

Learning disability

Introduction

Definition and classification of learning disability

Must fulfil 3 criteria:

- Intellectual impairment (IQ \leq 70)
- Social or adaptive dysfunction
- Onset in the developmental period (thus excluding people with dementia or other adult-onset diseases/injury affecting the brain)

Learning disability is a *descriptive diagnosis or concept*, not a disease or illness. It does not infer a particular aetiology. Social functioning is an integral part of the diagnosis. It is important to understand that it is different from mental illness – a person with a learning disability can also develop mental illness. Learning disability as a concept is also different from ‘learning difficulties’, which generally refers to specific learning problems (e.g. dyslexia), rather than a global impairment of intellect and function.

IQ (Intelligence Quotient)

IQ is used to classify level of learning disability

Mild learning disability	50 to 70
Moderate learning disability	35 to 50
Severe learning disability	20 to 35
Profound learning disability	<20

Note that IQ of 70 is 2 standard deviations below the mean, and is therefore considered statistically significant.

IQ may be an easily quantifiable way to classify learning disability, but there are problems with it. For example, the overall IQ score does not indicate individual strengths or weaknesses (e.g. verbal and motor skills). IQ also varies during development. IQ classification alone does not include the important area of social adaptation and functioning.

Prevalence

Statistically the prevalence of people with IQ<70 should be 2.5% (2SD from mean).

Actually the prevalence of people with learning disability is 1-2%, because of

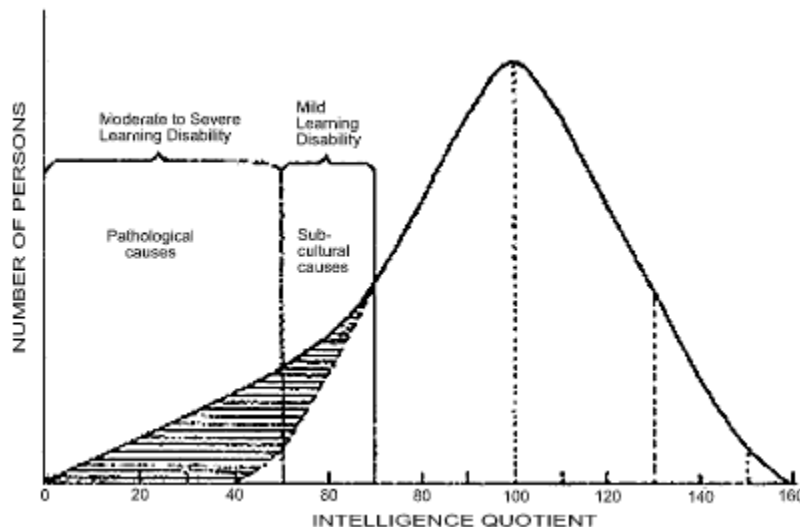
- *Differential mortality* (the more severe the degree of learning disability, the higher the mortality c.f. general population).
- *Diagnostic changes* with time, not all cases classified.
- The role of *functioning* – those with IQ<70 but no problems functioning within their environment would not be defined as having a learning disability.

Prevalence of people with IQ <50 is 0.35%.

IQ levels follow a normal distribution curve for IQs above 70. Below this the curve has an asymmetric distribution, with higher than expected numbers at IQ's of less than 70 and a small ‘bump’ at the lower end. The normal distribution represents the differences in IQ secondary to environmental and genetic effects right across the IQ range. The bump at the lower end is due to organic causes.

Aetiology

IQ levels follow a normal distribution curve for IQ's above 70. For lower levels of IQ the curve is skewed due to the addition of pathological (organic) causes of learning disability. The shaded area on the diagram below indicates the organic causes of learning disability.



Distribution of intelligence in the population from *Companion to Psychiatric Studies*. R Kendall & A Zeally Eds.

Mild learning disability

- These individuals form part of the lower end of the normal distribution curve for IQ. Generally can communicate with spoken language.
- Role of multifactorial genetic & environmental influences (which also control the variance of IQ above 70) traditionally thought most important in this group. Higher rates in social class IV & V, especially with large sibship, overcrowding & poverty point to 'sub-cultural causes' of learning disability in this group.
- But increasing evidence for organic involvement - recent studies show up to 45% have definite organic factors (subtle chromosome rearrangements, perinatal insults from toxins like alcohol).
- Presently no cause found >50%, 15-20% perinatal hypoxia, 10% congenital causes, 5% defined genetic cause.

Moderate to Profound Learning Disability

- Need greater degree of care, often including physical care to aid with feeding, continence etc.
- Can nearly always assume organic pathology for severe & profound learning disability: chromosomal 40%, genetic 15%, pre & peri-natal 10%, post-natal 10%, unknown 25%.
- Excess numbers at more severe levels ('bump' on IQ curve) is due to organic or pathological group with cluster of disorders of definable aetiology (genetic, environment – hypoxia, trauma, infections). Some will have syndromes like autism, cerebral malformation syndromes or cerebral palsy, without yet defined cause, but presumed definite biological factors in causation.

Examples of Specific Aetiologies

Vast number of different organic causes – anything that causes an insult to the brain in the developmental period. Common examples include:

- **Chromosomal abnormalities**, e.g. Down's Syndrome (trisomy 21), Prader-Willi Syndrome (microdeletion on chromosome 15).
- **Genetic disorders**, e.g. Fragile X Syndrome (X-linked), PKU (autosomal recessive), tuberous sclerosis (autosomal dominant).
- **Intrauterine & neonatal damage**, e.g. infections (rubella), foetal alcohol syndrome, prematurity, labour complications.
- **Developmental & anatomical abnormalities of CNS**, e.g. neural tube defects (learning disability usually secondary to hydrocephalus or infection), cerebral palsies & the profound & multiply handicapped.
- **Other later post-natal causes**, e.g. encephalitis, meningitis, trauma, hypoxia (for example, due to severe epilepsy).
- **Disorders of unknown, presumed biological origin**, e.g. Autism, Rett's syndrome.

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Why is it important to find the cause of a person's learning disability?

- Right of individual/family to know the cause of their learning disability.
- Enable awareness of associated physical and psychiatric conditions that may be treatable.
- To enable genetic counselling where appropriate.

Surprisingly few people with learning disability (especially adults) have ever been thoroughly investigated as to the cause. Paediatricians now usually investigate children with learning disability, but general practitioners and other hospital specialists may take the opportunity to do so. Psychiatrists frequently investigate and find causes for a person's learning disability relatively late in life.

Examples of Common Syndromes

Down's Syndrome

- Commonest chromosomal abnormality seen in people with learning disability and the commonest identifiable cause of learning disability.
- Usually moderate to severe learning disability.
- Typical facial appearance: small ears and eyes, small head, protruding tongue. Short stature.
- Common physical health problems: Many, including congenital heart defects, hypothyroidism, visual disorders (especially cataract and keratoconus), hearing impairment (middle ear infections, conductive hearing loss, early onset of presbycusis), obesity, epilepsy, degenerative spine disorders, atlanto-axial joint subluxation, periodontal and dermatological problems, diabetes mellitus, coeliac disease, hypotension, respiratory and other infections (to poor immunological defence mechanisms), leukaemia and obstructive sleep apnoea.
- Psychiatric problems: high rates of early onset dementia of Alzheimer's type and depression.

Fragile X

- Commonest inherited cause of learning disability.
- Usually borderline to moderate learning disability.
- Typical facial appearance: Large head circumference, long and prominent ears.
- Physical associations: Examples include mitral valve prolapse, lax joints, scoliosis, flat feet, testicular enlargement.
- Behavioural associations: poor concentration, hyperkinetic behaviour, avoidance of eye contact.

Services and Support for People with Learning Disability

Prevention of learning disability

- Early intervention for those at risk of mild learning disability (low birth-weight babies, children from disadvantaged families).
- Better obstetric care & control of neonatal infections (but this has also led to an increase in survival of small-for-dates deliveries, which are at risk of learning disability).
- Prevention of drug/alcohol abuse in pregnancy (foetal alcohol syndrome, teratogenic effects of substance misuse, HIV).
- Screening for heterozygotes/carriers in families with affected member and counselling by geneticists (ethical dilemmas as mostly no treatments available).
- Pre-natal diagnosis improving – but only 'treatment' yet available is termination.
- Newborn screening for reversible disorders – Guthrie test for phenylketonuria & other metabolic disorders, congenital hypothyroidism.
- Other – e.g. rubella immunisation, folate supplements in pregnancy.

Education and Social Care

In recent years, there has been an emphasis on inclusion and community integration of people with learning disability. This has been the principle underlying education, social care and health services. The closure programmes for the old large asylums are now largely complete. The previous 'medical model' of care for people with learning disability has generally been rejected.

Health professionals need to be aware of a large, diverse network of carers, volunteers and professional organisations involved in the care and support of people with learning disability. These include:

- Family and friends.
- Early intervention programmed (e.g. 'Sure Start')
- Voluntary befrienders.
- Advocates.
- Housing associations and agencies – from supported independent living to fully staffed residential accommodation.
- Teachers and allied professionals.
- Social workers – often coordinate 'care packages' for individuals.
- Respite providers – special units or in providers' own homes.
- Adult training centres.
- Supported work placements.

Health Service Provision

- GPs and primary health care teams deal with the majority of health problems themselves.
- Generic secondary physical health care for adults, other than that provided by community learning disability teams. There are often special needs clinics run by paediatricians for children.
- Community learning disability teams consist of community learning disability nurses, psychiatrists and a variety of other professionals such as occupational therapists, speech and communication therapists, dieticians, physiotherapists, psychologists and social workers. Usually provide services to a geographic area.
- Specialist in-patient units are available for a small minority of people with learning disability. These are usually run by psychiatrists and multidisciplinary colleagues and provide assessment/treatment for mental health problems and challenging behaviour. Some areas also provide specialist learning disability units for people with forensic problems, autism, and children and adolescents. There are also some continuing care beds for those with severe multiple disabilities.

The Role of Doctors in Treating People with Learning Disability

Learning disability is a common denominator in a wide variety of conditions. It can occur with or without any other mental or physical disorder. There has been an appropriate shift away from medicalising people with learning disability. So why should doctors consider people with learning disability as a group?

Medical Associations

People with learning disability as a group have higher rates of physical health problems and consequently higher morbidity and mortality rates. Conditions may result from the same underlying cause as the person's learning disability (e.g. cerebral palsy due to hypoxic brain damage). Higher morbidity and mortality rates also result from late presentation of illness, poor access to health services and screening, incomplete investigations of symptoms and delay in treatment.

In people whose functioning is already compromised, undiagnosed and untreated conditions can have a disproportionately high impact. Learning potential and the ability to live as independently as possible may be further impaired. Apart from specific physical illness in certain conditions e.g. valve disease in Down's syndrome, the prevalence of other illness is higher in people with learning disability as a whole, for example:

- Epilepsy – increased incidence with severity of learning disability. May be due to same underlying cause as the learning disability. Can further compromise cognitive ability through seizure activity or side effects of medication.
- Sensory impairments – adequate hearing and vision are important for psychomotor development, cognitive achievements and social/emotional development. Recognition and treatment of such impairments can significantly improve communication and learning ability. Earwax is particularly common and simple to treat.
- Obesity – may further increase stigma, as well as predispose to other health problems.
- Gastrointestinal problems – reflux oesophagitis, helicobacter pylori, carcinoma, constipation.
- Respiratory problems – chest infections particularly common.
- Cerebral palsy – especially in those with more severe learning disability.
- Orthopaedic problems – joint contractures, osteoporosis.
- Dermatological problems

Psychiatric Associations

Emotional & behavioural disorder is more common in children & adults with learning disability than in the general population. Specific psychiatric disorders are also more common, e.g. schizophrenia, bipolar affective disorder.

Why more psychiatric illness?

- The organic cause of the learning disability may also cause or predispose to psychiatric disorder (e.g. epilepsy, schizophrenia, Alzheimer's disease in Down's syndrome).
- Communication problems – inability to express feelings (e.g. after bereavement) may lead to depression/anxiety.
- Higher rates of social deprivation especially in mild learning disability group.
- Vulnerable to abuse.
- Low self esteem (stigma, dependence on others).
- Often multiple losses – changes of carer, home, etc. (may all occur together if living with parents until their deaths).

Presentation of psychiatric problems in people with Learning Disability

- People with mild learning disability may present in broadly similar way to the general population.
- Low IQ affects the symptom complex of common mental illnesses, e.g. less complex delusions in psychotic disorders. Where there is less verbal communication, observable signs are relied on more in making the diagnosis, e.g. weight loss, withdrawal, agitation, tearfulness in depression; behavioural disturbance in psychotic disorder.
- Those with lower IQ often present with 'challenging behaviour' - need to methodically look for cause (which may be physical, psychological or environmental/social).

Psychiatrists specialising in working with people with learning disability develop expertise in:

- Recognising presentation of psychiatric illness in this group as a whole.
- Knowledge of common patterns of behaviour ('behavioural phenotypes') or illnesses in particular syndromes.
- Behavioural management.
- Knowledge of appropriate treatment (more susceptible to side-effects of drugs).
- Multidisciplinary working with all agencies involved.
- Specialised areas such as forensic psychiatry, autism, and epilepsy.

Communication

People with learning disability may have problems in communicating with others due to:

- Intellectual impairment leading to problems comprehending and processing information.
- Sensory difficulties (hearing, vision).
- Problems in understanding social interaction (e.g. autism).
- Speech problems (e.g. articulation problems).
- Others not listening and valuing what they are trying to communicate.

Health professionals need to:

- Take time and have patience.
- Value what is being communicated.
- Recognise non-verbal cues.
- Find out about the person's alternative communication strategies if verbal communication is difficult (e.g. their typical non-verbal cues, use symbols, sign language).
- Explain things clearly in an appropriate way (verbally & with pictures etc).
- Be prepared to meet the person several times to build up rapport & trust.
- Use the knowledge and support of people's carers.

Accessing general health care

Considering the higher levels of physical health problems amongst people with learning disabilities, they are relatively low users of health services.

In order to access health care a person or their carer needs to be able to:

- Recognise symptoms.
- Realise their significance.
- Know who to report them to.
- Be able to communicate these symptoms effectively to health professionals.

If a person has problems with any of the above, conditions are at risk of being missed.

Health professionals need to be able to:

- Understand what person is communicating.
- Recognise different presentations.
- Make a diagnosis.
- Differentiate assent/dissent.
- Understand consent/capacity issues.
- Implement appropriate treatment.
- Identify side-effects.

Diagnostic overshadowing

This means when a person's presenting symptoms are put down to their learning disability, rather than seeking another, potentially treatable cause. For example, when a person presents with a new behaviour or existing ones escalate, doctors should consider:

- Physical problems – pain or discomfort, e.g. from ear infection, toothache, constipation, reflux oesophagitis, deterioration in vision or hearing.
- Psychiatric cause – depression, anxiety, psychosis, dementia.
- Social cause – change in carers, bereavement, abuse.

To enable people with learning disability to fully utilise generic health services, they may need extra support. Carers, community learning disability nurses and advocates may all play a role in this. Arranging longer clinic appointments or preliminary visits to hospitals can give extra time to aid communication, or reduce fear of the unknown. Some community learning disability teams carry out regular health screening. Books are available that use pictures to explain common health problems and procedures, such as breast screening ('Books Beyond Words' series, Gaskell Books).

Capacity and Consent issues

To give meaningful consent a person has to:

- Be appropriately informed about the decision in question.
- Have the ability (capacity) to be able meaningfully and freely to give or withhold consent.

A greater appreciation of personal autonomy has led to dilemmas for example, the unconscious patient who requires urgent medical care. In these urgent life-threatening cases treatment needs to proceed in the incapacitated persons "best interests".

When assessing a person with learning disability to consent for a particular investigation or intervention it must be remembered that this "capacity", if present is only valid for the procedure proposed. For example a person may have capacity to consent for a dental examination but not to surgery for bowel carcinoma (due to inability to understand the nature and likely outcomes of the more complex intervention). Capacity needs to be repeatedly assessed for each proposed intervention.

If a person does not have the capacity and is unable to consent to a plan of treatment, then no one else can give or withhold consent on that incompetent persons behalf. In this case if the treatment is to go ahead it must be in the persons "best interests". This is co-ordinated by the treating professional after widespread consultation, including getting the views of family, carers, advocates and other professionals involved.