Learning Disabilities
(Intellectual Disability)

February 2001

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Introduction

The original series Protocols for Investment in Health Gain were written in the early 1990s to suggest areas where the introduction, or more widespread use, of certain practices could lead to worthwhile improvements in health for the people of Wales. This revision has been prepared by reviewing the earlier Protocol for Investment in Health Gain: Learning Disabilities to provide some clear, updated statements with a precise indication of the strength of the evidence and its sources for each statement. The Bulletin also introduces new statements covering recent developments and subjects of important current interest.

The statements represent a methodical summary of the evidence in this area found through a formal literature search across a wide range of sources. The evidence has been critically appraised using internationally accepted methods, compiled into this document under the direction of a public health physician, and reviewed by a multidisciplinary team who are directly involved in patient care. The information in this document is also available electronically, via the NHS Cymru web (http://cymruweb.wales.nhs.uk/hebw) and the Internet (http://hebw.uwcm.ac.uk). Information on the methodology adopted (including a copy of the documentation), the formats in which the document is issued and details of other publications in the series are available on request.

The convention used in this document to indicate the type of evidence is:

- **Type I evidence** - at least one good systematic review (including at least one randomised controlled trial)
- **Type II evidence** - at least one good 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

Many health issues and interventions in the field of intellectual disability do not lend themselves to investigation by randomised controlled trial. Much of the quoted evidence is Type III, IV and V. There were very few randomised controlled trials to be found in the literature and even fewer systematic reviews. By valuing evidence from randomised controlled trials more highly than observational studies there is a danger that interventions with limited effectiveness might be judged to be more worthy than those based on observation. Similarly, those observational studies which clearly prove effectiveness (and make a randomised trial unethical) might be undervalued. Information assigned as Type V evidence may include expert opinion and important reports or recommendations which should rightly be highly regarded.

The health gain notation (used to indicate the potential benefit to health) is:

1. ‘beneficial’ - effectiveness clearly demonstrated
2. ‘likely to be beneficial’ - effectiveness not so firmly established
3. ‘trade-off between beneficial and adverse effects’ - effects weighed according to individual circumstances
4. ‘unknown’ - insufficient/inadequate for recommendation
5. ‘unlikely to be beneficial’ - ineffectiveness is not as clearly demonstrated as for 6
6. ‘likely to be ineffective or harmful’ - ineffectiveness or harm clearly demonstrated

It should be stressed that these gradings, while aiming to be impartial, represent only the best advice of the professionals involved in preparing the Bulletin. Where possible the health gain notation reflects both the type of evidence and the small size of some of the samples. Although the statements are deliberately brief, statistically significant quantitative information has been provided where possible. Issues of cost-effectiveness or cost-benefit are considered where evidence is available.

In keeping with the original Protocols, these revised documents are designed to assist Health Authorities in developing local strategies and in commissioning high quality health care. It is anticipated that they will also be of value to all professionals involved in delivering services for people with intellectual disability in keeping abreast of the large and increasing body of literature in this field. It should be stressed that the publications will act as a supplement to, not a substitute for, skills and experience. Some of the conclusions reached in this Bulletin will inevitably be controversial. Every effort has been made to include the best evidence within a subject area. Readers who are aware of any important studies that have been overlooked are encouraged to contact the project team.

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**Learning Disabilities Bulletin**

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**HEALTH EVIDENCE BULLETINS - WALES**
The prevalence rate in Western countries for moderate to severe intellectual disability is 30 per 1000 population. As such it is a common condition affecting at least 3% of the population and not surprisingly, people with intellectual disability have many of the conditions found in the general population. This Bulletin therefore considers a number of medical conditions with a known association with intellectual disability. The areas covered include:

- Epidemiology;
- Communication;
- Psychiatric illness;
- Behavioural disturbance;
- Forensics;
- Training staff;
- Hypothyroidism and epilepsy;
- Dementia and Down Syndrome;
- Drug therapy;
- Caring and carers

A deliberate decision has been taken to leave out a number of areas from the Bulletin due to difficulties either in identifying adequate literature in those areas or because the area has been covered in detail in another Bulletin. Areas omitted include ethics, medical conditions (other than epilepsy, thyroid disease and mental illness) and communication and special education in adults. Carers and caring is also covered in the Healthy Living Bulletin and cardiovascular disease is reviewed in the Cardiovascular Diseases Bulletin.

There is continuing confusion and complexity surrounding the classification of intellectual disability. With little agreement on terms, there is clearly a need for agreeing a common terminology. There are a range of general titles used in public phraseology depending upon local fashion, public confusion and prejudice, issues of perceived stigma, offensiveness and correctness. In this Bulletin we have used the term 'Intellectual Disability' except where another term is used as part of a formal reference. Terms include:

- Intellectual Disability: starting to appear in much of the UK and international literature and currently used by the International Association for Scientific Studies of Intellectual Disability and in the Journal of Intellectual Disability Research.
- Learning Disability: the 'official' term used in the UK.
- Mental Retardation: the 'official' term used in North America and the ICD-10 Mental Handicap: a term used historically in the UK.
- Mental Subnormality and Feeble-mindedness: now seriously outdated.
- Mental Impairment and Severe Mental Impairment: used in the UK 1983 Mental Health Act, although the Act is currently under revision.

Some authors propose a more innovative exploration of the potential of the multi-dimensional framework of the ICIDH-2. The current ICD-10 classification is limited, based upon assessment of IQ and an additional fourth character to identify the extent of impairment of behaviour:

- F70: Mild Mental Retardation, IQ 50-69.
- F71: Moderate Mental Retardation, IQ 35-49.
- F72: Severe Mental Retardation, IQ 20-34.
- F73: Profound Mental Retardation, IQ <20.
- F78: Other Mental Retardation.
- F79: Unspecified Mental Retardation.
- 0: no or minimal impairment of behaviour.
- 1: significant impairment of behaviour requiring attention or treatment.
- 8: other impairments of behaviour.
- 9: without mention of impairment of behaviour.

The statements made in this bulletin present a graded summary of the best available evidence of effectiveness across the spectrum of intellectual disability. The following information sources were systematically searched in the preparation of this Bulletin, according to the Project Methodology: Cochrane Library, Medline, Pre-Medline, Embase, CINAHL, PsychINFO, ASSIA, Caredata, Rehabdata, National Research Register, HealthPromis, the TRIP database, AMED, SIGLE. The full search strategies are available from the Project Office. The date of completion of the literature search for each chapter is given. Some additional evidence, published since the date of the systematic literature search, has been added on the basis of recommendations by Authors and/or the Review Groups where this lends support to an existing statement. The statements are intended to act as signposts to further sources of evidence, not as guidelines for the management of patients. It is hoped that this Bulletin will facilitate evidence-
based practice, which involves “integrating individual expertise with the best available external evidence from systematic research”\(^\text{12}\).

**Dr Laurence Hamilton-Kirkwood**, Team Leader.

December 2000.

3. Contact: Health Evidence Bulletins Wales, Department of Information Services, UWCM, Cardiff CF14 4XN.
4. This table is adapted from the Bandolier system (derived from the work at McMaster University, Canada) using the NHS Centre for Reviews and Dissemination criteria for a systematic review. See p.18 in ref.2 or http://www.jr2.ox.ac.uk/Bandolier/band6/6-5.html [accessed 8.12.00] and the Database of Abstracts of Reviews of Effectiveness (DARE) in the Cochrane Library.
1.1 Epidemiology

1.1a Classification of intellectual disability is a complex and confused area. There is still little agreement on terms and classes. There is a need for a general title for the field, a common terminology and taxonomy, and more innovative exploration of the potential of the multi-dimensional framework of the International Classification of Impairments, Disabilities and Handicaps (ICIDH).

1.1b ICIDH is under revision and is currently available in Beta-2 draft version.

1.1c The prevalence rate in Western countries for Severe Mental Retardation (IQ <50) is 3-4 per 1000 population. The prevalence of Moderate Mental Retardation (IQ 50-70) is 30 per 1000 population.

A typical District of 250,000 population would have 750 -1000 people with Severe Mental Retardation; 25-40 of these would have severe behavioural problems.

*These minor differences in prevalence rates reflect the confusion in terminology pointed out in 1.1a

The statements

The evidence

(Type V evidence - expert opinion with review of papers published during previous 12 months)

http://www.who.int/whosis/icidh/index.html [accessed 8.12.00]
(Type V evidence - expert opinion)

(Type IV evidence - systematic review of 43 prevalence studies)

(Type V evidence - expert opinion based on a survey of the field)
1.1d. The expected birth prevalence* of Down syndrome has increased from 1.67 (1996) to 1.84 per 1000 live births (1998). The observed birth prevalence has increased from 0.91 (1995) to 1.04 per 1000 live births (1998). Antenatal diagnosis has remained constant at 45-46%. This reflects a tendency for women to have babies at older ages and the greater use of antenatal screeningi. (UK)

*(assumes absence of antenatal screening and selective termination: to account for spontaneous foetal loss these figures also include 23% reduction in terminations after amniocentesis and 43% reduction in terminations after chorionic villus sampling)

1.1e. Down syndrome is the commonest known cause of mild and severe intellectual disability. Fetal Alcohol Syndrome is the second commonest known cause in many countriesi.

1.1f. Endemic cretinism caused by iodine deficiency is a common global cause of severe intellectual disability. Prevention with iodine is technically simple and cheapi. (Health gain notation – 1 “beneficial”)

1.1g. Extremely low birth weight children, even with optimal socio-economic environments, have a 50% chance of requiring special educational services and 20% are significantly disabled*. (USA)

*(presence of one or more severe functional disabilities including autism, cerebral palsy, mental retardation, borderline intelligence with global intellectual problems). See also1.2e.

1.1h. Between 5 and 34 years of age, the prevalence of severe intellectual disability is three times higher among the Asian community compared to the non-Asian community.

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**The statements**

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**The evidence**


i. Minns RA. Pre and perinatal conditions contributing to mental retardation. *Current Opinion in Psychiatry* 1997; 10(5): 354-59 (Type V evidence – expert opinion with review of 42 papers published during previous 12 months)


This document is a supplement to, not a substitute for, professional skills and experience. Users are advised to consult the supporting evidence for a consideration of all the implications of a recommendation.

1.1i. For most children with intellectual disability, between 30-60%, the cause is unknown.

1.1j. 80% of individuals with autism have significant intellectual disability. Conversely, autistic ‘traits’ are very common amongst people with intellectual disability; the full syndrome occurs in 17% overall and 27% of those with an IQ <50.

1.1k. Fragile X syndrome is not as common as was once thought with a revised prevalence of 1 in 5700 children of school age.

1.2 Risk Factors.

1.2a. Bacterial meningitis in childhood may result in a lower IQ score but rarely gives rise to significant intellectual disability (IQ≤70).

There is significant variation in likelihood of sequelae dependent upon the causative organism. Lower IQ score occurs with Neisseria meningitidis (2.1%), Haemophilus influenzae (6.1%) and Streptococcus pneumoniae (17%).

1.2b. Maternal smoking in pregnancy may be a preventable cause of intellectual disability (IQ≤70). Smoking one cigarette per day on five or more days per week in pregnancy may increase the risk of intellectual disability by 50%. There is evidence of a dose – response relationship.

(Health gain notation – 6 “likely to be harmful”)
1. Risk factors for intellectual disability need to be re-evaluated in the context of other important maternal, perinatal and neonatal characteristics. Characteristics associated with intellectual disability include low socio-economic status of the family (44-50% of intellectual disability) and low level of maternal formal education (20%). Other significant associations include maternal IQ ≤ 70, multiple births, low weight gain in pregnancy (<10 pounds), maternal anaemia and maternal urinary tract infection.

* This relationship does not exist for severe/profound intellectual disability.

1. Low Birth Weight (LBW) (birth weight ≤ 2500g) affects intellectual, educational and health outcomes.
Children with a birth weight ≤ 2500g have a higher rate of health and educational problems at seven years of age.
Children with a birth weight ≤ 1500g are more likely to have visited a GP, physiotherapist, speech therapist and eye specialist, and to require additional help in school.

1. Extremely Low Birth Weight (ELBW) (birth weight ≤ 800g) affects intellectual, educational and health outcomes. ELBW survivors are three times more likely to demonstrate intellectual disability at school age compared to full term babies with a birth weight ≥ 2500g. The most likely outcome for ELBW survivors at school age is a learning disorder, often multiple, or borderline intellectual functioning, combined with behavioural and motor risk factors rather than severe/multiple disability. But see also 1.1g.

1. Cerebral ventricular dilatation may be the strongest predictor of intellectual disability and cerebral palsy in ELBW babies.

The statements


1. Middle C, Johnson A, Alderdice F, Petty T, M acfarlane A. Birthweight and health and development at the age of 7 years. Child: Care, Health & Development 1996; 22(1): 55-71 (Type IV evidence - retrospective cohort study of 1169 UK children (aged 7) grouped by weight at birth)


1.3 Prevention

1.3a Surgical correction of single suture craniosynostosis in the first year of life may not prevent intellectual disability* or improve global cognitive functioning at a later stage. (* as defined by Bayley Scales of Infant Development and the McCarthy Scales of Children’s Abilities) (Health gain notation – 5 “unlikely to be beneficial”)

1.3b The reduction or cessation of smoking in pregnancy may reduce the likelihood of learning disability in offspring. See statement 1.2b. (Health gain notation – 2 “likely to be beneficial”)

i. Kapp-Simon KA. Mental development and learning disorders in children with single suture craniosynostosis. Cleft Palate - Craniofacial J 1998; 35(3): 197-203 (Type IV evidence - longitudinal evaluation of 84 consecutive children up to 50 months after initial assessment for surgery)

i. Drew CD, Murphy CC, Yeargin-Allsopp M, Decouflé P. The relationship between idiopathic mental retardation and maternal smoking during pregnancy. Pediatrics 1996; 97(4): 547-553 (Type IV evidence - case control study, 221 children with idiopathic intellectual disability and 400 controls from local schools)
2 COMMUNICATION

This document is a supplement to, not a substitute for, professional skills and experience. Users are advised to consult the supporting evidence for a consideration of all the implications of a recommendation.

2.1 Communication in Children

2.1a. Early Intervention has been shown to be important in fostering the development of communication skills in children.ii,iii,iv

(Health gain notation – 2 “likely to be beneficial”)

2.1b. An extensive range of interventions has been proposed for children with autistic spectrum disorders over recent years. A number of approaches have some evidence of their effectiveness but this is variable in quality. There is no study with rigorous control of variables such as intensity. Thus the effectiveness of any single approach cannot be precisely isolatedi.

(Health gain notation – 4 “unknown”)

The statements


(Type V evidence – expert overview of early intervention studies that concludes that there is a consensus from the research that early intervention makes a difference)


(Type III evidence - Meta-analysis examining six published studies showing positive outcomes with children with autism. None were randomised controlled trials)


(Type V evidence – expert review of interventions for children with an autistic spectrum disorder indicating sufficient consistent evidence to suggest that early intervention is effective)


(Type II evidence – randomised controlled trial of 20 mother-child dyads. Following 11 weeks of intervention the experimental group showed greater improvement in communication skills than the control group)

The evidence
This document is a supplement to, not a substitute for, professional skills and experience. Users are advised to consult the supporting evidence for a consideration of all the implications of a recommendation.

The statements

The evidence

2.2 Communication in Adults and Children

2.2a. Advice and Training for parents and carers on how to facilitate communication skills is important in enabling people with intellectual disability to communicate.


(Type III evidence - 78% of parents completing pre, post, and follow-up attitude questionnaires indicated that the group training enabled them to create a positive language-learning environment, and to continue as language facilitators after the training programme had ended)


(Type III evidence - 36 carer-client dyads were randomly assigned to 3 intervention approaches: direct therapy, teaching carers, and a combination approach. The combination approach demonstrated statistically significant improvements following intervention. There was not a 'no treatment' control group)


(Type III evidence - a case controlled cross sectional study of 102 adults with a profound intellectual disability. The evidence supports the use of a combined staff training and supervision format in communication intervention)

(iv) Hodgkinson P. Communication in ALD - What do carers think? Speech and Language Therapy in Practice 1998; Spring: 5-7

(Type IV evidence - Structured interviews of 12 staff support the need for an approach which combines a consultative role to carers with direct intervention)

2.2b. A primary focus of intervention should be to ensure:

- that a person with intellectual disability has maximum opportunities to communicate;
- that all communicative initiatives, responses to communication and interactions should be rewarding experiences.

(Health gain notation – 2 "likely to be beneficial")


(Type IV evidence - an observational single case study. The data presented is limited but supports the notion that the quality of communication improves when staff know how to enable a person with intellectual disability to communicate meaningfully)

(ii) van der Gaag AD. The view from Walter’s window; social environment and the communicative competence of adults with a mental handicap. Journal of Mental Deficiency Research 1989; 33: 221-7

(Type IV evidence - case controlled observational study of 120 learning disabled adults. Community settings were more effective in facilitating communication and clients in the community were significantly better communicators)


(Type IV evidence - cross sectional study suggesting that that the communicative environments of mentally handicapped persons become distorted and limit opportunities for communication)
This document is a supplement to, not a substitute for, professional skills and experience. Users are advised to consult the supporting evidence for a consideration of all the implications of a recommendation.

22c. **Intensive interaction** approaches are shown to be effective in promoting early communication skills in adults and children[1][2][3][4]. (Health gain notation – 2 “likely to be beneficial”)

**The evidence**

i. Nind M. Efficacy of intensive interaction: developing sociability and communication in people with severe and complex learning difficulties using an approach based on care-giver infant interaction. European Journal of Special Needs Education 1996; 11(1): 48-66 (Type IV evidence - observational study of six subjects suggesting that communication skills may be enhanced using intensive interaction based on typical care-giver - infant interaction)


iii. Prevezer W. Evaluation of an Alternative Approach. Musical Interaction with Children who were Considered Unable to Benefit from Conventional Speech and Language Therapy. Nottingham: Nottingham Community NHS Trust, 1994 (Type IV evidence - overview of 6 case studies)


22d. **Alternative** and **augmentative (AAC)** communication systems (high and low technology) may be helpful to adults and children with intellectual disability[1][2][3]. (Health gain notation – 2 “likely to be beneficial”)

**The evidence**

i. Jago JL, Jago AG, Hart M. An evaluation of the total communication approach for teaching language skills to developmentally delayed preschool children. Education and Training of the Mentally Retarded 1984; 19(3): 175-82 (Type III evidence - a case controlled study of 24 children supports the use of total communication with preschool, language delayed children)


This document is a supplement to, not a substitute for, professional skills and experience. Users are advised to consult the supporting evidence for a consideration of all the implications of a recommendation.

22a. Focussing on functional communication, social and pragmatic aspects of communication is effective whatever approach is used or whether AAC support is provided. (Health gain notation – 2 "likely to be beneficial")

i. Angelo DH, Goldstein H. Effects of a pragmatic teaching strategy for requesting information by communication board users. Journal of Speech and Hearing Disorders 1990; 55: 231-43

(Type IV evidence - observational study of 4 non-speaking children. Independent rater perceived significant differences when viewing before and after training video tapes)


(Type V evidence - expert opinion. A position paper which stresses the importance of teaching useful communication skills and in providing opportunities to express choice and preference)


(Type IV evidence - cross sectional study of 93 AAC users. Results indicated the importance of: aids being accessible; opportunities for a wide range of social contacts; training which focuses on the communication partner; useful vocabulary; and encouragement for spontaneous modes of communication)
This document is a supplement to, not a substitute for, professional skills and experience. Users are advised to consult the supporting evidence for a consideration of all the implications of a recommendation.

3.1 Psychiatric Illness

3.1a. Studies into the prevalence of psychiatric illness among adults with intellectual disability report a wide range, between 10% - 39%, depending on the sample selection; definition of psychiatric illness (some included and some excluded diagnoses such as behavioural disorders, pervasive developmental disorders, Rett syndrome and dementia); the diagnostic criteria used; and the diagnostic methods used.


ii. Göstason R. Psychiatric illness among the mentally retarded. A Swedish population study. Acta Psychiatrica Scandinavica, Supplementum 1985; 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 3 diagnostic criteria)

iii. Lund J. The prevalence of psychiatric morbidity in mentally retarded adults. Acta Psychiatrica Scandinavica 1985; 72(6): 563-70 (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)


3.1b. It is not clear whether or not the rate of psychiatric illness increases with the severity of intellectual disability.

It is difficult to decide with any degree of certainty whether mental illness in general but schizophrenia in particular is present in people with severe and profound intellectual disability.
This document is a supplement to, not a substitute for, professional skills and experience. Users are advised to consult the supporting evidence for a consideration of all the implications of a recommendation.

### 32 Psychiatric Illness in Elderly People

#### 3.2a Psychiatric morbidity

is found in 61.9% of adults with intellectual disability aged over 65 years. Among adults aged 50 years and over, the prevalence of psychiatric disorder excluding dementia is 11.4% and the prevalence of dementia is 11.4%.


Type IV evidence - cross sectional study of 134 people over 65 years of age with intellectual disabilities


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

### 33 Association of Psychiatric Illness with Other Disorders

#### 3.3a The rates of psychiatric illness as well as behavioural disorder in adults with intellectual disabilities and epilepsy are not significantly different from those in non-epileptic adults with intellectual disabilities.

i. Deb S. Mental disorder in adults with mental retardation and epilepsy. *Comprehensive Psychiatry* 1997; 38(3): 179-84

Type IV evidence - case-controlled study of 150 epileptic and an age-, sex- and IQ-matched control group of 150 non-epileptic adults with intellectual disabilities


Type IV evidence - cross-sectional study of 143 adults with intellectual disabilities and epilepsy

#### 3.3b The rate of functional psychiatric illness (excluding dementia and behavioural problems) in adults with severe intellectual disabilities but not Down syndrome is six times higher than adults with intellectual disabilities and Down syndrome.


Type IV evidence - cross-sectional study comparing 209 severely and 59 mildly intellectually disabled adults with Down syndrome with 477 severely and 1255 mildly intellectually disabled adults without Down syndrome


Type IV evidence - case-controlled study of 371 adults with Down syndrome compared with 371 matched adults with intellectual disabilities of causes other than Down syndrome
3.4 Prevalence of Specific Psychiatric Illnesses in Adults with Intellectual Disability

3.4a. Studies into the prevalence of specific psychiatric illnesses amongst adults with intellectual disability indicate that some conditions are more prevalent than others in people with intellectual disabilityi,ii,iii,iv.

The point prevalence of schizophrenia is reported as between 1.3% and 3.7%.

The point prevalence of affective disorders including depressive illness and mania are reported as between 1.2% and 6%.

The point prevalence of anxiety related neurotic disorders is found in around 16.4% adults (20-64 years).

3.4b. The prevalence of attention deficit hyperactivity disorder (ADHD) amongst adults with severe and profound intellectual disability (15%) is similar to children with severe intellectual disability (18%), but higher than in children with average intelligence (3%-5%).

3.4c. There is an association between depression and aggression with 40% of adults, adolescents and children with both intellectual disability and depression exhibiting aggressioni.

The statements

The evidence

(Type V evidence – expert review of 9 English language studies published between 1968 and 1985)

(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)

(Type IV evidence – cross-sectional study of a geographically defined total population of 2612 over a 25 year period)

(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)

(Type IV evidence - cross sectional study of 86 adults with severe to profound intellectual disability from a community setting, using the Conner’s (1990) Hyperactivity Index)

(Type IV evidence - cross sectional study of 528 adults, adolescents and children using Reiss scale)
This document is a supplement to, not a substitute for, professional skills and experience. Users are advised to consult the supporting evidence for a consideration of all the implications of a recommendation.

**3.5 Services for people with Psychiatric Illness and Intellectual Disability**

3.5a. Up to 5 years after **community resettlement**, people with intellectual disability show little change in the prevalence of psychiatric diagnoses or behavioural disturbance.

(Health gain notation – 4 “unknown”)

3.5b. **Outreach treatment** represents an effective and efficient alternative to hospital treatment for people with intellectual disability and psychiatric disorders

(Health gain notation – 2 “likely to be beneficial”)

**Assertive community outreach treatment** or intensive care (caseload, 10-15) significantly decreased the bed use and hospital admission in people with borderline intelligence and psychiatric illness when compared with those who received standard community care (caseload 25-35)

3.5c. 17% of all referrals to psychiatrists of people with intellectual disability were considered emergencies.

The majority of emergency referrals present as behavioural problems such as severe physical aggression and self-injurious behaviour.


(IV evidence - cross sectional study with 1 and 5 year follow ups of 74 adults with intellectual disability following resettlement in the community)

ii. van Minnen A, Hoogduin CAL, Broekman TG. Hospital vs. outreach treatment of patients with mental retardation and psychiatric disorders: a controlled study. Acta Psychiatrica Scandinavica 1997; **95**: 515-22

(II evidence - 28 week follow-up of 50 patients with intellectual disability referred for psychiatric admission: patients randomly allocated to outreach or hospital inpatient treatment)


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


(IV evidence - 12 month prospective study of referral patterns in a London borough)
4 BEHAVIOURAL DISTURBANCE

This document is a supplement to, not a substitute for, professional skills and experience. Users are advised to consult the supporting evidence for a consideration of all the implications of a recommendation.

4.1 Background

4.1a. Behavioural disturbance is referred to under a number of different terms – behavioural disorders, behavioural problems and challenging behaviour. Challenging behaviour was coined to reflect the fact that such behaviour is not intrinsic to the individual but should be viewed as a response to environmental, social, individual and historical characteristics. Problem behaviour is interpreted as a challenge to the system rather than a manifestation of an individual symptom or characteristic.\(^i\),\(^ii\),\(^iii\),\(^iv\).

4.1b. Recent studies suggest that between 12-17% of those administratively defined as having an intellectual disability will display challenging behaviour.\(^ii\),\(^iii\),\(^iv\),\(^v\).

Within a district of 500,000 general population, and assuming an administrative prevalence of intellectual disability of 0.45%, around 225-340 people will show challenging behaviour at any one point in time. Approximately 40-60% of these will show more severe problems.

Physical aggression, self-injury and destructiveness towards the environment tend to be the most commonly reported specific forms of challenging behaviour. Multiple forms are typically shown, and the behaviours concerned often appear to develop in childhood and remain highly persistent over time. Certain risk factors increase the likelihood of challenging behaviours. These include male gender; age between 15-35 years; having a more severe intellectual disability; having additional secondary impairments; reduced mobility and communication skills; and certain behavioural phenotypes.\(^i\),\(^ii\),\(^iii\),\(^iv\),\(^v\).

The statements

The evidence


4.1c Classification of behavioural disturbance:
One of the most enduring systems classifies aggressive behavioural problems into those which are directed towards other people (aggression to others), those which are directed towards objects in the environment (destructiveness) and self-injurious behaviour.

Non-aggressive behavioural disturbance include withdrawal, anxiety and phobic reactions. Self-injurious behaviour occurs in 14% of people with intellectual disability.

4.2 Assessment of Behavioural Disturbance

4.2a Assessment with a view to treatment should be wide ranging and comprehensive. Information should include cognitive abilities, communication abilities, perceptual and motor abilities, social skills, domestic skills, self-care skills, community skills, family history, living arrangements, health and medical status.

Functional analysis appears critical to effective behavioural intervention. The products of functional analysis include: a clear topographical description of the target behaviours (including classes or sequences that occur together); the identification of events that predict the occurrence and non-occurrence of the target behaviours over the person's day; the identification of events that maintain the behaviours (i.e. the purpose or function served by the behaviour); the development of hypotheses that link specific behaviours with specific triggers and consequences; and the collection of observational data to confirm or refute these hypotheses. Data for functional analysis may be gathered by interview, by observation and by setting up analogue assessments to test specific behaviour and environmental relationships.

Functional analysis enhances the success rates for behavioural intervention.

Reliability and validity is likely to be improved by the use of multiple methods.

The evidence

(Type IV evidence - review of challenging behaviour in a cohort of individuals with intellectual disability)

(Type V evidence - expert opinion)

(Type V evidence - expert opinion)

(Type I evidence - systematic review)

(Type I evidence - systematic review)

(Type IV evidence - experimental analysis of outcomes for different functional analysis methodologies)

(Type IV evidence - experimental analysis of outcomes for different functional analysis methodologies)
This document is a supplement to, not a substitute for, professional skills and experience. Users are advised to consult the supporting evidence for a consideration of all the implications of a recommendation.

4.2b ABC Analysis stands for antecedents – behaviour – consequences. Behavioural problems should always be assessed in the context of their environment. Antecedents should be assessed with a view to determining the extent to which these antecedents or stimuli control the instigation of the behaviour. There should be a topographical description of the behavioural problem including the frequency, duration and intensity of the behaviour. Finally the consequences of the behaviour should be assessed to gauge their effect on the maintenance or otherwise of the problem.

Antecedent analysis considers a behaviour in relation to the setting and situation in which it occurs, the time of day, week, month or year, immediate preceding activities or events, any interactions or people who are present at the time. Antecedent analysis also considers the same variables in relation to non-occurrence or absence of the behaviour. The distinction should be made between setting events (establishing operations such as tiredness, illness, mood states etc) that act by altering the value of reinforcers thereby setting the scene for challenging behaviours to occur; and specific antecedents (such as demands made on the individual) that actually trigger the behaviours of concern.

Consequence analysis considers the effects of a behaviour on the environment, the effects on other people, previous and current attempts to manage the behaviour and consequences which seem to increase or reduce the frequency, intensity or duration of the behaviour.

The evidence


**4.2c. Observation** of behavioural problems in relation to the environment is fundamental to the assessment of behavioural disturbance. The observational code should be relevant to the behaviour and environment in question, ensuring systematic objective reliable data recording. This data is crucial in developing functional analysis designed to identify controlling antecedents and consequences.

Observation will inform and guide appropriate treatment, and will also provide baseline information on the frequency, duration and intensity of the behaviour disturbance with a view to gauging the effectiveness of the intervention through re-assessment after the implementation of treatmenti,ii.

(Health gain notation – 2 “likely to be beneficial”)

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**4.2d. Continuous recording** of behaviour, its antecedents and consequences are the most valid methods of observation. This data will be comprehensive and relatively free from errors. However, since an individual may have to be observed over a lengthy period of time, continuous recording is mostly impractical because of the demands on the observersi,ii.

(Health gain notation – 2 “likely to be beneficial”)

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**4.2e.** Various observational methods have been assessed for comprehensiveness in relation to continuing recording. Methods include a record of the number of discreet events recorded, a measure of the total time spent engaged in the problem behaviour as a proportion of the total observation time, and various intervals of time sampling. This latter method allows the observer to watch the individual for a short period (for example, 10, 30 or 60 seconds) and then spend the next equivalent time interval recording whether or not the behaviour occurred and the circumstances surrounding the occurrence / non-occurrence of the behaviouri,ii.

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Related literature:

   (Type II evidence - randomised control trial comparison of time sampling methods)

   (Type V evidence - review and expert opinion)
42. Functional analysis assumes behavioural problems have a function for the individual. Description of the problem should include any antecedent stimuli or setting events; consequences or reinforcement contingencies which may be maintaining the problem; description of the personal, social and environmental impact of the disturbance; an analysis of the history of the problem and motivating factors. (Health gain notation – 2 "likely to be beneficial")

The most common behavioural processes are positive reinforcement. The disturbance results in social or other environmental reinforcement. The behaviour is likely to increase in frequency, duration or intensity when potential social contact is denied and reduce when this contact is available.

The second type of behavioural process is maintained by negative reinforcement (escape or avoidance). Behavioural disturbance increases where demands are placed on the individual and reduces when these demands are removed.

The third common behavioural disturbance is maintained by automatic reinforcement and appears uninfluenced by consequences. It occurs when there is little environmental stimulation and is controlled and maintained intrinsically by the automatic reinforcement provided by the behaviour.

Rating scales designed to identify these processes by considering events that affect frequency, duration or intensity of a behavioural disturbance are questionable and should not be used in the absence of additional information through interview of informants or observation.

The statements

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The evidence


Analogue assessments tests hypotheses by arranging artificial situations providing social reinforcement, tangible reinforcement, escape, avoidance or automatic reinforcement. Situations are individually tailored and the results can be used to identify appropriate individual treatment.

More recently studies have shown that less formal assessment including rating scales, informant interviews and some observational information can be valid and reliable ways of carrying out functional analysis\(^x\),\(^{xi}\),\(^{xii}\).

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\(^{xii}\) Yarbrough SC, Carr EG. Some relationships between informant assessment and functional analysis of problem behavior. The American Journal of Mental Retardation 2000; 105: 130-151 (Type IV evidence - well controlled study on 3 individual cases showing that informant assessment provided valid hypotheses about behavioural function only in situations likely to evoke challenging behaviour)
4.2 Functional analysis is now considered in relation to a wide range of issues including its effectiveness in relation to healthcare problems, delivery by parents and carers and behavioural classification.

4.3 Behavioural Treatments

4.3a Behavioural treatments derived from the fact that most behaviours are governed by their antecedents and consequences. Antecedent management includes consideration of stimulus control, setting events and establishing operations. Consequences can be environmental (increasing or decreasing behavioural disturbance) or internal to the person (automatic or intrinsic reinforcement). Behavioural treatments manipulate these relationships to improve the behavioural disturbance. Behavioural interventions appear to produce superior outcomes when compared to pharmacological interventions for challenging behaviour. (Health gain notation – 2 “likely to be beneficial”)

Both positive and negative reinforcement increase the future frequency, duration or intensity of behaviour. In positive reinforcement, a desirable outcome (eg. food, drink, attention, activity etc.) is presented contingent upon the target behaviour; in negative reinforcement, it is the contingent removal of an undesirable outcome (pain, demands, social attention) that achieves this effect.

Conversely, punishment decreases the future frequency, duration or intensity of behaviour. Positive punishment involves the contingent presentation of an aversive stimulus (eg pain, shock, forced exercise etc.), while negative punishment involves contingent removal of preferred stimuli (eg attention, activities, general access to positive reinforcement etc.).

The statements

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The evidence

i. Cone JD. Issues and functional analysis in behavioural assessment. Behaviour Research and Therapy 1997; 35: 259-75 (Type V evidence - expert opinion, review and synthesis of important studies)


Positive Behavioural Approaches

4.3b. **Positive behavioural** approaches aim to change challenging behaviours by focusing on environmental manipulation (thus preventing the occurrence of the behaviours) and by teaching alternative adaptive responses (for example, skills that serve the same function as the target behaviour). In contingency management terms, positive reinforcement based approaches are favoured and aversive approaches rejected for technical and ethical reasons.

Using a criterion of 90% reduction in challenging behaviour from baseline levels, positive interventions are successful approximately 52% of the time; using an 80% reduction criterion, this rate increases to 68%. Using the 80% criterion, separate success rates for antecedent/stimulus based interventions was almost 70% and for reinforcement based interventions almost 72%. Success rates are generally not influenced by whether or not those procedures are implemented alone or in combination. Success rates for studies that combine positive behavioural approaches with aversive procedures produce a success rate of almost 64% using a 90% suppression criterion. Caution is urged in interpreting the latter finding in that it is based on a small number of studies. Intervention for combinations of challenging behaviours rather than single behaviours tends to be less successful.

Interventions that involve systems/organisational change appear to be slightly more successful than those that do not (55% versus 42%). Some positive behavioural interventions may produce intervention effects that are at least as rapid as more aversive procedures.

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**The statements**

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**The evidence**


**4.3c. Functional communication training (FCT)**

assumes that behavioural disturbance, rather than being seen as an aberrant activity, should be viewed as a means of communicating the needs of that individual to others. FCT is effective in helping the individual learn alternative ways of communicating needs without recourse to behavioural disturbance. FCT therefore increases adaptive communication and decreases maladaptive communication\textsuperscript{i,ii}.

(Health gain notation - 2 “likely to be beneficial”)

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**4.3d. Recent studies have found that functional communication training on its own may be of limited effectiveness without concurrent extinction of the target behaviour.**

(Health gain notation - 5 “unlikely to be beneficial”)

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Functional communication training with extinction (not delivering a consequence for problem behaviour) resulted in 90% reductions of problem behaviour in 50% of clients\textsuperscript{i,ii,iii,iv}.

(Health gain notation - 2 “likely to be beneficial”)

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\textsuperscript{i.} Carr EG, Durand VM. Reducing behaviour problem through functional communication training. Journal of Applied Behaviour Analysis 1985; 18: 111-26

(Type IV evidence - well controlled case studies. This was one of the first papers to introduce the communication hypothesis and functional communications training)


(Type IV evidence - well controlled case studies)

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(Type IV evidence - well controlled case studies)


(Type IV evidence - well controlled case studies)

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(Type V evidence - expert opinion, review of important studies and description of treatment methods)

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(Type IV evidence - well controlled case studies)
4.3e. **Functional communication training** with mild punishment (e.g. time out in a room or a chair) directed at the target behaviour, resulted in 90% reduction in problem behaviour in 100% of clients.

(Health gain notation – 2 “likely to be beneficial”)

4.3f. **Extinction** is effective in reducing behavioural disturbance. By discontinuing any previous reinforcement of a behaviour, once the reinforcement is no longer available the behavioural disturbance reduces. No distinction is made between positive, negative, intrinsic or extrinsic reinforcement.

A serious side effect is the occurrence of extinction bursts where individuals respond with an increased frequency of the behaviour in order to gain (the now unattainable) reinforcement. In cases of aggressive, destructive or self-injurious behaviour this will present a danger to the individual and others. Extinction should normally only be considered and used in conjunction with other methods and for non-dangerous behaviours.

(Health gain notation – 3 “trade off between beneficial and adverse effects”)

4.3g. **Aversive Consequences**

There is extensive evidence that aversive consequences will produce short-term suppression of behavioural disturbance. Punishment techniques range from mild electric shock to cold water mist in the face and time out from positive reinforcement.

(Health gain notation – 2 “likely to be beneficial”)

These techniques do not carry the same requirements for functional analysis of the variables controlling the problem behaviour. They do not fit comfortably with the systematic, analytic and educational features of the majority of research on behavioural treatment. They also pose significant ethical and legal concerns.

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**The statements**


(Type IV evidence – a summary of data from well controlled case studies comparing functional communications training with extinction against functional communications training with mild punishment)

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**The evidence**


(Type V evidence - expert opinion with a review of some studies and methods)


(Type III evidence - systematic review of basic and applied research findings on variables that influence the direct and indirect effects of extinction. An expert evaluation of the general technology for the use of extinction)


(Type IV evidence – well controlled case studies)


(Type IV evidence – well controlled case studies)

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**Literature searches completed on 31.12.99**
4.3h Punishment does not institute alternative adaptive responses. Punishment may also have severe side effects in eliciting aggressive behaviours and avoidance or escape behaviours and it can serve as a negative modelling procedure\textsuperscript{ii,iii}.

Whereas more aversive procedures may produce greater levels of suppression (i.e. zero levels of target behaviour), the overall amount of change between baseline and treatment is not related to whether aversive or non-aversive approaches are used. The suppressive effect of more intrusive interventions is improved by the addition of Differential Reinforcement of Other Behaviour (DRO) to intervention packages\textsuperscript{iv}.

Despite their obvious effectiveness, concerns about the non-constructive nature of aversive interventions, together with their lack of social validity, has resulted in these approaches being described as “default” technologies. It has been suggested their use is only indicated when alternative positive approaches have failed or are not feasible and when the costs of not intervening are greater than the costs of using aversive procedures\textsuperscript{iii}.

4.3i. Time out, the loss of access to positive reinforcers to behaviour for a specific period of time, is effective in reducing future occurrences of abnormal behaviour. Time out is not simply removing an individual to a secluded setting (seclusion)\textsuperscript{ii}.

There is a distinction between “time in” and “time out”. The greater the reinforcing properties of the time in situation so will be the greater effectiveness of the time out situation. Time out may simply be ineligibility to access reinforcers for a short period of time. Time out procedures can increase the effectiveness of positive programming procedures when used in conjunction with them.

(Health gain notation – 2 “likely to be beneficial”)

See also 4.4i.

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\textsuperscript{i} Plummer S, Bare DM, LeBlanc JM. Functional considerations in the use of procedural time out and an effective alternative. J ournal of A pplied B ehaviour A nalys is 1977; 10: 689-706 (Type IV evidence - well controlled case study)


\textsuperscript{iv} Emerson E. Challenging B ehaviour. A nalys is and Intervention in People with L earning D ifficulties. C ambridge: Cambridge U niversity Press, 1985 (Type V evidence: expert opinion based on clinical evidence)
4.4 Skills teaching

4.4a. **Skills’ teaching** is effective in people with and without behavioural disturbance in adapting to and overcome problems in their lives. A crucial aspect of positive programming, skills’ teaching develops alternative functional behaviours that supplant behavioural problems and disturbances. Self-help skills, interpersonal skills, leisure skills, parenting skills and work skills can be developed using these methods.

A meta analysis of 73 studies found that social skills training produced improvements in a broad repertoire of social skills, had effects lasting several months and had a greater effect for developmentally disabled groups than other clients.

(Health gain notation – 2 "likely to be beneficial")

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**Behavioural Shaping, Prompting and Fading**

4.4b. **Shaping** is effective as an adjunct to training since subjects rarely achieve an adequate level of ability on their first attempt. Any approximation towards a reasonable level of skill is accepted by the therapist who then encourages subsequent attempts at improvement. The therapist may also establish one response and gradually shape it towards the desired end response.

In the early stages of training it is reasonable to **prompt** the desired response. Physical and verbal prompts can also help clients gain the confidence to begin a sequence of abilities.

An essential consideration when employing prompting is to build in the **fading** of prompts. It is unhelpful if the client becomes dependent on the therapists’ prompts, so the therapist should have a plan to begin fading not only therapeutic prompts, but other aspects of the therapeutic situation, so that the client is able to function independently.

(Health gain notation – 2 "likely to be beneficial")

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(Type I evidence - systematic review and meta-analysis of 73 studies)


(Type V evidence - expert opinion including review of important studies)
4.4c. Behavioural Chaining: is useful once a sequence or chain of skills has been established. The therapist focuses training at one end of the chain, the next step in the sequence is trained and linked to the previous one and so on until the whole complex ability has been learned. When training begins with the first aspect in the sequence this is called forward chaining; when it begins with the last it is backward chaining. (Health gain notation - 2 "likely to be beneficial")

4.4d. Role-play uses important stimuli from a real setting whilst remaining under therapist control. It is effective in allowing the group or individual to practice various skills concerned before going into the real setting. Role-play can involve a whole sequence of skills or one small aspect of a behavioural sequence. (Health gain notation - 2 "likely to be beneficial")

4.4e. Modelling allows a series of complex skills to be demonstrated without going into confusing explanations of how the skill is sequenced together. Modelling will then be combined with role-playing in an effort to help clients achieve the skill of the model. (Health gain notation - 2 "likely to be beneficial")

4.4f. Social problem solving is a useful process whereby skills for determining an effective action strategy in a given situation are taught to patients. (Health gain notation - 2 "likely to be beneficial") This is in addition to teaching the actual abilities to cope in specific situations. Results on the effectiveness of training social problem solving skills either on their own or as an adjunct to social skills training are equivocal.


iii. Baty FJ, Michie AM, Lindsay WR. Teaching mentally handicapped adults how to use a cafeteria. Journal of Mental Deficiency Research 1989; 33: 137-48 (Type IV evidence - case controlled study)

i. Lindsay WR, Michie AM. Teaching new skills. in Fraser W, Sines D, Kerr M (eds.). The Care of People with Intellectual Disabilities. 9th Edition. Oxford: Butterworth Heinemann, 1998 (Type V evidence - expert opinion including review of important studies)

i. Loumidis K, Hill A. Training social problem solving skills to reduce maladaptive behaviours in intellectual disability groups: the influence of individual difference factors. Journal of Applied Research in Intellectual Disabilities 1997; 10: 217-37 (Type II evidence - controlled group study involving 46 subjects in two groups. Also includes an extensive review of social problem-solving skills training)
Comprehensive systems based on behavioral principles

4.4. Skills Training Systems: Group comparison studies suggest that gains produced by social skills training are consistent and effective when compared with alternative group therapies and no treatment controls. Early intervention systems directed at improvements in cognitive academic and social skills have been employed for some time in children with autism. Recent studies enlisting parents to implement procedures at home have found positive resultsi,ii,iii,iv,v,vi.

(Health gain notation – 2 “likely to be beneficial”)

(Type III evidence - well designed controlled study comparing subjects receiving traditional psychotherapy and subjects receiving social skills training. Social skills training produced greater and more consistent improvements than psychotherapy)

(Type IV evidence - a series of cases seen in a group and employing a highly innovative and engaging approach to social skills training)

(Type II evidence - randomised controlled study comparing 29 subjects trained in community living skills using in vivo techniques, 13 using classroom techniques and 15 acting as a no treatment control. There was an overwhelming superiority in improvements for the skills training group)

(Type III evidence - a comparison of an experimental and control group with matched subjects. Significant improvements were found in the experimental cohort)

(Type III evidence - a matched group comparison showing significant improvements in the experimental cohort)

(Type V evidence - expert opinion based on a review of early intervention projects where the authors acknowledge undoubted treatment successes but recommend a healthy scepticism concerning unqualified endorsement)
Services for people with challenging behaviour and intellectual disability:
Effective behavioural support for persons with challenging behaviour can be provided within community settings. Dependence on institutional provision can be reduced as a consequencei,ii,iii,iv. (Health gain notation – 2 "likely to be beneficial")


iv. Mansell J. Specialised group homes for persons with severe or profound mental retardation and serious problem behaviour in England. Research in Developmental Disabilities 1994; 15: 371-388 (Type III evidence - well designed interventional study in 13 people with severe or profound intellectual disability and challenging behaviour)
In addition to having strategies for changing behaviour, carers supporting people who display dangerous behaviour will require strategies for managing these behaviours safely and effectively.

**Reactive behaviour management** strategies included distraction and diffusion, self-protective breakaway procedures and minimal restraint. Carers can effectively be taught a range of ethically acceptable techniques and their confidence increases as a result.\(^i\)

Training may also lead to a reduction in restraint, use of emergency medication, and injuries.\(^ii\)

(Health gain notation – 2 "likely to be beneficial")

It should be noted that all restraint procedures, however mild, carry the possibility of injury and should only be considered in conjunction with comprehensive policies for use and staff training.

The use of **seclusion** (the supervised containment of a person in a room to protect others from significant harm) for persons with a learning disability has been questioned. Its use may not bring about short-term improvements in behaviour as intended, may cause certain behaviours to worsen and elicit the appearance of new behaviours.\(^iv\)

(Health gain notation – 4 "unknown")

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4.4. In addition to having strategies for changing behaviour, carers supporting people who display dangerous behaviour will require strategies for managing these behaviours safely and effectively. Carers can effectively be taught a range of ethically acceptable techniques and their confidence increases as a result.\(^i\)

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(Health gain notation – 4 "unknown")
4.5 Relaxation

4.5a Abbreviated progressive relaxation (APR) and behavioural relaxation training (BRT) are effective when used in individuals with moderate, severe and profound intellectual disability. BRT works more rapidly and reliably than APR, and is an established treatment approach for individuals with anxiety, agitation, restlessness and disruptiveness\textsuperscript{i,ii,iii,iv}.

Because BRT is an essentially passive technique, it does not appear to have the drawbacks inherent in APR. There is no paradoxical tensing of muscles in order to relax, and there appear to be no side effects whereby clients misconstrue the demands of training, becoming more rather than less excited. It is an effective enabling technique to allow individuals to become more accessible to their environment.

(Health gain notation - 2 \textquotedblleft likely to be beneficial\textquotedblright)

4.5b Cue controlled relaxation (CCR) links the effects of relaxation to a cue word. In time the subject will be able to relax to the cue word only, rather than a time consuming relaxation procedure. CCR is effective at improving concentration and attention to an occupational task when used in individuals with moderate and severe intellectual disability\textsuperscript{ii}.

(Health gain notation - 2 \textquotedblleft likely to be beneficial\textquotedblright)

\textsuperscript{i.} Schilling D, Poppen R. Behavioural relaxation training and assessment. \textit{Journal of Behaviour Therapy and Experimental Psychiatry} 1983; \textbf{14}: 99-107 (Type II evidence - randomised controlled study comparing four forms of relaxation. BRT was consistently effective across all measures)

\textsuperscript{ii.} Lindsay W R, Baty F J, Michie A M, et al. A comparison of anxiety treatments with adults who have moderate and severe mental retardation. \textit{Research on Developmental Disabilities} 1989; \textbf{10}: 129-40 (Type II evidence - randomised controlled trial comparing BRT and APR against a control condition. Subjects in both conditions showed significant improvements over control subjects with BRT using more rapid improvement)

\textsuperscript{iii.} Lindsay W R, Morrison F M. The effects of behavioural relaxation on cognitive performance in adults with severe intellectual disabilities. \textit{Journal of Intellectual Disabilities Research} 1996; \textbf{40}: 285-290 (Type II evidence - randomised controlled trial. Subjects receiving BRT showed more significant improvements in cognitive functioning than control subjects on all tasks except those involving long term memory)


\textsuperscript{i.} Wells K C, Turner S M, Bellack A S, et al. Effects of cue control relaxation on psychomotor seizures: an experimental analysis. \textit{Behaviour Research and Therapy} 1978; \textbf{16}: 51-3 (Type IV evidence - well designed case-study employing multiple measure across various conditions)

\textsuperscript{ii.} Lindsay W R, Fee M, Michie A M, et al. The effects of cue control relaxation on adults with severe mental retardation. \textit{Research in Developmental Disabilities} 1994; \textbf{15}: 425-37 (Type IV evidence - well designed series of case controlled studies for the sequence of introduction on a range of variables)
4.6 The Snoezelen environment

4.6a. The Snoezelen environment was developed in Holland to induce meditation, tranquillity and relaxation. It has been adopted in the UK for people with multiple handicap and severe or profound intellectual disability. The environment is artificial with a variety of soothing stimuli for all the senses including lights, textures, sounds and smells.

The original developers have explicitly resisted empirical assessment of the environment. Recent studies have found that, with individuals who have severe and profound intellectual disability, snoezelen is no less effective than relaxation techniques on a variety of measures. It was found to be more effective than other “alternative” or “complementary” techniques such as hand massage or physical activity. It is considerably more expensive than relaxation techniques.i,ii.

A controlled trial demonstrated a decrease in aggressive responding in anger management and control conditions with no significant differences in the conditionsiii.

While some studies have noted short term positive effects, one controlled study found no generalised effect outside the immediate results of the Snoezelen environmentiv. (Health gain notation – 4 “unknown”)

4.7 Psychotherapy

4.7a. Reports of psychoanalytically informed psychotherapies have begun to appear over the last 10 years. In general these reports do not provide outcome data but where they do, the results are generally positivev,vi. (Health gain notation – 4 “unknown”)


ii. Lindsay WR, Pitcaithly D, Geelen N, et al. A comparison of the effects of four therapy procedures on concentration and responsiveness in people with profound learning disabilities. Journal of Intellectual Disability Research 1997; 41: 201-7 (Type III evidence - controlled crossover group comparison employing 8 subjects who each received treatments involving Snoezelen, aromatherapy, relaxation and physical exercise)

iii. Martin NT, Gaffan EA, Williams T. Behavioural effects of long term multi-sensory stimulation. British Journal of Clinical Psychology 1998; 37: 69-82 (Type II evidence – 27 adults with severe or profound intellectual disability were included in a randomised controlled trial employing a double cross-over design. The Snoezelen environment was compared with an ordinary environment)

i. Frankish P. Meeting the emotional needs of handicapped people: a psychodynamic approach. Journal of Mental Deficiency Research 1989; 33: 407-14 (Type IV evidence - case studies of 7 intellectually disabled children and adults)

4.8 Cognitive Therapy

4.8a. There is a body of evidence, based on single case studies, demonstrating the effectiveness of cognitive therapy for people with mild intellectual disability and borderline intelligence\textsuperscript{i,ii}.

These reports maintain the essential principles, components and procedures of cognitive therapy (setting an agenda, developing an awareness of the role of underlying beliefs in determining thought, establishing the relationship between thoughts, feelings of anxiety and behaviour, monitoring automatic thoughts, challenging maladaptive beliefs and developing adaptive automatic thoughts) but simplify them considerably to allow for the linguistic and intellectual deficits of clients. As yet there have been no controlled treatment trials investigating long-term outcome.

(Health gain notation - 2 “likely to be beneficial”)  

4.8b. Anger management training is an essentially cognitive therapy. The way in which an individual misconstrues a situation and their personal feelings of arousal is germane to the development of anger responses. The techniques help individuals appraise and reappraise situations for the extent to which they are in reality anger arousing, to identify their own emotions more accurately, to role-play adaptive ways of behaving in these situations and to practise them in real life\textsuperscript{i,ii,iii}.

One case-controlled study demonstrated a decrease in aggressive responding in anger management and control conditions with no significant differences between the conditions. A further controlled trial demonstrated significant reductions in expressed anger for subjects receiving anger management training which maintained up to one year\textsuperscript{iv}.

(Health gain notation – 2 “likely to be beneficial”)

London: Routledge, 1997  
(Type V evidence - expert opinion and review of clinical case studies)

(Type V evidence - expert opinion and presentation of over 50 case studies)

\textbf{i.}  Benson BA, Rice CJ, M iranti SV. Effects of anger management training with mentally retarded adults and group treatment. J ournal of Consulting and Clinical Psychology 1986; 54: 728-9  
(Type II evidence - although a controlled trial it lacks a no treatment control condition)

(Type V evidence - expert opinion and case studies)

(Type IV evidence - case controlled study)

(Type III evidence - an intervention study without randomisation comparing 25 individuals who completed anger management training with 19 individuals from a waiting list control. The interventions produced significant reductions in expressed anger which were maintained at 6 and 12 month follow-up)
5.1  Behavioural Disturbance

5.1a. 26.5% of people with an intellectual disability can be treated effectively, 47.1% fairly effectively, 23.5% with intermittent success and 2.9% cannot be treated.

Pharmacological procedures are the least effective interventions. Response contingent procedures are significantly more effective than other procedures.
(Health gain notation – 2 “likely to be beneficial”)

Internally maladaptive and socially disruptive behaviors can be treated more effectively than externally destructive behaviors.
(Health gain notation – 2 “likely to be beneficial”)

Undertaking functional analysis prior to treatment is significantly associated with treatment success.
(Health gain notation – 2 “likely to be beneficial”)

5.1b.  There is no good evidence that antipsychotic medication helps in managing behaviour, or harms people with intellectual disability. Further research is required.
(Health gain notation – 4 “unknown”)

(Type II evidence - systematic review and meta-analysis of 482 empirical studies (no randomised controlled trials) on treatment of problem behaviors of individuals with mental retardation)

(Type I evidence - systematic review of 500 citations with 3 sound randomised controlled trials)
5.1c. It is possible to reduce or even withdraw neuroleptics in severely and profoundly intellectually disabled adults\textsuperscript{a,ii,iii}.

At least 60% of patients on long-term treatment can eventually be managed without psychoactive medication. Transient behavioural deterioration during drug reduction may be prolonged (up to two years), but then returns to baseline without further pharmacological intervention.

Although 40% demonstrate persistent deterioration following drug withdrawal and require some type of psychoactive medication, very few will need to recommence neuroleptics. Those that do tend to be older and well controlled but receiving higher baseline doses of neuroleptics.

(Health gain notation – 3 “trade off between beneficial and adverse effects”)

5.1d. Using multiple assessment measures, Clomipramine was effective in reducing the frequency and intensity of one or more stereotyped behaviour, hyperactivity and irritability.

60% showed significant improvement, 30% had treatment limiting side effects and 10% showed no improvement.

(Health gain notation – 2 “likely to be beneficial”)

5.1e. There is limited evidence that Risperidone is effective in improving a range of behavioural disturbances in people with varying levels of intellectual disability. Further research is needed.

(Health gain notation – 4 “unknown”)

The evidence


(Type IV evidence - naturalistic study of 23 severely and profoundly intellectually disabled adult male patients undergoing slow “diagnostic” neuroleptic taper)


(Type III evidence - non-controlled study of 123 patients who had reduction of their antipsychotic medication)


(Type II evidence - randomised controlled trial of 67 subjects in whom antipsychotic medication was used to treat behavioural problems. A 25% reduction in dosage per month led to one third of patients successfully coming off the medication and a further 19% having the dose reduced by half. Environmental factors influenced success in drug reduction rather than subject characteristics)


(Type II evidence - double blind placebo controlled cross-over study involving 10 severely or profoundly intellectually disabled adults)


(Type IV evidence - retrospective audit of seventeen long term inpatients with mild to severe intellectual disability; did not use recognised audit tool)
5.1f Antidepressants with a specific 5-HT action should be used with caution in people with intellectual disability and autism. They may cause agitation, physical aggression and SIB and sleep disturbance related to the role of serotonin in autistic symptoms. (Health gain notation – 5 "unlikely to be beneficial")

In autistic adults Fluvoxamine, a selective serotonin reuptake inhibitor, results in a significant reduction in repetitive thoughts and behaviours, aggression and maladaptive behaviours compared to placebo. Fluvoxamine had a 53% response rate.

(Health gain notation – 2 "likely to be beneficial"

5.2 Self-Injurious Behaviour

5.2a There is growing evidence that serotonergic drugs are effective in the reduction of self-injurious (SIB) and aggressive behaviour in people with intellectual disability. Sertraline, a serotonin reuptake inhibitor, has been shown to be more effective (when measured by Clinical Global Impressions) and have fewer side effects than traditional neuroleptics. Further trials are recommended.

(Health gain notation – 4 "unknown")

The statements

The evidence


(Type IV evidence - case study of two patients)


(Type II evidence randomised controlled trial of 30 adults with autism. Mean dose of Fluvoxamine, 277 ±42mg, was compared to placebo. A significant improvement with Fluvoxamine was seen after 4 weeks. Robust study design with standardised rating scales and thorough statistical analysis)


(Type I evidence – systematic review showing a 35-75% reduction in self-injurious behaviour in adults with severe and profound intellectual disability after treatment with oral Naltrexone)


(Type II evidence – double-blind, placebo controlled crossover study of 8 men and 2 women with self-injurious behaviour treated with Naltrexone following plasma measurement of beta-endorphin. The study showed that an elevated beta-endorphin level indicated a good response to Naltrexone, suggesting that such individuals may be addicted to endogenous opiates released following self-injury)


(Type II evidence – double-blind randomised controlled trial of 8 adults with severe or profound intellectual disability treated with 50 mg and 100 mg of Naltrexone. Both doses significantly reduced self-injurious behaviour. Naltrexone showed no side effects at either dose including no effect on sleep pattern)


(Type III evidence - open trial, nine consecutively admitted adult intellectually disabled outpatients presenting with target behaviours of self-injury and/or aggression. Six were mildly or moderately mentally retarded by DSM-III-R criteria; five had comorbid autistic disorder)
5.2b. There is limited evidence that **Naltrexone** may be effective in reducing self-injurious behaviour (SIB) in adults with profound intellectual disability and that this benefit might be both durable to environmental changes and continue long after cessation of therapy. (Health gain notation – 4 “unknown”)

5.2c. There is limited evidence that **Risperidone** is effective in the management of aggressive and self-injurious behaviour (SIB) in people with moderate to profound intellectual disability. Side effects are mild and primarily those of sedation and restlessness. (Health gain notation – 4 “unknown”)

5.3 Attention-Deficit Hyperactivity Disorder

5.3a. **Amphetamine** is effective in the treatment of attention-deficit hyperactivity disorder (ADHD) in children.

It has been shown to reduce inattention, hyperactivity and other disruptive behavioural problems and tends to improve results in the Wechsler Intelligence Scale for Children. Measured benefits last at least one year and adverse effects are few and mild. (Health gain notation – 2 “likely to be beneficial”)

5.3b. There is limited evidence that both **Methylphenidate** and **Fenfluramine** may be effective treatments for attention-deficit hyperactivity disorder (ADHD) in children with intellectual disability or borderline IQ. (Methylphenidate: Health gain notation – 4 “unknown”)

**Fenfluramine is a controversial drug.** At higher doses (>1.5mg/kg/day) it causes drowsiness, dizziness and anorexia. There are reports of possible neurotoxicity with long-term depletion of serotonin in laboratory models. **FENFLURAMINE HAS BEEN WITHDRAWN IN THE UK DUE TO REPORTS OF LINKS WITH VALVULAR HEART DISEASE** (Fenfluramine: Health gain notation – 6 “likely to be harmful”)

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The statements

5.2b. There is limited evidence that **Naltrexone** may be effective in reducing self-injurious behaviour (SIB) in adults with profound intellectual disability and that this benefit might be both durable to environmental changes and continue long after cessation of therapy. (Health gain notation – 4 “unknown”)

5.2c. There is limited evidence that **Risperidone** is effective in the management of aggressive and self-injurious behaviour (SIB) in people with moderate to profound intellectual disability. Side effects are mild and primarily those of sedation and restlessness. (Health gain notation – 4 “unknown”)

The evidence


i. Aman MG, Kern RA, Osborne P, et al. Fenfluramine and methylphenidate in children with mental retardation and borderline IQ: clinical effects. American Journal of Mental Retardation 1997; 101(5):521-34 (Type II evidence - double blind placebo-controlled crossover study of M ethylphenidate (0.4 mg/kg/day) and different doses of Fenfluramine (1.0, 1.5, or 2.0 mg/day) in 35 children aged 5-14 years with mental retardation or borderline IQ and ADHD)
6a. The relationship between behavioural disturbance and forensic problems in people with intellectual disability is subtle. There is no doubt that many behaviour problems in people with severe and profound intellectual disability would be construed as offences in more able individuals.

One of the determining characteristics of an ‘offence’ is that the perpetrator is aware of behaviour that is socially sanctioned or censured. Even when someone with mild intellectual disability may understand the nature of the offence, the criminal justice response and the response of carers is diverse across cases and situations.

General methodological difficulties with work in this area are that offenders with intellectual disability are only mentioned as part of larger offender cohorts. Where studies are specifically directed towards offenders with intellectual disability, many studies are small in subject numbers.

6b. An important practical problem encountered when considering the population of individuals who offend is one of identifying those people with a intellectual disability and ensuring they are offered the additional safeguard of having an Appropriate Adult present during police interviews.

i. Clare I C H., Murphy G H. Working with offenders or alleged offenders with intellectual disabilities. In E Emerson, C Hatton, J Bromley and A Caine (eds.) Clinical Psychology and People with Intellectual Disabilities. Chichester: Wiley, 1998 (Type V evidence - expert opinion and review of some important cases)

ii. Swanson CK, Garwick GB. Treatment for low functioning sex offenders: group therapy and interagency co-ordination. Mental Retardation 1990; 28:155-61 (Type V evidence - expert opinion based on a description of a group therapy treatment service)

iii. Johnston SJ, Halstead S. Forensic issues in intellectual disability. Current Opinion in Psychiatry 2000; (in press) (Type V evidence - systematic review of studies over the previous year. Does not include a randomised control trial but reviews important issues and evidence relating to the current status and use of high security, medium security and community provision; issues of consent, capacity, competence, assessment and treatment effectiveness)

The statements

6c. It is generally considered that the prevalence rates for offenders with intellectual disability may be higher than those in the general population. This is especially true for sexual offences and arson.

6d. Well controlled studies have found prevalence rates for individuals with intellectual disability to be slightly higher in offender populations than in the general population. There is a vast predominance of males amongst offenders with intellectual disability.

The evidence

(Type III and V evidence - review of legislation, secure hospital provision and prison services up until 1973. Detailed review of statistics from 1950-1970 relating to criminals with a psychiatric diagnosis)

(Type IV evidence - review of treatment work with men who have committed sex offences. Treatments included plethysmographic assessment and electrical aversion; number of participants not noted)

(Type V evidence - expert opinion based on an extensive review of clinical cases. 12-13% of offenders in the New South Wales prison population were assessed as having intellectual disability)

(Type III evidence - a study of 72 arsonists examined before trial they found that 11% fell into the range of intellectual disability)

(Type III evidence - out of 57 adults and 47 juvenile arsonists they found intellectual disability to be the diagnosis in just over 10% of both groups)

i. MacEachron AE. Mentally retarded offenders: prevalence and characteristics. American Journal of Mental deficiency (American Journal on Mental Retardation) 1979; 84: 165-76
(Type III evidence - review of 3938 adult male offenders. Offending rates were only slightly higher than in the general population)

(Type IV evidence - review of 8 observational studies between 1975 & 1985 involving adults with intellectual disability in both the hospital and community settings)
The way in which intelligence is measured and the use of different population bases leads to a wide variability in prevalence statistics across studies. For these reasons there is little substantive evidence to support any link between the presence of intellectual impairment and a predisposition to criminal behaviour. Different studies report prevalence rates for intellectual disability within prison populations varying from around 2% to around 40%.

In-patient treatment has been provided for individuals who have shown offending behaviours of violence, fire-setting, sexual abuse and also for individuals who have offended and have concurrent intellectual disability and psychiatric illness. Reports on the short-term outcome for individual case studies have been encouraging. (Health gain notation – 2 "likely to be beneficial")

Study of longer term outcomes indicates that 84.2% of treated patients return to community based resources. Arsonists are over-represented in those with a poorer outcome.

The evidence


ii. Murphy GH, Clare ICH. MIETS: A service option for people with mild mental handicaps and challenging behaviour or psychiatric problems. Assessment, treatment and outcome for service users and service effectiveness. Mental Handicap Research 1991; 4: 180-206 (Type IV evidence - review of a series of cases)


6. Reviews of high security provision indicate that offenders with intellectual disability have the longest duration of stay and are the most difficult to discharge because of the lack of availability of suitable discharge resources. More recently, clinicians and researchers have begun to address the problem of treatment in the community for individuals who have offended. Without necessarily admitting clients for in-patient treatment, several reports have suggested the feasibility of such treatment (Health gain notation – 2 "likely to be beneficial")

(Type III evidence - review of referrals and admissions to 3 high security hospitals)

(Type III evidence - systematic review and analysis of the case registers of 3 high security hospitals over a 10 year period from 1986-95)

(Type IV evidence - detailed process study of 6 cases. No re-offending is reported 4 years following the initial conviction although the authors did not feel confident about re-offending data in one case)

(Type IV evidence - review of four case studies: no re-offending reported 3 years following initial conviction)

(Type IV evidence - a detailed process study of four cases. No reoffending reported four years following initial conviction)

(Type IV evidence - case study review of a series of cases)

(Type IV evidence - description of a problem solving intervention with 13 adult male sex offenders aged 17-43 years. Most subjects achieved more community access)
6. Characteristics and predisposing factors. Sex offenders with intellectual disability may have a high incidence of family psychopathology, low specificity for age and sex of the victim, psychosocial deprivation, behavioural disturbances at school, psychiatric illness, social naivety, poor ability to form normal sexual and personal relationships, poor impulse control and low self-esteem.

Experience of sexual and physical abuse in childhood is associated with offending in adulthood although it is neither a necessary or sufficient cause of adult offending including sexual offending.

Sex offenders with learning disability have a greater tendency to offend against male children and younger children.

(Type III evidence - review of 47 male patients referred for antisocial sexual behaviour)

(Type V evidence - expert opinion based on clinical evidence and experience)

(Type IV evidence - investigation into adults charged with offences and/or leaving custody. Only two subjects with a self-reported intellectual disability actually had an IQ below 70)

(Type V evidence - expert opinion based on clinical evidence)

(Type IV evidence - an analysis of the characteristics of 75 men who had allegedly perpetrated some form of sexual abuse)

(Type IV evidence - study comparing the abuse histories of 46 sexual offenders with 48 non-sexual offenders)

(Type IV evidence - study, by interview, clinical chart information, phallometric tests and self-administered questionnaire, of 991 male sexual offenders)
This document is a supplement to, not a substitute for, professional skills and experience. Users are advised to consult the supporting evidence for a consideration of all the implications of a recommendation.

6. Studies have found re-offending rates of untreated offenders of between 40 and 70% \(^{i, ii}\).

The risk of recidivism is highest during the year immediately following discharge \(^{iii}\).

A range of studies have found re-offending rates following treatment to be between 20 and 55% depending on the type of treatment and the offence \(^{iv}\).

Outcomes for individuals treated for 2 or more years would appear to be superior to outcomes for individuals treated for less than one year \(^{v, iv}\). (Health gain notation – 2 "likely to be beneficial")

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i. Scorzelli JF, Reinke-Scorzelli M. Mentally retarded offenders: a follow-up study. Rehabilitation Counselling Bulletin 1979; September: 70-73 (Type III evidence - review of 135 offenders with intellectual disability found that 68% had a prior history of arrest)


iv. Thomas DH, Singh T. Offenders referred to a learning disability service: a retrospective study from one county. British Journal of Learning Disabilities 1995; 23: 24-7 (Type IV evidence - 3 year follow-up of 20 offenders treated in a community based service: 50% of subjects re-offended and appeared before a court)

v. Brier N. Targeted treatment for adjudicated youths with learning disabilities: effects on recidivism. Journal of Learning Disabilities 1994; 27: 215-22 (Type III evidence - controlled group study comparing 73 offenders who completed the treatment requirements with 85 who did not and a further matched group of 34 untreated subjects. Over an average follow-up period of 20 months, the completors had a significantly lower recidivism rate (12%) relative to the non-completers (40%) and the matched controls (38%))

vi. Lindsay WR, Smith AHW. Responses to treatment for sex offenders with a learning disability: a comparison of men with one year and two year probation sentences. Journal of Intellectual Disabilities Research 1998; 42: 346-53 (Type III evidence - comparison of two groups of sex offenders, significant differences were found between the groups with the greater and most durable changes occurring in the group treated for a longer duration)
6. **Representation, consent and competence.**

People with an intellectual disability may be disadvantaged by the judicial process because of lack of appropriate support and appropriate legal representation from early stages in the process.\(i, ii, iii, iv, v\)

\[\begin{align*}
&ii. \ \text{Hayes SC. Prevalence of intellectual disability and local courts. *Journal of Intellectual and Developmental Disability* 1997; 22: 71-85 (Type IV evidence - review of 208 individuals appearing before 6 courts. The review emphasises the need for policies safeguarding the rights of people with intellectual disability in the criminal justice system)} \\
&iii. \ \text{Gudjonsson G, MacKeith J. Learning disability and the Police and Criminal Evidence Act 1984. Protection during investigative interviewing: a video recorded false confession to double murder. *Journal of Forensic Psychiatry* 1994; 5: 35-49 (Type IV evidence - careful case study illustrating the psychological processes causing an individual to confess falsely to a double murder)} \\
&iv. \ \text{Kebbell MR, Hatton C. People with mental retardation as witnesses in court: a review. *Mental Retardation* 1999; 37: 179-87 (Type I evidence - comprehensive review which concludes that in general people with intellectual disability can provide accurate accounts of evidence. Cross-examination methods may lead to memory distortions. Resulting errors could lead to false conviction or acquittal)} \\
&v. \ \text{Everington C, Fulero SM. Competence to confess: measuring understanding and suggestibility of defendants with mental retardation. *Mental Retardation* 1999; 37: 212-20 (Type III evidence - group comparison of individuals with and without intellectual disability. Participants with intellectual disability were significantly less able to comprehend their Miranda rights. They were also more likely to respond to suggestive questioning)}
\end{align*}\]
The frequency of most common medical conditions is similar in adults with intellectual disability and the general population. There is however an increased frequency of thyroid disorders, non-ischaemic heart disorders, and sensory (visual and hearing) impairment. \textsuperscript{(i), (ii)}.

Compared with non-Down syndrome adults with intellectual disability, people with Down syndrome have a significantly lower rate of hypertension. \textsuperscript{(i)}.

People with intellectual disability have lower levels of arthritis and back pain compared to the general population. \textsuperscript{(i)}.

People with intellectual disability have higher levels of impaired vision compared to the general population. \textsuperscript{(i)}.


People with intellectual disability have higher levels of impaired hearing (14.2%) compared to the general population (11.7%).

People with intellectual disability have fewer natural teeth (less than 20 including teeth which have been capped or filled) compared to the general population.

One third of children with mild, moderate and severe intellectual disability have high levels of poor oral hygiene, but this is no higher than the general population.

Children with borderline or mild intellectual disability have the poorest dental condition and worst dental care with the lowest level of restorative care (restorative index 43.9%). 22% do not brush daily and over 90% do not receive any help with toothbrushing from their parents or carers.

Oral health care guidelines for people with a learning disability are due for publication in February 2001.

People with intellectual disability are less likely to smoke (19.2% smokers / ex-smokers) compared to the general adult population (63.1% smokers / ex-smokers).

7.2 Hypothyroidism

Subclinical hypothyroidism* is present in one third of children and adults with Down syndrome. Clinically manifested hypothyroidism can be detected in 2.3% of children with Down syndrome.

*(high basal level TSH, low total or free T4)


**Oral Health Care Guidelines for People with a Learning Disability will be available from the Royal College of Surgeons (http://www.rceng.ac.uk) or the British Society for Disability and Oral Health (http://wwwbsdh.org.uk).**
7.2b. The presence of antithyroid antibody is higher amongst people with Down syndrome (18%) when compared with a control group of non-Down syndrome subjects (5.8%).

Hepatitis B and Autoimmune Thyroiditis

7.2c. The frequency of thyroiditis in people with Down syndrome who are also carriers of HBsAg is three times higher than in people with Down syndrome who are non-carriers. They should be regularly monitored for the development of thyroid disease. This does not apply to non-Down syndrome people with intellectual disability.

Screening for Thyroid Function

7.2d. People with Down syndrome should be screened annually for thyroid function. Antithyroid antibodies are rare in children under 8 years of age but titres subsequently increase with age.iii.

The evidence


iii. Rooney S, Walsh E. Prevalence of abnormal thyroid function tests in a Down’s syndrome population. Irish Journal of Medical Science 1997; 166(2): 80-2 (Type IV evidence - observational study of 36 subjects)
7.3 Epilepsy

7.3a. 14-24% of people with an intellectual disability are affected by epilepsy.

The frequency of life-time history of epilepsy ranges from 7-15% of people with mild to moderate intellectual disability, 45-67% of people with severe intellectual disability and 50-82% of people with profound intellectual disability. The prevalence also varies according to the age of patients and the aetiology of intellectual disability.

The cumulative incidence of epilepsy at 22 years of age is much higher among those intellectually disabled people who also have cerebral palsy (38%) compared to those who do not (15%).

7.3b. False positive and false negative diagnoses of epilepsy in people with intellectual disability are possible.

The statements

The evidence


iv. Shepherd C, Hoskins G. Epilepsy in school children with intellectual impairment in Sheffield: the size and nature of the problem and its implications in service provision. Journal of Mental Deficiency Research 1989; 33: 511-14 (Type IV evidence - a study of the prevalence and nature of epilepsy in all children between the ages of 5 and 16 years in Sheffield, UK)

v. Deb S. Epidemiology and treatment of epilepsy in patients who are mentally retarded. CNS Drugs 2000; 13(2): 117-28 (Type V evidence - expert opinion based on a review of observational, experimental and randomised controlled trials)

This document is a supplement to, not a substitute for, professional skills and experience. Users are advised to consult the supporting evidence for a consideration of all the implications of a recommendation.

### The statements

#### 7.3c. For people with intellectual disability multiple seizure types are common and up to three-quarters of patients remain refractory to treatment. Tonic-clonic seizures are the most common type of epilepsy in people with an intellectual disability (60%). Complex partial seizures are the second most common type (20%). Compared with the general population, seizure types such as absences (typical and atypical), myoclonus, tonic and atonic are more common among adults with an intellectual disability.

### The evidence

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

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

### Assessment

#### 7.3d. EEG remains difficult to perform in a proportion of people with an intellectual disability. EEG abnormality is detected in a high proportion of adults with an intellectual disability and epilepsy (>90%). In most cases EEG abnormality is non-specific such as excessive background slow wave. In 50% of cases the abnormality includes epileptiform changes in the EEG.

(Type IV evidence - cross sectional study of 100 adults with intellectual disability and epilepsy, randomly selected from hospital and community settings)

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

### Treatment and Care

#### 7.3e. Reducing polypharmacy use of antiepileptic drugs in institutionalised people with intellectual disability is associated with a reduced frequency of seizures and a reduced frequency of behavioural disorders and improved quality of life. Large scale studies including community based cohorts are required to confirm this apparent benefit.

(Health gain notation – 2 “likely to be beneficial”)

(Type IV evidence - prospective cohort study of 244 institutionalised patients)

(Type IV evidence - open prospective study of 44 institutionalised patients)

#### 7.3f. Phenobarbitone is no longer a drug of choice for people with intellectual disability. It can cause cognitive deficiency and serious behavioural disturbances.

(Health gain notation – 6 “likely to be ineffective or harmful”)

(Type V evidence – expert opinion based on a review of experimental and observational studies, but no randomised controlled trials)
This document is a supplement to, not a substitute for, professional skills and experience. Users are advised to consult the supporting evidence for a consideration of all the implications of a recommendation.

### 7.3a. Phenytoin
Phenytoin can not be recommended as a first line drug for treatment of epilepsy in people with intellectual disability due to its potentially severe adverse effects on the central nervous systemi. (Health gain notation – 6 "likely to be ineffective or harmful")

### 7.3b. Sodium Valproate
Sodium Valproate is a drug of first choice for a broad range of epileptic seizures in people with intellectual disability. These include: generalised tonic-clonic, tonic, clonic, atonic (drop attacks), myoclonic, absence and partial seizures with or without secondary generalisationi. (Health gain notation – 2 "likely to be beneficial")

CARBAMAZEPINE MAY MAKE ABSENCE, MYOCLOMNIC AND ATONIC (DROP ATTACKS) SEIZURES WORSE.

### 7.3c. Carbamazepine
Carbamazepine is a drug of first choice for a range of epileptic seizures in people with intellectual disability. These include partial seizures with or without secondary generalisation and generalised tonic-clonic seizures. (Health gain notation – 3 "trade-off between beneficial and adverse effects")

### 7.3d. Oxcarbazepine
Oxcarbazepine is similar to Carbamazepine in its indication, but has fewer adverse effects. It is useful in the treatment of epilepsy in people with intellectual disability including partial seizures with or without secondary generalisation and generalised tonic-clonic seizures. (Health gain notation – 2 "likely to be beneficial")

### 7.3e. Lamotrigine
Lamotrigine is an effective broad-spectrum antiepileptic treatment with minimal adverse effects in people with intellectual disability. It is effective in generalised tonic-clonic, partial seizures with or without secondary generalisation, Lennox-Gastaut and West Syndromei. (Health gain notation – 2 "likely to be beneficial")

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**Literature searches completed on 19.4.99**
7.3. Vigabatrin, Gabapentin, Topiramate and Tiagabine are effective as add-on therapies in the treatment of partial seizures in people with intellectual disability.
(Health gain notation – 2 “likely to be beneficial”)

The evidence

(Type III evidence - evaluation study in a group of patients with refractory epilepsy; 36 patients with intellectual disability and 75 patients with normal intelligence)

(Type III evidence - non-randomised trial of 26 children and adolescents with intellectual disability and 6 with normal intelligence, all suffering from refractory partial seizures)

(Type V evidence - expert opinion based on a review of experimental and observational studies)

(Type V evidence - expert opinion based on a review of experimental and observational studies)

(Type IV evidence - prevalence study of visual defects in 33 patients with epilepsy taking vigabatrin (31 assessed) and 16 unexposed control patients with epilepsy)

7.3m. Zonisamide is effective in the treatment of partial and generalised seizures in people with intellectual disability.
(Health gain notation - 2 “likely to be beneficial”)

The evidence

(Type III evidence - non-randomised controlled trial of 115 children with epilepsy (66 with intellectual disability and 49 with normal intelligence))
7.3a. Benzodiazepines including clobazam, nitrazepam and clonazepam can be used as short-term adjuncts in the treatment of epilepsy in people with intellectual disability. Long term use of benzodiazepines is not recommended due to the effects on tolerance and symptoms associated with withdrawal. (Health gain notation – 3 “trade off between beneficial and adverse effects”)

7.3b. Intranasal and buccal spray of Midazolam may have significant advantages over rectal diazepam as a rescue medication for prolonged recurrent seizures or seizures associated with hypoxia in people with intellectual disability. Intranasal midazolam is as effective as intravenous diazepam in the treatment of prolonged febrile seizures in children. Buccal midazolam is at least as effective as rectal diazepam in the acute treatment of children and adolescents. (Health gain notation – 2 “likely to be beneficial”)

7.3c. A substantial proportion of epileptic patients with an intellectual disability improve following neurosurgery with some becoming totally seizure free. (Health gain notation – 2 “likely to be beneficial”)


7.3e. Scheepers M, Scheepers B, C lough P. M idazolam via the intranasal route: an effective rescue medication for severe epilepsy in adults with a learning disability. Seizure 1998; 7: 509-12 (Type IV evidence – case studies of 3 patients with epilepsy)

7.3f. Lahat E, Goldman M, Barr J, Bistrüter T, Berkovitch M. Comparison of intranasal midazolam with intravenous diazepam for treating febrile seizures in children: prospective randomised study. British Medical Journal 2000; 321: 83-86 http://www.bmj.com/cgi/content/full/321/7253/83 [accessed 8.12.00] (Type II evidence – randomised controlled trial (intranasal midazolam or intravenous diazepam) of 47 children, aged 6 months to 5 years, with prolonged febrile seizures)

7.3g. Scott R C, Besag F M C, N eville B G R. Buccal midazolam and rectal diazepam for treatment of prolonged seizures in childhood and adolescence: a randomised trial. Lancet 1999; 353: 623-626 (Type II evidence – randomised controlled trial of young people, aged 5-19, treated with buccal midazolam (40 episodes in 14 patients) or rectal diazepam (39 episodes in 14 patients))

This document is a supplement to, not a substitute for, professional skills and experience. Users are advised to consult the supporting evidence for a consideration of all the implications of a recommendation.

# 8 DEMENTIA IN DOWN SYNDROME

## 8.1 Background

### 8.1a. The quoted prevalences of dementia in people with Down 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.


(Type V evidence – expert review of 14 studies)

### 8.1b. Between 31% and 78.5% of adults 65 years or older with intellectual disability but without Down syndrome show Alzheimer’s neuropathology.


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


(Type IV evidence – observational autopsy study of people with intellectual disability – 15 Down syndrome and 18 non-Down syndrome)


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

### 8.1c. Almost all adults over the age of 40 years with Down syndrome display Alzheimer’s neuropathology.


(Type IV evidence – review of case-reports including 398 cases altogether)
8.1d. Among people with intellectual disability, the only known risk factors for the development of Alzheimer’s disease are increasing age and Down syndromei,ii.

It is not clear what effect, if any, possible risk factors as seen in the general population – family history, low educational level, head trauma, cardio-vascular disease, stroke, diabetes, apolipoprotein E-4, major depressive episode – have on dementia in people with Down syndromei,ii,iii,iv.

8.2 Assessment

8.2a. The diagnosis of dementia in people with intellectual disability, especially in the early stages, is made difficult by the lack of reliable and standardised criteria and diagnostic procedures.

Neuropsychological testsi and informant-based questionnaires such as the Dementia Questionnaire for Persons with Mental Retardation need further evaluation before they could be accepted for day-to-day clinical assessmenti.ii.iii.iv.

(Health gain notation – 4 “unknown”)


ii. Evenhuis HM. Further evaluation of the Dementia Questionnaire for Persons with Mental Retardation (DMR). Journal of Intellectual Disability Research 1996; 40(4): 369-73 (Type IV evidence - 5 year longitudinal follow-up of 33 elderly institutionalised persons (aged 70 and over) and 45 institutionalised persons with Down syndrome (aged 35 and over) with no dementia in the diagnosis at initial evaluation)

iii. Deb S, Braganza J. Comparison of rating scales for the diagnosis of dementia in adults with Down’s syndrome. Journal of Intellectual Disability Research 1999; 43(5): 400-7 (Type IV evidence – a study of the Clinician’s diagnosis (ICD-10), the Dementia Questionnaire for Persons with Mental Retardation (DMR), the Dementia Scale for Down Syndrome (DSDS) and the Mini Mental State Examination (M M SE) in 62 adults with Down syndrome – 26 demented and 36 non-demented according to clinician’s diagnosis)
This document is a supplement to, not a substitute for, professional skills and experience. Users are advised to consult the supporting evidence for a consideration of all the implications of a recommendation.

9a. **Staff** are supporting clients with a range of challenging behaviours, including **aggression** and **sexually inappropriate behaviour** in community settings, and such staff are likely to be the victims of **assault**.

i. Harris, P. The nature and extent of aggressive behaviour amongst people with learning difficulties (mental handicap) in a single health district. *Journal of Intellectual Disability Research* 1993; 37: 221-42
(Type IV evidence - survey of the prevalence rates different categories of challenging behaviour including: aggression, self-injury, stereotyped and withdrawal behaviours. Study covered one health district and included 78 services. Based on base population data available, the overall prevalence rate of aggression was estimated at 17.6%)

(Type V evidence - expert committee analysis of the approaches of a number of services, service commissioners and providers to challenging behaviour, with a recommendation that models of care should centre around community-based residences)

(Type IV evidence - survey covering three geographical areas and including 53 social care and 40 health staff working in intellectual disability services, and control group (n=30) who did not. Study found 73% of former group had experienced workplace assault, compared with 7% of the latter group)

(Type IV evidence - survey of 81 social care staff and 15 health staff. The latter group were employed to provide medium security accommodation to sex offenders. 59% of the former group supported a client who had committed a sexual offence or displayed an offending type behaviour. The range of behaviours was similar to that displayed by clients supported by the former group)
Well trained and skilled staff are identified as essential in the provision of quality community services. Both health and social care staff have been found to be lacking knowledge about the basic defining features of an intellectual disability and their duty of care to intervene if clients put themselves or others at risk.

Literature searches completed on 31.12.99
This document is a supplement to, not a substitute for, professional skills and experience. Users are advised to consult the supporting evidence for a consideration of all the implications of a recommendation.

**The statements**

9d. Staff have been found to be **lacking knowledge, training** and **confidence** in managing challenging behaviour and sexually inappropriate behaviour.

9e. Staff behaviour has been shown to affect the occurrence or non-occurrence of challenging behaviour.

**The evidence**


(ii) Evidence - review of observational and experimental studies: staff behaviour maintains challenging behaviours by negative or positive reinforcement.


(Type IV evidence - survey of 70 staff supporting clients with an intellectual disability. Staff reported colleagues often reacted inappropriately to displays of challenging behaviour e.g. by showing fear and disgust)


(Type IV evidence: questionnaire examining the knowledge of 95 health and social care staff working in intellectual disability services about the factors important in managing challenging behaviour).


(Type IV evidence - survey of 50 social care staff. The minority had received training in prevention and management and levels of confidence varied widely)


(Type IV evidence - randomised controlled survey of 81 social care and 15 health staff supporting clients with a forensic or sexual offence background. Few had received formal training in dealing with prevention and management of offending type behaviour)
9f. Training may range from time-limited formal and informal training courses\(^i\)\(^ii\)\(^iii\) to ongoing input\(^iv\).

(Type V evidence - example of formal National Vocational Qualifications (NVQs) for staff working in intellectual disability services)

(Type IV evidence - review of some formal training courses available to staff working with clients with an intellectual disability)

(Type II evidence - randomised control study: the impact of a one day training course on the knowledge of 59 health and social care staff, compared to a control group of 73 untrained staff)

(Type III evidence - interventional study evaluating the impact of a psychologist providing continuing advice to staff working with a client who exhibited high rates of self-injury)

9g. A substantial number of staff have not received training\(^i\)\(^ii\) or report that it is inadequate to meet the needs of their job\(^i\).

(Type IV evidence - survey of 299 staff in six health districts in the West Midlands. 12% of managers and 17% of care staff had received induction training: 75% of care staff in private establishments, 32% of local authority care staff, 37% of voluntary sector care staff and 4% of health care staff had not attended any courses in the previous five years)

(Type IV evidence - review of formal training courses available to staff working with clients with an intellectual disability. Both staff and managers of social care services felt that the training that they had received was inadequate in relation to that required for their job)
This document is a supplement to, not a substitute for, professional skills and experience. Users are advised to consult the supporting evidence for a consideration of all the implications of a recommendation.

**The statements**

9. Some **training** has been found to be effective in increasing staff **knowledge** and impacting on **practice**. (Health gain notation - 2 "likely to be beneficial")

**The evidence**


(Type II evidence - randomised control study examining the impact of a one day training course on the knowledge of 59 health and social care staff, compared with a control group of 73 untrained staff. Training was found to significantly increase knowledge for up to one year)


(Type III evidence - intervention study of the impact of two types of training on staff (n=83) attitudes and knowledge. Those trained in non-aversive techniques showed increased understanding of complexity of causes for challenging behaviour and skills based treatment approaches)


(Type III evidence - intervention study over a 5 year period which found clinically significant changes in terms of reduction in use of restraint, medication and in rates of staff and client injury)


(Type III evidence - interventional study evaluating the impact of a psychologist giving ongoing consultation regarding functional assessment and adhering to behavioural principles to staff working with a client who exhibited high rates of self-injury. This led to a significant decrease in the client’s behaviour.)

9. **Training** has not always been found to be cost-effective or to have long-term benefits.


(Type V evidence - expert opinion based on a review of early literature in relation to effectiveness of staff training. Concludes that staff training alone is not an adequate solution to deficits in staff performance)


(Type V evidence - expert opinion based on a literature review, concluding that staff training alone is not a sufficiently powerful factor to achieve enduring change)
Evaluating the effectiveness of staff training is difficult. The outcome of training can be affected by a number of factors; the social, organisational and political context that the staff work in (e.g. if changes in staff attitudes, knowledge and behaviour are not supported in the work environment they are unlikely to be maintained over time); staff characteristics (e.g. experience, skills and knowledge); the perceived quality, relevance and applicability of the training itself.

It is important to establish clearly the type and nature of the training needs of the service in question and to establish which goals the training is designed to meet and which outcome measures will be used to evaluate effectiveness. Five different types of outcome measures used in this field are outlined.

**Subjective** i.e. what staff report about the usefulness, validity, and quality of the training;

**Cognitive** i.e. knowledge gain;

**Behavioural** i.e. the impact of training on behaviour;

**Client centred** i.e. the impact on clients;

**Organisational** i.e. the impact on the work organisation.

There is currently insufficient evidence to unequivocally establish the effectiveness of staff training alone in improving staff practice in managing challenging behaviour.

(Health gain notation - 4 "unknown")
This document is a supplement to, not a substitute for, professional skills and experience. Users are advised to consult the supporting evidence for a consideration of all the implications of a recommendation.

The term Carer is taken to mean informal caregivers, rather than those providing care in the formal sector on an organised and paid basis.

The ‘hard evidence’ base for carers specifically in the area of intellectual disability is limited. However, it is clear that carers and the people they care for are individuals with a vast range of preferences and needs. Just as one carer may find caring a stressful experience, another may find it enriching and rewarding. It is difficult to draw conclusions concerning interventions that work. The main message from the many research publications is the need for flexibility, i.e. the service system (health, social services and voluntary) should be ‘carer sensitive’. Health service providers should listen and respond to the expressed preferences of the people to whom they are providing services. A related chapter, covering the evidence for supporting carers in general, is available in the Healthy Living Bulletin (http://hebw.uwcm.ac.uk/healthyliving/chapter6.html, accessed 8.12.00).

101 Epidemiology and Background

101a. In the 1998 Welsh Health Survey, one in thirteen (7.5%) of adults reported that they were carers and one in ten of those cared for had intellectual disability.

102 Gain and Strain in Caregiving

102a. Informal carers of learning disabled adults reported 40% more limiting health disorders than the general population, with depression almost four times more common among female carers (standard morbidity prevalence ratio for depression = 3.7, 95% CI, 2.4-5.7).
In a large Dutch study, the significant predictors of caregiving burden were time demands, child physical health, child adaptation skills, (less) service use and parents (lower) educational level. Service needs of parents varied according to the age range of the child. The major needs mentioned were:

- Age 0-9: Information and help with developmental issues, medical/dietician/speech therapy advice, in-home respite;
- Age 10-19: Information and help with development and future residential living, leisure activity and respite care, dental help;
- Adults: Information and concrete help re future out-of-home placements, information and advice from physicians.

The presence of significant behaviour problems is more important than disability type in determining most forms of parental stress, and predicts mental health services use. Parents of children with intellectual disability worried most about providing ongoing care into adulthood. Single mothers were not more stressed but used more services than 2-parent families.

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**The statements**

10.2b. In a large Dutch study, the significant predictors of caregiving burden were time demands, child physical health, child adaption skills, (less) service use and parents (lower) educational level. Service needs of parents varied according to the age range of the child. The major needs mentioned were:

- Age 0-9: Information and help with developmental issues, medical/dietician/speech therapy advice, in-home respite;
- Age 10-19: Information and help with development and future residential living, leisure activity and respite care, dental help;
- Adults: Information and concrete help re future out-of-home placements, information and advice from physicians.

10.2c. The presence of significant behaviour problems is more important than disability type in determining most forms of parental stress, and predicts mental health services use. Parents of children with intellectual disability worried most about providing ongoing care into adulthood. Single mothers were not more stressed but used more services than 2-parent families.

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**The evidence**


(Type IV evidence – population-based study in the Netherlands of 2,573 parents of children with intellectual disability living at home – A 77% response rate to the distribution of 3327 questionnaires)

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(Type IV evidence – study, in the U.S., of mothers and fathers of 231 children (aged 6-18 years) with intellectual disability with (n=46) or without (n=46) behaviour problems, or chronic illness, and a nondisabled behavior-problems sample (n=66). No details were given as to how the sample was recruited)
This document is a supplement to, not a substitute for, professional skills and experience. Users are advised to consult the supporting evidence for a consideration of all the implications of a recommendation.

**The statements**

10.2d. Caregivers report the existence of pervasive rewards and gratifications, as well as stresses, as part of the caregiving experience, and these stem from various sources.

The collection of six predictors of burden and satisfaction findings indicate that greater support from the adult child to the caregiver resulted in greater satisfaction and less burden. Adaptive and maladaptive behaviours and caregiving assistance all predicted caregiving satisfaction but only maladaptive behaviours predicted caregiving burden. The study also found that increased levels of support by the caregiver to the adult with intellectual disability was associated with greater caregiving satisfaction.

In contrast, another study found that neither help given, nor help received demonstrated significant effects on either positive well-being, negative well-being or subjective burden.

10.2e. The marital status of the caregiver, the level of retardation, frequency of maladaptive behaviours, and health status of the adult family member with mental retardation; and the number and level of services needed were factors in the stress experienced by families.

10.2f. Parental responses indicate that caretakers of adult children with intellectual disability are worried and feel responsible for their care regardless of residential placement. There were no statistically significant differences between carers of adult children living in-home and out-of-home in terms of social burdens, gratifications (eg intimacy), emotional burdens or objective burdens.

**The evidence**


ii. Heller T, Miller AB, Factor A. Adults with mental retardation as supports to their parents: effects on parental caregiving appraisal. *Mental Retardation* 1997; 35(5): 338-346


(The above studies used volunteer rather than representative samples, and cross-sectional rather than longitudinal study designs. Studies ii and iii were based in the United States)
10.3 Support and Respite Care

10.3a. The leading unmet needs reported by informal carers were for financial help, long-term social support, respite care and housing adaptations.

10.3b. Younger caregivers are more predisposed toward seeking outside help and have higher expectations of the service system than older caregivers. Overall there were no differences in the number of support services received. However, younger caregivers reported significantly more unmet service needs and rated significantly more of them as a critical or an emergency need. Older caregivers were more likely to seek spiritual support and the younger caregivers more apt to mobilize their families to acquire and accept help.

10.3c. From a longitudinal study of older mothers caring for an adult with intellectual disability, the authors made the following conclusions:

- Support groups were not beneficial although some groups may provide a forum for social contact and information exchange
- Mothers whose friendship networks were composed largely of close friends who were also coping with a relative with disabilities, reported greater increases in burden than those with more diverse networks
- Mothers who were pessimistic about their son’s or daughter’s future had increasing levels of burden and depressive symptoms over time. Service providers could encourage these parents to make arrangements to help them reduce their fears and pessimism


(Type IV evidence - cross-sectional study based on the Leicestershire Learning Disabilities Register to describe the prevalence of disabilities, felt needs, and use of services for 2,117 learning disabled adults, and to compare outcomes of morbidity, stress, and satisfaction among 982 informal and residential carers)


(Type IV evidence - interview data from a convenience sample of 105 families of adults (aged 20+) with intellectual disability living in Minnesota, US)


(Type IV evidence - longitudinal study, over three years, of volunteer mothers (aged 55 years and older) of adults with mental illness (n=73) or intellectual disability (n=288) living in the US)
Interview data showed that support services for adult offspring were positively received by parents in the way they interrupted, what would otherwise be, a continuous cycle of care. However, for some parents the nature of the provision was such that support services heightened rather than moderated their sense of living a restricted lifestyle. The authors noted that the challenge for service providers and planners is to find innovative responses to these parents by allowing a greater flexibility of provision.

The paradox is that, greater support could lead to an increased demand for residential provision, as parents sought to emulate their non-carer peers.

From a systematic review of the literature, the major recommendations to support informal carers of people with intellectual disability and challenging behaviour were that:

- stress management techniques were effective for use both routinely and at times of high arousal
- respite care services which genuinely provide respite and are able to offer support both on a planned and crisis basis are valuable
- information should be provided (eg regarding the child’s disability, challenging behaviour, support services and welfare benefits)

(Health gain notation - 2 "likely to be beneficial")
Community Support Teams (CST) for people with intellectual disability and challenging behaviour report that they serve the majority of their clients (at least two thirds) with some degree of success. However, from an analysis of recent cases, the majority of cases successfully closed were closed due to such factors as the acceptance of recommendations or the person being referred on to another service after the completion of assessment rather than reduction in challenging behaviour (which accounted for only 36% of closures). It was recommended that, due to the potential bias of positive reporting from Teams, the actual rather than reported outcomes of CST cases should also be examined.

One small study in the UK suggested that carers were satisfied with the Adult Learning Disability Community Support Team (CST). The most popular response was that a mild improvement in the referred problem (38%) or in life-quality (48%) had resulted from the referral. Areas of improvement suggested by carers included the provision of more information regarding the team’s role and more written information for referred cases.

A three year study found that adults with intellectual disability were more likely to move away from the parental home if their mothers were older and in poorer health. Following the move to out-of-home, 93.6% of the sample had at least weekly contact with the mother and 48.4% of mothers continued to provide help with at least one caregiving task.

The evidence


10.3. A review of respite services for people with intellectual disability concluded that, although quantifiable results from research studies are limited, expressed satisfaction with respite and felt relief are sufficient criteria to justify the need. However it is generally considered that respite can influence the quality of life of carers (and users of the services) in both negative and positive ways. To provide a benefit, services must be flexible and responsive to users so that they feel confidence in the care provided.

(Health gain notation - 4 "unknown")

10.3i. In a questionnaire study of families of children with an intellectual disability, respite care was generally perceived as an inadequate service, although 88% of respondents had no knowledge of the existence of respite-care services.

Mothers found a pilot summer respite scheme for children with intellectual disability (aged 6-18 years) very useful and expressed felt needs for substantially more such provision both during the summer and at other times.

(Health gain notation - 4 "unknown")


(Type V evidence - expert summary from a non-systematic review of the literature)


(Type IV evidence - results of interviews with mothers of 18 children who attended the scheme in the Medway, U K)
This document is a supplement to, not a substitute for, professional skills and experience. Users are advised to consult the supporting evidence for a consideration of all the implications of a recommendation.

The statements

10.3. A review of research approaches and service design for people with intellectual disability and their carers noted the following research gaps in the literature:

- A dearth of longitudinal studies and therefore deficits in evidence about caring and adaptation over the life course, over the course of disability, and how carers manage service transitions
- A failure to fully involve carers and families in setting the research agenda
- A concentration on the perspectives of mothers and caregiving dyads (e.g., mother-child, carer-professional) to the exclusion of the broader family relationships
- The lack of a balanced examination of stress, uplift and effective coping in families (i.e., considering both the strain and gain of caregiving)
- A failure to include both user and carer views in research. The chances of obtaining the views of people with intellectual disability (including profound disability) should be maximised, for example via generation of research questions by users and differing communication techniques. Improved outcomes for carers should be accompanied by gains for their dependents.

The evidence

   (Type IV evidence – descriptive systematic review of the literature)
104 Carers Entitlements

104a. The Carers (Recognition and Services) Act 1995 gives people who provide or intend to provide “a substantial amount of care on a regular basis” the right to request an assessment from social services. The implementation of this Act is patchy ii,iii. Carers are not always informed of their rights (53% in one survey) ii and many Social Services staff are unclear about entitlement and lack relevant training iii. Assessments are not always carried out. Some carers are offered very sensitive and practical support, others almost nothing. However, when assessments are undertaken, carers report satisfaction both with the process and the results ii,iii. Carers should be entitled to expect at least an annual discussion of what they need, what is available, the help they are receiving and the care provided iv.

i. Carers (Recognition and Services Act) 1995 (c.12) (Type V evidence – influential report)

ii. Carers National Association. Still battling? The Carers Act One Year On. London: CNA, 1997 (Type IV evidence – a questionnaire survey (1655 replies) from 12,000 members of the Carers National Association) and 50 follow-up interviews)


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