicit the standards in assessments and interventions required (Vanhaecht, Panella, van Zelm, & Sermeus, 2010). The pathways will look at those who enter into the pathway at the point of referral and identify the assessments required and the appropriate intervention taking an individualised approach. For those using inpatient services, it will be important to specifically focus on progress through and discharge out of services.

Recent reports around poor care of people with intellectual disability in hospital settings will drive the agenda further to enhancing community services for those presentencing with challenging behaviours (Department of Health, 2013).

Service models of today and in the future will develop care pathways around the clinical need of the individual supported by the best evidence base available. For example, adult services for people with intellectual disability may have a number of defined care pathways to provide for those with neurodevelopmental disorders such as autism spectrum disorder, those with severe mental illness such as schizophrenia, those presenting with substance misuse (Taggart & Chaplin, 2013) and those with challenging behaviour. The focus will not only be on improving the assessment and diagnostic skills of clinicians through standardised tools and training but also on ensuring a consistent approach across services using measures of outcomes to show improved well-being (Kobler, 2012).

Future changes to the classification system to DSM-V and ICD-11 will not only change terminology to intellectual development disorders but also our approach to diagnosis, for example, in those with autism spectrum disorders.

Services in these difficult financial times will need to show cost-effectiveness and efficiency in the delivery of services with proven outcomes for users of the service (Lake, Balogh, & Lunsky, 2013). Therefore we will need to have measures that are validated and reliable in showing good outcomes such as quality-of-life measures, well-being measures and clinical response to interventions (Kobler, 2012). These interventions will need to be driven by current evidence base, but we need more evidence on psychological interventions (Sturmey & Hamelin, 2013) specifically in combination with pharmacological interventions. We need to see improvement in assessment and diagnostic leads to a more personalised approach to recommending medication, psychological and environmental interventions.

The past decade has seen policies that have encouraged choice, independence and improved access to services. The past 40 years for many countries has seen the closure of large hospitals and the development of community-based services (O'Hara et al., 2013). However for many countries such policy and service developments may take many years to achieve.

The whole field of medicine is on a path of increasing specialisation, and for the psychiatry of intellectual disability, this may require the development of experts in particular areas at a national, regional or each local level for those with genetic conditions such as Prader-Willi or Down syndrome (Paschos, Bass, & Strydom, 2013). For others this may require a specialisation in those with severe mental illness such as schizophrenia or those with dementia (Hemmings, 2013; Strydom & Sinai, 2013). The development of a critical group of professionals with expertise around a defined spectrum of psychiatric conditions may only be possible in countries with highly developed specialist mental health services for people with intellectual disability. For other countries access to a mental health professional with expertise in intellectual disability may need a different approach with a development of professionals working in the mainstream services for the wider population with specialist services being developed and accessed at regional or state level.

It may be with improved and defined care pathways that more people with intellectual disability and psychiatric disorders can access the care and treatment required to improve their outcome and ultimately their quality of life. Clinicians and researchers in the psychiatry of intellectual disability will find the summary of the current evidence base provided in this book hopefully informative but more importantly will encourage reflection on their current practice to drive improvements in practice and research in the future.

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