

CLINICAL FOCUS – PRIMARY CARE

Intellectual disability – the essentials for GPs

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Abstract

In this article we introduce the key features and health considerations associated with intellectual disability. Although roughly 1% of the population has an intellectual disability, their needs are frequently overlooked, both in medical training and in clinical practice. This contributes to worse clinical outcomes and excess mortality. General Practitioners have an important role in the co-ordination and delivery of high-quality care to this group.

Key words

(MeSH) Intellectual disability; delivery of healthcare; primary healthcare

Introduction

Intellectual disability is a lifelong condition that affects learning, communication, and a variety of adaptive daily living skills.¹ Intellectual disability is synonymous with learning disability, the term that tends to be used in UK clinical services. An older term, mental retardation, is now seen as stigmatising and has largely been abandoned.

Intellectual (or learning) disability is sometimes confused with learning *difficulty*, which is used to describe specific disorders that can interfere with learning (such as dyslexia or dyspraxia), without significant global impairment of intelligence. The other major qualification for intellectual disability is that the onset must be within the developmental period (i.e. <18 years). Thus, intellectual disability is distinguished from cognitive impairment secondary to acquired brain injury (>18 years), severe mental illness, and neurodegenerative disorders such as dementia.

Box 1 – Criteria for diagnosis of intellectual disability

Criterion	Notes
Significant impairment in cognitive function	Measured by formal IQ testing e.g. using the WAIS. Full-scale IQ < 70 is necessary though not sufficient for diagnosis
Significant impairment in adaptive function	Assessed indirectly (via third-party reports) and directly (by performance on standardised tests)
Onset in developmental period	Although milder forms are not always recognised in early life, there must be a history of problems extending back to childhood

WAIS – Wechsler Adult Intelligence Scale

The presentation of intellectual disability varies substantially. Intellectual disability is often sub-classified into mild, moderate, severe, profound (box 2). These classifications, based on recorded intelligence quotient (IQ), have been criticised as being somewhat arbitrary and provide little detail about individual strengths and the supports that a person may need.

Box 2 – Classification of intellectual disability

Degree of intellectual disability	IQ range	Possible presentation
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Mild (85% cases)	50-70	Conversational language. Struggle academically but usually able to read, write, count. Usually live independently and may work. Require support with more complex demands and at times of stress.
Moderate (10% cases)	35-49	Language ability varies and understanding may be over-estimated. Limited achievement in school. Likely to need support in ADLs. Supervised practical work may be possible.
Severe (4% cases)	20-35	Single words or no speech. May communicate with 'objects of reference', by gestures or expression. Can learn routines. Require assistance for basic tasks and self-care. Highly-supported accommodation.
Profound (1% cases)	<20	Require intensive support. High rates of medical co-morbidity (e.g. epilepsy) and physical disability that need ongoing nursing and therapy.

ADLs – activities of daily living

Over the past generation there has been a radical shift in attitudes towards people with intellectual disability both within the medical community and in society at large. This has been reflected in how people with intellectual disability live; large residential institutions have closed and community living is now the norm. People with learning disability may live alone, with family, in supported placements, or small residential homes.

It is estimated that about 1% of the population has an intellectual disability.² Causes vary widely and in many cases the cause is unknown (Box 3). As genetic testing becomes more powerful and less expensive, previously undiscovered genetic contributions to intellectual disability will increasingly be recognised. People with more severe intellectual disability, or a clear syndromic cause, will probably be diagnosed in childhood. Those with more mild degrees of intellectual disability may not be formally recognised as such, although there will often have been a history of poor educational attainment and other difficulties (e.g. in social domains).

Box 3 – causes of intellectual disability

Idiopathic

Unknown cause, may cluster in families

Genetic / chromosomal

Chromosomal abnormalities – e.g. Down's syndrome

Gene mutations – e.g. Fragile X syndrome

Antenatal

Maternal TORCH infection

Teratogens (alcohol, illicit drugs, valproate)

Perinatal

Hypoxia

Metabolic – hypoglycaemia, severe jaundice

Infancy / childhood

Biological – CNS infection or injury

Psychosocial – abuse and neglect

TORCH – toxoplasmosis, other, rubella, cytomegalovirus, herpes

There are few validated brief screening tests for intellectual disability, although certain indicators might suggest its presence. Asking someone about their early development (special schooling, extra support in classes, exams taken and results) and current functioning (e.g. if they can tell the time or read a newspaper, how they manage at home, if they can travel to new places independently, and their ability to budget and manage money) can provide pointers. Probing answers and asking for examples will be important to avoid gaining a false impression due to acquiescence, a tendency to answer with simple 'yes/no' responses, and attempts to minimise difficulties. More formal assessment can be carried out by the local multi-disciplinary specialist learning disability team. A full assessment is likely to take several sessions to complete and usually requires detailed collateral information including a thorough family and developmental history. It is worthwhile contacting specialist services in advance of a referral as processes and criteria can vary locally. It is also worth noting that in order for the results of an assessment to be valid it is best performed outside times of acute illness or in the context of significant change in life circumstances.

Diagnosing intellectual disability is important. As we will describe, people with intellectual disability have a range of additional health needs and suffer inequities across their use of healthcare. Having a diagnosis can guide both population and individual efforts to proactively manage health and inform certain adjustments to ensure access to appropriate care. Having a diagnosis can also provide access to other services and opportunities for people with intellectual disabilities.

People with intellectual disability access mainstream services for most of their health needs. Community Learning Disability Teams are locality-based multi-disciplinary teams that provide specialist healthcare and support (such as psychiatrists specialising in intellectual disability, learning disability nurses, and allied health professionals). Composition and governance of these services varies greatly; in some cases an NHS Mental health Trust or Community Healthcare Trust will lead, others are run as partnerships between healthcare and council Local Authorities. A recent development has been delivery of services by third sector providers and charities. Knowing what service is available locally and how to contact them is important for those working in primary care.

Health of people with intellectual disability

People with intellectual disability are more likely to have physical and mental health problems than the general population. This is often associated with the underlying cause of the intellectual disability but other reasons for the excess morbidity are modifiable and related to lifestyle factors (diet, exercise) and the social disadvantages that are prevalent in this group (e.g. poverty, isolation, and loneliness).

The presentation of ill-health in people with intellectual disability may be atypical because of poor health literacy, communication difficulties, and the complicating effect of multi-morbidity. Presentations, even of acute illness, can be non-specific (e.g. distress, sleep disturbance, and behaviour that challenges) and people may seek help relatively late in the course of an illness. Good communication and clinical skills, and an ability to think laterally are important in consultations. Change in usual behaviour or functioning is always significant.

In general terms, prevention and management of health conditions follows guidelines recommended for people without intellectual disability; NICE guidelines, for example, apply equally to this group. Some of the more common presentations in primary care are discussed below.

Respiratory disease is the most common cause of death in people with intellectual disability who have significantly higher rates of aspiration pneumonia (often linked with dysphagia or reflux disease) and inhalation of foreign objects. People with intellectual disability are eligible for a seasonal 'flu vaccination, although there is sometimes confusion about this and uptake has been poor (https://www.gov.uk/government/uploads/system/uploads/attachment_data/file/649902/supporting_people_with_learning_disabilities_get_flu_injection.pdf).

Gastro-intestinal problems are common. There is an increased incidence of *H. pylori* infection (and re-infection), particularly amongst those who live in shared accommodation or attend congregate activities or day centres. Gastro-oesophageal reflux disease may affect up to half of people with intellectual disability yet is frequently overlooked. Chronic constipation due to the combined effects of medication, physical inactivity, and a low-fibre diet is very common. Constipation needs to be actively considered as it can present with minimal signs and symptoms but may progress to intestinal obstruction and death (<https://www.gov.uk/government/publications/reasonable-adjustments-for-people-with-learning-disabilities/constipation> and <http://www.bmj.com/content/340/bmj.c222>).

The prevalence of epilepsy in people with intellectual disability is approximately 20 times higher than in the non-intellectually disabled population. Furthermore, the prognosis is worse, with higher rates of complex seizure types, treatment-resistance and sudden unexpected death in epilepsy (SUDEP). Where seizures are frequent, the effect of aura and post-ictal phases needs to be considered in the evaluation of behaviour change. Secondary care services for people with intellectual disability and epilepsy may be provided by neurologists or in some cases specialist psychiatrists.

With the increases in life-expectancy of people with intellectual disability witnessed over the past several decades, the incidence of age-associated diseases such as cancer is rising. The pattern of cancer types seems to differ in people with intellectual disability compared with the general population, with a greater proportion of gastro-intestinal malignancies. People with Down syndrome have a unique pattern and increased risk of leukaemia but decreased risk of most solid tumour types; as yet this association is unexplained. Uptake of cancer screening programmes by people with intellectual disability is generally poor and needs to be improved (<https://www.nationalelfservice.net/publication-types/qualitative/practical-recommendations-to-improve-uptake-in-cancer-screening-services-by-people-with-learning-disabilities/>).

There is a higher prevalence of physical disability including cerebral palsy, spinal problems and mobility issues, particularly in those with severe-profound intellectual disability. Contractures, poorly fitted adaptations and pressure sores are important concerns in this group.

Sensory impairments are common in people with intellectual disability, and often relatively easy to rectify. Up to 40% of people with intellectual disability have vision. Hearing impairments are equally as common and can compound communication difficulties, especially if aids are not used.

Attention should be paid to oral hygiene and regular dental review. People with intellectual disability are more likely to have filled teeth and require extractions. Poorly fitting dentures can cause pain along with problems with eating and speaking. Referral to specialist dental services might be more appropriate for those with complex needs or who require oral or intravenous sedation to tolerate dental procedures.

People with intellectual disability commonly face difficulties in expressing their sexuality and may experience opposition to sexual activity. Education and awareness is vital to enable people with intellectual disability (who have capacity to establish intimate relationships) to do so in a safe way, and can reduce the risk of unwanted pregnancy or sexually-transmitted infection. Sexual and reproductive healthcare must be provided without discrimination.

It pays to be mindful that genetic conditions are associated with increased risk of particular physical and mental illnesses. Down's syndrome (trisomy 21) is the most common genetic cause of intellectual disability and has many associated co-morbidities (box 4). Useful further information on less common genetic conditions is available at <https://rarediseases.info.nih.gov/diseases>.

Box 4 – medical associations with Down's syndrome

Condition	Notes
Dementia	Alzheimer's type. Early onset (40s onwards). Atypical presentation (early frontal symptoms e.g. behaviour change)
Congenital heart disease	Up to 50% affected. Most common are septal defects. Advances in surgical treatment have led to drastically improved life expectancy
Leukaemia	Children significantly more likely to develop megakaryoblastic leukaemia and acute lymphoblastic leukaemia
Thyroid disorder	Hypothyroidism may be congenital or acquired
Gastro-oesophageal reflux disease	One of a range of gastro-intestinal disorders that are common in Down's syndrome
Coeliac disease	Prevalence may be up to 10%
Hearing loss	Conductive type, often caused by 'glue-ear'
Vision problems	Reduced detail (visual acuity) persists even where refractive errors (short- or long-sightedness) are corrected. Older are prone to cataracts

Neurodevelopmental conditions

The prevalence of autism spectrum disorder (ASD) and attention-deficit hyperactivity disorder (ADHD) are both highly heritable neurodevelopmental disorders that are common in people with intellectual disability.

ASD is characterized by impaired social and interaction, and repetitive behaviours, preference for routine, and highly-focused and sometimes obscure interests. Onset is typically before three years of age, although in common with intellectual disability itself, those with a more mild degree may not be formally diagnosed. People with autism spectrum disorder can have a wide range of intellectual ability.

People with ASD commonly experience difficulty with cognitive and behavioural flexibility, altered sensory sensitivities, and emotional regulation. They have higher rates of mental health problems, particularly anxiety disorders, and sleeping and eating issues. Following the Autism Act of 2009, each area should have a clear and consistent pathway for ASD assessment and diagnosis. However some of the recently-established teams that provide this service routinely exclude people with intellectual disability; in these cases the local Community Learning Disability Team may take responsibility.

ADHD is characterized by core symptoms of hyperactivity, impulsivity and inattention. Although commonly regarded as a disorder of childhood, the disorder can often persist beyond the age of 18. ADHD can be particularly difficult to diagnose in people with intellectual disability where norms of behavior and function are difficult to determine and where many are already supported in activities where symptoms would traditionally be more apparent. Benchmarking against people of a similar developmental level is necessary.

Treatment with stimulants (such as methylphenidate) is usually considered first line treatment for ADHD in adults that is associated with moderate-severe impairment. Pharmacotherapy should only be started under the guidance of a specialist as part of a comprehensive treatment programme that considers psychological, behavioural and educational needs. Prescribing and ongoing monitoring are dictated by drug type and should be carried out under locally-agreed shared care arrangements.

Mental health

Most mental health disorders are more common in people with intellectual disability than in the general population but they can be difficult to diagnose owing to altered expression of symptoms, especially in people with more severe intellectual disability who have limited verbal communication. There are modified diagnostic criteria and rating scales/checklists available which often place emphasis on observable or biological signs rather than subjective personal accounts (e.g. the Diagnostic Criteria for psychiatric disorders for adults with Learning Disabilities, DC-LD, based on the ICD-10).

Physical signs of anxiety (e.g. rapid breathing, muscle tension, tremor, and motor agitation) can be observed in people with intellectual disability, but psychological symptoms might be harder to elicit.

Depression in people with mild intellectual disability is diagnosed as in the general population. In those with severe intellectual disability physical signs such as weight loss, change in sleep pattern, or social withdrawal tend to be more prominent. There might also be atypical indicators such as self-injury or aggression, uncharacteristic incontinence or screaming. Poor sleep, motor hyperactivity, sexual disinhibition and aggression can signal mania.

Psychosis is three times more common in people with intellectual disability but difficult to recognise when people are less able to communicate complex internal experience and phenomena. People with intellectual disability can experience the full range of psychotic symptoms but delusions will tend to be less complex. There is also a risk of misattributing developmentally-normal behaviours to mental illness. For example, self-talk or having imaginary friends is common in some people with intellectual disability and does not necessarily indicate a response to hallucinations.

Intellectual disability seems to be an independent risk factor for the development of dementia, which may develop at an earlier age than in the general population. People with Down syndrome are at significantly increased risk of developing Alzheimer's disease; by the age of 60 years up to half of people will warrant a diagnosis. Cognitive decline, especially in

its early stages, can easily be missed in this group due to pre-existing deficits and unclear baseline level of skills and functioning. After reversible causes of decline have been excluded (e.g. hypothyroidism), consider dementia if there are persistent changes in the person's behaviour, personality, or a loss of skills and need for more prompting.

Management of mental disorders in people with intellectual disability parallels management in the general population, although the evidence base for many interventions is less robust. Where pharmacological treatment is used, be aware that people with intellectual disability have potential for greater sensitivity to side effects and idiosyncratic responses to medication. The mantra 'start low and go slow' is often observed. Psychological therapies should be available for people with intellectual disability (where appropriate) although are likely to need adaptation in order to reflect limitations in communication and understanding. Mainstream adult Improving Access to Psychological Therapies (IAPT) services should be able to provide therapy to people with mild-moderate intellectual disability.

Challenging behaviour

People with intellectual disability, or more likely their carers, may consult primary care due to the presence of problem behaviours. These behaviours, also known as 'challenging behaviours' are culturally abnormal behaviours of an intensity, frequency or duration that threatens the physical safety of the person or others, or which is likely to seriously limit use or deny access to ordinary community facilities.³ Challenging behaviours encompass a range of presentations including self-injury, aggression, inappropriate sexual behaviours, and property destruction, amongst others.

A significant minority of people with intellectual disability display challenging behaviour. It is associated with individual factors (higher rates in young adulthood, boys/men, those with autism spectrum disorder, and those with more severe communication deficits) as well as systemic factors such as environments that are poorly-organised, lacking in suitable stimulation, or unable to respond well to an individual's needs. If not managed appropriately there is risk of harm to the person or those around them, including through the use of restrictive practices (such as mechanical restraints or segregation). If persistent or severe, the

behaviours may result in a requirement to change accommodation or even admission to hospital.

It is important to recognise that challenging behaviour is a description and not a diagnosis. Challenging behaviour can also be an expression of physical illness or of altered psychological states that require intervention. It may serve a function for the individual, for example, producing sensory stimulation, attracting attention, avoiding demands, or gaining access to a desired item. Given the wide list and inter-dependence of contributing factors a holistic bio-psycho-social approach should be adopted.

The complexity of challenging behaviour presentations often warrants referral to the local Community Learning Disability Team (case scenario 1). The role of the General Practitioner is likely to include initial information-gathering and investigation of possible physical causes (e.g. untreated pain, acute infection) and to consider any systemic factors that may be contributing (e.g. change in environment or care givers). Antipsychotic drugs, recommended only when alternative forms of intervention have been ineffective or where the risks are severe, should be instituted by a specialist and kept under regular review.⁴ Some community learning disability teams have an Intensive Support Team which people can be referred to when there is acute risk and the prospect of hospital admission.

‘Transforming Care’ is the current UK Government policy to ensure the best possible care for people with intellectual disability. A major focus of the programme is improving and expanding community provision and returning those people who are currently in psychiatric hospital back to their area. This cohort often present with highly complex needs and challenging or offending behaviours and it is vital that General Practitioners are able to work closely with specialist services and social care providers.

Case scenario 1

Joseph is a 29 year old man with severe intellectual disability, autism spectrum disorder, and history of anxiety. He resides at home with his aging mother, who helps him with most of his

activities of daily living. His verbal communication is limited to a few words and he generally makes his needs and wishes known by pointing at objects.

When his mother required hospitalisation for a sudden illness, Joseph moved to emergency respite accommodation. Since moving, staff report frequent challenging behaviour with loud vocalisations and head banging. The respite staff act on Joseph's obvious distress by contacting his GP who confirms this represents a significant change in behaviour. Thankfully Joseph's GP has a good working knowledge of intellectual disability. She uses her communication skills to facilitate Joseph's co-operation with a full physical examination. As no obvious physical cause was discovered, Joseph's case was discussed with the Community Learning Disability Team.

A multi-disciplinary assessment (including the psychiatrist, psychologist, and speech and language therapist) was undertaken which informed the development of a holistic care plan and behaviour support guidelines for staff to follow. This included explaining to Joseph in accessible way why he had moved from home, and practical advice on responding to episodes of challenging behaviour. Various means of enhancing communication were adopted including visual planners and choice boards. His behaviour gradually improved.

Abuse

People with learning disability are at increased risk of abuse in all its forms. They may be dependent on others for various aspects of their care, be socially naïve, and unable or afraid to report when something is wrong. Bullying and financial exploitation may be perpetrated by those who feign friendship with the person (so-called 'mate crime'). It is important to be alert to possible signs of abuse and to flag concerns using safeguarding processes where abuse or neglect is suspected.

Health inequalities

People with intellectual disability are four times as likely to die of preventable causes compared with the general population. 'Death by Indifference', a seminal report published in

2007 by the learning disability charity Mencap, highlighted the stories of six individuals whose deaths were potentially avoidable and attributed to failings in the NHS.⁵

Several years later the Confidential Inquiry of Premature Deaths in People with Learning Disability found that men with intellectual disability died on average 13 years younger than their non-intellectually disabled counterparts.⁶ For women the mortality gap was 20 years. Over one-third of deaths were judged to be avoidable, given the provision of high-quality healthcare. Particular factors identified as contributing to untimely deaths of people with intellectual disability included significant delay in diagnosis, further investigation and specialist referral; a lack of coordinated care between disease pathways and service providers; lack of reasonable adjustments to facilitate care.

Health promotion messages might not be accessible to people with intellectual disability. We know that people with intellectual disability are less likely to eat a balanced healthy diet and achieve recommended exercise levels, and more likely to be underweight or overweight.

Overt discrimination against people with intellectual disability is prohibited by law but the negative effect of implicit attitudes and assumptions that may be present in health care settings (e.g. about quality of life) and that can influence clinical decision-making are difficult to judge. Diagnostic overshadowing occurs when a presentation is attributed to the intellectual disability, rather than a potentially-treatable cause. This can result in missed opportunities for intervention and under-treatment.

The Learning Disabilities Mortality Review (LeDeR) Programme was established in 2015 to promote and implement a new review process for deaths in people with intellectual disability. Anyone, including General Practitioners, can report a death of a person with intellectual disability to the investigators. The aim of the programme is to understand the circumstances leading to deaths and whether identify learning points that will lead to policy and practice improvements (<http://www.bristol.ac.uk/sps/leder/>).

Getting primary care right for people with intellectual disability

Qualitative research shows that people with ID have mixed experiences of accessing primary care services⁷, highlighting that good practice can be achieved but is inconsistent. The systems and practice changes that might be helpful to people with intellectual disability have broad appeal and are also likely to improve experience for other patient groups (e.g. those with dementia, sensory impairments, or reduced literacy).⁸

General Practitioners are key players in responding to the health inequalities experienced by people with intellectual disability on both an individual and population level. A number of practical actions to improve health outcomes for individuals with intellectual disability are supported by evidence and should be incorporated into standard care.

General practices should keep a record of patients who have intellectual disability which can be used to anticipate needs, plan on a population scale, communication between different healthcare providers and primary/secondary care, identifying those who qualify for directed health interventions. Ensuring that a person with intellectual disability has a suitable Read code can ensure that the individual is automatically added to the register on the electronic notes system makes it easier to interrogate the electronic notes system to gather the list of people with intellectual disability.

The Equality Act 2010 places a legal responsibility on all public sector bodies, including NHS providers to make 'reasonable adjustments' for any individual with a disability, including those with intellectual disability (see box 5).⁹ The Act specifies that the provider must anticipate and not merely respond to people with additional needs, meaning that services must proactively consider changes that need to be made. A small adjustment in how care is provided can improve the experience of care, often increasingly the likelihood of a successful encounter and reducing the likelihood of behaviour which challenges. If you are aware of an adjustment which is particularly helpful for one of your patients, it is helpful to share this information with other professionals, for example when making a referral to secondary care.

Box 5 – Examples of some reasonable adjustments for people with intellectual disability accessing primary care services

Removing physical barriers to access
Longer appointment times
Offering home visits
Adapting communication (see box 6)
Easier process to make appointments
See at beginning/end of clinic when quieter
Close partnership working with paid or family carers (where applicable)
Relevant training for all staff
Continuity of care / named GP
Having a practice lead for intellectual disability

A growing evidence base attests to the value of a primary care annual health checks for people with intellectual disability. Annual health checks enable the regular screening and identification of common and serious unmet health needs. They increase the opportunity for both early intervention and health promotion, which may help to reduce the level of the health inequalities and avoidable deaths.¹⁰ Additionally they have been shown to provide a beneficial educational support for carers and relatives and can also help improve the engagement of individuals with intellectual disability in their local health economy. The use of a standardised health assessment instrument such as the 'Cardiff Health Check' tool is advised (available at: https://www.google.co.uk/url?sa=t&rct=j&q=&esrc=s&source=web&cd=1&ved=0ahUKEwii55_g1fZAhXMFsAKHRgIBXgQFggNMAA&url=http%3A%2F%2Fwww.easyhealth.org.uk%2Fsite%2Fdefault%2Ffiles%2FCardiff_Health_Check.pdf&usg=AOvVaw0f0ga7oUhIRYbc4Az5q5tf)

A key component of an annual health check is a comprehensive medication review. People with intellectual disability often receive complex drug regimens and may lack understanding of indications and side-effects. The annual health check offers a chance to review the potential for drug interactions, duplication of therapy, and continued need for medication. People with ID are often more susceptible to side effects and are often prescribed medication off licence or for unclear indications.

In light of data demonstrating consistently high levels of psychotropic medication use for people with intellectual disability (often in the absence of an underlying diagnosis of mental illness)¹¹ there has been a recent focus on reducing the use of this class of drugs. Resources are available from NHS England (<https://www.england.nhs.uk/wp-content/uploads/2017/07/stomp-gp-prescribing-v17.pdf>) and additional advice can be obtained from specialist teams.

The results from the annual health check should be used to inform and populate a personalised Health Action Plan for the forthcoming year. Several templates are available, and some clinical record systems include embedded forms that can be printed and completed during the appointment (<http://www.derbyshirehealthcareft.nhs.uk/services/learning-disabilities/annual-health-check/health-action-plan/>). A Health Action Plan supports an individual with intellectual disability to make decisions about their health, including identifying health needs and supporting decisions about how to maintain and promote good health. The Plan is ideally completed and owned by the person with intellectual disability, where they are able. There will be space for input from a variety of professionals.

If it is anticipated that time will be spent in secondary care then a hospital passport can also be developed.¹² A hospital passport is a quick and easy reference guide to help health care professionals deliver high quality personalised care by providing them with ready access to essential information about an individual with an intellectual disability. In addition to containing information about their physical health it is likely to contain information about someone's likes, dislikes and communication preferences and should help the relevant professionals to make reasonable adjustments in their provision of care. The hospital passport can be developed by the person with learning disability in conjunction with carers and input from the secondary care team.

As with all clinical encounters, it is advisable for a clinician to be mindful of the Mental Capacity Act.¹³ If you are unclear as to whether an individual with intellectual disability has capacity to consent please do assess and document your clinical opinion with regards to their capacity. As is outlined in the Act, an assessment needs to confirm the presence of a disorder of mind or brain (i.e. intellectual disability) and consider if the individual is able to understand,

retain, weigh up and communicate information pertinent to the decision. For many decisions it can be appropriate to continue to recommend treatment as usual in a person's best interests. However if the decision is significant, complex or involves considerable risks, it is advisable to arrange for a best interests meeting with relevant parties. Community Learning Disability Teams are usually happy to provide advice and input in this area. Whilst the opinion of family and friends should be sought, it is pertinent to remember that they cannot make a decision on behalf of their relative unless they are a Court of Protection appointed deputy for that particular decision. It is advisable to refer to your local Mental Capacity Act advocacy services to ensure that the individual with intellectual disability's voice is heard during any discussions.

For those with complex health needs a multidisciplinary team approach may be required. Examples of excellent care outcomes occur when a team forms around an individual to provide relevant input, for example an oncologist, GP, palliative care nurse, and learning disability specialist nurse may work across traditional boundaries to ensure to provide the best quality of care to someone with a terminal illness (case scenario 2).

Paid and family carers often have a wealth of information and are important sources of information. They are often keen to be involved and have an important role to play in augmenting communication, improving understanding, and monitoring health conditions.

In addition, carers and individuals with intellectual disability themselves are often well placed to comment on broader strategic decisions based on their lived experience of healthcare services. Including this group in consultations via the patient and carer participation group is important to a practice wanting to provide best care.

Finally, being a carer can be a difficult and lonely role. Local Authorities have a statutory duty under the Care Act to offer a Carer's Assessment where it is requested. This can identify areas of unmet need and determine eligibility for additional support. Carer's networks and group meetings often exist locally and can be a further source of information and support.

Case scenario 2

Amba is a 44 year old lady with moderate intellectual disability. Her carers noticed change in bowel habit and weight loss and supported Amba to her GP who recognised ‘red flag’ signs and referred her to a specialist. The learning disability liaison nurse at the hospital was informed in advance that Amba would be attending for appointments and various investigations. The nurse liaised with the clinicians and radiography department to ensure that reasonable adjustments were undertaken to make the process as comfortable as possible for Amba. When Amba had surgery, she took her hospital passport which helped the ward team to provide personalised care. The psychologist at the local community learning disability team spent some time with Amba and used adapted easy-read resources to help her make sense of her situation, mitigating the psychological effect of the diagnosis and subsequent treatment. Amba made a full recovery and returned to her previous activities.

Box 6 – tips for effective communication with people with intellectual disability

- Keep the person with intellectual disability the focus of the interaction
- Give extra time where possible. A double appointment can help
- Invite a carer to the appointment (where applicable) and gather collateral information
- Consider non-verbal modes of communication – incorporating simple signs, actions, or objects might be useful
- Keep questions short and use common words, avoiding jargon (https://www.gmc-uk.org/learningdisabilities/Jargon_Buster_A4_chart.pdf_47935778.pdf)
- Repeat key points
- Check understanding and summarise your discussion at the end of the consultation
- Accessible information leaflets (such as those available at <http://www.easyhealth.org.uk/categories/health-leaflets/>) can be used to improve information exchange and can be taken away by the person with intellectual disability
- A ‘distress thermometer’ is an example of a communication aid that can be used to identify distress coming from any source.¹⁴

Key points

- People with intellectual disability have impairment in general mental abilities that affects independence in a range of daily activities.
- People with intellectual disability comprise a highly heterogeneous group. People with intellectual disability should be appropriately supported to live active and meaningful lives in line with their aspirations and abilities.
- People with intellectual disability often have additional health needs. The General Practitioner has a vital role to play in optimising the care that people with intellectual disability receive.
- Making reasonable adjustments in the delivery of care on an individual and group level can reduce inequities. Keeping a register of people with intellectual disability and providing annual health checks are important initiatives.
- Community learning disability teams provide advice and specialist expertise. Close liaison working between those involved is important and should include the views and knowledge of family members and carers.

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Single Best Option questions

1. All of the following are needed for a diagnosis of intellectual disability except...

1. Significantly impaired intellectual function on psychometric testing
2. Impaired adaptive function
3. Onset of difficulties in childhood or adolescence
4. Evidence of risk to others
5. An intelligence quotient (IQ) of less than 70

Explanation:

Intellectual disability is defined by three core criteria; significantly impaired intellectual functioning (equating to a full-scale IQ of <70), impaired adaptive function (e.g. in personal care, money management, social judgement, and interpersonal skills). Onset of the difficulties must be under the age of 18 years. Evidence of risk to others is not a requirement.

2. All of the following can cause of intellectual disability except...

1. Maternal alcohol dependence in pregnancy
2. Patau's syndrome
3. Extreme neglect in early infancy
4. Lead poisoning in childhood
5. Chronic schizophrenia

Explanation:

Excess alcohol use in pregnancy can cause Fetal Alcohol Syndrome, associated with a range of intellectual, behavioural, and physical manifestations. Patau's syndrome is trisomy 13 and usually associated with severe intellectual disability; only a small proportion survive past the age of 1 year. Extreme neglect in early infancy can retard normal development to such a degree that a diagnosis of intellectual disability can be made. Lead is a neurotoxin and

ingestion can lead to intellectual disability. Although chronic schizophrenia can be associated with significant cognitive impairment, the onset of symptoms is usually beyond the developmental period.

3. Which of the following intelligence quotient (IQ) range is associated with moderate intellectual disability?

1. IQ 50-70
2. IQ <20
3. IQ 70-85
4. IQ 35-49
5. IQ 20-60

Explanation:

IQ classification for intellectual disability is as follows: IQ 50-70 = mild intellectual disability, IQ 35-49 = moderate intellectual disability, IQ 20-34 = severe intellectual disability, IQ <20 = profound intellectual disability. 95% of the population have an IQ between 70 and 130.

4. Which of the following is not true about mental disorder in people with intellectual disability

1. Assessment should only be conducted by specialist psychiatrists
2. Collateral information is useful in understanding the full picture
3. Change in behaviour from baseline is significant
4. Adapted diagnostic criteria can be used
5. Mental disorder can be difficult to recognise in people with limitations in communication

Explanation:

Although mental illness can manifest differently in people with intellectual disability, specialists are not always needed to recognise signs and symptoms, especially in those with mild intellectual disability where presentation of illness manifests in a similar way to those without intellectual disability.

5. People with Down's syndrome have an increased risk of the following conditions, except...

1. Gastro-oesophageal reflux disease
2. Hypothyroidism
3. Myopia
4. Congenital heart defects
5. Breast cancer

Explanation:

People with Down's syndrome are at increased risk of a range of physical illnesses. Particularly common are gastrointestinal disorders, hypothyroidism (congenital or acquired), eye conditions (refractive errors, strabismus, cataracts). Congenital heart defects occur in approximately 50%. Breast cancer and some other solid tumours are less common in people with Down's syndrome.

6. Which of the following is not a reasonable adjustment?

1. Giving a longer appointment to someone with communication difficulties
2. Providing step-free access to the surgery
3. Refusing a request from someone with autism to wait in a quiet area because everyone must be treated the same
4. Offering 'easy-read' information
5. Making a home visit where someone finds it difficult to get to appointments

Explanation:

The Equality Act (2010) demands that reasonable adjustments are made so that people with additional needs are not disadvantaged when accessing services. It is wrong to interpret this as meaning that everyone must be treated in an identical manner.

7. Which of the following is not true regarding the Mental Capacity Act?

1. People with capacity are free to make unwise decisions
2. It should be assumed that people have capacity, unless demonstrated otherwise
3. For those lacking capacity, a decision is made in their best interests
4. Family members always have the final say in decisions where someone lacks capacity
5. Capacity is decision-specific

Explanation:

Capacity to make decisions is assumed unless demonstrated otherwise. In order to have capacity to make a decision, an individual must be able to understand, retain, weigh up, and communicate their decision. If a person lacks mental capacity in any one (or more) of these areas, a best interests decision must be made. This should involve stakeholders, but unless family members are Court of Protection appointees for that decision, they will not have the deciding say in the case of disagreement.

8. Which of the following is true

1. People with intellectual disability cannot engage with psychological therapies
2. People with intellectual disability should generally be given higher doses of psychotropic drugs
3. NICE guidelines are not relevant to the care of people with intellectual disability
4. IAPT services should make reasonable adjustments, including training practitioners, to ensure that people with mild-moderate intellectual disability can access the service

5. People with intellectual disability experience very low rates of mental illness

Explanation: Increasing Access to Psychological Therapies (IAPT) treatment should be available to people with intellectual disability with common mental disorders. People with intellectual disability have generally higher rates of mental illness than those in the general population.

9. Which of the following is not typically seen in Down syndrome?

1. Wide gap between first and second toes
2. Single palmar crease
3. Delayed puberty
4. Epicanthic folds and Brushfield spots

5. Tall stature

Explanation:

All of the characteristics listed are typical of Down syndrome. Epicanthic folds are skin of the upper eyelid covering the medial corner of the eye. Brushfield spots are white or greyish spots around the periphery of the iris. People with Down's syndrome are usually of short stature.

10. A 22 year old man attends the clinic. He has recently started work in a bakery but is struggling with the job and has had two warnings from his manager. You suspect he may have an undiagnosed intellectual disability. Which of the following is true?

1. Moderate intellectual disability is the most frequent type of intellectual disability
2. People with intellectual disability must have attended a special school
3. Intellectual disability can affect adaptive function across a range of areas
4. An adult with intellectual disability must be incapable of living alone
5. People with intellectual disability are a danger to others

Explanation:

In the majority of cases intellectual disability is of a mild degree. People with intellectual disability may attend mainstream schools, especially if given additional support. It is important to challenge prejudices and stigma surrounding intellectual disability. Many people with the condition live independently with minimal support. Vulnerability, rather than dangerousness, is more common in this group.

Extended Matching Questions

Theme: Mental disorders

- a) Alcohol dependence
- b) Alzheimer's dementia
- c) Attention deficit hyperactivity disorder
- d) Bipolar disorder
- e) Depression
- f) Factitious disorder
- g) Generalised anxiety disorder
- h) Psychosis

Please give the single most likely diagnosis from the list of options given. Each option may be used once, more than once, or not at all.

1. A 58 year old man with mild intellectual disability due to Down's syndrome developed new-onset epileptic seizures last year. Over the past 6 months carers report disinhibited behaviour, stubbornness and occasional verbal aggression. He has lost skills, especially in finding his way around the local community.

2. A 22 year old man with autism and mild learning disability who left college last year and recently moved from the family home to supported accommodation. He has difficulty sleeping, is irritable, often complains of pins-and-needles and dizziness, and sometimes has a fine tremor.

3. A 26 year old woman with mild intellectual disability lost her job as a waitress two months ago following poor attendance and frequent lateness. Her mother reports she has stopped seeing friends, is not eating well and has lost weight, is lethargic and frequently tearful.

4. A 15 year old boy with mild intellectual disability attends mainstream school but is struggling in class and has been suspended on two occasions. In clinic he appears restless, fidgety, and quickly frustrated.

5. A 22 year old man with mild intellectual disability and heavy cannabis use. Neighbours report erratic behaviour and he is brought to A&E by the police. On assessment he presents as perplexed and frightened.

Answers

1. b

2. g

3. e

4. c

5. h

Explanation:

1. People with Down syndrome are prone to the early onset of Alzheimer's dementia. New-onset seizures and behavioural changes are often seen.

2. People with autism spectrum disorder have high rates of co-morbid anxiety. In this case, there are clear precipitants including loss of routine and change in environment. The patient demonstrates biological signs of anxiety.

3. These symptoms are recognisable as classic of depression.

4. Attention-deficit hyperactivity disorder is more common in people with intellectual disability. This often causes problems in schools. The hyperactivity may be obvious during appointments.

5. Cannabis use is a risk factor for the onset of psychotic disorders.

Theme: Presentations to primary care

- a) Arrange to review in 2 weeks' time
- b) Perform full physical examination
- c) Prescribe benzodiazepine
- d) Contact local safeguarding team to discuss concerns
- e) Refer to local community learning disability team
- f) Consider reducing anti-psychotic medication

- g) Refer to the Community Learning Disability Team
- h) Consider requesting a Mental Health Act assessment

Select the most appropriate next step from the list above. Each option may be used once, more than once, or not at all.

1. An 18 year old man with moderate intellectual disability and autism spectrum disorder presents with carers who report a two day history of poor sleep, agitation, and head banging.
2. A 50 year old man with severe intellectual disability who lives in a residential home. At annual review his carers report no problems with behaviour. He has been taking regular risperidone following a single incident of aggression 6 years ago.
3. The parents of a 32 year old lady with mild intellectual disability and bipolar disorder report that she has reduced sleep, increased energy, is leaving the house at night, and has become sexually disinhibited, placing herself at risk of exploitation. She has had several previous admissions and has stopped maintenance mood stabilising medication.
4. A 40 year old man with profound intellectual disability and who is non-verbal attends the surgery for an annual health check with a carer. Physical examination reveals clusters of bruises across the upper arms outer thighs. There is nothing in his medical history to suggest a cause and the carer is unable to explain the findings.
5. A 44 year old lady with mild intellectual disability and good general health consults with a one week history of feeling down and more tired than usual. She has no other symptoms and a supportive family.

Answers

1. b
2. f
3. h
4. d
5. a

Explanations:

1. Physical illness must always be considered in behaviour change. Diagnostic overshadowing occurs where the presentation is presumed to be a consequence of the intellectual disability and potentially-treatable causes are overlooked.
2. Medication review is important to avoid deleterious consequences of prescribed drugs in this group.
3. Increased energy with reduced need for sleep, over-activity, excitement, disinhibition, and irritability are symptoms indicating a manic relapse of bipolar disorder, especially when considered in the context of medication recently having been stopped.
4. Health professionals must be alert for signs suggestive of abuse.
5. Active monitoring may be appropriate for those with mild depression and where risks are judged to be low.

Theme: Genetic causes of intellectual disability

- a) Fragile X syndrome
- b) Down's syndrome
- c) Prader-Willi syndrome
- d) Velo-cardio facial syndrome
- e) Klinefelter's syndrome
- f) Turner's syndrome
- g) Capgras syndrome
- h) Smith-Magenis syndrome

Each option may be used once, more than once, or not at all.

1. Also known as DiGeorge syndrome or 22q11.2 deletion syndrome
2. Characterised by excessive and insatiable appetite and food-seeking behaviour
3. 45,XO karyotype
4. The most common inherited form of intellectual disability

5. High rates of autism spectrum disorder, social anxiety, and attention-deficit hyperactivity disorder

Answers

1. d

2. c

3. f

4. a

5. a

Explanations

1. Velo-cardio facial syndrome is also known as DiGeorge syndrome (after Angelo DiGeorge, who first described the condition) or 22q11.2 deletion syndrome (related to the segment of genetic material on the long arm of chromosome 22 that is deleted).

2. Prader-Willi syndrome is a genetic condition that involves excessive hunger, hyperphagia and weight gain.

3. Turner's syndrome is caused by a missing X chromosome. It is estimated that between 1 in 2000 and 1 in 5000 females are affected.

4. Fragile X syndrome is the most common inherited form of intellectual disability. Down's syndrome is the most common genetic cause of intellectual disability but is rarely inherited.

5. Those with fragile X syndrome often have impairments in social interaction and difficulties forming friendships. ADHD is the most common psychiatric diagnosis in people with fragile X syndrome.