# Epidemiology

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## Introduction

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

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

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

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

## History of Intellectual Disability

### Intellectual Disability in Ancient History

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

### Rehabilitation and Reintegration

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

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

## Eugenics and Segregation

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

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

## Psychological Testing

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

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

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

### Deinstitutionalisation

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

### International Agreements

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

## Diagnosis and Classification of Intellectual Disability

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

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

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

## Epidemiology of Intellectual Disability

### Prevalence

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

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

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

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

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

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

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

## Factors That Affect Assessment of ID

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

### Study Design

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

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

### Characteristics of the Study Population

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

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

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

### Instruments Being Used

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

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

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

## Early Epidemiological Studies on ID

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

## More Recent Epidemiological Studies

### Prevalence Estimates

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

### Incidence Estimates

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

### Mortality Estimates

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

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

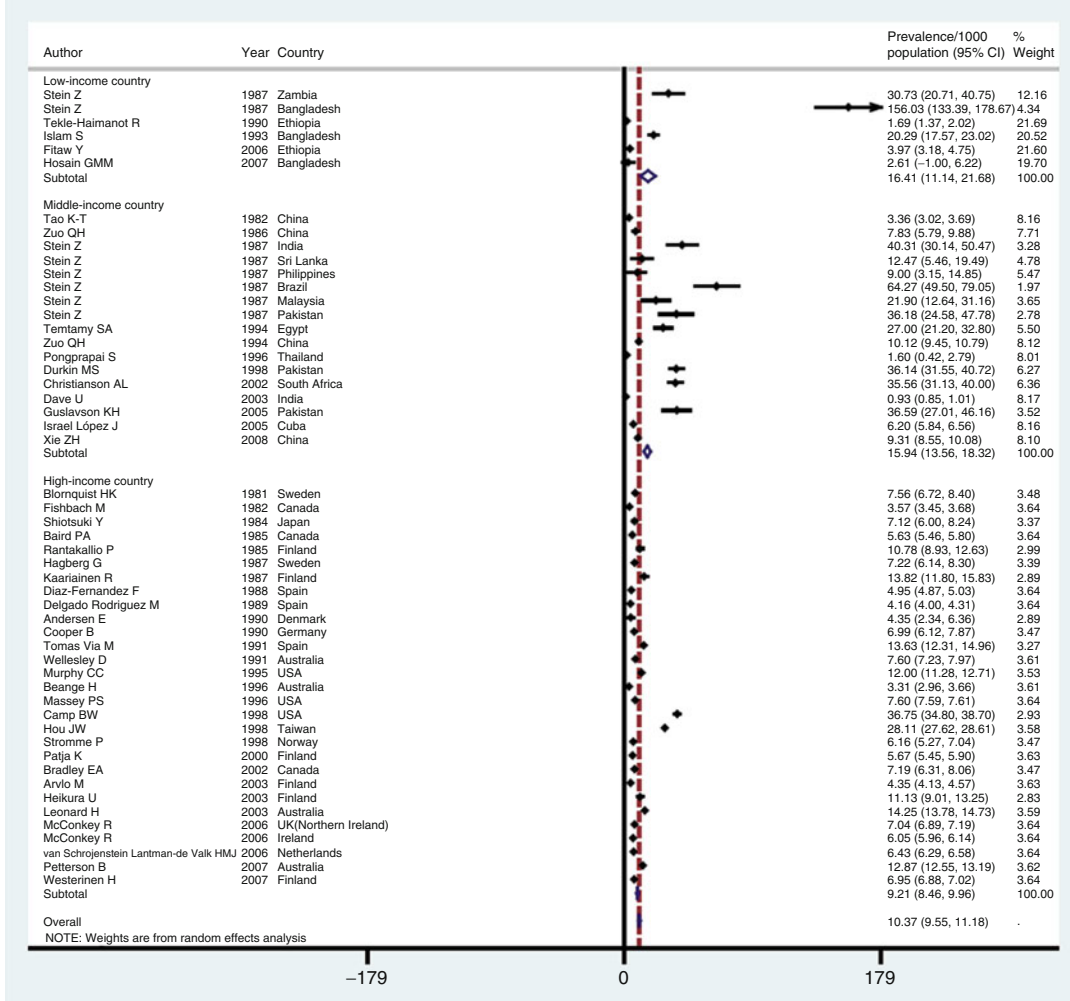


**Fig. 2.1** Forest plot of studies on prevalence of intellectual disability by income group of country ($N=2$) (Maulik et al. (2011))

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

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

## Aetiological Factors and Physical and Psychological Conditions Associated with ID

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

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

## Health Services Epidemiology

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

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

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

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

## Gaps in Knowledge and Future Research

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

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

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

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

## Conclusion

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

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

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