



**Practice Guidelines**

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**for the Assessment**

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**and Diagnosis of**

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**Mental Health**

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**Problems in Adults**

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**with Intellectual**

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**Disability**

Shoumitro Deb,  
Tim Matthews,  
Geraldine Holt  
& Nick Bouras

**MH MR**

**Practice Guidelines for the Assessment  
and Diagnosis of Mental Health Problems  
in Adults with Intellectual Disability**

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Shoumitro Deb, Tim Matthews, Geraldine Holt & Nick Bouras

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for The European Association for Mental Health in Mental Retardation

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# C ONTENTS

<b>About the editors</b>	<b>vii</b>
<b>Members of the Practice Guidelines Panel</b>	<b>viii</b>
<b>Preface</b>	<b>ix</b>
<b>Introduction</b>	<b>1</b>
<b>Section 1</b> <i>Epidemiology</i>	<b>5</b>
<b>Section 2</b> <i>Assessment procedure</i>	<b>21</b>
<b>Section 3</b> <i>Particular psychiatric disorders</i>	<b>31</b>
<b>Appendix</b>	<b>103</b>



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The following contributors have reviewed the *Practice Guidelines for the Assessment and Diagnosis of Mental Health Problems in Adults with Intellectual Disability* and made comments on the initial draft of this publication. They were identified from their recent publications, as holders of research grants and members of the European Association MH-MR. Of the 38 professionals to whom we sent the Practice Guidelines, 30 replied (80%).

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# PREFACE

In the last decade the professional knowledge concerning the problems of mental health among persons with intellectual disability has significantly grown. Behavioural and psychiatric disorders in these individuals can cause serious obstacles to their social integration

Clinical experience and research show that the existing diagnostic systems of DSM-IV and ICD-10 are not fully compatible when making a psychiatric diagnosis in people with intellectual disability. This may be one of the reasons why the evidence-based knowledge on the assessment and diagnosis of mental health problems in people with intellectual disability is still scarce.

This is the reason for the European Association for Mental Health in Mental Retardation (EAMHMR) supporting the current project to produce a series of Practice Guidelines for those working with people with intellectual disability to encourage and promote evidence-based practice. The first publication of the series is entitled *Practice Guidelines for the Assessment and Diagnosis of Mental Health Problems in Adults with Intellectual Disability*.

This work was undertaken very skillfully by Dr Shoumitro Deb, Dr Tim Matthews, Dr Geraldine Holt and Professor Nick Bouras. Comments were received from an international panel of experts in the field of mental health and intellectual

disability.

On behalf of the Executive Committee of the EAMHMR I would like to thank all contributors. I believe that their hard work will influence the quality of care provided to people with intellectual disability and mental health problems and will further evidence-based practice and research.

**Anton Dosen**

*President*

*European Association for Mental Health in Mental Retardation*

# INTRODUCTION

This document is the first in a series of practice guidelines that the European Association for Mental Health in Mental Retardation (EMHMR) wishes to develop. These guidelines are based on the current evidence on the subject, and consensus opinion from clinicians working in this field.

Unlike the procedure used in the *Health Evidence Bulletins Wales – Intellectual Disability* (Hamilton-Kirkwood *et al*, 2001), we have not critically appraised the evidence in this document but used the following convention (Cochrane Library, 2001) to categorise the type of evidence:

- 'Type I evidence' – good systematic review and meta-analysis (including at least one randomised controlled trial)
- 'Type II evidence' – randomised controlled trial
- 'Type III evidence' – well designed interventional studies without randomisation
- 'Type IV evidence' – well designed observational studies
- 'Type V evidence' – expert opinion, influential reports and studies

Most of the comments received from members of the expert consensus panel have been incorporated in the text. Because of

lack of information we could not categorise all evidence according to the convention mentioned above. The list of evidence in this document is by no means an exhaustive one, and not all references listed are quoted in the text. Some references are taken from the *Health Evidence Bulletins Wales – Intellectual Disability* (Hamilton-Kirkwood *et al*, 2001) and some are cross-references cited in other papers.

This document is focused on the description of psychiatric illnesses that could affect adults with intellectual disability. The issues related to the diagnosis of psychiatric illness among children with intellectual disability are different, and therefore have not been covered in this document. Aspects of treatment, pervasive developmental disorders and behaviour problems are also not covered. These topics will be the subject of future guidelines.

Recently, the *American Journal of Mental Retardation* (Aman *et al*, 2000) published consensus guidelines for the diagnosis, assessment and treatment of mental illness in people with intellectual disability. These guidelines, however, do not describe in detail various symptoms that are associated with different psychiatric illnesses. In this document we have described various features of mental illnesses in adults with an intellectual disability in a descriptive fashion so that the readers find it easy to read and could use this as a reference point.

Whilst the guidelines in this document are primarily intended to help psychiatrists and psychologists specialising in psychiatric diagnosis and treatment of adults who have intellectual disability, it is our hope that other professionals and carers working with adults with intellectual disability could also use this document as a point of reference. Hopefully, this will raise awareness

about adults with intellectual disability who develop signs of psychiatric illness, and consequently promote referral to specialists for further assessment and treatment.

The guidelines in this document should not be confused with diagnostic criteria such as DCR-10 (Diagnostic Criteria for Research-10th revision) (WHO, 1992); DSM-IV-TR (Diagnostic and Statistical Manual – 4th text revision) (American Psychiatric Association, 2000); or the recently published DC-LD (Diagnostic Criteria – Learning Disability) (Royal College of Psychiatrists, 2001). The latter has recently been developed as a specific tool for use in people with intellectual disability. Neither should these practice guidelines be confused with diagnostic instruments such as the Psychiatric Assessment Schedule for Adults with Developmental Disability (PAS-ADD) (Moss *et al*, 1993).

While this document makes reference to standardised classification systems – especially the International Classification of Diseases – 10th revision (ICD-10) (WHO, 1992) – it is clear that these systems are not always useful in intellectual disability. Some features might not always be apparent, or be difficult to elicit, making diagnosis difficult, particularly in those who have severe intellectual disability, and impaired communication. This document tries to describe clinical features of psychiatric illness, with especial reference to where these may differ from their presentation in the general, non-intellectually disabled population. Perhaps these guidelines might be useful in further development of the ICD system and particularly for developing a special ICD-11 for adult people with intellectual disability.

The guidelines start with a brief account of the prevalence studies of psychiatric illness among adults with an intellectual

disability. Then some important principles in the assessment of people with an intellectual disability are discussed. This discussion is followed by a section presenting particular psychiatric disorders in this group, with particular reference to how the intellectual disability may affect the manifestation of mental health problems in this population. Finally, the **Appendix** lists current diagnostic systems and rating scales that have been developed for use specifically with people with intellectual disability.

# SECTION 1

## *Epidemiology*

Intellectual disability, or ‘mental retardation’ as it is described in ICD-10 (WHO, 1992) and the Diagnostic and Statistical Manual – IVth revision text review (DSM-IV-TR) (APA, 2000) – is not a psychiatric illness, despite being part of the psychiatric classification system. The quoted prevalence of psychiatric illness among adults with intellectual disability varies widely between 10% and 39% (Corbett, 1979; Jacobson, 1982; Eaton & Menolascino, 1982; Lund, 1985; Göstason, 1985; Inverson & Fox, 1989; Reiss, 1990; Borthwick-Duffy & Eyman, 1990; Bouras & Drummond, 1992; Hagnell *et al*, 1993; Borthwick-Duffy, 1994; Cooper, 1997; Roy *et al*, 1997; Deb *et al*, 2001a). This fourfold discrepancy in the quoted prevalence rate is caused by methodological difficulties, particularly in the areas of sampling error and case ascertainment.

Up until recently, most prevalence studies of psychiatric illness among adults with intellectual disability included primarily people from institutions or from a clinic population, therefore causing sampling bias. Case ascertainment has also been a problem because of the difficulty of detecting adults with mild intellectual disability in the population. Many studies were based on case-notes scrutiny. Direct patient interviews were seldom used. Even where direct patient interviews *were* used, these often depended on screening instruments such as the

Psychopathology Instrument for Mentally Retarded Adults (PIMRA) (Matson *et al*, 1984), Reiss Scale (Sturmey *et al*, 1995), Mini-Psychiatric Assessment Schedule for Adults with Developmental Disabilities (Mini-PAS-ADD) (Prosser *et al*, 1998), and PAS-ADD checklist (Moss *et al*, 1998), therefore increasing the chance of detecting a higher rate of psychiatric illness in the study population.

Some studies, however, used direct patient interviews using instruments such as PAS-ADD (Moss *et al*, 1993) or Medical Research Council-Handicap and Behaviour Schedule (MRC-HBS) (Wing, 1980) and made psychiatric diagnoses according to the DSM-III (APA, 1980) criteria. The difficulty of diagnosing psychiatric illness using these criteria in adults who have severe and profound intellectual disability is well known.

Some authors included personality disorder, behavioural disorders, autism, attention deficit hyperactivity disorder (ADHD), Rett syndrome, dementia, and pica in their overall diagnosis of psychiatric illness. This caused wide discrepancy in the quoted prevalence rate.

It appears that if diagnoses like behavioural disorder, personality disorders, autism, and ADHD are excluded, the overall rate of psychiatric illness in adults with intellectual disability does not differ significantly from that in the non-intellectually disabled general population, (Deb *et al*, 2001a). Compared with the general population, there seems to be a higher rate of schizophrenia among adults who have mild to moderate intellectual disability (Turner, 1989; Doody *et al*, 1998; Copper, 1997; Deb *et al*, 2001a). However, if behaviour disorders are included within psychiatric diagnoses, the rate of psychiatric illness seems significantly more prevalent among

adults with intellectual disability compared with the general population (Meltzer *et al*, 1995; Deb *et al*, 2001b). Whether or not behavioural disorders are included in the overall diagnosis of psychiatric illness, behavioural problems are common causes for psychiatric referrals (Kohen, 1993; Deb, 2001a).

Studies show controversial evidence as to whether or not psychiatric illness is more common among severely, compared with mildly, intellectually disabled adults. Göstason (1985) and Lund (1985) both showed higher rates of psychiatric illness among more severely intellectually disabled adults, whereas, Inverson and Fox (1989), Jacobson (1982), and Borthwick-Duffy and Eyman (1990) all showed a higher prevalence of psychiatric illness among adults with a milder degree of intellectual disability. Corbett (1979) found no relationship either way. The problem may lie in the fact that the authors used the same psychiatric diagnostic criteria for adults with all degrees of severity of intellectual disability, which may not be appropriate.

Because of the heterogeneity in abilities and communication skills among adults with intellectual disability, it is difficult to use one standardised criterion for psychiatric diagnosis across the whole spectrum. While psychiatrists tend to use syndromic classification (a cluster of symptoms or behaviours that relate with each other), some believe that the approach of behavioural classification that is more prevalent in the clinical psychology literature may be more appropriate for use in adults with severe intellectual disability.

## Risk factors for psychiatric morbidity

It is important to detect relevant predisposing, precipitating and perpetuating risk factors associated with psychiatric illness in adults with intellectual disability.

### Biological factors

- **Genetic liability:** Fragile X syndrome; Prader-Willi syndrome; Williams syndrome; Rett syndrome; Lesch-Nyhan syndrome; Cornelia de Lange syndrome; cri-du-chat syndrome and so on, are shown to be associated with certain 'Behavioural Phenotypes' (O'Brien & Yule, 1995; Deb, 1997a; Deb & Ahmed, 2000) and velo-cardio-facial syndrome with schizophrenia (Murphy *et al*, 1999).
- **Structural abnormality in the frontal lobe** can cause apathy, social withdrawal and disinhibition.
- **Interaction between the environment and existing physical disabilities** such as spasticity, or mobility problems; sensory deficits in hearing and vision; or speech and language difficulties may indirectly cause psychopathology.
- 14–24% people with intellectual disability have a **lifetime history of epilepsy** (Deb, 2000). Epilepsy could predispose to psychopathology in adults with intellectual disability (Deb, 1997b; Deb & Joyce, 1998; Deb *et al*, 2001b).
- **Abnormal thyroid function test** can be detected among one third of children and adults with Down's syndrome (Deb, 2001b). Thyroid disorder could predispose to psychopathology in adults who have intellectual disability.
- Prescribed and non-prescribed drugs can cause psychopathology.

## Psychological factors

- impaired intelligence
- impaired memory due to the dysfunction of the temporal lobes of the brain
- impaired sense of judgement and lack of initiative caused by damage to frontal lobes
- lower thresholds for stress tolerance
- poor self-image
- immature psychological defence mechanism such as 'regression' when under stress
- inability to solve problems using abstract thinking
- learned dysfunctional or abnormal coping strategies (manifestation of anger under stressful situations)
- lack of emotional support

## Social factors

- under- or over-stimulating environment
- conflicts with family members or residents or staff members
- issues around the lack of social support
- difficulties in developing fulfilling relationships
- problems in finding employment
- physical and psychological abuse
- lack of appropriate social exposure and patronisation by others

- lack of integration within the wider society, stigmatisation, and discrimination
- bereavement and other life events
- changes in the immediate environment, with the family and carers
- carer stress

It is worth remembering that in diagnosing psychiatric illness, it is important to differentiate which symptoms could be part of such an illness, and which can be explained by the intellectual disability. Signs and symptoms (which are common in psychiatric illness) – such as social withdrawal, excessive agitation, lack of concentration, stereotyped movement disorders, abnormal sleep, and certain other behaviours – can be the expression of underlying brain damage rather than symptoms of an illness (*‘diagnostic overshadowing’*; Reiss & Sysko, 1993).

It is therefore important to establish a ‘baseline’ of what the subject was like, and look for any change from this. For example, a patient may have a longstanding history of difficulty getting to sleep. However, carers have noticed in recent months that she has also been waking more early than usual, which is a change from baseline, and could be a symptom of a psychiatric illness (*‘baseline exaggeration’*; Sovner & Hurley, 1989).

**EVIDENCE**

AACAP Official Action (1999) Practice parameters for the assessment and treatment of children, adolescents, and adults with mental retardation and comorbid mental disorders. *Journal of American Academy of Child and Adolescent Psychiatry* **38** (12) S5–S31.

Aman, M. G., Alvarez, N., Benefield, W., Crismon, M. L., Green, G., King, B. H., Reiss, S., Rojahn, J. & Szymanski, L. (Eds) (2000) Expert Consensus Guideline Series: Diagnosis and assessment of psychiatric and behavioral problems in mental retardation. *American Journal on Mental Retardation* **105** (3 – May) 159–169.

(Type V evidence: expert opinion.)

American Psychiatric Association (1980) *Diagnostic Statistical Manual* (3rd Revision, DSM-III). APA, Washington DC.

(Type V evidence: consensus opinion)

American Psychiatric Association (1994) *Diagnostic Statistical Manual* (4th Revision, DSM-4). APA, Washington DC.

(Type V evidence: consensus opinion.)

American Psychiatric Association (2000) *Diagnostic Statistical Manual* (4th Revision, Text Review DSM-4 TR). APA, Washington DC.

(Type V evidence: consensus opinion.)

Amir, R. E., Van den Veyver, I. B., Wean, M. *et al* (1999) Rett syndrome is caused by mutations in X-linked MECP-2, encoding methyl-CpG-binding protein 2. *Nature Genetics* **23** 185–188.

Ballinger, B. R., Ballinger, C. B., Reid, A. H. & McQueen, E. (1991) The psychiatric symptoms, diagnosis and care needs of 100 mentally handicapped patients. *British Journal of Psychiatry* **158** 255–259.

Borthwick-Duffy, S. A. (1994) Epidemiology and prevalence of psychopathology in people with mental retardation. *Journal of Consulting and Clinical Psychology* **62** (1) 17–27.

(Type IV evidence: review of eight observational studies published between 1975 and 1985 involving adults with intellectual disability in both hospital and community settings. In this paper authors have also reviewed studies on children with intellectual disability.)

Borthwick-Duffy, S. A. & Eyman, R. K. (1990) Who are the dually diagnosed? *American Journal of Mental Retardation* **94** 586–595.

(Type IV evidence: cross-sectional study of psychological symptoms among 78,603 adults with intellectual disability. Information was collected from California Developmental Disability register's case-records.)

Bouras, N. & Drummond, C. (1992) *Behaviour and psychiatric disorders of people with mental handicaps living in the community*. *Journal of Intellectual Disability Research* **36** 349–357.

(Type IV evidence: cross-sectional study of 318 adults with intellectual disability referred to a Community Learning Disability Team in London, using DSM-III-R criteria.)

Campbell, M. & Malone, R. P. (1991) Mental retardation and psychiatric disorders. *Hospital and Community Psychiatry* **42** 374–379.

Charlot, L. R. (1998) Developmental effects on mental health disorders in persons with developmental disabilities. *Mental Health Aspects of Developmental Disabilities* **1** (2) 29–38.

Clarke, D. J., Cumella, S., Corbett, J., Baxter, M., Langton, J., Prasher, V., Roy, A., Roy, M. & Thinn, K. (1994) Use of ICD-10 Research Diagnostic Criteria to categorise psychiatric and behavioural abnormalities among people with learning disability: The West Midlands trial. *Mental Handicap Research* **7** 273–285.

Clarke, D. J. & Gomez, G.A. (1999) Utility of modified DCR-10 criteria in the diagnosis of depression associated with intellectual disability. *Journal of Intellectual Disability Research* **43** 413–420.

Cochrane Library (2001) Issue 2. Update Software, Oxford.

Collacott, R. A., Cooper, S-A. & McGrother, C. (1992) Differential rates of psychiatric disorders in adults with Down's syndrome compared with other mentally handicapped adults. *British Journal of Psychiatry* **161** 671–674.

(Type IV evidence: cross-sectional study of 371 adults with Down's syndrome compared with 371 matched adults with intellectual disabilities of causes other than Down's syndrome.)

Cooper, S.-A. (1997) Psychiatry of elderly compared to younger adults with intellectual disability. *Journal of Applied Research in Intellectual Disability* **10** (4) 303–311.

(Type IV evidence: cross-sectional study of 134 people over 64 years of age and 73 adults between 20 and 64 years of age with intellectual disability. Assessments were carried out using Present Psychiatric State for Adults with Learning Disabilities [PPS-LD].)

Corbett, J. A. (1979) Psychiatric morbidity and mental retardation. In: F. E. James and R. P. Snaith (Eds) *Psychiatric Illness and Mental Handicap* pp11–25. London: Gaskell Press.

(Type IV evidence: cross-sectional study of psychiatric morbidity in a population-based sample of 140 children and 402 adults with intellectual disability in London)

Deb, S. (1997a) Behaviour Phenotype. In: S. G. Read (Ed) *Psychiatry in Learning Disability* pp93–116. London: W. B. Saunders.

Deb, S. (1997b) Mental disorder in adults with mental retardation and epilepsy. *Comprehensive Psychiatry* **38** (3) 179–184.

(Type IV evidence: case controlled study of 150 adults with learning disability and epilepsy and an age-, sex- and IQ-matched control group of 150 people without epilepsy disability.)

Deb, S. (2000) Epidemiology and treatment of epilepsy in patients who are mentally retarded. *CNS Drugs* **13** (2) 117–128.

(Type IV evidence: review of literature.)

Deb, S. (2001a) Epidemiology of psychiatric illness in adults with intellectual disability. In: L. Hamilton-Kirkwood, Z. Ahmed and S. Deb *et al* (Eds) *Health Evidence Bulletins – Learning Disabilities (Intellectual Disability)* pp14–17. [wttp://hebv.uwcm.ac.uk](http://hebv.uwcm.ac.uk)

(Type IV evidence: review of literature.)

Deb, S. (2001b) Medical conditions in people with intellectual disability. In: L. Hamilton-Kirkwood, Z. Ahmed and S. Deb *et al* (Eds) *Health Evidence Bulletins – Learning Disabilities (Intellectual Disability)* pp48–55. [wttp://hebv.uwcm.ac.uk](http://hebv.uwcm.ac.uk).

(Type IV evidence: review of literature.)

Deb, S. & Ahmed, Z. (2000) Special conditions leading to mental retardation. In: M. G. Gelder, N. Adreassen & J. J. Lopez-Ibor Jr (Eds) *New Oxford Textbook of Psychiatry* pp1953–1963. Oxford: Oxford Press.

(Type IV evidence: review of literature.)

Deb, S. & Joyce, J. (1998) Psychiatric illness and behavioural problems in adults with learning disability and epilepsy. *Behavioural Neurology* **11** (3) 125–129.

(Type IV evidence cross-sectional study of 143 adults with intellectual disability and epilepsy.)

Deb, S., Thomas, M. & Bright, C. (2001a) Mental disorder in adults who have intellectual disability. 1: Prevalence of functional psychiatric illness among a 16–64 years old community-based population. *Journal of Intellectual Disability Research* (in press).

(Type IV evidence: cross-sectional observational study of the rate of functional psychiatric illness among a population-based sample of 101 adults with intellectual disability.)

Deb, S., Thomas, M. & Bright, C. (2001b) Mental disorder in adults who have intellectual disability. 2: The rate of behaviour disorders among a 16–64 years old community-based population. *Journal of Intellectual Disability Research* (in press).

(Type IV evidence: cross-sectional observational study of the rate of behavioural

disorders among a population-based sample of 101 adults with intellectual disability.)

Deb, S. & Weston, S. N. (2000) Psychiatric illness and mental retardation. *Current Opinion in Psychiatry* **13** 497–505.

(Type IV evidence: review of literature.)

Doody, G. A., Johnstone, E. C., Sanderson, T. L., Cunningham Owens, D. G. & Muir, W. J. (1998) ‘Propfschizophrenie’ revisited: Schizophrenia in people with mild learning disability. *British Journal of Psychiatry* **173** 145–153.

(Type IV evidence: a case control study of adults with mild learning disability with and without schizophrenia, and a control group of non-intellectually disabled adults with schizophrenia. The study assessed various aspects of demographical variables and symptomatology among the three groups.)

Dorn, M. B., Mazzacco, M. M. & Hagerman, R. J. (1994) Behavioral and psychiatric disorders in adult male carriers of Fragile X. *Journal of American Academy of Child and Adolescent Psychiatry* **33** 256–264.

Dosen, A. (2000) Psychiatric and behavioural disorders among mentally retarded adults. In: M. Gelder, J. Lopez-Ibor Jr. & N. Andreasen (Eds) *New Oxford Textbook of Psychiatry* pp1972–1978. Oxford: Oxford University Press.

Eaton, L. F. & Menolascino, F. J. (1982) Psychiatric disorders in the mentally retarded: types, problems and challenges. *American Journal of Psychiatry* **139** 1297–1303.

(Type IV evidence: cross-sectional study of 798 participants in community-based intellectual disability programme.)

Fox, R. A. & Wade, E. J. (1998) Attention deficit hyperactivity disorder among adults with severe and profound mental retardation. *Research in Developmental Disabilities* **19** (3) 275–280.

(Type IV evidence: cross-sectional study of 86 adults with severe to profound intellectual disability from a community setting using the Connor’s Hyperactivity Index.)

Gecz, J., Barnett, S., Liu, J. *et al* (1999) Characterisation of the human glutamate receptor subunit 3 gene (GRIA3), a candidate for bipolar disorder and non-specific X-linked mental retardation. *Genomics* **62** 356–368.

Gedye, A. (1998) *Behavioral Diagnostic Guide for Developmental Disabilities*. Vancouver BC, Canada: Diagnostic Books.

Glick, M. (1998) A developmental approach to psychopathology in people with mental retardation. In: J. Burack, R. Hoddap & E. Zigler (Eds) *Handbook of Mental Retardation and Development* pp563–582.

Cambridge: Cambridge University Press.

Göstason, R. (1985) Psychiatric illness among the mentally retarded: a Swedish population study. *Acta Psychiatrica Scandinavica* **318** 1–117.

(Type IV evidence: cross-sectional study of 51 severely and 64 mildly intellectually disabled adults and 64 control cases. Assessed using Comprehensive Psychopathological rating Scale (CPRS) and DSM III diagnosis.)

Gothelf, D., Frisch, A., Munitz, H. *et al* (1999) Clinical characteristics of schizophrenia associated with velo-cardio-facial syndrome. *Schizophrenia Research* **35** 105–112.

Hagnell, O., Ojesjo, L., Otterbeck, L & Rorsman, B. (1993) Prevalence of mental disorders, personality traits and mental complaints in the Lundby study. *Scandinavian Journal of Social Medicine* **21** (Suppl. 50) 1–76.

(Type IV evidence: cross-sectional study of a geographically defined total population of 2672 adults over a 25-year period.)

Hamilton-Kirkwood, L., Ahmed, Z., Deb, S. *et al* (2001) *Health Evidence Bulletin Wales: Learning Disabilities (Intellectual Disability)*. [wttp://hebw.uwcm.ac.uk](http://hebw.uwcm.ac.uk). Cardiff, Wales: NHS.

(Type IV evidence: critical review of literature.)

Hampson, R. M., Malloy, M. P., Mors, O. *et al* (1999) Mapping studies on a pericentric inversion (18) (p11.31 q21.1) in a family with both schizophrenia and learning disability. *Psychiatric Genetics* **9** 161–163.

Holden, P. & Neff, J. A. (2000) Intensive outpatient treatment of persons with mental retardation and psychiatric disorder: a preliminary study. *Mental Retardation* **38** (1) 27–32.

(Type III evidence: non-randomised controlled study of 28 adults with intellectual disability and severe psychiatric disorder.)

Holland, A. J. & Koot, H. M. (1998) Mental health and intellectual disability: An international perspective. *Journal of Intellectual Disability Research* **42** 505–512.

(Type V evidence: review article.)

Iverson, J. C. & Fox, R. A. (1989) Prevalence of psychopathology among mentally retarded adults. *Research in Developmental Disabilities* **10** 77–83.

(Type IV evidence: cross-sectional study using PIMRA among a random sample of 165 adults receiving intellectual disability services in the Midwestern USA.)

Jacobson, J. W. (1982) Problem behavior and psychiatric impairment within a developmentally disabled population: I Behavior frequency. *Applied Research in Mental Retardation* **3** 121–139.

(Type IV evidence: cross-sectional study of psychological symptoms using case

records of 27,023 of people with intellectual disability known to the New York Developmental Disability system.)

Jacobson, J. W. (1990) Do some mental disorders occur less frequently among persons with mental retardation? *American Journal of Mental Retardation* **94** 596–602.

Kohen, D. (1993) Psychiatric emergencies in people with a mental handicap. *Psychiatric Bulletin* **17** 587–589.

(Type IV evidence: 12 month prospective study of referral pattern in a London borough.)

Kon, Y. & Bouras, N. (1997) Psychiatric follow-up and health services utilisation for people with learning disabilities. *British Journal of Developmental Disabilities* **43** (1) 20–26.

(Type IV evidence: longitudinal study with one and five year follow ups of 74 adults with intellectual disability following resettlement in the community.)

Lund, J. (1985) The prevalence of psychiatric morbidity in mentally retarded adults. *Acta Psychiatrica Scandinavica* **72** 563–570.

(Type IV evidence: cross-sectional cohort study of 302 adults with intellectual disability, identified from the Danish National Register. It also draws comparisons with eight previous cross-sectional studies.)

Matson, J. L., Kazdin, A. E. & Senatore, V. (1984) Psychometric properties of the Psychopathology Instrument for Mentally Retarded Adults. *Applied Research in Mental Retardation* **5** 81–90.

(Type IV evidence: observational study.)

Meltzer, H., Gill, B., Petticrew, M. & Hinds, K. (1995) *The prevalence of psychiatric morbidity among adults living in private households: OPCS survey of psychiatric morbidity in Great Britain, report 1*. London: HMSO.

(Type IV evidence: cross-sectional study of psychiatric illness using Clinical Interview Schedule-Revised, among 10,108 adult general population in England and Wales.)

Mental Health-Special Interest Group (MH-SIRG); International Association for the Scientific Study of Intellectual Disability (IASSID) (2000) *Mental health and intellectual disabilities: addressing the mental health needs of people with intellectual disabilities* (in press).

(Type V evidence: expert opinion.)

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## SECTION 2

### *Assessment procedure*

The assessment will detect most information when it is done in a systematic and comprehensive way. There may already be a lot of background information about the patient, from previous assessments and records.

There are various standardised psychiatric histories. A brief outline of useful areas in the history is discussed below. It should be noted that this is not a comprehensive list.

#### History taking

##### Family history

- intellectual disability, psychiatric illness, neurological (epilepsy, dementia) or other relevant (such as physical) illness
- the quality of important relationships between the patient and other family members

##### Personal and developmental history

- information about the pregnancy and birth, and progressing up to the present day
- developmental years, including milestones
- family's management of a child with an intellectual disability
- education and job history

- relationship with others at school and at employment or day placements
- personality and behaviour prior to the development of the psychiatric illness
- psychosexual history
- notable life events, especially loss, abuse, and changes in placement or carers
- the highest level of functioning that the patient reached should be mentioned

### Medical history

- cause of the intellectual disability (including genetic cause), if known
- past and present physical illnesses (eg epilepsy, sleep apnea, thyroid disorder etc)
- past and present physical disability (eg limb weakness, spasticity etc)
- impairment in vision, hearing, speech or mobility should be mentioned
- recurrent physical illnesses (eg chest infection, toothache, constipation etc)

(Note: Establish how the individual communicates pain or any other bodily discomfort.)

### Psychiatric history

- a previous history of contact with services, and diagnoses (as previous diagnosis could be wrong, it is better, if possible to find out the exact clinical picture of the previous illness)
- risk assessment (risk to the person and others)

## Social history

- current and previous level of functioning in different areas of adaptive behaviours
- current and previous social circumstances (eg marital and employment status etc)
- current and previous living arrangements (eg group home, family home etc)
- current and previous social support (eg quality and quantity of carer support, daytime activities, social and leisure activities etc)

(Note: One good way of gathering information on the above areas may be through an individual's daily/weekly routine.)

## Drug history

- past and present medication (psychotropic and other) (including dosage)
- drug adverse effects
- recent change in medication
- substance and alcohol use (if relevant)
- known allergy

## Forensic history

- past and present history of problem with the law both in patients and in their friends and relations

## History of the presenting complaint

(This is described in detail in the next section.)

## Setting for the assessment

- Interviews can occur in a wide variety of settings (eg subjects' homes, day centres, outpatient clinics and so on).
- As much as possible, subject should be seen in a surrounding that is familiar to her/him.
- There is flexibility in the choice of the setting of assessment.
- A clinician may need to see a subject in different settings.
- It is helpful if the assessor is familiar to the subject.
- Where family or carers attend with the subject, they should know the subject well and have a good relationship with them.
- Issues about confidentiality need to be borne in mind (eg the subject may wish the carer not to be present in the interview).
- There should be flexibility in the length of interview (eg several shorter interviews).
- It is important to gather information from as many sources as possible (including written records).

In the general population, making a psychiatric diagnosis depends primarily on the account given by the subject. Many psychiatric diagnoses rely on patients describing quite complicated internal, subjective feelings or cognitions (such as thought broadcasting, obsessions or derealisation etc). Whilst many subjects with a mild intellectual disability will be able to describe such phenomena, those with a more severe intellectual disability may either not have had such experiences or be able to adequately describe them. It is important at the outset to assess the subject's communicating abilities, hearing, vision, memory, and ability to concentrate, as these will all affect their responses during

the assessment.

## Interview techniques

- The interviewer should, if possible, think about how he or she will **structure** the interview prior to starting it.
- Asking some general, easy questions at the start of the interview will help put the subject at ease.
- Assessment of the subject's **communication ability** is necessary at the outset of the assessment. (Some subjects may show discrepancy between their verbal and non-verbal performance; some may appear superficially able because of an apparent discrepancy between their comprehensive and expressive speech.)
- **Visual aids:** In certain cases it may be appropriate to use pictures, drawings or picture books.
- As much as possible, **speak to and involve the subjects themselves** during the assessment.
- **Avoid the use of leading questions**, where possible.
- **Use appropriate language** (eg simple phrases, short sentences; avoid double clauses, metaphors, idioms, and words, phrases or expressions that the subject may not understand, which includes medical jargon).
- The interviewer may need to **repeat questions**. Where there is doubt that the question has been understood, the interviewer could ask the subject to repeat the question back to them, or explain what has been asked.
- **Minimise suggestibility**

## Asking questions

- **Leading questions** are not useful. For example:  
*'You don't like living there, do you?'* should be avoided.  
A better question may be *'What is it like in your home?'*
- Where possible, **open questions** should be used. For example:  
*'How are you feeling today?'*  
*'What do you like doing at the (day) centre?'*  
*'Tell me about...'*
- **Closed questions** may sometimes be necessary to clarify a particular issue. Where closed questions need to be used, the interviewer should check for suggestibility. This includes cross-questioning techniques, such as asking for examples and contradictory questioning. For example, if the question *'Do you feel sad?'* were asked, then the interviewer could ask: *'What do you do when you feel sad?'* or *'Do you feel happy?'* (contradictory question)  
Ideally, when using a contradictory question, it should be asked later on in the interview.
- Another method of avoiding closed questions is to use questions with multiple choices.
- Stop every so often, and ask the patient to feedback to you things that you have said to them, to ensure comprehension.

## Patient observation

- In some cases, it may be necessary to **ask an observer**, such as the carer, about any changes shown in the subject's mental state. Ideally, the observer should have known the subject over a period of time so that he or she can describe the new symptoms or changes in the quality or intensity of existing symptoms.
- It may also be necessary to ask a carer or family member to **monitor for a period of time**, certain variables such as sleep, appetite and weight, level of activity, particular behaviours and so on, to aid in the diagnosis. Where this is done, clear instructions – so that the information is accurately and consistently documented – will aid in the diagnosis.
- **Direct observation of the patient is always necessary.**
- **Physical examination** (where relevant).
- Certain investigations such as thyroid function test, EEG, brain scan etc (if relevant).
- **Multi-professional assessment** (nurse, medic, psychiatrist, general practitioner, clinical psychologist, neuropsychologist,

## EVIDENCE

behaviour therapist, occupational therapist, speech therapist, social workers, music therapist, dietitian etc).

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# SECTION 3

## *Particular psychiatric disorders*

In this section the following disorders are considered:

- Schizophrenia
- Delusional disorder
- Schizoaffective disorder
- Affective disorders including:
  - Depressive disorder
  - Bi-polar affective disorder (mania and hypomania)
  - Dysthymia
  - Cyclothymia
- Anxiety disorders such as:
  - Generalised anxiety disorder
  - Phobic anxiety disorder
  - Panic disorder
  - Obsessive Compulsive Disorder (OCD)
- Other 'neurotic' and stress-related disorders, including:
  - Acute stress reactions
  - Post-traumatic stress disorder
  - Adjustment disorders

- Somatoform (hypochondriacal) disorders
- Eating disorders
- Sleep disorders
- Psychosexual dysfunction
- Disorders due to psychoactive substance use
- Dementia
- Delirium
- Personality disorders

## Schizophrenia

Schizophrenia has a point prevalence in the general population of about 0.4% (Meltzer *et al*, 1995), and in the population with intellectual disability of about 3% (range between 1.3% and 3.7%) (Deb, 2001a). It is characterised by particular, fundamental distortions in thinking, perception, mood and behaviour. The course is variable, but in many individuals, the disorder follows a chronic relapsing and remitting course.

First episodes of schizophrenia often, but not always, present with an acute picture, characterised by what are called ‘positive’ symptoms. These include hallucinations, delusions and thought disorder. In some individuals, the disorder will develop into a more chronic picture, characterised by apathy, lack of communication, social withdrawal, and blunted mood (negative

symptoms).

**In ICD-10, schizophrenia is subdivided into a number of categories:**

### **Paranoid schizophrenia**

- delusions and hallucinations are prominent

### **Hebephrenic schizophrenia**

- affective changes are prominent
- speech is often incoherent
- disorganised thoughts
- mannerisms, social withdrawal and other behavioural changes
- delusions and hallucinations are not prominent

### **Catatonic schizophrenia**

- stupor (marked decrease in reactivity to the environment and reduction of spontaneous movements and activities) is characteristic
- mutism
- purposeless motor activities
- posturing, negativism, rigidity, waxy flexibility, maintenance of limbs and body in externally imposed positions
- command automatism (automatic compliance with instructions)

### **Simple schizophrenia**

- slow progressive development
- odd conducts
- inability to meet society's demands
- gradual decline in total performance
- negative features such as blunting of affect and loss of initiation

**Note:** It may not be easy to distinguish between these subtypes in adults with intellectual disability (DC-LD, 2001).

Several authors (Reid, 1972; Royal College of Psychiatrists, 2001) have noted the difficulty in diagnosing schizophrenia in those with a moderate or more severe intellectual disability. The diagnosis is based upon the presence of a number of complex subjective symptoms (delusions, thought broadcasting etc), and thus a certain level of communicative ability is needed to describe such symptoms to an interviewer. This compares with the affective disorders, where theoretically, diagnosis is even possible in profound intellectual disability, due to observable behavioural elements of such disorders.

The aetiology of schizophrenia in intellectual disability is probably similar to that of the general population, but the higher prevalence suggests that this population have an increased risk. Part of this may be through increased rates of obstetric complications (O'Dwyer, 1997) and genetic risk factors (Doody *et al.*, 1998).

## Clinical features of schizophrenia

### Abnormal thought process

- delusions (characteristically paranoid)
- delusions of control ('passivity' phenomenon)
- thought 'interference'
- thought insertion
- thought withdrawal
- thought broadcasting
- delusional perception
- thought disorder (characteristically disorganised thought)

## Abnormal perception

- auditory hallucinations (characteristically third person)
  - thought echo
  - running commentary

## Abnormal mood

- incongruous mood (sometimes labile mood) (characteristic in the acute stage)
- flattened mood (affect) (characteristic in the chronic stage)

## Abnormal behaviour

- bizarre behaviour
- homicidal and suicidal behaviour
- catatonic behaviour (sometimes)
- negative symptoms (sometimes)
- impaired personality (in the chronic form)
- impaired social function (in the chronic form)

## Abnormal thought process

Delusions are false, fixed, unshakeable beliefs, and held with conviction, despite evidence to the contrary. They are not in keeping with a person's social, religious and cultural background. Classically, delusions in schizophrenia are delusions of control (also known as passivity), where a person believes that some external force controls their body or mind. However, other types of delusions commonly occur, including persecutory and grandiose. ICD-10 states that where delusions of other than

delusions of control are present, they must be ‘culturally inappropriate and completely impossible’ to make a diagnosis.

Related to this, a delusional perception is where an ordinary perception suddenly leads to an abnormal belief that is totally unconnected, for example *‘I saw a black cat run across the road, and that means that I am the King of Belgium’*.

In thought interference, patients believe that their thoughts are being ‘tampered’ with. This includes thought insertion (something or somebody directly putting thoughts into their head, and disrupting their train of thought), thought withdrawal (their own thoughts being taken out suddenly), and thought broadcasting (their own thoughts are apparent to others as soon as they think them, which may be via such mediums as the TV or radio).

Thought disorder is manifested through disordered speech, due to an underlying disorganisation in the thought processes. In severe cases, speech can be totally incomprehensible, known as a ‘word salad’. For example, *‘This is my cup door train went in going round can have to make’*. Thought disorder may contain examples of neologisms (non-existent words), for example ‘solix’.

Delusions (or ideas) of reference are often seen as soft psychotic signs. Here, for example, a person thinks wrongly that his/her name has been mentioned in the media or newspaper. If a person with intellectual disability reports that his/her name has been mentioned in the media or in the newspaper it is worth checking that indeed that is not the case. If the subject says that people in the street look at them, this may be true (if the person has a specific disability or manifests abnormal behaviour).

To elicit paranoid delusions the subject may be asked whether they feel someone is trying to harm them or plotting

against them. It is however possible for an adult with intellectual disability to think that someone is trying to harm them if they do not get on with care staff or a relative or one of their peers. This is not a psychotic symptom. The same can apply to passivity phenomenon (delusion of control by others).

In grandiose delusions the subject matter can be quite simple in the case of an adult with intellectual disability. For example, instead of thinking they can control the world they might think they can read a book (which is a false belief) ('psychosocial masking').

It is important to distinguish 'psychotic-like' symptoms from true 'psychotic' symptoms in adults with intellectual disability. Certain types of 'fantasy thinking' can be part of the symptomatology of autistic spectrum disorder. However, it is also important to recognise that adults who show autistic features may also show genuine psychotic symptoms.

**Here are some examples of questions that are included in the PAS-ADD (Moss *et al*, 1993) interview schedule for the purpose of eliciting psychotic features in an adult who have intellectual disability.**

*'Do you ever have thoughts in your mind that are not your own?'*

If so, *'Where do they come from?'* and *'How do they get there?'*

*'Do people know what you are thinking?'* and *'How does that happen?'*

*'Do thoughts leave your head?'*

If yes, *'Does it feel like something is sending them out?'*, *'Is something taking your thoughts out?'*, *'Do they go outside your head?'*, *'Are you losing your thoughts to the outside?'*

*'Do you feel that someone has taken you over?'*

If so, *'Who has taken you over?'*, *'What does that feel like?'*,  
*'Do you have to do what they want you to do?'*, *'Do you feel that you  
are a robot?'*

*'Do you hear yourself saying things that you do not know?'*

*'Do you hear yourself saying things that you did not mean to say?'*

*'Does the voice seem to come from your own mouth?'*

*'Is anything like hypnotism or telepathy going on?'*

*'Do X-rays, radio waves or machines affect you?'*

If so, *'In what way?'*

*'Do you feel upset or puzzled by these strange feelings?'*

The above are examples only, and in many cases it may be necessary to modify or simplify the language used in these questions.

## Abnormal perception

The third person auditory hallucinations (voices talking among themselves about the subject) are more characteristic of schizophrenia than second person auditory hallucinations (the voice speaks to the subject directly). Visual hallucinations can be presenting symptoms of schizophrenia in adults who have intellectual disability, however these hallucinations are more characteristic of organic psychoses (eg drug induced, seizure activity related etc).

**Here are some examples of questions that could be used to elicit abnormal perception in a person who has intellectual disability.** (See also PAS-SAD; Moss *et al*,1993)

*'Have you ever had any strange feelings on your skin?'*

*‘Have you ever smelt anything strange that no-one else could smell?’*

*‘Have you ever had any strange experiences?’*

*‘Have you ever heard any strange noises that no-one else could hear?’*

*‘Have you ever heard voices when there was nobody around?’*

If so, *‘Where does the voice come from?’*, *‘Does it come from outside or inside your head?’*

*‘Can you hear the voice as clearly as you can hear my voice?’*

*‘When do you hear the voice?’*, *‘Do you only hear it when you are going to sleep?’*

*‘Is it a male or a female voice?’*, *‘Could you tell whose voice it is?’*

*‘Do you hear one voice or more than one voice?’*

*‘Do the voices speak among themselves or do they speak to you?’*

*‘What does the voice say?’*

Determine whether the hallucination is mood congruent (characteristic of depression) or incongruent (could be characteristic of schizophrenia).

Some adults may look as if they are hallucinating when they speak to themselves, speak or look at an imaginary person, or speak to an object such as a tree or a table. These behaviours could be indirect evidence of hallucinations but these behaviours could also be part of the subject’s long-standing abnormal behavioural repertoire. They could be caused by the underlying brain abnormalities. In the absence of other diagnostic features, a diagnosis of schizophrenia should not be made in a subject with intellectual disability on the basis only of these indirect features of hallucinations.

## Abnormal mood

In the acute phase the subject may show incongruent mood in the form of inappropriate laughter etc. Anxiety is often a presenting feature in the acute phase. In the chronic phase the mood tends to become either flattened or depressed. Both anxiety and depressed mood could also be precipitated by the antipsychotic drugs that are used to treat schizophrenia.

## Abnormal behaviour

In the acute stage bizarre behaviour, excessive agitation, aggression and self-harm may be present. The subject may act under the influence of their delusions or hallucinations. In the chronic phase apathy, lack of motivation, and social withdrawal are common. These features, along with stereotyped movement disorders should be distinguished from the similar features that are associated with autistic spectrum disorders and other developmental disorders. Antipsychotic medications can cause both agitation (akathisia) and negative symptoms.

Other abnormal behaviours that are not so common include ‘catatonic’ symptoms such as:

- a) negativism (the subject does the opposite to what they are asked)
- b) ambitendency (actions are started then reversed; for example, the subject begins to take your hand, but then withdraws; or they are unable to complete movements, such as passing through a door, where they will continually advance then withdraw)
- c) forced grasping (ie taking your hand repeatedly, does so even when asked not to)

- d) echopraxia (imitates movements)
- e) waxy flexibility (a particularly unusual muscle tone with a 'waxy' feel to it)
- f) opposition (movement in any direction is countered by equal resistance in the opposite direction).

Odd postures and slowness are common in catatonia. The disorder needs to be differentiated from movement disorders, adverse effects from medication (especially neuroleptics), and the bizarre movements sometimes seen in autistic disorders.

Negative symptoms are much more observable, and therefore easier to elicit. It is important to remember that 'negative' symptoms (like catatonic symptoms) may be due to other causes such as depression or medication, and a standardised diagnosis of schizophrenia cannot rely on negative symptoms alone.

### Guidelines for the diagnosis of schizophrenia

- Sensory impairments, such as vision and hearing, are common in those with intellectual disability. They should be checked, as they may be a cause, or aggravating factor in schizophrenia (or other psychoses).
- Schizophrenia is difficult, if not impossible to diagnose in those with an IQ of below about 45.
- People with intellectual disability may have what appear to be 'psychotic' symptoms, but which in fact are related to their developmental level, or to other disorders. One example would be 'imaginary friends'.
- Interviewers need to be aware that people with limited verbal communication might use behaviour to communicate through means that appear 'odd' or unusual.

- An important thing to look for is a change in a person's behaviour or level of functioning. Symptoms that are part of the intellectual disability, or due to a disorder such as autism will tend to be longstanding, and constant in quality, rather than a change from a previous baseline.
- Be aware that people with intellectual disability may have difficulty recognising what are their own thoughts, and may attribute them to others, without this being thought interference.
- Also be aware that other people, carers, family, professionals, may well have a great deal of 'control' over what the individual is able to do, and thus the patient may feel that other people control them.
- In asking about auditory hallucinations, it may be helpful to ask on several different occasions to check for consistency in the presentation of the 'voices'.
- Be aware that 'negative' symptoms may be due to other causes, ie medication, or unstimulating environments.
- Make an assessment for adverse effects of drug treatment (particularly antipsychotics).
- Make an assessment for exclusion of other differential diagnoses.

## Differential diagnosis

- Psychotic depression
- Delusional disorders
- Schizo-affective disorder
- Organic conditions, particularly complex partial seizures

- Adverse effects of drugs and alcohol
- Substance misuse
- Withdrawal of drugs or alcohol
- Underlying brain damage
- Autism and other developmental disorders
- Learned behaviours and habits
- Unstimulating environment
- Acute stress reaction

## Delusional disorder

In this disorder, there is usually a single delusion, or set of related delusions, that tend to be persistent. Other symptoms such as auditory hallucinations are usually fleeting or absent. There is not the overall, pervasive change in personality seen in people with schizophrenia. The delusions are often resistant to treatment.

There are case reports of people with intellectual disability with delusional disorders. The literature is too limited to comment on whether delusional disorder presents differently, but the general guidelines above on diagnosing schizophrenia will be useful in assessing apparent psychotic symptoms.

## Schizo-affective disorder

This disorder is characterised by the presence of schizophrenic and affective symptoms. Both sets of symptoms must be prominent to

make the diagnosis. In ICD-10, diagnosis is based on a slightly modified set of schizophrenic symptoms, with a concurrent episode of mania, hypomania or depression of at least moderate severity.

The presence of mood-incongruent psychotic symptoms in affective disorders by themselves do not justify a diagnosis of schizo-affective disorder.

The literature on intellectual disability and schizo-affective disorder is sparse, but there are some case reports. Issues with making the diagnosis in those with intellectual disability are likely to be the same as that for schizophrenia. The readers are referred to the diagnostic criteria suggested in the DC-LD (Royal College of Psychiatrists, 2001).

Verhoeven and colleagues (1998) reported cycloid psychosis (ICD-10, F23.0: acute polymorphic psychotic disorder without symptoms of schizophrenia) in Prader-Willi syndrome.

## EVIDENCE

Deb, S. (2001a) Epidemiology of psychiatric illness in adults with intellectual disability. In: L. Hamilton-Kirkwood, Z. Ahmed, S. Deb *et al* (Eds) *Health Evidence Bulletins – Learning Disabilities (Intellectual Disability)* pp 14–17. <http://hebw.uwcm.ac.uk>. Cardiff: NHS.

(Type IV evidence: review of literature.)

DesNoyers Hurley, A. (1996) The misdiagnosis of hallucinations and delusions in persons with mental retardation: A neurodevelopmental perspective. *Seminars in Clinical Neuropsychiatry* **1** 122–133.

Doody, G. A., Johnstone, E. C., Sanderson, T. L., Cunningham Owens, D. G. & Muir, W. J. (1998) ‘Propfschizophrenie’ revisited. Schizophrenia in people with mild learning disability. *British Journal of Psychiatry* **173** 145–153.

(Type IV evidence: a case control study of adults with mild learning disability with and without schizophrenia, and a control group of non-intellectually disabled adults with schizophrenia. The study assessed various aspects of

demographical variables and symptomatology among the three groups.)

Gothelf, D., Frisch, A., Munitz, H. *et al* (1999) Clinical characteristics of schizophrenia associated with velo-cardio-facial syndrome. *Schizophrenia Research* **35** 105–112.

Hampson, R. M., Malloy, M. P., Mors, O. *et al* (1999) Mapping studies on a pericentric inversion (18) (p11.31 q21.1) in a family with both schizophrenia and learning disability. *Psychiatric Genetics* **9** 161–163.

Hasan, M. K. & Mooney, R. P. (1979) Three cases of manic-depressive illness in mentally retarded adults. *American Journal of Psychiatry* **136** (8) 1069–1071.

(Type V evidence: review of three cases. Although the title of the paper relates to affective disorders, the third patient in the paper is described as having a schizo-affective disorder.)

Heaton-Ward, A. (1977) Psychosis in mental handicap. *British Journal of Psychiatry* **130** 525–533. (Type V evidence: expert opinion.)

Hucker, S. J., Day, K. A., George, S. & Roth, M. (1979) Psychosis in mentally handicapped adults. In: F. E. James and R. P. Snaith (Eds) *Psychiatric Illness and Mental Handicap* pp27–35. London: Gaskell Press.

James, D. H. & Mukherjee, T. (1996) Schizophrenia and Learning Disability. *British Journal of Learning Disabilities* **24** 90–94.

(Type V evidence: review article.)

Meadows, G., Turner, T., Campbell, L., Lewis, S. W., Reveley, M. A. & Murray, R. M. (1991) Assessing schizophrenia in patients with mental retardation: a comparative study. *British Journal of Psychiatry* **158** 103–105.

(Type IV evidence: case control study comparing schizophrenia in people with mild intellectual disability against those with no intellectual disability. Classical symptoms of schizophrenia occurred in both groups, though certain symptoms were less common in the group with intellectual disability.)

Meltzer, H., Gill, B., Petticrew, M. & Hinds, K. (1995) *The prevalence of psychiatric morbidity among adults living in private households: OPCS survey of psychiatric morbidity in Great Britain, report 1*. London: HMSO.

(Type IV evidence: cross-sectional study of psychiatric illness using Clinical Interview Schedule – Revised, among 10108 adult general population in England and Wales.)

Mohl, P. C. & McMahon, T. (1980) Anorexia nervosa associated with mental retardation and schizo-affective disorder. *Psychosomatics* **21**

602–3, 606.

(Type V evidence: comment on a case report.)

Moss, S. C., Patel, P., Prosser, H., Goldberg, D., Simpson, N., Rowe, S. & Luccino, R. (1993) Psychiatric morbidity in older people with moderate and severe learning disability (mental retardation). Part 1: Development and reliability of the patient interview (The PAS-ADD). *British Journal of Psychiatry* **163** 471–480.

(Type IV evidence: development of a semi-structured psychiatric interview for use in people with intellectual disability.)

O'Dwyer, J. M. (1997) Schizophrenia in people with intellectual disability: the role of pregnancy and birth complications. *Journal of Intellectual Disability Research* **41** 238–251.

(Type IV evidence: case-control study comparing a group of people with schizophrenia and intellectual disability against a group with intellectual disability only.)

Reid, A. H. (1972) Psychoses in Adult Mental Defectives: II Schizophrenic and Paranoid Psychoses. *British Journal of Psychiatry* **120** 213–8.

(Type IV evidence: observational study.)

Reid, A. H. (1989) Schizophrenia in mental retardation: clinical features. *Research in Developmental Disabilities* **10** 241–9.

(Type IV evidence: review article.)

Royal College of Psychiatrists (2001) *DC-LD: Diagnostic Criteria for Psychiatric Disorders for use with Adults with Learning Disabilities/Mental Retardation*. London: Gaskell Press.

(Type V evidence: consensus document.)

Sharp, C. W., Muir, W. J., Blackwood, D. H. R., Walker, M., Gosden, C. & StClair, D. M. (1994) Schizophrenia and mental retardation associated in a pedigree with retinitis pigmentosa and sensorineural deafness. *American Journal of Medical Genetics* **54** 354–360.

Szymanski, L. S. (1980) Individual psychotherapy with retarded persons. In: L. S. Szymanski and P. E. Tanguay (Eds) *Emotional Disorders of Mentally Retarded Persons: Assessment, treatment, and consultation* pp 131–147. Baltimore: University Park Press.

Turner, T. H. (1989) Schizophrenia and mental handicap: an historical review, with implications for further research. *Psychological Medicine* **19** (2) 301–314. (Type IV evidence: review article.)

World Health Organisation (1973) *Report of the International Pilot Study of Schizophrenia, Vol 1*. Geneva: WHO.

(Type IV evidence.)

Verhoeven, W. M. A., Curfs, L. M. G. & Tuinier, S. (1998) Prader-

Willi syndrome and cycloid psychosis. *Journal of Intellectual Disability Research* 42 455–462.

## Affective disorders

### Depressive disorder

The point prevalence of depressive disorder in the general population is around 2% (Meltzer *et al*, 1995), with a lifetime prevalence of 6–17%. Symptoms of depression and anxiety can be detected in up to 15% of the general population at one point in time. The point prevalence of depressive disorder in adults with intellectual disability varies between 1.3–3.7% (Bouras & Drummond, 1992; Collacott *et al*, 1992; Patel *et al*, 1993; Cooper, 1997; Deb, 2001a; Deb *et al*, 2001a). Some studies showed a higher prevalence of depression among adults with Down's syndrome (Collacott *et al*, 1992) but others showed an opposite result (Haveman *et al*, 1994).

The course of depression can be recurrent or chronic but in some cases complete remission is possible. The aetiology of depression in intellectual disability is not well established, but it is likely that factors associated in the general population are relevant. The risks are likely to be higher by the additional difficulties that those with intellectual disability have, such as brain damage, higher rates of physical illness, and poorer social support.

### Clinical features of depressive disorder

- severe, persistent low mood that is pervasive across all situations and unresponsive to circumstances, even those that would normally lift a person's mood

- can be precipitated by a life event
- a marked loss of interest in all activities ('anhedonia')
- feelings of excessive tiredness
- loss of energy and initiative

### 'Biological features'

- sleep disturbance (usually insomnia in the form of early morning awakening, however, in atypical cases excessive sleep can be a feature) (early insomnia and interrupted sleep may also be present)
- changes in appetite (usually decrease but increase in some atypical cases)
- significant weight loss not due to dieting or any other physical illness (increase in weight in atypical cases)
- psychomotor retardation or agitation
- decreased libido
- diurnal variation in mood (mood being worse in the morning)

### 'Cognitive features'

- loss of self-esteem or confidence
- lack of purpose in life (life is not worth living, there is no future)
- feelings of guilt or self-reproach
- suicidal thoughts or behaviour
- difficulty with concentration
- marked slow thought processes
- ruminative thoughts (pre-occupation with depressive thoughts)

- speech is slow and lacks content

### Appearance

- depressed look (may be crying)
- restless and agitated
- poor eye contact with the interviewer
- evidence of self-neglect (or sometimes self-harm)
- stooped posture
- slow and sometimes awkward movements

### Psychotic features (in severe cases)

- auditory hallucinations (often derogatory and in second person) or delusions (nihilistic)

ICD-10 requires symptoms to be present for at least two weeks. Severity is graded according to how many symptoms are present.

In mild to moderate intellectual disability, the individual may be able to describe the above symptoms, and a diagnosis can be made according to strict criteria, such as those in ICD-10 (see criteria suggested in the DC-LD, Royal College of Psychiatrists, 2001). The presentation can also be atypical, for example with prominent somatic complaints.

It is important to exclude causes of depression, such as physical illness, medication (especially neuroleptic medication), and to look for environmental factors that could perpetuate the illness (for example, ongoing conflicts with another resident at home). Dementia may have a similar initial presentation.

## Guidelines for the diagnosis of depression

- Assessment should include:
  - physical examination and appropriate investigations
  - medication history (neuroleptics, antihypertensives, steroids etc) is important
  - adverse effects of drugs (including anti-depressants).
- Assessment to exclude other differential diagnoses.
- Risk assessment (for both self-harm, self-neglect, and harm to others) is important.
- In those with mild to moderate intellectual disability, standardised diagnostic criteria such as ICD-10 may be appropriate.
- Depression may present atypically (eg hypersomnia and increased appetite).
- In those with a more severe disability, diagnosis will depend more on behavioural symptoms. Depression should be suspected where there is a change or onset in behaviour disturbance, associated with some of the 'biological' symptoms, but not necessarily enough to meet standard diagnostic criteria. However, other causes of behavioural disturbances should always be excluded first.

## Deterioration in skills

- It may be useful, in situations where there is doubt about the diagnosis in those with a severe to profound disability, for family and carers to carefully record on a daily basis, over a

period (for example, a week/month) behaviours that would be suggestive of depression, such as sleep disturbance, withdrawal, loss of communication (however limited), level of behaviour disturbance, and how the person appears in mood (for example, 'looks sad').

- Where diagnosis continues to be in doubt, a trial of anti-depressant medication could be considered, although a positive response does not prove the diagnosis.

## Differential diagnosis

- psychosis
- organic disorders (eg hypothyroidism, Addison's disease, epilepsy)
- drug treatment (eg antipsychotics, steroids, antihypertensives, neuroleptics)
- alcohol and substance misuse
- alcohol and substance withdrawal
- life events (eg bereavement, change in or loss of a carer, change in accommodation) and acute stress
- environmental factors
- use of regression as a psychological defence mechanism for coping with stress

## EVIDENCE

### ■ dementia

Benson, B. A. & Ivins, J. (1992) Anger, depression and self-concept in adults with mental retardation. *Journal of Intellectual Disability Research* **36** 169–175.

Benson, B. A., Reiss, S., Smith, D. C. & Laman, D. (1985) Psychosocial correlates of depression in mentally retarded adults: II. Poor social skills. *American Journal of Mental Deficiency* **89** 657–659.

Berman, A. L. (1992) Treating suicidal behavior in the mentally retarded: The case of Kim. *Suicide and Life-threatening Behavior* **22** 504–506.

Bonell-Pascual, E., Huline-Dickens, S., Hollins, S. *et al* (1999) Bereavement and grief in adults with learning disabilities: A follow-up study. *British Journal of Psychiatry* **175** 348–350.

Bouras, N. & Drummond, C. (1992) Behavioural and psychiatric disorders of people with mental handicaps living in the community. *Journal of Intellectual Disability Research* **36** (4) 349–357.

(Type IV evidence: study of 318 individuals in the community, and the level of psychiatric and behavioural disorder.)

Carlson, G. (1979) Affective psychoses in mental retardates. *Psychiatric Clinics of North America* **2** 499–510.

Collacott, R. A., Cooper, S.-A. & Lewis, K. R. (1992) Differential rates of psychiatric disorders in adults with Down's syndrome compared with other mentally handicapped adults. *British Journal of Psychiatry* **161** 671–674.

(Type IV evidence: case-control study of 371 adults with Down's syndrome compared with 371 matched adults with intellectual disability who did not have Down's syndrome.)

Cooper, S.-A. (1997) Psychiatry of elderly compared to younger adults with intellectual disability. *Journal of Applied Research in Intellectual Disability* **10** (4) 303–311.

(Type IV evidence: cross-sectional study of 134 people over 64 years of age and 73 adults between 20 and 64 years of age with intellectual disability. Assessments were carried out using Present Psychiatric State for Adults with Learning Disabilities [PPS-LD].)

Cooper, S.-A. & Collacott, R. A. (1996) Depressive episodes in adults with learning disabilities. *Irish Journal of Psychological Medicine* **13** 105–113.

(Type V evidence: review of the literature.)

Davis, J. P., Judd, F. K. & Herman, H. (1997) Depression in adults with intellectual disability. Part 1: A review. *Australian and New Zealand Journal of Psychiatry* **32** 232–242.

Day, K. (1990) Depression in moderately and mildly mentally handicapped people. In: A. Dosen & F. Menolascino (Eds) *Depression in Mentally Retarded Children and Adults* pp129–154. Leiden: Logon Publications.

Deb, S. (2001a) Epidemiology of psychiatric illness in adults with intellectual disability. In: L. Hamilton-Kirkwood, Z. Ahmed, S. Deb *et al* (Eds) *Health Evidence Bulletins – Learning Disabilities (Intellectual Disability)* pp14–17. [wttp://hebw.uwcm.ac.uk](http://hebw.uwcm.ac.uk). Cardiff: NHS.

(Type IV evidence: review of literature.)

Deb, S., Thomas, M. & Bright, C. (2001a) Mental disorder in adults who have intellectual disability. 1: Prevalence of functional psychiatric illness among a 16–64 years old community-based population. *Journal of Intellectual Disability Research* (in press).

(Type IV evidence: cross-sectional observational study of the rate of functional psychiatric illness among a population-based sample of 101 adults with intellectual disability.)

Dosen, A. & Gielen, I. (1993) Depression in persons with mental retardation; assessment and diagnosis. In: R. Fletcher and A. Dosen (Eds) *Mental Health Aspects of Mental Retardation* pp70–97. New York: Lexington Books.

Evans, K. M., Cotton, M. M., Einfeld, S. L. & Florio, T. (1999) Assessment of depression in adults with severe or profound intellectual disability. *Journal of Intellectual and Developmental Disability* **24** (2) 147–160.

Haveman, M. J., Maaskant, M. A., van Schroyenstien, H. M., Urlings, H. F. J. & Kessels, A. G. H. (1994) Mental health problems in elderly people with and without Down's syndrome. *Journal of Intellectual Disability Research* **38** (3) 341–355.

(Type IV evidence: cross-sectional study comparing 209 severely and 59 mildly intellectually disabled adults with Down's syndrome with 477 severely and 1255 mildly intellectually disabled adults without Down's syndrome.)

Kazdin, A. E., Matson, J. L. & Senatore, V. (1983) Assessment of depression in mentally retarded adults. *American Journal of Psychiatry* **140** 1040–1043.

Kearns, A. (1987) Cotard's syndrome in a mentally handicapped man.

*British Journal of Psychiatry* **150** 112–114.

Levitus, A. S. & Gilson, S. F. (1990) Toward the developmental understanding of the impact of mental retardation on assessment of psychopathology. In: E. Dibble and D. G. Gray (Eds) *Assessment of Behavior Problems in Persons with Mental Retardation Living in the Community* pp71–106. Rockville, MD: National Institute of Mental Health.

Lindsay, W. R., Michie A. M., Baty, F. J., Smith, A. H. W. & Miller, S. (1994) The consistency of reports about feelings and emotions from people with intellectual disability. *Journal of Intellectual Disability Research* **38** 61–66.

Marston, G. M., Perry, D. W. & Roy, A. (1997) Manifestations of depression in people with intellectual disability. *Journal of Intellectual Disability Research* **41** 476–480.

(Type IV evidence – case – control study comparing individuals with depression and intellectual disability against those with intellectual disability alone. Diagnosis in those with severe intellectual disability relied more on behavioural 'depressive equivalents', such as aggression, screaming and self-injury.)

Meins, W. (1993) Prevalence and risk factors for depressive disorders in adults with intellectual disability. *Australia and New Zealand Journal of Developmental Disabilities* **18** 147–156 (cited in Prasher, 1999).

Meins, W. (1995) Symptoms of major depression in mentally retarded adults. *Journal of Intellectual Disability Research* **39** 41–45.

Meins, W. (1995) Are depressive mood disturbances in adults with Down's syndrome early signs of dementia? *Journal of Nervous and Mental Disorders* **183** 663–664.

Meins, W. (1996) A new depression scale designed for use with adults with mental retardation. *Journal of Intellectual Disability Research* **40** 220–226.

Meltzer, H., Gill, B., Pettecrew, M. & Hinds, K. (1995) *The prevalence of psychiatric morbidity among adults living in private households: OPCS survey of psychiatric morbidity in Great Britain, report 1*. London: HMSO.

(Type IV evidence: cross-sectional study of psychiatric illness using Clinical Interview Schedule-Revised, among 10108 adult general population in England and Wales.)

Moss, S., Emerson, E., Kiernan, C., Turner, S., Hatton, C. & Alborz, A. (2000) Psychiatric symptoms in adults with learning disability and challenging behaviour. *British Journal of Psychiatry* **177** 452–456.

Nezu, C. M., Nezu, A. M., Rotherburg, J. L., DelliCarpini, L. & Groag,

I. (1995) Depression in adults with mild mental retardation: are cognitive variables involved? *Cognitive Therapy and Research* **19** 227–239.

Pary, R. J., Loschen, E. L. & Tomkowiak, S. B. (1996) Mood disorders and Down's syndrome. *Seminars in Clinical Neuropsychiatry* **1** 148–153.

Patel, P., Goldberg, D. & Moss, S. (1993) Psychiatric morbidity in older people with moderate and severe learning disability II: The Prevalence Study. *British Journal of Psychiatry* **163** 481–491.

(Type IV evidence: cross-sectional study of 105 people with intellectual disabilities aged 50 years and over, using PAS-ADD diagnostic interview.)

Pawlarczyk, D. & Beckwith, B. E. (1987) Depressive symptoms displayed by persons with mental retardation: a review. *Mental Retardation* **25** 323–330.

Prasher, V. P. (1995a) Age-specific prevalence, thyroid dysfunction and depressive symptomatology in adults with Down's syndrome and dementia. *International Journal of Geriatric Psychiatry* **10** 25–31.

Prasher, V. (1999) Presentation and management of depression in people with learning disability. *Advances in Psychiatric Treatment* **5** 447–454.

(Type V evidence: review article on the presentation and management of depression in intellectual disability.)

Prout, H. T. & Schaefer, B. M. (1985) Self-report of depression by community-based mentally retarded adults. *American Journal of Mental Deficiency* **90** 220–222.

Reiss, S. & Benson, B. A. (1985) Psychosocial correlates of depression in mentally retarded adults: I. Minimal social support and stigmatisation. *American Journal of Mental Deficiency* **89** 331–337.

Reiss, S. & Rojahn, J. (1993) Joint occurrence of depression and aggression in children and adults with mental retardation. *Journal of Intellectual Disability Research* **37** (3) 287–294.

(Type IV: cross-sectional study of 528 adults, adolescents and children using Reiss scale.)

Reiss, S. & Sysko, J. (1993) Diagnostic overshadowing and professional experience with mentally retarded persons. *American Journal on Mental Deficiency* **47** 415–421.

Royal College of Psychiatrists (2001) DC-LD: Diagnostic Criteria for Psychiatric Disorders for use with Adults with Learning Disabilities/Mental Retardation. London: Gaskell Press.

(Type V evidence: consensus document.)

Sovner, R. & Hurley, A. D. (1983) Do the mentally retarded suffer from affective illness? *Archives of General Psychiatry* **40** 61–67.

Sovner, R. & Lowry, M. (1990) A behavioral methodology for diagnosing affective disorders in individuals with mental retardation. *Habilitative Mental Health Care Newsletter* **9** (7) 55–61.

Szymanski, L. S. & Biederman, J. (1984) Depression and anorexia nervosa of persons with Down syndrome. *American Journal of Mental Deficiency* **89** 246–251.

Tsiouris, J. A. (2001) the diagnosis of depression in persons with severe/profound intellectual disabilities. *Journal of Intellectual Disability Research* **45** 115–120.

Warren, A. C., Holroyd, S. & Folstein, M. F. (1989) Major depression in Down's syndrome. *British Journal of Psychiatry* **155** 202–205.

Wolkowitz, O. M. (1990) Use of the dexamethasone suppression test with mentally retarded persons: review and recommendations. *American Journal on Mental Retardation* **94** 509–514.

## Hypomania and mania

In hypomania, there is a consistent elevation of mood, lasting at least several days. It is usually associated with feelings of wellbeing, increased energy and appetite, and a decreased need for sleep. Subjects may be over-talkative, over-familiar and inappropriately sociable. They may talk about or describe great plans or projects they have decided to start, but rarely carry these out. The symptoms, however, are not marked enough to cause severe disruption to a person's functioning. In the early stage the subject may become usefully productive (in writing or drawing etc). They may also spend an excessive amount of money without any regard to their financial position.

Mania occurs where symptoms are severe enough to cause disruption, or there is the presence of psychotic symptoms, such as grandiose delusions. Patients with mania display marked disturbance of speech ('pressure of speech'), and thoughts (flight of ideas or the patient describing their thoughts as 'racing'). They may be distractible, constantly changing plans, and show reckless or foolhardy behaviour, including sexually indiscreet behaviour and social disinhibition. People with mania may become irritable or aggressive if challenged about their grandiosity or behaviour.

Single episodes of mania or hypomania are unusual. Recurrent episodes, often with interspersed episodes of depression form the diagnosis of bi-polar affective disorder (see below).

In people with an intellectual disability, it is possible that the mood may be predominantly irritable rather than elated. It may be associated with aggression. An early study (Reid, 1972) noted that pressure of speech was seen more commonly than flight of ideas. Delusions and hallucinations were present,

but not convincingly diagnosed in those with a moderate or greater disability. They tended to be simpler in nature. Therefore, a person with intellectual disability who cannot drive may have a grandiose delusion that he can drive a car, whereas a non-intellectually disabled person may believe he is the king of the country.

Other symptoms, reported in case studies (Hassan & Mooney, 1979), include aggression, destructive behaviour, restlessness, intensified or rambling speech, echolalia, both increased and decreased appetite, crying and overactivity.

Mixed affective states appear to be a commoner presentation of bipolar disorder in this group (Berney & Jones, 1988). Patients may have lability of mood, pressure of speech but no motor overactivity, and describe both grandiose and persecutory delusions at the same time. A family history of bipolar disorder may be present, and on occasions precipitants to the illness can be found.

Several authors suggested that mania can be diagnosed in adults with a severe or profound intellectual disability (Reid, 1972), as many of the symptoms can be elicited from accounts from family and carers, and direct observation. These include marked irritability or elation, decreased sleep, increased (or decreased) appetite, overactivity, overfamiliarity with others (especially compared to a baseline), easy distractibility and sexual disinhibition. Changes in behaviour such as aggression

and destructiveness are not specific enough to bipolar disorder.

### Differential diagnosis

- schizophrenia
- depressive disorder
- acute anxiety state
- medical conditions (eg hyperthyroidism, Cushing's disease, epilepsy)
- adverse effects of drugs (eg antipsychotics, SSRIs, antihypertensives)
- alcohol and substance misuse
- alcohol and substance withdrawal
- environmental factors

### Bi-polar affective disorder

This disorder is characterised by recurrent episodes of mania or hypomania that are sometimes mixed with episodes of depression. The disorder has a lifetime rate of around 1% in the general population. Rapid cycling disorder refers to a subtype of bipolar disorder, where there are four or more episodes in a 12-month period. This type of bipolar disorder usually has a poor response to lithium treatment. Rapid cycling disorder has been described in those with intellectual disability (Vanstraelen & Tyrer, 1999) and may be more common in this population.

In a study of bipolar disorder (Reid, 1972), the author noted that several patients with depressive episodes had attempted suicide. Risk assessment is an important part of the investigation

of possible bipolar disorder, especially in depressive episodes. Some patients with intellectual disability appear to show cyclical changes in their behaviour, which can be associated with altered mood. Deb & Hunter (1991b) described cyclical behaviour and mood changes among 4% of adults with intellectual disability and epilepsy and a similar proportion (4%) of adults with intellectual disability who did not have epilepsy. There may be other factors to account for this, which could include physical factors (changes around periods in women, epilepsy that shows ‘clustering’, etc), and various environmental factors.

## EVIDENCE

Berney, T. P. & Jones, P. M. (1988) Manic-depressive disorder in mental handicap. *Australia and New Zealand Journal of Developmental Disabilities* **14** (3–4) 219–25.

(Type IV evidence: study of eight cases found higher rates of rapid cycling, mixed affective states and lithium resistance.)

Cooper, S.-A. & Collacott, R. A. (1993) Mania and Down’s syndrome. *British Journal of Psychiatry* **162** 739–743.

Deb, S. & Hunter, D. (1991b) Psychopathology of people with mental handicap and epilepsy. II: Psychiatric illness. *British Journal of Psychiatry* **159** 826–830.

Gecz, J., Barnett, S., Liu, J. *et al* (1999) Characterisation of the human glutamate receptor subunit 3 gene (GRIA3), a candidate foe bipolar disorder and non-specific X-linked mental retardation. *Genomics* **62** 356–368.

Glue, P. (1989) Rapid cycling bipolar disorders in the mentally retarded. *Biological psychiatry* **26** 250–256.

Hassan, M. K. & Mooney, R. P. (1979) Three cases of manic-depressive illness in mentally retarded adults. *American Journal of Psychiatry* **136** (8) 1069–1071.

(Type V: opinion based on three case reports.)

Lowry, M. A. & Sovner, R. (1992) Severe behavior problems associated with rapid cycling bipolar disorder in two adults with profound mental retardation. *Journal of Intellectual Disability Research* **36** 269–281.

Parry, A., Hurley, A. & Pary, R. J. (1999) Diagnosis of bipolar disorder in persons with developmental disabilities. *Mental Health Aspects of Developmental Disabilities* **2** (2) 37–49.

Raghavan, R., Day, K. A. & Perry, R. H. (1996) Rapid cycling bipolar affective disorder and familial learning disability associated with temporal lobe (occipitotemporal gyrus) cortical dysplasia. *Journal of Intellectual Disability Research* **39** 509–519.

Reid, A. H. (1972) Psychoses in Adult Mental Defectives: I. Manic Depressive Psychosis. *British Journal of Psychiatry* **120** 205–12.

Reid, A. H. & Naylor, G. J. (1976) Short-cycle manic depressive psychosis in mental defectives: a clinical and psychological study. *Journal of Mental Deficiency Research* **20** 67–76.

Ruedrich, S. (1993) Bipolar mood disorders in person with mental retardation: assessment and diagnosis. In: R. J. Fletcher and A. Dosen (Eds) *Mental Health Aspects of Mental Retardation* pp111–129. New York: MacMillan.

Sovner, R. (1986) Limiting factors in the use of DSM-III criteria in mentally retarded persons. *Psychopharmacology Bulletin* **22** 1055–1059.

Sovner, R. & Hurley, A. D. (1983) Do the mentally retarded suffer from affective illness? *Archives of General Psychiatry* **40** 61–67.

Szymanski, L. S. & Biederman, J. (1984) Depression and anorexia nervosa of persons with Down syndrome. *American Journal of Mental Deficiency* **89** 246–251.

Vanstraelen, M. & Tyrer, S. P. (1999) Rapid cycling bi-polar affective disorder in people with intellectual disability: a systematic review. *Journal of Intellectual Disability Research* **43** 349 – 359.

(Type V evidence: opinion based on review on several cases.)

## Persistent mood disorders: Dysthymia and Cyclothymia

### *Dysthymia*

Dysthymia is a chronic state of low mood, lasting several years, but which is not sufficiently severe to make a diagnosis of depressive disorder. People with dysthymia do have a higher risk, however, of developing a depressive disorder during this state.

There are single case and small group studies of dysthymia in intellectual disability in the literature (eg Masi *et al*, 1999). Dysthymia may show fewer disturbances of biological features such as sleep and appetite compared with a depressive disorder.

### *Cyclothymia*

Cyclothymia describes a persistent instability of mood, with periods of low mood, and elation. These episodes are not severe enough to make a diagnosis of bi-polar affective disorder. The literature on cyclothymia in intellectual disability is sparse.

These disorders are likely to be difficult to diagnose in those with intellectual disability.

## EVIDENCE

Göstason, R. (1985) Psychiatric illness among the mentally retarded. *Acta Psychiatrica Scandinavica* (Suppl. 318) 71.

(Type V evidence: comment on two case reports of dysthymia.)

Jancar, J. & Gunaratne, IJ. (1994) Dysthymia and Mental Handicap. *British Journal of Psychiatry* **164** 691–693.

Masi, G., Mucci, L., Favillia, L. & Poli, P. (1999) Dysthymic disorder in adolescents with intellectual disability. *Journal of Intellectual Disability Research* **43** 80–87.

(Type IV evidence: observational study of dysthymic disorder in a group of

adolescents with intellectual disability.)

## Anxiety disorders

### Generalised anxiety disorder (GAD)

**Generalised anxiety disorder** is equally common, and according to some studies more common, among adults with intellectual disability as in the general adult population (Raghavan, 1997).

In this condition anxiety is generalised and persistent but not restricted to, or even strongly predominating in, any particular environmental circumstances (ie it is ‘free-floating’). The dominant symptoms are variable but typically include complaints of persistent nervousness, trembling, muscular tensions, sweating, light-headedness, palpitations, dizziness, and epigastric discomfort. Fears that the patient or a relative may shortly become ill or have an accident are often expressed (ICD-10). Adults with intellectual disability may be able to describe a persistent generalised anxiety or tension. Signs such as persistent irritability, difficulty getting to sleep or somatic complaints may be noticed. The presentation again may be through behaviour disorder.

A comparison study (Masi *et al*, 2000) of generalised anxiety disorder in those with intellectual disability against those with normal intelligence suggested that GAD can be identified in those with a mild disability. Symptoms were similar, although there was an increase in brooding, somatic complaints and sleep disorder. There were high rates of co-morbid depression. Other possible diagnoses, if there is marked irritability associated with overactivity, are **mania** or **hypomania**.

Physical illness and medication may be an underlying cause,

and should be ruled out if possible. Co-morbidity with other psychiatric illnesses such as depression is common and should be ruled out.

## EVIDENCE

Lindsay, W. R., Baty, F. J., Michie A. M. & Richardson, I. (1989) A comparison of anxiety treatments with adults who have moderate and severe mental retardation. *Research in Developmental Disabilities* **10** 129–140.

Masi, G., Favilla, L. & Mucci, M. (2000) Generalised anxiety disorder in adolescents and young adults with mild mental retardation. *Psychiatry* **63** 54–64.

McNally, R. J. (1991) Anxiety and phobias. In: J. L. Matson & J. A. Mulick (Eds) *Handbook of Mental Retardation* pp413–423. Elmsford, New York: Pergamon Press.

McNally, R. J. & Ascher, L. M. (1987) Anxiety disorders in mentally retarded people. In: L. Michlson & L. M. Ascher (Eds) *Anxiety and Stress Disorders: Cognitive Behavioural Assessment and Treatment*. New York: Guilford Press.

Ollendick, T. H. & Ollendick, D. G. (1982) Anxiety disorders. In: J. L. Matson & R. P. Barrett (Eds) *Psychopathology in the Mentally Retarded*. New York: Grune & Stratton.

Poindexter, A. R. (1996) *Assessment and Treatment of Anxiety Disorders in Persons with Mental Retardation*. Kingston, New York: National Association for the Dually Diagnosed.

Raghavan, R. (1998) Anxiety disorders in people with learning disabilities: A review of the literature. *Journal of Learning Disabilities for Nursing, Health and Social Care* **2** (1) 3–9.

## Phobic anxiety disorders

These are a group of disorders in which excessive anxiety occurs, largely in particular situations or circumstances that would not normally be seen as anxiety provoking. People with these disorders will tend to avoid the situation that causes the anxiety if at all possible. They may also show signs of anxiety if exposed to that situation, such as sweating, tremor and palpitations (**autonomic symptoms**), but in severe cases even thinking about the situation may bring on anxiety. People with phobias tend to recognise that their fears are unreasonable.

One study showed a higher rate of specific phobic disorders among adults with intellectual disability (Deb *et al*, 2001a). People with intellectual disability may show external signs of anxiety, but may not be able to describe their inner agitation or fear. They may also be more prone to experience anxiety in situations that would not normally be seen as anxiety provoking.

When anxiety cannot be expressed, it may present as a change in behaviour, especially in those with more severe disability. Functional analysis may be of use in identifying potential triggers. A good history and analysis may identify a triggering factor in the onset of the phobia. A study looking into the assessment of anxiety (Matson *et al*, 1997) found that only the behavioural symptoms associated with anxiety could reliably be assessed. However, a disturbance of behaviour in a particular situation may not be due to an anxiety disorder, but to other environmental factors. These could include dislike of another person in the environment, or not wanting to do a particular act that the subject is asked to carry out.

Anxiety may occur in **autistic disorders**, particularly in individuals who are routine-bound and ritualistic. It may occur

when routines are changed (for example, walking a different way to the shops). This would be considered part of the autistic disorder, and not a new anxiety disorder.

ICD-10 divides phobic disorders into three categories – agoraphobia, social phobia, and specific phobias (such as the fear of spiders). For the disorder to be diagnosed, the phobia must have an impact on the person's life that prevents or restricts what they are able to do.

### *Agoraphobia*

Agoraphobia includes a number of defined phobias, which include fears of being in crowds or public places, travelling alone or away from home (ICD-10). Panic disorder may also occur (see below), as can **depressive and obsessional symptoms**. People with agoraphobia may be able to avoid their phobic situations and thus experience little anxiety, albeit with a restricted lifestyle.

### *Social phobias*

Social phobias occur when people have a fear of attention from other people in social situations. This may include the worry that they will behave in an embarrassing way in settings such as speaking in public or being part of a group. Theoretically, they can occur in people with an intellectual disability, but the evidence is limited. There are some case reports in the literature.

### *Specific phobias*

Specific phobias tend to be restricted to certain situations,

such as a proximity to certain animals, insects (eg spiders), the sight of blood, darkness, etc. To be classified as a phobia, the situation must cause marked anxiety and/or avoidance. Some phobias are age dependent (for example, young children are often scared of the dark). People with an intellectual disability may show similar fears dependent on their developmental level (Sternlicht, 1979).

There are case reports in the literature on the presentation and treatment of specific phobias in those with intellectual disability.

### Guidelines of the diagnosis of specific phobias

- Phobic disorders are difficult to diagnose, except possibly in those with mild intellectual disability. Some signs of anxiety may be identifiable, though.
- Specific phobias to ordinary things such as dogs, water, storms, stairs, crowds can be found in this population. However, they should be carefully distinguished from reasonable fears of these things.
- Behavioural analysis may identify triggers to behavioural disturbance, but this does not prove that underlying anxiety causes the behaviour.
- There is a high level of co-morbidity in the general population suffering with phobic disorders, particularly depression, and this may be the case for those with intellectual disability

**EVIDENCE**

as well. The assessment should therefore look for evidence of this.

Bodfish, J. W. & Madison, J. T. (1993) Diagnosis and fluoxetine treatment of compulsive behaviour disorder of adults with mental retardation. *American Journal of Mental Retardation* **98** (3) 360–367.

(Type III evidence: an open trial of fluoxetine in ten individuals with 'compulsive behaviour disorder', against six individuals without.)

Carminati, C. & Yogwa, S. D (1998) Obsessive compulsive disorders in a young man with a mild mental retardation: Importance of a longitudinal study of the symptoms and of an integrated treatment. *European Journal on Mental Disability* **15** (19) 25–34.

(Type V evidence: case report.)

Chambliss, D. L. (1995) The relationship of severity of agoraphobia to associated psychopathology. *Behaviour, Research and Therapy* **23** 305–310.

Malloy, E., Zealberg, J. J. & Paolone, T. (1998) A patient with mental retardation and possible panic disorder. *Psychiatric Services* **49** 105–106.

(Type V evidence: case report.)

Masi, G., Favilla, L. & Mucci, M. (2000) Generalised anxiety disorder in adolescents and young adults with mild mental retardation. *Psychiatry* **63** (1) 54–64.

(Type IV evidence: observational study identifying GAD in three groups, adolescents and young adults with intellectual disability, children without intellectual disability, and adolescents without intellectual disability.)

Matson, J. L., Smirolfo, B. B., Hamilton, M. & Baglio, C. S. (1997) Do anxiety disorders exist in persons with severe and profound mental retardation? *Research in Developmental Disabilities* **18** 39–44.

McNally, R. J. & Calamari, J. E. (1989) Obsessive-compulsive disorder in a mentally retarded woman. *British Journal of Psychiatry* **155** 116–117.

(Type V evidence: case report.)

Sternlicht, M. (1979) Fears of institutionalised mentally retarded

adults. *Journal of Psychology* **101** 67–71.

(Type IV evidence: study comparing the fears in people with intellectual disability against a control group.)

## Panic disorder

In panic disorder, there are recurrent attacks of severe anxiety (panic), which are not limited to particular situations, and therefore occur ‘out of the blue’. The symptoms of anxiety present during an attack include palpitations, chest pain, choking feelings, dizziness, abdominal discomfort, and feelings of unreality. There is extreme anxiety, with the subject feeling as though they are about to die, go mad, or lose control.

Panic disorder may occur in an adult with intellectual disability. Some of the signs associated with a panic attack may be observable eg sudden onset, external signs of anxiety such as shaking and sweating. Some may present with blackouts. Again though, in those with limited communication, the presentation may be with disturbed behaviour.

**EVIDENCE**

Malloy, E., Zealberg, J. J. & Paolone, T. (1998) A patient with mental retardation and possible panic disorder. *Psychiatric Services* **49** 105–106.

## Obsessive Compulsive Disorder (OCD)

This disorder is characterised by recurrent obsessions (thoughts, images or ideas), with or without compulsions (repetitive acts which have no useful function). According to ICD-10, the obsessions or compulsions must have all the following characteristics for a diagnosis to be made:

- The obsessions or compulsions must be recognised by the patient as originating in the subject's mind.
  - They must be repetitive and unpleasant, and seen as excessive and unreasonable.
  - The subject will attempt to resist 'thinking' the obsession or carrying out the compulsion. The degree of resistance may be minimal in long-standing disorders.
  - Experiencing the obsession or carrying out the compulsion is not pleasurable, although it may bring temporary relief.
  - The obsessions or compulsions must cause distress or interfere with the subject's social functioning.
- The symptoms must not be due to another mental disorder, such as a psychotic disorder or an affective disorder.

In the general population, the point prevalence is around 1% (Meltzer *et al*, 1995). Studies on populations with intellectual disability have described rates between 1% and 3.5% (Deb,

2001a; Vitiello *et al*, 1989).

Obsessions need to be distinguished from obsessional traits in which the person tends to be over-organised and meticulous in their habits, but with the traits usually having some purpose, and without them causing distress to that person or others. They also need to be distinguished from repetitive questions (repetitive speech and echolalia) that some subjects ask and repetitive ruminations of a particular thought. These may occur in especially anxious patients with limited verbal skills or in those with an autistic disorder.

It may be difficult to elucidate the presence of obsessions in a person with an intellectual disability. They may be unable to recognise it as coming from their own mind, and resistance may not occur. Anxiety is not always a recognised feature.

Compulsive behaviours are common in adults with intellectual disability, and their cause is not always apparent. They occur in a range of psychiatric disorders, including schizophrenia, affective disorders and autism. They also need to be distinguished from stereotyped behaviour and movement disorders that are caused by the underlying brain damage in many adults with intellectual disability.

Whilst there are case reports of people with intellectual disability who present with ‘classical’ OCD (McNally & Calamari, 1989), other authors have suggested that the diagnosis should be considered in patients with intellectual disability, with some of the signs, but with the emphasis being on the behavioural, externally observable components of the disorder, rather than internal states and anxiety (Vitiello *et al*, 1989). The term ‘compulsive-behaviour disorder’ has been suggested in these circumstances (Bodfish & Madison, 1993), and there

## EVIDENCE

is evidence that some people with mainly ‘compulsive behaviour’ symptoms respond to the standard treatments used in OCD (Barak *et al*, 1995). The readers are referred to the DC-LD (2001) criteria.

Barak, Y., Ring, A., Levy, D., Granek, I., Szor, H. & Elizur A. (1995) Disabling compulsions in 11 mentally retarded adults: an open trial of clomipramine SR. *Journal of Clinical Psychiatry* **56** (10) 459–61.

Bodfish, J. W., Crawford, T. W., Powell, S. B., Parker, D. E., Golden, R. N. & Lewis, M. H. (1995) Compulsions in adults with mental retardation: prevalence, phenomenology, co-morbidity with stereotypy and self-injury. *American Journal on Mental Retardation* **100** 183–192.

Bodfish, J. W. & Madison, J. T. (1993) Diagnosis and fluoxetine treatment of compulsive behaviour disorder of adults with mental retardation. *American Journal of Mental Retardation* **98** (3) 360–367.

Carminati-G. G. & Yogwa, S. D. (1998) Obsessive-compulsive disorders in a young man with a light mental retardation: Importance of a longitudinal study of the symptoms and of an integrated treatment. *European Journal of Mental Disability* **5** (19) 25–35.

Deb, S. (2001a) Epidemiology of psychiatric illness in adults with intellectual disability. In: L. Hamilton-Kirkwood, Z. Ahmed, D. Allen, S. Deb *et al* (Eds) Health Evidence Bulletins-Learning Disabilities pp14–17. Cardiff: NHS. [wttp://.hebw.uwcm.ac.uk](http://.hebw.uwcm.ac.uk)

Gedye, A. (1992) Recognising obsessive-compulsive disorder in clients with developmental disabilities. *The Habilitative Mental Health Care Newsletter* **11** 73–74.

King, B. H. (1993) Self-injury by people with mental retardation: a compulsive behavior hypothesis. *American Journal on Mental Retardation* **98** 93–112.

McNally, R. J. & Calamari, J. E. (1989) Obsessive-compulsive disorder

in a mentally retarded women. *British Journal of Psychiatry* **155** 116–117.

Vitiello, B., Spreat, S. & Behar, D. (1989) Obsessive-compulsive disorder in mentally retarded patients. *Journal of Nervous and Mental Disease* **177** (4) 232–236.

## Other 'neurotic' and stress-related disorders

In ICD-10, a variety of disorders are grouped with anxiety disorders and obsessive-compulsive disorder under this heading. The literature on the presence of these disorders in intellectual disability is sparse, and diagnosis may not be possible. However, as with anxiety disorders, some of the more persistent disorders in this group show a high level of co-morbidity, which it may be possible to detect and therefore treat.

This document does not discuss neurasthenia (chronic fatigue syndrome) or dissociative (conversion) disorders, which may well occur in this group, but on which there is little research.

## Reactions to stress

This includes brief reactions to stress, as well as post-traumatic stress disorder.

Acute stress reactions are short-lived disorders that occur in response to marked physical or mental stress. They usually present with symptoms of mood disturbance, changes in level of activity and behaviour disturbance, and subside within hours or days.

People with intellectual disability are often subject to short-lived stresses. Acute stress reactions may theoretically present with the above symptoms, or brief (ie hours to days) disturbances in behaviour.

## Post-Traumatic Stress Disorder (PTSD)

PTSD is a disorder where the response to a stressful event or situation is more delayed, and the symptoms more long lasting. The event or situation has to be particularly threatening or catastrophic in nature (one that would cause marked distress in most people, such as a serious road accident involving fatalities).

Typical features include:

- Episodes of reliving the experience in flashbacks (intrusive memories or images), dreams or nightmares.
- Avoidance of situations similar to, or reminding the subject of the traumatic event.
- Associated symptoms such as difficulty remembering important aspects of the event, or evidence of increased arousal (sleep difficulties, irritability, poor concentration, hyper-vigilance or exaggerated startle response).
- There may be associated depressive symptoms including suicidal ideation.
- Stressors in the lives of persons with intellectual disability can be long-lasting because of the person's inability to avoid the stressors.

The literature on PTSD in adults with intellectual disability is limited. It is likely to occur, as people with intellectual disability do experience traumatic events, and may be at a higher risk for some traumatic experiences (eg sexual abuse) than the general population. It may be that the disorder can occur in this population group following what could be seen as relatively less traumatic events. The diagnosis relies on the subject being able to describe quite complex internal states. Those who are

## EVIDENCE

unable to communicate their experiences may manifest behavioural problems. PTSD should be considered where there are changes in a person's behaviour, mood or level of functioning following a marked traumatic event.

Co-morbidity, especially depressive disorder, is common.

Ryan, R. (1994) Post-traumatic stress syndrome: Assessing and treating the aftermath of sexual assault. *The NADD Newsletter* **3** 5–7.

Ryan, R. (1995) Post-traumatic stress disorder in persons with developmental disabilities. *Community Mental Health Journal* **30** (1) 45–54.

(Type IV evidence: observational study describing 51 individuals with intellectual disability who met DSM-III-R criteria for PTSD. The author notes that those who received recommended treatment for the disorder improved.)

## Adjustment disorders

According to the ICD-10 criteria these are states of subjective distress and emotional disturbance, usually interfering with social functioning and performance, arising in the period of adaptation to a significant life change or a stressful life event. In the case of an adult with a learning disability they may be vulnerable to even minor life events.

A wide range of life events may cause adjustment disorders including a move to another home, changes in carers, other residents, changes in occupation or day activities, or bereavement or illness in the family. Adjustment disorders tend to present with symptoms of anxiety or depression, which may manifest as a behaviour disorder in adults with intellectual disability. The symptoms, however, do not meet criteria for a depressive disorder or anxiety disorder.

## *Somatoform disorders (hypochondriacal disorder)*

In somatoform disorders, there is an ongoing belief that the subject has some physical disorder (in the absence of such evidence). The subject will frequently present to doctors, sometimes with actual somatic complaints, despite previous reassurance and lack of evidence of physical illness on examination and investigation. In some people, the belief that they have some physical illness (often a serious illness, such as cancer)

reaches delusional intensity.

Many adults with intellectual disability will complain to their carers of aches and pains in the body, on a regular basis. It may be difficult to ascertain whether this amounts to a diagnosis of somatoform disorder or whether these are merely the manifestation of the subject seeking some form of attention or reassurance from others. It is important though, to take such complaints seriously. Somatic complaints are a common feature of depression in this group (Prasher, 1999).

**EVIDENCE**

It is not known how common the disorder is in people with intellectual disability. There are some single case reports in the literature that suggests that the disorder does occur.

Brooke, S., Collacott R. A. & Bhaumik, S. (1996) Monosymptomatic hypochondriacal psychosis in a man with learning disabilities. *Journal of Intellectual Disability Research* **40** (1) 71–74.

(Type V evidence: case report.)

Prasher, V. (1999) Presentation and management of depression in people with learning disability. *Advances in Psychiatric Treatment* **5** 447–454.

## Eating disorders

Disorders of appetite and weight are common in people with intellectual disability. In Prader-Willi syndrome compulsive over-eating occurs, which can lead to extreme obesity and associated illnesses such as diabetes.

A significant number of people with intellectual disability are on medication that affects appetite and weight. This includes neuroleptics, antidepressants, and some anticonvulsants.

### Anorexia nervosa and bulimia nervosa

The central features in anorexia nervosa are:

- deliberate weight loss
- behaviour to maintain the low weight such as self-induced vomiting, starving or excessive exercise
- distorted sense of body image (the subject may believe herself fat despite evidence to the contrary)
- disturbance of the hypothalamic-pituitary axis (amenorrhea in females).

The characteristics of bulimia nervosa are: – a pre-occupation with weight control, repeated episodes of over-eating, followed by vomiting or the use of purgatives. There is an overconcern with body shape and weight. People with bulimia are usually of

around normal weight. There may be a history of anorexia preceding this disorder.

Other psychiatric disorders are commonly present, especially depression.

There are reports in the literature of specific eating disorders, such as anorexia nervosa and bulimia nervosa, occurring in those with intellectual disability. One case report (Thomas, 1994) notes the typical presentation of symptoms. Thomas

**EVIDENCE** suggests that in those with a more severe intellectual disability, subjects may not 'show sophisticated ploys to reduce weight or express body image distortion'.

In intellectual disability eating disorders can also present as a behavioural disorder such as 'pica'.

Clarke, D. J. & Yapa, P. (1991) Phenylketonuria and anorexia nervosa. *Journal of Mental Deficiency Research* **35** 165–170.

Cottrell, D. J. & Crisp, A. H. (1984) Anorexia nervosa in Down's syndrome: a case report. *British Journal of Psychiatry* **145** 195–196.

Danford, D.E. & Huber, A. M. (1982) Pica among mentally retarded adults. *American Journal of Mental Deficiency* **87** 141–146.

Holt, G. M., Bouras, N. & Watson, J. P. (1988) Down's syndrome and eating disorders: a case study. *British Journal of Psychiatry* **152** 847–848. (Type V evidence: case report.)

O'Brien, G. & Whitehouse, A. M. (1990) A psychiatric study of deviant eating behaviour among mentally handicapped adults. *British Journal of Psychiatry* **157** 281–284.

(Type IV evidence: observational study looking at the link between psychiatric

disorders and eating behaviours.)

Szymanski, L. S. & Biederman, J. (1984) Depression and anorexia nervosa of persons with Down syndrome. *American Journal of Mental Deficiency* **89** 246–251.

Thomas, P.R. (1994) Anorexia nervosa in people with severe learning disabilities. *British Journal of Learning Disabilities* **22** 25–26.  
(Type V evidence: case report and review.)

## Sleep disorders

Problems with sleep are common in those with an intellectual disability, and are often long-standing and persistent. Certain syndromes such as Prader-Willi (Clarke and Boer, 1998) have a link with sleep disorders. Sleep apnoea is common in certain conditions associated with intellectual disability such as Down's syndrome. Sleep apnoea may also manifest as behavioural disorders and may remain unrecognised.

Disorders of sleep are commonly due to an underlying disorder or problem. This could include physical disorders causing pain or discomfort, epilepsy, psychiatric disorders such as depression, medication, and environmental factors (noise, warmth of room, etc).

Investigation of sleep problems should include a comprehensive assessment to rule out the above. This may include the

## EVIDENCE

use of functional analysis. Other sleep related disorders that may occur in adults with intellectual disability are nocturnal seizure activities and ‘parasomnias’.

Brylewski, J. & Wiggs, L. (1999) Sleep problems and daytime challenging behaviour in a community-based adult sample of adults with intellectual disability. *Journal of Intellectual Disability Research* **43** 504–512.

Clarke, D. J. & Boer, H. (1998) Problem behaviors associated with deletion in Prader-Willi, Smith-Magenis, and cri du chat syndromes. *American Journal on Mental Retardation* **103** 264–271.

Clarke, D. J., Waters, J. & Corbett, J. A. (1989) Adults with Prader-Willi syndrome: abnormalities of sleep and behaviour. *Journal of the Royal Society of Medicine* **82** 21–24.

Clift, S., Dahlitz, M. & Parkes, J. D. (1994) Sleep apnoea in the Prader-Willi syndrome. *Journal of Sleep Research* **3** 121–126.

Espie, C. A. & Tweedie, F. M. (1991) Sleep patterns and sleep problems amongst people with mental handicap. *Journal of Mental Deficiency Research* **35** 25–36.

Ferri, R., Curzi-Dascalove, L., DelGracco, S., Elia, M., Musumeci, S. A. & Stefanini, M.C. (1997) Respiratory patterns during sleep in Down’s syndrome: importance of central apnoeas. *Journal of Sleep Research* **6** 134–141.

Lancioni, G. E., O’Reilly, M. F. & Basili, G. (1999) Review of strate-

gies for treating sleep problems in persons with severe or profound mental retardation or multiple handicaps. *American Journal on Mental Retardation* **104** (2) 170–186.

(Type V evidence: a review looking at behavioural strategies in managing sleep disorders.)

Mixer, R. C., David, D. J., Perloff, W. H., Green, C. G., Pauli, R. M. & Popic, P. M. (1990) Obstructive sleep apnoea in Apert's and Pfeiffer's syndromes: more than a craniofacial abnormality. *Plastic and Reconstructive Surgery* **86** 457–463.

## Psychosexual function

### EVIDENCE

There are some case reports of individuals with moderate to severe intellectual disability, and psychosexual disorders, but the literature in this area is sparse. However, there is a considerable literature on the forensic aspect of deviant sexual behaviour in adults who have mild intellectual disability.

Day, K. (1997) Sex offenders with learning disabilities. In: S. Read (Ed) *Psychiatry in Learning Disability* pp278–306. London: W.B. Saunders Company.

Fegan, L. & Rauch, A. (1993) *Sexuality and People with Intellectual Disability* (2nd edition). Baltimore: Paul H. Brookes Publishing

Company.

McCurry, C., McClellan, J., Adams, J., Norrei, M., Storck, M., Eisner, A. & Breiger, D. (1998) Sexual behavior associated with low verbal IQ in youth who have severe mental illness. *Mental Retardation* **36** 23–30.

Van Dyke, D. C., McBrien, D. M. & Mattheis, P. J. (1996) Psychosexual behavior, sexuality and management issues in individuals with Down's syndrome. In: J. A. Rondal, J. Perera *et al* (Eds) *Down's Syndrome: Psychological, psychobiological, and socio-educational perspectives* pp 191–203. London: Whurr Publishers.

## Mental and behavioural disorders due to psychoactive substance misuse

There is some research on alcohol and drug use in those with intellectual disability. Studies show a lower level of use compared with the general population (Christian & Poling, 1997). The opportunity to use and misuse substances is more likely in those with mild and moderate disability, who have greater independence and access. Much of the literature is on the use of alcohol. This suggests that the rate of problems in those who actually do drink is higher compared to the general population. Managing alcohol related disorders in this group may be more difficult for a variety of reasons, including:

- the cognitive impairment present in those with intellectual disability – exacerbating cognitive deficits due to excess alcohol use
- the increased presence of physical disorders, especially epilepsy, with the effect heavy alcohol use has on the management of epilepsy.

## EVIDENCE

Assessments should include routine questions about substance use, in those people with a less severe degree of disability, especially in those with high levels of independence, or where there is a suspicion of substance misuse (physical signs). Family and carers may be aware of substance use where a patient denies such use.

Christian, L. A. & Poling, A. (1997) Drug abuse in persons with mental retardation: a review. *American Journal on Mental Retardation* **102** 126–136.

(Type V evidence: a review of the current literature on the level of substance

misuse in this population.)

Clarke, J. J. & Wilson, D. N. (1999) Alcohol problems and intellectual disability. *Journal of Intellectual Disability Research* **43** 135–139.  
(Type V evidence: review of case reports.)

Degenhardt, L. (2000) Interventions for people with alcohol use disorders and intellectual disability: a review of the literature. *Journal of Intellectual and Developmental Disability* **25** 135–146.  
(Type V evidence: review article.)

Krishef, C. H. (1986) Do the mentally retarded drink? A study of their alcohol usage. *Journal of Alcohol and Drug Education* **31** 64–70.

## Dementia

This is a condition where there is a progressive deterioration in higher cognitive abilities, often associated with changes in personality, as a result of a degenerative process in the brain.

The condition occurs in clear consciousness.

The symptoms of cognitive decline include changes in memory (especially new learning), orientation, comprehension and planning, language, visuospatial abnormalities and judgement. The disorder may initially present with changes in emotion, social behaviour and motivation, before the cognitive changes are seen.

### Main types of dementia

- Alzheimer's disease (Temporoparietal predominance) (may account for half or more of all dementias. Specific link with Down's syndrome)
- Vascular dementia (increasingly recognised, may co-exist with Alzheimer's disease. Presentation can be acute and steplike)
- Lewy body dementia

- Pick's disease (Frontotemporal predominance) (usually commences in middle age, frontal lobe symptoms predominate)
- Huntington's disease (Subcortical predominance)
- Parkinson's disease

Dementia is an acquired condition, usually of later life, as opposed to an intellectual disability, which is a developmental disorder. People with intellectual disability will already have some degree of cognitive impairment. It is important to have a baseline of a person's best cognitive ability, in order to identify a decline in ability that may indicate dementia.

A few cases of dementia have an underlying treatable cause, such as hydrocephalus or hypothyroidism. Most cases of dementia, however, are progressive.

## Epidemiology of dementia

Dementia is largely a disease of old age. In the general population, the prevalence is about 2% in persons aged 65–70, and approximately 20% in those over the age of 80 (Royal College of Physicians, 1981).

In the population with intellectual disability, dementia occurs earlier and at higher rates. Community-based studies have found rates of 14% (Lund, 1985, in people over 45 years), 11% (Patel *et al*, 1993, in people over 50 years), and 22% (Cooper, 1997, in people over 64 years).

## Clinical features of dementia

In ICD-10, the diagnosis relies on the following general criteria:

- Evidence of **decline in memory, especially in learning new information**. Evidence for this can come from the history (patient or informant) or neuropsychological tests. In severe dementia, there will be a loss in previously acquired memories. Patients may show disorientation in familiar surroundings, forget things they previously knew (such as some or all of their address). More distant memories tend to be spared for longer, and a patient may regress to an earlier period in their life (for example saying that they are still living at home with their parents).
  - A **decline in other cognitive abilities**, such as judgement, thinking, planning and organisation. Such abilities may already be impaired in a person with intellectual disability. To make a diagnosis of dementia there should be evidence of loss of ability, ie skills that the individual could previously do, are now lost, such as the ability to dress themselves, or a loss of communication skills.
  - Happens in **clear consciousness**.
  - **Changes in emotions, social behaviour or motivation**. This may manifest itself as behaviour disturbance, withdrawal or apathy. In persons with intellectual disability this should be distinguished from the other causes of dementia.
  - Cognitive symptoms eventually affect a person's **social skills and adaptive behaviour**.
- The features ideally need to have been present for at least six months.
  - **Other clinical features include psychiatric symptoms**. Depressive symptoms commonly occur. (Indeed, a depressive disorder is a differential diagnosis, as well as being a possible

co-morbid illness). Psychotic symptoms tend to occur in the middle stages of the illness.

There are a variety of rating scales that have been developed for specific use in dementia and intellectual disability (see **Appendix**). A number of practice guidelines have also been developed (Janicki *et al*, 1995; Aylward *et al*, 1997; Zigman *et al*, 1997).

### The course of the illness

The early stages of dementia are usually characterised by impairment of memory, especially new learning, but later on acquired memories are lost, particularly more recently laid down memories. There may be episodes of disorientation. Language ability and learnt skills are usually less affected at this stage. Associated symptoms may occur, such as emotional change, a loss of motivation, and changes in social behaviour. In some adults with intellectual disability ‘sleep cycle reversal’ and ‘time disorientation’ are early manifestations of dementia.

In the middle stages of the disease, dyspraxias and agnosias may occur, which in a person with intellectual disability, can present as loss of skills. Some primitive reflexes may reappear. Psychiatric symptoms may occur at this stage.

In the latter parts of the illness, increasing muscle tone occurs. Subjects may develop epileptic seizures. Other reflexes such as the sucking and grasping reflexes reappear. The patient becomes incontinent (if previously continent), and there is a loss of most motor functions. With these impairments, the risk of infection is increased, and patients can develop broncho-pneumonia, which they commonly die from.

## Down's syndrome and Alzheimer's disease

The link between Down's syndrome and Alzheimer's disease has been well established (Oliver & Holland, 1986). Almost all people with Down's syndrome show neuropathological evidence of the changes associated with the disease from the age of 40 (Mann, 1988). However, in contrast the clinical presentation is variable, and far from universal over the age of 40. The quoted prevalence of clinical dementia in people with Down's syndrome are: 0–4% under 30 years of age; 2–33% for 30–39 years of age; 8–55% for 40–49 years of age; 20–55% for 50–59 years of age; 29–75% for 60–69 years of age (Zigman *et al*, 1997).

Between 31% and 78.5% of adults 65 years or older with intellectual disability but without Down's syndrome show Alzheimer's neuropathology (Barcikowaska, 1989; Cole *et al*, 1994; Popovich *et al*, 1990).

The only risk factors among people with intellectual disability for developing Alzheimer's dementia are increasing age and Down's syndrome. It is not clear what effect possible risk factors seen in the general population (eg family history, low educational level, lower IQ, head trauma, cardio-vascular disease, stroke, diabetes, Apolipoprotein E4, Presenilin-1 (PS-1) polymorphism, major depressive episode etc) have on dementia in people with Down's syndrome (Zigman *et al*, 1997; Tsolaski *et al*, 1997; Rubinsztein *et al*, 1999; Deb *et al*, 1998 & 2000).

The course of presentation is similar to that found in the general population (Prasher, 1995). However, one study has reported features similar to that related to frontal lobe dysfunction, such as change in personality and behaviour in the early stages of dementia in adults with Down's syndrome (Holland *et*

*al*, 1999). Age related cognitive decline has also been described in adults with Down's syndrome (Devenny *et al*, 1996).

The diagnosis of dementia in people with (particularly severe and profound) intellectual disability, especially in the early stages, is made difficult by the lack of reliable and standardized criteria and diagnostic procedures. Neuropsychological tests and observer rated scales such as the Dementia Questionnaire for Persons with Mental Retardation (DMR) (Evenhuis, 1996) and the Dementia Scale for Down Syndrome (DSDS) (Gedye, 1995) need further evaluation before they can be accepted for day-to-day clinical assessment (Aylward *et al*, 1997; Deb & Braganza, 1999).

## Guidelines on assessment and diagnosis of dementia

- An important part of the assessment is to establish a 'baseline' – in order to determine the highest level of functioning that the patient had, and determine which cognitive deficits may be a longstanding result of the intellectual disability.
- In patients with severe or profound disability, it may be difficult to notice cognitive decline. The assessor should look for evidence of loss of skills, even where such skills are limited (for example, a patient who was previously able to feed themselves, but now is unable).
- Appropriate investigations should be done to rule out treatable causes of dementia.

## EVIDENCE

- It may be necessary to defer a diagnosis, given that decline in dementia may be gradual, and a period of observation over time needed to observe the decline. This is important, given that a diagnosis of dementia has many important implications for care and management.
- Be aware of co-morbid illness, and differential diagnoses, especially depression.

Aylward, E. H, Burt, D. B., Thorpe, L. U., Lai, F. & Dalton, A. (1997) Diagnosis of dementia in individuals with intellectual disability. *Journal of Intellectual Disability Research* **41** (2) 152–64.

(Type V evidence: consensus expert opinion suggesting standardized criteria for diagnosis.)

Barcikowska, M., Silverman, W., Zigman, W. *et al* (1989) Alzheimer-type neuropathology and clinical symptoms of dementia in mentally retarded people without Down syndrome. *American Journal of Mental Retardation* **93** (5) 551–557.

(Type IV evidence: post-mortem findings of 70 people aged over 65 years, with intellectual disability but without Down syndrome.)

Cole, G., Neal, J. W., Fraser, W. I. & Cowie, V. A. (1994) Autopsy findings in patients with mental handicap. *Journal of Intellectual Disability Research* **38** 9–26.

(Type IV evidence: autopsy study of people with intellectual disability – 15 with Down syndrome and 18 without Down's syndrome.)

Collacott, R. A. (1992) The effect of age and residential placement on adaptive behaviour of adults with Down's syndrome. *British Journal of Psychiatry* **161** 675–679.

(Type IV evidence: observational study.)

Cooper, S-A. (1997) Psychiatry of elderly compared to younger adults with intellectual disabilities. *Journal of Applied Research in Intellectual Disability* **10** (4) 303–311.

(Type IV evidence: cross-sectional; study of 134 people over 65 years of age with intellectual disability, and 73 people aged 20–64 years with intellectual disability.)

Cooper, S-A. (1997) High prevalence of dementia among people with learning disabilities not attributable to Down's syndrome. *Psychological Medicine* **27** 609–616.

Cosgrave, M. & Lawlor, B. (1999) Dementia in Down's syndrome. *CNS* **2** 17–19.

(Type V evidence – review article of the aetiology and diagnosis of dementia in Down's syndrome.)

Cosgrave, M. P., McCarron, M., Anderson, M., Tyrrell, J., Gill, M. & Lawlor, B. A. (1998) Cognitive decline in Down syndrome: a validity/reliability study of the Test for Severe Impairment. *American Journal on Mental Retardation* **103** 139–197.

Cosgrave, M. P., Tyrrell, J., McCarron, M., Gill, M. & Lawlor, B. A. (1999) Determinants of aggression, and adaptive and maladaptive behaviour in older people with Down's syndrome with and without dementia. *Journal of Intellectual Disability Research* **43** (5) 393–399.

Dalton, A. J. (1992) Dementia in Down syndrome: methods of evaluation. In: L. Nadel and C. J. Epstein (Eds) *Alzheimer's Disease and Down Syndrome* pp51–76. New York: Wiley Liss.

Deb, S. & Braganza, J. (1999) Comparison of rating scales for the diagnosis of dementia in adults with Down's syndrome. *Journal of Intellectual Disability Research* **43** (5) 400–407.

(Type IV evidence: study compares clinicians' diagnosis (ICD-10) of dementia with that according to DMR, DSDS, and Mini Mental State Examination (MMSE) among 62 adults with Down's syndrome, 26 of whom had dementia.)

Deb, S., Braganza, J., Norton, N. *et al* (2000) Apolipoprotein E e4 allele influences the manifestation of Alzheimer's disease in adults with Down's syndrome. *British Journal of Psychiatry* **176** 468–472.

(Type IV evidence: case control study of ApoE genotypes among 24 adults with dementia and 33 non-demented adults with Down's syndrome, aged 35 years and over, and an additional group of 164 non-intellectually disabled adults. This study also includes meta-analysis of nine studies.)

Deb, S., Braganza, J., Owen, M. *et al* (1998) No significant association between PS-1 intronic polymorphism and dementia in Down's syndrome. *Alzheimer's Report* **1** (6) 365–368.

(Type IV evidence: case control study of PS-1 genotype among demented and non-demented adults with Down's syndrome and a non-intellectually disabled control group.)

Deb, S., de Silva, P. N., Gemmell, H. G., Besson, J., Ebmeier, K. P. & Smith, S. (1992) Alzheimer's disease in adults with Down's syndrome: the relationship between regional blood flow deficits and dementia. *Acta Psychiatrica Scandinavica* **86** 340–345.

Devenny, D. A., Silverman, W. P., Hill, A. L., Jenkins, E., Sersen, E. A. & Wisniewski, K. E. (1996) Normal ageing in adults with Down's syndrome: a longitudinal study. *Journal of Intellectual Disability Research* **40** 208–221.

Fenner, M. E., Hewitt, K. & Torpy, D. M. (1987) Down's syndrome: intellectual and behavioural functioning during adulthood. *Journal of Mental Deficiency Research* **31** 241–249.

Gedye, A. (1995) *Dementia Scale for Down syndrome – Manual*. Vancouver, Canada: Gedye Research & Consulting.

Gedye, A. (1998) Neuroleptic-induced dementia documented in four adults with mental retardation. *Mental Retardation* **36** (3) 182–186.

Haxby, J. V. (1989) Neuropsychological evaluation of adults with Down's syndrome: patterns of selective impairment in non-demented old adults. *Journal of Mental Deficiency Research* **33** 193–210.

Holland, A. J., Hon, J., Huppert, F. A., Stevens, F. & Watson, P. (1998) Population-based study of the prevalence and presentation of dementia in adults with Down's syndrome. *British Journal of Psychiatry* **172** 493–498.

Janicki, M. P., Heller, T., Seltzer, G. & Hogg, J. (1995) *Practice Guidelines for the Clinical Assessment and Care Management of Alzheimer and other Dementias among Adults with Mental Retardation*. Washington DC: American Association on Mental Retardation.

Lund, J. (1985) The prevalence of psychiatric morbidity in mentally retarded adults. *Acta Psychiatrica Scandinavica* **72** (6) 563–570.

(Type IV evidence – case-control cohort study of 302 adults with intellectual disability identified from the Danish National Register. It also draws comparisons with eight previous cross-sectional studies.)

Mann, D. M. A. (1988) Alzheimer's disease and Down's syndrome. *Histopathology* **13** 125–137.

(Type V evidence: review of case reports including 398 cases altogether.)

Moss, S. & Patel, P. (1993) The prevalence of mental illness in people with intellectual disability over 50 years of age and the diagnostic importance of information from carers. *Irish Journal of Psychological Medicine* **14** 110–129.

Moss, S. & Patel, P. (1995) Psychiatric symptoms associated with dementia in older people with learning disability. *British Journal of Psychiatry* **167** 663–667.

(Type IV evidence – looking at the non-cognitive features of 105 people with intellectual disability using the PAS-ADD.)

Moss, S. & Patel, P. (1997) Dementia in older people with intellectual disability: symptoms of physical and mental illness, and levels of adaptive behaviour. *Journal of Intellectual Disability Research* **41** 60–69.

Morris, J. C., Heyman, A., Mohs, R. C., Hughes, J. P., van Belle, G., Fillenbaum, G., Mellits, E. D. & Clarke, C. (1989) The consortium to establish a registry for Alzheimer's disease (CERAD). Part 1: Clinical and neuropsychological assessment of Alzheimer's disease. *Neurology* **39** 1159–1165.

National Institute for Clinical Excellence (NICE) (2001) Guidance on the use of donepezil, rivastigmine and galantamine for the treatment of Alzheimer's disease. *Technology Appraisal Guideline* no. 19 (January). London: NICE (NHS).

Oliver, C. & Holland, A. J. (1986) Down's syndrome and Alzheimer's disease: a review. *Psychological Medicine* **16** 307–22.

Patel, P., Goldberg, D. & Moss, S. (1993) Psychiatric morbidity in older people with moderate and severe learning disability. II. The prevalence study. *British Journal of Psychiatry* **163** 481–491.

(Type IV evidence: cross-sectional study of 105 people with intellectual disabilities aged 50 years and over, using PAS-ADD diagnostic interview.)

Popovich, E. R., Wisniewski, H. M., Barcikowska, M. *et al* (1990) Alzheimer's neuropathology in non-Down's syndrome mentally retarded adults. *Acta Neuropathologica* **80** 362–367.

(Type IV evidence: autopsy study of 385 non-Down syndrome people with intellectual disability.)

Prasher, V. P. (1995a) Age-specific prevalence, thyroid dysfunction and depressive symptomatology in adults with Down's syndrome and dementia. *International Journal of Geriatric Psychiatry* **10** 25–31.

(Type IV evidence: observational study.)

Rubinsztein, D. C., Hon, J., Stevens, F. *et al* (1999) ApoE genotypes and risk of dementia in Down syndrome. *American Journal of Medical Genetics* **88** (14) 344–347.

(Type IV evidence: case-control study of the ApoE genotypes among 20 demented and 25 non-demented adults with adults with Down syndrome. Also a meta-analysis of six studies.)

Royal College of Physicians (1981) Organic mental impairment in the elderly: implications for research, education and the provision of services. *Journal of the Royal College of Physicians* **15** 141–167.

Tsolaki, M., Fountoulakis, K., Chantzi, E. *et al* (1997) Risk factors for clinically diagnosed Alzheimer's disease: a case-control study of Greek population. *International Psychogeriatrics* **9** (3) 327–341.

(Type III evidence: case-control study of 65 patients with Alzheimer's disease and

69 age-matched controls.)

Wisniewski, K. E., Wisniewski, H. M. & Wen, G. Y. (1985) Occurrence of Alzheimer's neuropathology and dementia in Down syndrome. *Annals of Neurology* **17** 278–282.

Zigman, W., Scupf, N., Haveman, M. *et al* (1997) The epidemiology of Alzheimer's disease in mental retardation: results and recommendations from an international conference. *Journal of Intellectual Disability Research* **41** (1) 76–80.

(Type V evidence: expert review of 14 studies and consensus opinion.)

Zigman, W. B., Schupf, N., Lubin, R. *et al* (1987) Premature regression of adults with Down's syndrome. *American Journal of Mental Deficiency* **92** 161–168.

## Delirium (Acute/sub acute confusional state)

Delirium is a confusional state caused by an underlying medical condition.

### Clinical features of delirium

- **Appearance and behaviour:** overactive and agitated or underactive and drowsy or mixed features, worse at night. Also features of underlying medical condition
- **Mood:** anxious or irritable or depressed or perplexed or mixed, tends to vary
- **Thought:** speech reduced, form muddled, content – ideas of reference or delusions
- **Speech:** mumbling and incoherent
- **Perception:** visual illusions or misinterpretations or hallucinations, less common in other modalities
- **Cognition:** abnormalities in all areas; memory-recognition, retention, recall-all impaired, orientation disturbed, concentration

impaired; mild cases may only have slow task performance or wandering of attention

■ **Insight:** impaired

(Gill & Mayo, 2000)

## Causes of delirium

**I:** Infections: intracerebral (encephalitis, meningitis), extracerebral (chest, UTI)

**W:** Withdrawal: alcohol, sedatives

**A:** Acute metabolic: hypoglycaemia, diabetic coma, renal or hepatic failure

**T:** Trauma: head injury, burns, heat stroke

**C:** CNS pathology: space occupying lesion, infection, epilepsy (status and post ictal state), Wernicke's and other encephalopathy

**H:** Hypoxia (acute myocardial infarction, carbon monoxide poisoning, respiratory failure, hanging, poisoning with overdose)

**D:** Deficiency: thiamine, vitamin B12, folate etc

**E:** Endocrine: over or underactivity of thyroid, parathyroid, adrenal glands

**A:** Acute vascular: transient ischaemic attack (TIA), stroke, hypertensive encephalopathy, shock

**T:** Toxins or drugs (legal or illicit)

**H:** Heavy metals, eg lead, mercury

(I WATCH DEATH mnemonic: Gill & Mayo, 2000–modified from Wise & Trzepacz, 1994)

Many medical conditions are common among adults with intellectual disability (Beange *et al*, 1995, Webb & Rogers, 1999; Deb 2001b). Many medical conditions including adverse

**EVIDENCE** Education can cause behavioural problems in adults with intellectual disability (Kalachnik *et al*, 1995; Peine *et al*, 1995; Bosch *et al*, 1997). Epilepsy is a common medical condition that affects 14–24% of adults with intellectual disability. There is a considerable body of literature on epilepsy in intellectual disability and its association with psychopathology. It is not within the remit of this document to go into a detailed discussion on this subject, however for further reading the readers are referred to a recent review on the subject by Deb (2000).

Beange, H., McElduff, A. & Baker, W. (1995) Medical disorders of adults with mental retardation: a population study. *American Journal on Mental Retardation* **99** 595–604.

Bosch, J., Van Dyke, D. C., Smith, S. M. & Pulton, S. (1997) Role of medical conditions in the exacerbation of self-injurious behaviour: an exploratory study. *Mental Retardation* **35** 124–130.

Deb, S. (2000) Epidemiology and treatment of epilepsy in patients who are mentally retarded. *CNS Drugs* **13** (2) 117–128.

(Type V evidence: literature review.)

Deb, S. (2001b) Medical conditions in people with intellectual disability. In: L. Hamilton-Kirkwood, Z. Ahmed, D. Allen, S. Deb *et al* (Eds) *Health Evidence Bulletins-Learning Disabilities* pp48–55. Cardiff: NHS Wales. <http://hebw.uwcm.ac.uk>

Gill, D. & Mayou, R. (2000) Delirium. In: M.G. Gelder, J. J. Lopez-Ibor Jr. and N. C. Andreasen (Eds) *New Oxford Textbook of Psychiatry* pp382–387. Oxford: Oxford University Press.

Kalachnik, J. E., Hanzel, T. E., Harder, S. R. *et al* (1995) Anti-epileptic drug behavioral side effects in individuals with mental retardation and the use of behavioral measurement techniques. *Mental Retardation* **33** 374–382.

Peine, H. A., Darvish, R., Adams, K. *et al* (1995) Medical problems, maladaptive behaviours and the developmentally disabled. *Behavioral Intervention* **10** 119–140.

Webb, O. J. & Rogers, L. (1999) Health screening for people with intellectual disability: the New Zealand experience. *Journal of Intellectual Disability Research* **43** (6) 497–503.

Wise, M. G. & Trzepacz, P. (1994) Delirium. In: J. R. Rundell and M. G. Wise (Eds) *Textbook of Consultation-liaison Psychiatry*. Washington DC: American Psychiatric Press.

## Personality disorders

‘Personality’ refers to those longstanding qualities of an individual that are manifest in the way that s/he behaves in a wide variety of circumstances. These qualities or characteristics are usually seen as present from adolescence, stable, and easily recognisable to those who know that individual.

Personality disorders refer to ‘*characteristic and enduring patterns of inner experience and behaviour as a whole (which) deviate markedly from the culturally expected and accepted range (or ‘norm’)*’. These aspects relate to cognition (thought, perception of others), affectivity (emotions), control over impulses and needs, and manner of relating to others (from ICD-10 diagnostic criteria). They tend to persist throughout adult life, although may become less marked in middle age.

Personality disorder remains a controversial area, especially in its relation to psychiatric illness. Assessment and treatment of an individual with a psychiatric illness and a personality disorder is that much more difficult.

In the general population, one study found the prevalence

of personality disorder to be 10 – 13% (Weissman, 1993). This compares with studies in the population with an intellectual disability, where a prevalence of 22–27 % (Corbett, 1979; Eaton & Menolascino, 1982, Reid & Ballinger, 1987) has been found. However, these figures should be interpreted with caution because of the difficulties associated with making this diagnosis in an adult who has intellectual disability.

- 1 There is the issue of whether particular characteristics or behaviours are due to a person's intellectual disability, or a personality disorder, as both are developmental issues.
- 2 The diagnosis of personality disorder is based in part on recognising that an individual's 'inner experiences' deviate from the norm. It may not be possible to assess this in someone with a severe or profound intellectual disability.
- 3 No validated personality disorder diagnostic scale exists for use in people with intellectual disability, although Reid and Ballinger (1987), Khan *et al* (1997), and Deb and Hunter (1991c) used the Standardised Assessment of Personality (SAP) (Mann *et al*, 1981).
- 4 Both Corbett (1979), and Deb and Hunter (1991c) showed considerable overlap between the diagnosis of personality disorders and behavioural disorders in adults who have intellectual disability. Therefore, it is essential to draw a proper distinction between behaviour disorder and personality disorder in persons with intellectual disability.
- 5 There are difficulties in using the standard personality disorder categories in adults who have intellectual disability. For example, a study by Corbett (1979) found that 25% could be diagnosed with a personality disorder according to ICD-8 criteria. However, the majority did not meet a

definite category, but were described as ‘*immature/irritable*’ or ‘*impulsive*’ (some of these patients also had anxiety-related disorders). The classifications do not however recognise categories such as ‘immature/irritable’.

Sixth, Bear and Fedio (1977) described personality traits such as ‘*humorless sobriety*’, ‘*hypergraphia*’ (excessive writing tendency), ‘*religiosity*’, ‘*circumstantiality*’ and so on, that are associated with complex partial seizures. These personality categories are not recognised in any of the current psychiatric classification systems such as the ICD-10 or DSM-IV-TR, yet epilepsy is common in adults with intellectual disability, and so are the epilepsy specific personality disorders (Deb, 2000; Deb & Hunter, 1991c).

Making a diagnosis of a personality disorder in someone with a severe intellectual disability is difficult, if not impossible. It may only be possible to give a good description of the behaviours observed in that individual, and assess the circumstances around them to see if any understanding can be made of them.

### Table of personality disorders

ICD-10	DSM-4
Paranoid	Paranoid
Schizoid	Schizoid Schizotypal*
Dissocial	Antisocial
Emotionally unstable - impulsive - borderline	Borderline

## EVIDENCE

	Histrionic
	Narcissistic
Anankastic	Obsessive-compulsive
Anxious (avoidant)	Avoidant
Dependent	Dependent
Other	Other

\*Schizotypal disorder is classified in ICD-10 under Schizophrenia and related disorders.

Bear, D. M. & Fedio, P. (1977) Quantitative analysis of interictal behavior in temporal lobe epilepsy. *Archives of Neurology* **34** 454–467.

Corbett, J. A. (1979) Psychiatric morbidity and mental retardation. In: F. E. James and R. P. Snaith (Eds) *Psychiatric Illness and Mental Handicap* pp11–25. London: Gaskell Press.

Dana, L. (1993) Personality disorder in persons with mental retardation. In: R. Fletcher and A. Dosen (Eds) *Mental Health Aspects of Mental Retardation* pp130–140. New York: Lexington Books.

Deb, S. & Hunter, D. (1991) Psychopathology in mentally handicapped patients with epilepsy. III: Personality Disorders. *British Journal of Psychiatry* **159** 830–834.

Eaton, L. F. & Menolascino, F. J. (1982) Psychiatric disorders in the mentally retarded: types, problems, and challenges. *American Journal of Psychiatry* **139** (10) 1297–1303.

Goldberg, B., Gitta, M. Z. & Puddephatt, A. (1995) Personality and trait disturbances in an adult mental retardation population: significance for psychiatric management. *Journal of Intellectual Disability Research* **39** (4) 284–294.

Khan, A., Cowan, C. & Roy, A. (1997) Personality disorders in people with learning disabilities: a community study. *Journal of Intellectual Disability Research* **41** (4) 324–330.

(Type IV evidence: study of a community sample, using the Standardised

Assessment of Personality (SAP). The study found that 31% of individuals could be given a diagnosis of personality disorder.)

Mann, A. H., Jenkins, R., Cutting, J. C. & Cowen, P.J. (1981) The development of a standardized measure of abnormal personality. *Psychological Medicine* **11** 839–847.

Oldham, J. M., Skodol, A. E., Kellman, H. D. *et al* (1992) Diagnosis of DSM-III-R Personality disorders by two structured interviews: patterns of co-morbidity. *American Journal of Psychiatry* **149** 213–222.

# APPENDIX

## *List of rating scales*

Here we have listed only those instruments that have been published and were used among adults with intellectual disability. The list is in no way exhaustive, and some scales listed here are mentioned in the evidence below. Some of these scales are adaptations from well-known scales in use in general psychiatry, whilst others are completely new. Some scales that are used in adults with intellectual disability have been adapted from a children's version.

Rating scales can be used to screen for the possible presence of psychiatric disorder (in general, or for a particular disorder). They are sometimes used as diagnostic tools, and some scales are designed to monitor the course of symptoms (during treatment), or adverse effects (side-effects).

A rating scale needs to demonstrate good reliability (consistent measurement across different settings), and good validity (the items actually measure what they are supposed to be measuring). Many rating scales listed here have not had their reliability and validity properly tested for use among adults with intellectual disability.

**EVIDENCE**

Aman, M. A. (1991) Review and evaluation of instruments for assessing emotional and behavioural disorders. *Australian and New Zealand Journal of Developmental Disabilities* **17** (2) 127–145.

Aylward, E. H. & Burt, D. (1998) *Test battery for the diagnosis of dementia in individuals with intellectual disability*. Report of the Working Group for the Establishment of Criteria for the Diagnosis of Dementia in Individuals with Intellectual Disability. Washington DC: American Association on Mental Retardation.

Borthwick-Duffy, S. A. (1990) Application of traditional measurement scales to dually diagnosed populations. In: E. Dibble and D. B. Gray (Eds) *Assessment of Behavior Problems in Persons with Mental Retardation Living in the Community* pp147–157. Rockville, MD: National Institute of Mental Health (Information Resources and Inquiries Branch).

Collacott, R. A. (1989) Rating scales used in the diagnosis and assessment of mental handicap. *Current Opinion in Psychiatry* **2** 613–617.

Hogg, J. & Ryanes, N. V. (1988) *Assessment in Mental Handicap: a guide to assessment practices, tests and checklists*. London: Croom Helm.

*Mental Measurements Yearbooks* (2001) Buros Institute, The University of Nebraska, Lincoln, Nebraska. [www.unl.edu/buros](http://www.unl.edu/buros)

O'Brien, G. (1995) Measurement of behaviour. In: G. O'Brien and W. Yule (Eds) *Behavioural Phenotypes* pp45–58. Cambridge: Cambridge University Press.

Sturme, P., Reed, J. & Corbett, J. (1991) Psychometric assessment of psychiatric disorders in people with learning difficulties (mental handicap): a review of measures. *Psychological Medicine* **21** 143–155.

The Penrose Society (1994) *Psychiatric Instruments and Rating Scales: A selected bibliography of mental measurements for the study of intellectual disability*. London, unpublished document.

The Royal College of Psychiatrists (1994) *A Selected Bibliography*. The Royal College of Psychiatrists, London, Occasional Paper (OP 23).

## Scales for the diagnosis/screening of psychiatric illness

### Affective rating scale

Wiesler, N. A., Campbell, G. J. & Sons, W. (1988) Ongoing use of an affective rating scale in the treatment of a mentally retarded individual with a rapid-cycling bi-polar affective disorder. *Research in Developmental Disabilities* **9** 47–53 (cited in Collacott, 1989).

### Assessment of Dual Diagnosis (ADD)

Matson, J. L. & Bamburg, J. (1998) Reliability of the Assessment of Dual diagnosis (ADD). *Research in Developmental Disabilities* **20** 89–95.

### Beck Depression Inventory (BDI)

Kazdin, A.E., Matson, J.L. & Senatore, V. (1983) Assessment of depression in mentally retarded adults. *American Journal of Psychiatry* **140** 1040–1043.

Beck, A. T., Ward, C. H., Mendelson, M., Mock, J. & Erbaugh, J. (1961) An inventory for measuring depression. *Archives of General Psychiatry* **4** 561–571.

### CANDID

Xenitidis, K., Thornicroft, G., Leeds, M. *et al* (2000) Reliability and validity of the CANDID – a needs assessment instrument for adults with learning disabilities and mental health problems. *British Journal of Psychiatry* **176** 473–478.

### Clinical Interview Schedule (CIS) – Mental Handicap Version

Ballinger, B. R., Armstrong, J., Presley, A. J. & Reid, A. H. (1975) Use of a standardized psychiatric interview in mentally handicapped patients. *British Journal of Psychiatry* **127** 540–544.

Goldberg, D. B., Cooper, B., Eastwood, M. R., Kedward, H. B. & Shepherd, M. (1970) A standardized psychiatric interview for use in community surveys. *British Journal of Preventive and Social Medicine* **24** 18–23.

### Diagnostic Assessment of the Severely Handicapped (DASH) Scale

Matson, J. L., Gardner, W. I., Coe, D. A. & Sovner, R. (1991) A scale for evaluating emotional disorders in severely and profoundly mentally retarded persons: development of the Diagnostic Assessment for the Severely Handicapped (DASH) Scale. *British Journal of Psychiatry* **159** 404–409.

### Diagnostic Assessment of the Severely Handicapped (DASH) II Scale

Matson, J. L., Rush, K. S., Hamilton, M., Anderson, S. J., Bamburg, J. W., & Baglio, S. (1999) Characteristics of depression as assessed by the Diagnostic Assessment for the Severely Handicapped-II (DASH-II). *Research in Developmental Disabilities* **20** (4) 305–313.

### Diagnostic Criteria for Research-10th Version (DCR-10) (modified)

Clarke, D. J. & Gomez, G. A. (1999) Utility of modified DCR-10 criteria in the diagnosis of depression associated with intellectual disability. *Journal of Intellectual Disability Research* **43** (5) 413–420.

### Hamilton Depression Scale – Mental Handicap Version

Sireling, L. (1986) Depression in mentally handicapped patients: diagnostic and neuroendocrine evaluation. *British Journal of Psychiatry* **149** 274–278.

### Hamilton Rating Scale for Depression

Hamilton, M. (1960) A rating scale for depression. *Journal of*

*Neurology, Neurosurgery and Psychiatry* **23** 56–62.

### Learning Disability version of the Cardinal Needs Schedule (LDCNS)

Raghavan, R., Marshall, M., Lockwood, A. & Duggan, L. (2001) *The Learning Disability version of the Cardinal Needs Schedule (LDCNS): a systematic approach to assess the needs of people with dual diagnosis*. Unpublished version ([r.raghavan@bradford.ac.uk](mailto:r.raghavan@bradford.ac.uk)).

Marshall, M., Hogg, L. I., Gath, D. H. & Lokwood, A. (1995) The Cardinal Needs Schedule: A modified version of the MRC Needs for Care Assessment Schedule. *Psychological Medicine* **25** 605–617.

### Mental Retardation Depression Scale

Meins, W. (1993) Prevalence and risk factors for depressive disorders in adults with intellectual disability. *Australia and New Zealand Journal of Developmental Disabilities* **18** 147–156.

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**I**n the last decade the professional knowledge concerning the problems of mental health among persons with intellectual disability has grown significantly. Behavioural and psychiatric disorders can cause serious obstacles to individual's social integration.

Clinical experience and research show that the existing diagnostic systems of DSM-IV and ICD-10 are not fully compatible when making a psychiatric diagnosis in people with intellectual disability. This may be one of the reasons why the evidence-based knowledge on the assessment and diagnosis of mental health problems in people with intellectual disability is still scarce.

This is the reason for the European Association for Mental Health in Mental Retardation (MH-MR) supporting the current project to produce a series of Practice Guidelines for those working with people with intellectual disability, to encourage and promote evidence-based practice. This is the first publication of the series.