Section 1 Understanding Intellectual Disability

Chapter 1

Epidemiology of Intellectual Disability

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1.1 Introduction

This chapter examines the epidemiology of intellectual disability (ID). Epidemiology is 'the study of the occurrence and distribution of health-related states or events in specified populations, including the study of the determinants influencing such states, and the application of knowledge to control the health problem' (Porta, 2008). Epidemiological research in ID is complicated not only by the stigma associated with many historical terms used to denote ID, but also by the heterogeneity of different presentations of disability represented under this single entity (Leonard & Wen, 2002). This chapter discusses first the historical and current perspectives of ID; then terminology, classification and diagnosis; prevalence and incidence; aetiology; and finally concludes with morbidity and mortality.

1.2 Perspectives of ID

A contextual approach is required when considering the epidemiology of ID. Unlike other health conditions, such as communicable disease which may more traditionally be the focus of epidemiological study, the issue of what constitutes a 'true case' of a person with an ID (caseness) has been conceptualised using various historical approaches. Schalock (2013) identifies four of these approaches:

(1) Social criterion: prior to the introduction of formal diagnostic criteria for ID, individuals were identified on the basis of their behaviour, or more specifically their 'failure' to socially adapt to their environment (Greenspan, 2003, 2006a, 2006b).
(2) Clinical criterion: 'the medical model' classified individuals with ID on the presence of specific symptoms and syndromes with a focus on heredity, organicity and pathology. The model sought to 'cure' disability and actively pursued prevention through segregation policies (Drum, 2009; Devlieger; Rusch & Pfeiffer 2003).
(3) Intellectual criterion: psychometric testing heralded the classification of individuals on the basis of intelligence as measured by the IQ test (Devlieger, 2003).
(4) Dual criterion: in 1959 the American Association on Mental Deficiency (AAMD; now AAIDD, the American Association on Intellectual and Developmental Disabilities) combined both intellectual and social criteria by defining ID as deficits originating in the developmental period in intellectual functioning with co-morbid difficulties in 'maturation, learning and social adjustment', collectively referred to as 'adaptive behaviours'.

The addition of 'adaptive behaviour' as a criterion for ID was a response to ongoing dissatisfaction with IQ as the sole indicator of ID, notably due to its failure to measure

social and practical skills (Schalock et al., 2010). Originally defined as 'the effectiveness with which the individual copes with the nature and social demands of his environment' (Heber, 1959: 61), adaptive behaviour is currently conceptualised as a multidimensional concept comprising conceptual skills (e.g. linguistic and numeric skills), social skills (e.g. interpersonal skills, following rules) and practical skills (e.g. daily living and occupational skills). The addition of adaptive behaviour in the classification criteria of ID reflects efforts by the disability movement to move ID from a medical diagnostic position where ID was considered an 'absolute trait' of the individual to an ecological position where ID is influenced by the development of adaptive skills (AAIDD, 2010).

Advancing this ecological position, AAIDD developed a support needs model of disability where ID is understood as neither fixed nor dichotomised but rather as fluid and changing depending on the individual's functional limitations and the supports available within the person's environment. 'Supports' are defined as resources and strategies that promote personal development and enhance functioning, while 'support need' is identified as a psychological construct referring to both the pattern and intensity of support required for an individual with disability to participate in activities associated with 'normative human functioning' (Thompson et al., 2009). While both adaptive behaviour and support needs address typical performance on everyday tasks, they differ in focus whereby adaptive behaviour assesses the individual's level of performance or mastery on a given task, while support needs focuses on the type and intensity of support that an individual requires to successfully participate in an activity (Buntinx, 2015); 'put another way, if supports were removed, people with ID would not be able to function as successfully in typical activities and settings' (AAIDD, 2010: 113). Both adaptive behaviour and support needs are deemed relatively recent constructs which remain highly influential progressing the understanding, diagnosis and classification of ID from an intellectual limitation to an issue concerning the whole person in his or her life situation (Buntinx, 2015).

1.3 Terminology, Classification and Diagnosis

Current classification systems of ID are dominated by three systems: the fifth edition of the American Psychiatric Association's Diagnostic and Statistical Manual of Mental Disorders, known as DSM 5 (APA, 2013); the 10th edition of the World Health Organization's International Classification of Diseases, known as ICD-10 (WHO, 1992); and the 11th edition of the American Association on Intellectual and Developmental Disabilities' manual (AAIDD, 2010). Less utilised for diagnostic purposes is the World Health Organization's International Classification of Functioning, Disability and Health, known as ICF (WHO, 2001). Within the WHO family of classifications, health conditions are classified using ICD-10, while associated functioning and disability are classified using ICF (WHO, 2001). For diagnostic purposes, three criteria are common across current classification systems: deficits in intellectual functioning, deficits in adaptive behaviour, and onset during the developmental period. Deficits in intellectual functioning, typically an IQ score of 70 or less, and deficits in adaptive behaviour are objectively assessed as scoring two standard deviations below the mean on standardised psychometric tests. Intellectual functioning and adaptive behaviour are correlated but are not deemed causally associated (Tasse et al., 2016). Guidance on assessing ID is available from the British Psychological Society.[1]

[1] www.rcpsych.ac.uk/pdf/ID%20assessment%20guidance.pdf [accessed 11 August 2018].

Preparations for the 11th edition of WHO's ICD led to the establishment of a Working Group charged with securing international evidence-based consensus on the name, definition, subtypes and architecture of ID (Bertelli et al., 2016). The Working Group comprised various stakeholders including APA, AAIDD, the World Psychiatric Association (WPA) and the International Association for the Scientific Study of Intellectual and Developmental Disabilities (IASSIDD) (Salvador-Carulla et al., 2011). Despite ongoing debate of these definitional issues in the field (Switzky & Greenspan, 2006), the establishment of this international Working Group provided the first expert multidisciplinary discourse on the topic in forty years (Bertelli et al., 2016).

Driving much of the discussion is the apparent inconsistency among classification systems whereby ID is conceptualised as a health disorder by ICD and DSM and as a disability by ICF and AAIDD. Extreme positions in this discourse may conceptualise ID solely as a disability thereby calling for its exclusion from ICD, an action which may potentially render it invisible in many health monitoring systems. At the other extreme, conceptualising ID solely as a health condition is inconsistent with the social and biopsychosocial model of disability adopted by many jurisdictions in terms of legislation, policy and practice (Bertelli et al., 2016).

The Working Group agreed on the term 'Intellectual Developmental Disorder' (IDD) for ICD-11, which is classified within the parent category of 'Neurodevelopmental Disorder'. Intellectual Developmental Disorder is formally defined as 'a group of developmental conditions characterized by significant impairment of cognitive functions which are associated with limitations of learning, adaptive behaviour and skills' (Bertelli et al., 2016: 7). Intellectual Developmental Disorder is described as an early cognitive meta-syndrome analogous to the later-life syndrome of dementia. Conditions classified as Intellectual Developmental Disorders result from significant interference with brain development up to adolescence. Within the WHO family of classifications, Intellectual Developmental Disorder is thus coded within ICD-11, while ID is conceptualised as its functioning and disability counterpart and is coded within ICF. Reflecting this new landscape, APA replaced the term 'Mental Retardation' as used in the fourth edition of DSM (DSM IV) with the term 'Intellectual Disability (Intellectual Developmental Disorder)' (ID/IDD) when publishing the 5th edition (DSM 5) in 2013. The addition of 'Intellectual Developmental Disorder' within DSM 5 specifically aimed to harmonise DSM terminology with that proposed for ICD-11. Similarly, APA changed the classification of ID/IDD from Developmental Disorder in DSM IV to Neurodevelopmental Disorder in DSM 5, once again in order to harmonise with ICD.

Mindful of the degree of heterogeneity in limitations of intellectual functioning and adaptive behaviour among individuals with ID, both DSM and ICD employ a classification of clinical severity using four categories; mild, moderate, severe and profound. Within ICD-10 these categories are based on IQ score (IQ between 50 and 69 is classified within the mild range, 35 to 49 as moderate, 20 to 34 as severe and below 20 as profound). While DSM IV similarly used IQ scores to classify severity, DSM 5 introduces a new classification of mild, moderate, severe and profound ID based on deficits in adaptive behaviours, specifically in the areas of conceptual, social or practical skills. The rationale for employing adaptive behaviour rather than intellectual functioning is 'because it is adaptive functioning that determines the level of support required – moreover, IQ measures are less valid in the lower end of the IQ range' (APA, 2013: 33).

In addition to these four levels of severity, preparations for ICD-11 initiated a discussion of individuals with 'borderline' intellectual functioning, typically defined as individuals who

score within the 70–84 IQ range. These individuals are not covered by DSM and ICD (Salvador-Carulla et al., 2011). Described as 'an invisible clinical entity', borderline intellectual functioning is defined as a heterogeneous group of specific neurodevelopmental syndromes, disorders or diseases, distinct from ID (Hassiotis, 2015), which should be prioritised for further development of definition, measurement and intervention (Salvador-Carulla et al., 2011).

1.4 Prevalence and Incidence

A meta-analysis of fifty-two worldwide studies undertaken as part of the Global Burden of Disease study (Gustavsson et al., 2011) estimated the prevalence of ID at 1% globally (95% CI 0.95–1.12), with a moderately higher prevalence among males than females (Maulik et al., 2011). Globally, higher prevalence rates are reported in low-income countries (1.64%) when compared with middle-income (1.59%) and high-income countries (0.92%). The higher prevalence observed in lower-income countries is attributed to environmental factors such as malnutrition, iron deficiency and poor-quality perinatal services (Bertelli et al., 2009).

In general, the global range of estimates is extremely wide. Of the fifty-two studies reported, the lowest prevalence estimate was 0.93 per 1,000 reported from a large screening study of 550,000 residents in Mumbai, India (Dave et al., 2005). In contrast, the highest prevalence estimate was 156.03 per 1,000 reported in one of eight developing regions in a comparative study (Stein et al., 1987). This level of disparity among prevalence estimates is, according to some commentators, 'almost certainly due to methodological flaws' (Tharper et al., 2015: 720). A number of factors are however known to influence prevalence estimates of ID: higher prevalence estimates are reported in lower-income countries; where samples are drawn from urban slums/mixed rural–urban districts; among children and adolescent populations; among population-based screening studies; and in studies where psychometric scales are employed (Maulik et al., 2011).

Epidemiological community surveys reveal the vast majority of persons with ID have 'mild' ID (85%), with only a minority having an ID within the range of 'moderate' (10%), 'severe' (4%), or 'profound' (2%) (King et al., 2009). Prevalence estimates of the severity of ID may however be hindered by a lack of available data. An extensive record linkage study of all individuals born in Western Australia from 1983 to 1992, for example, reported estimates combining mild-to-moderate ID (10.6 per 1,000) and combining severe-to-profound ID (1.4 per 1,000) as no further level of detail was available from the educational sources from which the data were obtained (Leonard et al., 2003).

Higher prevalence estimates reported among studies of child and adolescents when compared with adults are commonly observed and reflect a 'transition cliff' where the age-specific prevalence of ID ascertained from administrative databases abruptly declines during transition to adulthood. UK data, for example, reported a significant decline in prevalence from 40–50 per 1,000 for children to 6–7 per 1,000 in adulthood among individuals with ID identified through use of public services (e.g. education, social care, health services) (Emerson & Glover, 2012). This decline is observed for those with milder ID, but not among those with more severe levels of ID. The transition cliff identifies a 'hidden population' of adults with mild-to-moderate levels of ID who are essentially lost from administrative databases in the transition to adulthood. In contrast, the administrative prevalence of persons with severe or profound ID remains relatively stable from childhood

through to adulthood, presumably as these individuals tend to remain in services from childhood and onward throughout adulthood (Emerson & Glover, 2012). A similar pattern is observed in an Irish national database of all persons within the state who are in receipt of, or registered as in need of ID services (Doyle & Carew, 2016). The administrative prevalence of mild ID reported nationwide for 2015 was low at 1.99 per 1,000, a reflection of the fact that many individuals with mild ID, either by choice or for eligibility reasons, are neither in receipt of nor on a waiting list for ID services. While little is known of this hidden population, they are deemed to comprise a vulnerable group who report poorer health outcomes, lower occupational prestige and greater likelihood of being involved in the criminal justice system (Emerson, 2011; Farrington et al., 2007; Wells et al., 2003).

Incidence studies of ID are scarcer than prevalence studies (McKenzie et al., 2016), largely due to the methodological challenges they present (Hatton, 2012). Most incidence studies focus on children with ID (Maulik & Harbour, 2010). A comprehensive incidence study was conducted as part of the Rochester Epidemiology Project, a pioneering record linkage study of all residents of Olmstead, Minnesota, established in 1966 (Rocca et al., 2012). Using a cohort of all children born in the region from 1976 to 1980, the cumulative incidence of ID at 8 years of age was 9 per 1,000, with little variation between boys (8.3 per 1,000) and girls (10 per 1,000). A gender difference was however noted by severity of ID; cumulative incidence for severe ID was higher in girls, while cumulative incidence for mild ID was higher in boys (Katusic et al., 1996). Incidence patterns over time were examined in Northern Finland among a cohort of individuals born in 1966 who were compared with a cohort born twenty years later. No overall difference in incidence was reported over this time period (Heikura et al., 2003). Marked differences were however observed in the incidence of ID by severity, with the incidence of mild ID increasing by 50% over the twenty years, while the incidence of moderate and severe ID decreased 55% and 34% respectively. Incidence levels for profound ID remained relatively stable (Heikura et al., 2003). The authors comment that these temporal changes are likely to reflect true differences in incidence rates resulting from a complex array of medical, social and educational changes over the twenty-year period. More recently, analysis of the Danish Civil Registration System, which provides continual medical information on all 5.6 million residents in Denmark, identified a cumulative incidence rate of ID at 50 years of 1.58% for males and 0.96% for females (Pederson et al., 2014), figures which are within the range of the 1% estimated for prevalence (Maulik et al., 2011).

1.5 Aetiology

The causal factor for ID remains unknown in almost half of all cases (Maulik et al., 2011). Where known, traditional conceptualisations propose two broad causal factors of ID: aetiology due to biological origin and aetiology due to sociocultural/familial factors (Grossman, 1983; Hatton, 2012; Ziegler, 1967). This dichotomy is deemed to reflect differing developmental pathways (Hodapp et al., 1990) with differing levels of impairment: biological causes being linked with severer levels of impairment and sociocultural/familial aetiologies being associated with milder levels of impairment (Simonoff, 2015). This 'two-group' theory is now deemed overly simplistic given the widely differing level of intellectual impairment among individuals with the same biological aetiology (e.g. Down syndrome) and among individuals with the same sociocultural/familial aetiology (e.g. social deprivation) (Simonoff, 2015).

AAIDD expanded the concept of aetiology to a multifactorial construct comprising four 'categories of risk': biomedical, social, behavioural and educational (Schalock et al., 2010). Each category of risk occurs over the lifetime of the individual, classified according to three time periods: prenatal, perinatal and postnatal. Prenatal risk factors may include chromosomal disorder (biomedical), domestic violence (social), parental alcohol use (behavioural) and lack of preparation for parenthood (educational). Of particular interest are recent biomedical advances in genetics which have led to the identification of a number of behavioural phenotypes for ID including Down syndrome, fragile X, Prader–Willi syndrome, Rett syndrome and tuberous sclerosis complex. By definition, behavioural phenotypes identify a 'heightened probability or likelihood that people with a given syndrome will exhibit certain behavioural and developmental sequelae relative to those without the syndrome' (Dykens, 1995: 523). Increased incidence of early onset dementia such as Alzheimer's disease, for example, is well evidenced among people with Down syndrome (Bush & Beail, 2004). While behavioural phenotypes are clearly useful in predicting the support needs of individuals with specific chromosomal disorders (Bertelli et al., 2009; Hatton, 2012), caution is required to avoid self-fulfilling prophecies or diagnostic overshadowing where the identification of a behavioural phenotype is equated with the inevitability of specific behaviours (Hatton 2012).

Perinatal risk factors for ID may include birth injury (biomedical), lack of access to perinatal care (social), parental abandonment (behavioural) and lack of medical referral for intervention services at discharge (educational) (Schalock et al., 2010). Postnatal aetiologies may include malnutrition (biomedical), family poverty (social), child abuse and neglect (behavioural) and delayed diagnosis (educational) (Schalock et al., 2010).

The precise impact of many risk factors for ID, however, remains poorly understood (Hatton, 2012). The relationship between socio-economic disadvantage and ID illustrates the complexity and bidirectionality of risk factors (Simonoff, 2015); poverty is a risk factor for exposure to both environmental and psychosocial hazards associated with ID (Emerson, 2012; Emerson & Hatton, 2007); having an ID is a risk factor for under- and unemployment and associated poverty (Siperstein et al., 2013); caregiving for a family member is associated with increased risk of poverty due to additional costs of transport, childcare, etc., and reduced rates of maternal employment (Parish et al., 2004; Shahtahmasedi et al., 2011). While numerous and complex, the aetiology of ID is deemed preventable in a substantial proportion of cases (Gustavsson et al., 2011).

1.6 Morbidity

ID is associated with significant health problems (Gustavsson et al., 2011) which tend to differ by severity rather than type of ID (McLaren & Bryson, 1987). A pioneering population-based Dutch study highlighting the disparity in health conditions between people with and without ID identified those with ID having on average 2.5 times more health problems than the general population, with significantly higher rates observed across a number of somatic conditions including multiple congenital abnormalities, epilepsy, musculoskeletal conditions, visual difficulties, deafness and obesity (van Schrojenstein Lantman de Valk et al., 2000).

While people with ID were traditionally reputed to necessarily have poorer physical health (Krahn et al., 2006), the observed disparity in health status with the general

population is now conceptualised as resulting from a myriad of factors including genetics (e.g. thyroid problems associated with Down syndrome), social circumstances (e.g. social isolation, inadequate attention by care providers to health needs), environmental factors (e.g. exposure to contaminants, residential settings that promote inactivity), individual behaviours that contribute to co-morbidities due to inadequate access to health promotion (e.g. nutrition) and inadequate access to healthcare (Krahn et al., 2006). Disparities of access to routine healthcare are well evidenced; people with ID report lower rates of screening for cancer, blood pressure, vision, health and cholesterol (Chan et al., 1999; Iezzoni et al., 2000; Ramierz et al., 2005). These challenges of health status and access to healthcare have been reported as a 'cascade of disparities' (Krahn et al., 2006) which are deemed 'health inequities' (Havercamp & Scott, 2015) defined as 'unnecessary and avoidable but, in addition, are also considered unfair and unjust' (Whitehead, 1990: 5).

Adults with ID also experience high rates of mental ill-heath (Cooper & van der Speck, 2009). A systematic review of sixteen papers from 2003 to 2013 reported prevalence estimates of psychiatric disorders ranging from 13.9% to 74% (Buckles et al., 2013) with the authors noting that the higher end estimate may be atypical given the study identified 'psychiatric symptoms' in a population over 65 years (Strydom et al., 2005). More commonly, studies report estimates within the 30–40% range (Cooper & van der Speck, 2009; Enfield et al., 2006; Morgan et al., 2008) but observe marked variation by level of ability. A study of 113 adults with ID in residential care, for example, of whom 88% were classified within the severe and profound range, identified 83% to have a psychiatric diagnosis (Felstrom et al., 2005). A large population-based study of 1,023 adults with ID, of whom just 36% had severe-to-profound ID, used the same diagnostic measure as the study above and identified 35% of the sample as having psychiatric co-morbidity. The most common psychiatric conditions reported in this population-based study were problem behaviours (18.7%), affective disorder (5.7%) and autistic spectrum disorder (4.4%). Factors found to be associated with the presence of these conditions included having profound or severe ID, experiencing life events in the preceding twelve months, higher contact with a physician in the previous twelve months, being female, being a smoker and living with paid support. In contrast to the general population, no association was reported between these conditions and living in a deprived area, not having a daytime occupation, epilepsy and marital status (Cooper et al., 2007). The findings from this study are considered particularly robust given the use of multiple assessments, comparison of multiple criteria findings and classification of findings based on severity of ID (Buckles et al., 2013).

1.7 Mortality

Compared to the general population, people with ID have shorter life expectancy and increased risk of early death (Hollins et al., 1998; McGuigan et al., 1995). For those with mild ID, however, life expectancy is approaching that of the general population (Patja et al., 2000; Puri et al., 1995). The pattern of differing mortality by level of disability is illustrated by the survival probabilities of 8,724 individuals included on a database managed by the Disability Services Commission of Western Australia (Bittles et al., 2002); median life expectancy declined from 74.0 for persons with mild ID, to 67.6 for those with moderate ID, to 58.6 for those with severe ID.

A similar association with level of ID is observed regarding causes of mortality. A comprehensive Finnish study examining cause of death followed 2,369 individuals with

ID over a thirty-five-year period. Of the 1,095 individuals who were deceased at follow-up, the cause of death for those with milder ID mirrored that of the general population, but this trend diminished for those with more severe levels of disability (Patja et al., 2001). Across all levels of disability, vascular disease was the most common cause of death, followed by respiratory diseases and cancer.

Attempts to reduce health inequalities are required to reduce the mortality of people with ID from conditions with potentially preventable causes (Tyrer et al., 2009). This inequity was sharply placed in focus by a UK confidential inquiry into premature deaths of individuals with ID (Heslop et al., 2014). The most common underlying cause of death for 247 persons with ID was heart and circulatory disorders, followed by cancer. On examination, almost half of these deaths (48%) were deemed avoidable, defined as preventable through good-quality healthcare or public health intervention. The report concludes that 'despite numerous previous investigations and reports, many professionals are either not aware of, or do not include in their usual practice, approaches that adapt services to meet the needs of people with learning [intellectual] disabilities' (Heslop et al., 2014: 5).

1.8 Conclusion

This chapter has provided an overview of the epidemiology of ID. Two recurrent themes emerge. First, that the level of ID rather than type is a key factor impacting on individuals' somatic and psychological health, as well as influencing their life expectancy. Second, inequities pervade the literature. ID is preventable in a substantive proportion of cases; a 'cascade of disparities' is observed with regard to health status; and almost half of all deaths in a confidential inquiry were deemed avoidable. As people with ID and their advocates make strident efforts to secure their inclusion in society, it is imperative that these inequities are addressed to ensure a level playing field for all.

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