Mental Health, Intellectual and Developmental Disabilities and the Ageing Process

Vee P. Prasher
Philip W. Davidson
Flavia H. Santos
Editors

Second Edition

Springer

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iassidd
International Association for the Scientific Study of Intellectual and Developmental Disabilities

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This book is dedicated to the many thousands of people with IDD who are ageing with compromised psychiatric or behavioural health. Some I was able to help, many I was not.

VPP. To my father Bhopinder Partap (1930–2018) who at age 19 years left India in 1951 to come and settle in the UK. As the 'First Punjabi in Derby', he was an inspiration to his family, friends and community. Much loved and much missed.

FHS. To Helena, Tania and Ana Luisa. The indissociable part of me which is brighter, empowering and genuine.

Foreword

The growing life expectancy of adults with intellectual and developmental disabilities comes with both challenges and opportunities in providing the supports and services needed and addressing their age-related physical and mental health changes. In addition to the challenges of increased chronic conditions with ageing and unique neurodevelopmental issues for some subgroups of people with intellectual and developmental disabilities are the health care disparities and social losses they often experience. On the other hand, with age are also opportunities for meaningful self-determined lives.

This book addresses a critical aspect, that of mental health and behavioural issues in these adults. As noted in the book, adults with intellectual and developmental disabilities have higher rates of mental health disorders than the general population and for many of the diagnoses the rates are even higher for the older adults. Yet even the rates reported in the research are likely to be underestimates as there are many barriers to obtaining accurate diagnoses and prevalence rates. A landmark conference, the *Healthy Aging: Adults with Intellectual Disabilities,* held by the World Health Organization (WHO) and the International Association for the Scientific Study of Intellectual and Developmental Disabilities (IASSIDD) 20 years ago, noted the importance of addressing the mental health and behavioural aspects of this population. That conference's goals were (a) to examine the general health status of adults with intellectual disabilities, (b) identify the conditions that support their longevity and promote healthy ageing and (c) propose health and social inclusion promotion activities that would universally foster sound health and improve quality of life. The findings and conclusions of four task forces resulted in four special reports (physical health, women's health, mental and behavioural health, and ageing and social policy) and a series of three books, including the third book *Mental Health, Intellectual Disabilities and the Aging Process* [1]. The present book builds on this work and includes updates, progress, and new issues and approaches emanating in the last two decades.

In addressing mental health and ageing in intellectual and developmental disabilities the field is recognising the importance of taking a life course approach that goes beyond the medical model to examine not only the biological and

neurodevelopmental aspects but also the socio-environmental aspects. In addition there is a growing emphasis on respecting human rights and self-determination as noted in the *United Nations Convention on the Rights of Persons with Disabilities* (CRPD) (2006), which declares that all people with disabilities should have the full and equal enjoyment of all human rights, fundamental freedoms and inherent dignity. Hence, to promote healthy ageing and mental well-being, it is important to understand and respect the lived experience and the desires and preferences of people with intellectual and developmental disabilities. Secondly, a need exists to consider the social determinants in their lives such as the economic, cultural, housing, employment, and health care access factors impacting them.

Furthermore, as they age these adults are likely to face not only ageism but also ableism, which can cause further stigma and discrimination. Ableism is the discrimination of and social prejudice against people with disabilities based on the belief that typical abilities are superior. At its heart, ableism is rooted in the assumption that disabled people require 'fixing' and defines people by their disability. For people with intellectual disabilities this also results in under-recognition of their mental health conditions and other health conditions due to assumptions that symptoms are just manifestations of the disability. Ageism refers to negative stereotypes about ageing, such as assuming that older persons are unable to change their behaviour or that they inevitably experience cognitive and physical decline. Hence, mental health problems—such as cognitive impairment or psychological disorders caused at least in part by complex pharmacological treatments—often go unrecognised and untreated. The combination of these negative assumptions results in people with intellectual and developmental disabilities still generally regarded as a devalued class and often disadvantaged when attempting to access or secure social and health services.

This book brings the expertise of international researchers to provide the latest information on the scientific and medical advances that can benefit the mental health of people with life-long disabilities. It focuses on several key issues faced by this population and ways to address them. In addition to the lack of consistent and precise data on the prevalence rates of people with intellectual and developmental disabilities as they age there is a need for more robust studies on the efficacy and effectiveness of physical health and behavioural health care interventions. In general, mental health interventions can be divided into individual treatments (such as medication or individual therapy) or systemic approaches that provide supportive environments and policies. There is a fairly strong literature demonstrating the effectiveness of behaviourally based treatment, with the focus largely on developing behavioural plans to promote compliance or the use of medications to control behaviours, often with not enough attention to potential underlying mental health or medical conditions making recovery unlikely. An emerging literature is exploring psychotherapy interventions for both challenging behaviours and mental illness for people with intellectual and developmental disabilities, including dialectic behaviour therapy and cognitive behaviour therapy, though more data is needed to determine effectiveness.

Furthermore, a need exists for systemic practices to bridge the health and behavioural health services and fields. The intellectual and developmental disabilities field has not yet embraced the recovery model—a process of change through which individuals improve their health and wellness, live a self-directed life and strive to reach their potential, which is now promoted in the mental health field.

In order to truly understand the mental health experiences of people ageing with intellectual and developmental disabilities with mental and behavioural challenges, they need to have input in the research about them and in the type of interventions they receive. Persons with intellectual and developmental disabilities have not been well represented in health research, partly due to their difficulties in communicating and participating in traditional assessments. The reliance solely on proxy respondents to represent their needs is not only contrary to a person-centred approach but is also inadequate for subjective measurements. However, even in instances where research incorporates self-report data, the variability in processing abilities of people with intellectual disabilities can complicate data collection.

Advancement in the field will depend on strengthening the bridges between the intellectual and developmental disabilities, the mental health and the gerontology fields in research, practice and policies. The goals in all three of these fields intersect to improve the health and quality of life of people ageing with intellectual and developmental disabilities with mental health and behavioural challenges.

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Abbreviations

AD	Alzheimer's disease
ADHD	Attention deficit hyperactivity disorder
APA	American Psychiatric Association
ASD	Autism spectrum disorder
AT	Assistive technology
BPSD	Behavioural and psychological symptoms of dementia
CBT	Cognitive behavioural therapy
CT	Computerised tomography
DAD	Dementia in Alzheimer's disease
DC-LD	Diagnostic Criteria for Psychiatric Disorders for Use with Adults with Learning Disabilities/Mental Retardation
DLD	Dementia Questionnaire for Learning Difficulties
DMR	Dementia Questionnaire for Mentally Retarded Persons
DS	Down syndrome
DSDS	Down Syndrome Dementia Scale
DSM	Diagnostic and Statistical Manual of Mental Disorders (several versions)
EEG	Electroencephalography
GERD	Gastroesophageal reflux disease
IASSIDD	International Association for the Scientific Study of Intellectual and Developmental Disabilities
ICD	International Classification of Diseases and Related Health Problems (several versions)
IDD	Intellectual and developmental disabilities
IQ	Intelligence Quotient
MDT	Multidisciplinary team
MMSE	Mini Mental State Examination
MRI	Magnetic resonance imaging
NICE	The National Institute for Health and Care Excellence
OCD	Obsessive compulsive disorder

PAS-ADD	Psychiatric Assessment Scale for Adults with Developmental Disability
SIB	Severe Impairment Battery
WHO	World Health Organisation

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Part I
Clinical Issues

Chapter 1
Introduction

Vee P. Prasher, Philip W. Davidson, and Flavia H. Santos

Clinical experience and research findings have now established that older adults with intellectual and developmental disabilities (IDD) are at a high risk for mental and behavioural health problems. Prevalence rates for psychiatric and behavioural disorders among older persons with IDD, can be difficult to assess accurately, but problems can be as high as for persons without IDD and be comparable to the younger IDD population. The reasons for these trends are not yet fully understood, but it seems evident that cognitive, physical and social factors interact together to affect both the mental and behavioural health. Further, although specific risk factors for each outcome have yet still to be understood, the inter-play between physical health and mental and behavioural disorders, well known in general medical and psychiatric practice, seems even more important in affecting behavioural and psychiatric outcomes in older persons with IDD. Although a wide spectrum of challenging behavior and mental illnesses seen in younger adults with IDD can continue to varying degrees into older age, a number of disorders are first evident in older age e.g. dementia. As per the IDD population as a whole, historically, providers and policy makers have given very little attention to mental health of older persons with IDD. It was not until the beginning of the deinstitutionalisation movement that

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occurred in most developed countries in the latter decades of the twentieth century, that progress was made in understanding the overlap between mental health and getting old in the IDD population. Since then relatively good effort has been made to understand causes, presentation, treatment of mental and behavioural problems in the ageing IDD population.

Effective psychopharmacologic and behavioural intervention methods to permit older persons with IDD who were affected by behavioural and psychiatric difficulties to live and be integrated into community settings are now available. Progress is being made in linking psychiatric diseases to specific neurotransmitters and new medications are now being made available to affect neurochemical corrections. Subsequent use of these medications is slowly infiltrating the pharmacopeia for older persons with IDD and are replacing less effective medications while reducing the likelihood of unacceptable side effects and the problems inherent in polypharmacy. Alongside pharmaceutical developments behavioural psychology research is bringing forth treatment approaches for individuals with IDD that effectively reduces severe behavioural morbidity that previously would have jeopardised independent living in the community for affected persons and relegated them to lifelong institutional care. Further, improvements in social care, independence and self-advocacy are enabling a more person centred approach to be delivered.

These successes have led to a greater number of older adults with IDD living and functioning successfully in the community. By the year 2040, the number of persons with IDD age 65 years and older-who now represent about 12% of the IDD population- will grow to about 25%. Additionally, about the same proportion of these persons who now have what we may term a ‘dual diagnosis’ (i.e. both an intellectual and psychiatric impairment) will retain that status into older adulthood and old age. Unfortunately, our scientific and treatment literature still has limited information for us to draw upon to make the appropriate decisions about best practices as more elderly individuals with dual diagnosis present for services.

Goals of the Book

This book reviews the up-to-date scientific and practice literatures pertinent to mental health and behavioural disorders in older adults with IDD. Clinical issues and best therapeutic practices are highlighted along with suggestive models for both community services and professional education in the field. This work has grown out of conferences sponsored by the International Association for the Scientific Study of Intellectual and Developmental Disabilities (IASSIDD), a non-governmental member of the World Health Organisation (WHO), that address the health and mental health practice and policy needs of older persons with IDD. Previous goals addressing biobehavioural issues resulting from the 1999 WHO Conference on

Healthy Aging: Adults with Intellectual Disabilities (Adapted from [1]) concerning mental health and behavioural issues in older persons with IDD are as follows:

- To improve the understanding of normal psychological functioning throughout the lifespan
- To improve knowledge and awareness of age-related stressors and their impact
- To understand and appreciate the social, cultural, environmental and developmental context of behaviours and their functions
- To improve the detection and holistic assessment of mental disorders such as depression, anxiety and dementia
- To increase mental health knowledge and skills in professionals, carers and families
- To develop living environments that are responsive to the mental health needs
- To promote mental health and minimize negative outcome of mental health problems
- To increase mental health services and supports in communities
- To collaborate with older people with IDD and their support system in developing culturally sensitive, humane and minimally restrictive mental health interventions with an integrated bio-psychosocial orientation.
- To improve the quality of life in older people with IDD and mental health problems
- To develop a research agenda that will provide evidence concerning each goal for all nations.

Findings from work undertaken by IASSIDD regarding the problems people face as a result of dual diagnosis ([2], p. 7) were as follows:

'Most research in the area of mental or behavioural disorders or problems has had treatment as its focus. Much less has been done about the causes and risk factors of such disorders and their prevention. Almost all of the data available come from populations of persons with IDD from nations with established market economies, where research funding has been most available and there has been a critical mass of workers who specialised in this field. For instance, prevalence data for psychiatric and behavioural disorders may differ between nations with established market economies and developing nations and treatment outcomes may vary where the cultural ethos may inhibit referrals and special resources or services are limited. Improved health status and prevention in developing nations, the principal goal of WHO, must depend on identification of special issues pertaining to developing nations and application of techniques that permit information to be gathered free of cultural or other restraints.

Well-controlled research in mental and behavioural disorders as they occur in persons with IDD is limited. Most of the work over the past 30 years addresses treatment issues; fewer focused on diagnosis or aetiologic factors, or prevention. Only a small number address basic mechanisms. These disappointing data probably reflect

several things, including a well-known lack of a research focus or funding. As a consequence, there are limited numbers of scientists in the field and a lack of programmatic efforts in research centres addressing any relevant issue related to IDD. Without specific attention from health planners and ministerial level policy makers, as well as a critical mass of investigators working on a common problem in programmatic ways, little converging data can emerge and, quite likely, few if any major discoveries will appear quickly.

> Promising lines of inquiry relate to both treatment strategies and biological determination and regulation of behaviour. Rigorous methodologies are available to undertake controlled or randomised clinical trials for behavioural and pharmacologic interventions. Recent advances in molecular genetics and neuropharmacology provide new opportunities for linking severe behavioural and psychiatric disorders to brain neurochemistry. The field must move toward a research focus that includes a better balance of studies of basic mechanisms, translational and clinical outcome studies.

The principal aim of this book is to review and update what is known in the areas of mental health and behavioural issues in older persons with IDD with a view to improving clinical and research practice. We have organised the book into three broad sections. Part 1 addresses clinical specific issues, Part 2 looks at treatment and interventions, and Part 3 considers service system issues. In Part 1, the chapters by Wark (Chap. 2) and Mulryan and colleagues (Chap. 3) explore and evaluate prevalence of behavioural and mental health disorders and the range of psychopathology that may be seen in older persons with IDD. As these authors point out, estimating prevalence is fraught with pitfalls, starting with our lack of any common agreement on what constitutes a behavioural disorder and on the complexities of recognizing psychopathology, which are often more difficult in adults with IDD than in the general population. Chapter 4 (Bertelli and Bianco) and Chap. 5 (Gomiero and colleagues) highlight the complex issues of assessing mild and major neurocognitive disorders in persons with IDD. The next five chapters (Chaps. 6–10) address specific issues rarely discussed at any length in the literature on older adults with IDD: depression (Prasher and colleagues), interaction between mental and physical health (Herrera and Sulkes), emotional health and Down syndrome (Patti and colleagues), mental health in persons with autism (Oakes) and finally in Part 1, Guerin and colleagues' chapter on end of life, bereavement and grief (Chap. 10) is particularly important in relation to older adults with or without IDD; however, in the case of adults with IDD it is even more important, as the lack of typical ability to communicate often coincides with the growing probability of experiencing loss and manifesting problematic behaviours. In extreme instances, unfocused grief may then present as a more complex mental illness. Models for tackling this common and quite treatable outcome are discussed.

Part 2 of this book explores various traditional treatment and interventions, including psychotherapy (Chap. 11; Weber and Streicher), psychotropic medications (Chap. 12; Evans and Trollor) and anti-dementia medications (Chap. 13; Prasher and colleagues). In each case, the authors emphasize recognised approaches that have evolved for addressing the mental health needs of adults with IDD as they age. To complement these explorations, Cea and colleagues in Chap. 14 discuss

treatment ethics, an issue of growing concern where vulnerable populations are concerned and when the usual medications used to treat persons without IDD may not always have the expected result in older adults with IDD. Chapters by McCallion and Ferretti on the impact of mental illness on carers (Chap. 15) and by McGrew on the role of assistive technology (Chap. 16) round off these explorations. The final chapter (Chap. 17) of Part 2 examines successful and positive ageing in older adults with IDD (Santos and Heyn).

Rounding off the book, we have Part 3 on service system issues. Factors affecting biobehavioural and dual diagnosis issues are very complex, ranging from a lack of understanding of the factors contributing to behaviour, to a deficit in well-trained personnel for assessment, treatment and care, and to a lack of available community-based systems of care that are accessible to persons who pose special challenges. Such deficiencies in community care are found worldwide, equally affecting both developed and developing nations. In some instances the deficiencies are the result of a lack of public realisation of the need or commitment to specialised services for older people with IDD; in other instances they may be the result of financial constraints and the paucity or lack of allocation of scarce public resources to these types of services. In other areas they may be the result of the lack of sophistication of mental health professionals and care providers, where the special needs or situations of older people with IDD go misunderstood and confounded with archaic psychiatric treatment and hospitalisation practices. Lastly, they may be due to the confusion around behavioural presentations and the misattribution of the diagnosis of mental illness to rather mundane and treatable maladaptive behaviours.

Hemmings and Bouras describe innovative models for community services for older adults with IDD (Chap. 18), the living arrangements of older persons with IDD (Linehan) and impact on families of ageing in a relative with IDD (Watchman) are discussed in Chaps. 19 and 20 respectively. Van Heumen and Schippers address quality of life issues (Chap. 21) and by including a chapter on Eugenics by Reinders and colleagues (Chap. 22) we aim to help put to rest the idea of social engineering as an acceptable way of excluding the more disabled individuals from society.

Final Thoughts

The scope of this book is mainly limited to older adults with IDD, but we accept the myriad issues affecting adults in their mature years, do overlap broader age-span issues (when appropriate). We believe that, with the exception of organic age-associated conditions such as dementia and depression, mental or behavioural difficulties occurring in older age may, and almost always do, have origins in earlier years.

We have used the term ‘intellectual and developmental disabilities’ throughout the text, recognizing that our target population encompasses a diverse group of individuals with chronic conditions that are associated with mental and/or physical impairments. More than just cognitive deficits. We remain mindful that in some

countries other terms are used (such as mental retardation, learning disability, mental handicap, intellectual disability) may still have prominence. We consider these terms synonymous. We have asked our contributors to use these terms with care and within the context of their targeting meaning.

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Chapter 2
Prevalence of Mental, Behavioural, and Neurobehavioural Disorders

Stuart Wark

Introduction

Since the start of the twentieth Century, a combination of enhanced health-care provision, the development and implementation of new life-saving technologies, and a greater appreciation for the rights for people with individuals with intellectual and developmental disabilities (IDD) has seen the life expectancy of this cohort increase dramatically [1–3]. Improved knowledge and the timely provision of health services across the lifespan has resulted in many individuals with complex care needs surviving childhood [4] and now living into adulthood and beyond, even with severe physical and mental health co-morbidities [5]. It is recognised that, in developed countries all around the world, people with intellectual disabilities are now living longer than at any time in recorded history [6–9].

The improvements in early medical diagnosis and treatment, when combined with appropriate ongoing care management, has seen progress that now means that a person with a mild IDD in the United States has a life expectancy that exceeds 70 years [10]. If people who have a specific genetic disability, such as Down, Angelman or Prader–Willi syndrome, as well as those individuals with complex and severe associated health co-morbidities, are removed from the total figures, the life expectancy of an Australian individual with IDD now approximates that of the mainstream [11]. Similarly, a United States study estimated the difference in lifespan between the standard population and a person with a mild IDD to be only 2% [12].

The present cohort of people with IDD is the first that has lived into chronological old age in large numbers, and can also reasonably expect to outlive their parents

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[8]. While it may seem obvious that older people with IDD will face additional problems when compared to the mainstream ageing community, it should not be automatically assumed that every individual will experience significantly worse health or social problems [13]. Nonetheless, the mere extension of life does not necessarily equate to a high quality of life if health issues are not appropriately treated [14], and there is limited data and information regarding the mental health of people with an IDD as they grow older. This lack of knowledge in turn limits the capacity of support services to effectively meet emerging and changing mental health needs.

It is important to recognise that many older people with IDD may be just as 'healthy' as other older persons without long-standing disabilities. However, service providers and government bureaucracy will often argue whether the lifelong IDD or a newly emerging ageing-related problem is the predominant issue that requires support [15], and this approach can lead to a focus upon areas of weakness, rather than the continuing strengths of the individual. It can result in a distorted view that older people with IDD are more dependent and require a greater level of care than is actually needed. The concentration upon the care needs of the individual, rather than upon the maintenance of existing skills and abilities, can lead to a situation in which people with IDD are prematurely viewed as being non-productive and unnecessarily reliant upon external assistance as they grow older [16]. With this in mind, there do remain a number of both internal and external issues that impact upon the capacity of people with IDD to be successfully supported as they grow older. One of these factors is the co-morbidity of mental and behavioural disorders in this cohort, and this issue is the focus of this chapter.

Moving Away from the Biomedical Model: An Acknowledgement of Change in Approach

There are a variety of different theoretical perspectives for considering the concept of health in general and, more specifically, the issue of disability. Historically, there has been a strong focus upon medical models and individual rehabilitation in mainstream society; however, concurrently with the emergence of ageing as a significant support issue, the past three decades have seen the proposal of an increasing number of social models of disability [17]. Disability can be defined within a variety of potentially disparate contexts including public health, economics, politics, history and feminism. The various social models of disability facilitate a greater examination of environmental, community and cultural aspects to disability than the previous medically orientated frameworks.

The past 100 years have seen a major shift in thinking with respect to the perception of disability. It has been previously argued that the presence of IDD denies the individual 'personhood' as a result of a perceived inability for self-awareness. This premise was the basis of many now condemned practices, such as widespread

compulsory sterilisation and even euthanasia [18]. Through much of the past century, the dominant conceptualisation of disability saw it related to a diagnostic medical model that was premised around deficit. This approach has been more recently challenged by the alternate view that disability is instead a reflection of human diversity [19] and is impacted by a variety of other factors.

The World Health Organisation (WHO) had initially defined health in 1948 as "a state of complete physical, mental and social well-being and not merely the absence of disease or infirmity" ([20], online). This definition effectively categorized the presence of a disability as inferring the point when health ceased. The diagnostic medical approach to disability was based upon the premise of an underpinning medical problem within the individual. It had an internal focus, with the belief that the person with a disability was in some way defective when compared to wider societal norms. This deficit or problem could then be addressed through medical diagnosis and subsequent intervention [19]. This framework is predominantly derived from the work of Parsons in the 1960s, who argued that an illness provided the individual with an opportunity to ignore or discard the normal socially enforced beliefs of the society. However, with this reduced expectation was an associated insistence upon compliance with curative medical interventions [21]. People with IDD would appear to present a significant problem to this paradigm, as the likelihood of medicine providing a cure for their lifelong problems was minimal. It was argued that the diagnostic medical framework was applicable to individuals suffering from a temporary or acute episode of illness, but not necessarily appropriate to people who were older or who had a permanent disability [22].

Over time, WHO and others started to re-conceptualize the understanding of health to incorporate both medical and social aspects [23] that better accommodated the rights of people with disability. These changes in perception were associated with the possible application of a number of different theoretical models and frameworks, ranging from diagnostic medical perspectives through to a social constructionist perspective [24]. A movement away from a purely medical model can be seen through changes to the research focus of the IDD sector. For example, in the 1970s, 42% of papers in the *Journal of Intellectual and Developmental Disability* had a medical focus. In the following decades, this figure dropped to just 7% of articles being framed within the medical model [25].

This was a welcome shift in approach. It facilitated a greater emphasis on the individual and their specific needs, rather than just being seen simplistically in terms of a medical diagnosis. However, the older diagnostic medical model is not necessarily redundant, and an understanding of population health is still important if we are to better support individuals with their health-care needs. It is acknowledged that social models are more commonplace in IDD research today, but the focus of this chapter is on the epidemiology of mental and behavioural disorders of older people with IDD. In its simplest state, epidemiology can be considered to be study of the disease patterns in either entire populations, or more commonly, in specifically defined sub-groups of an entire population. In practice, this means examining factors such as incidence (number of new cases of the disease), prevalence (number of people currently with the disease), the distribution of the disease, any possible

controls for the disease, and consideration of any other health determinants that may contribute towards these factors. Epidemiology can assist us to understand the distribution of certain disease, what factors may contribute towards increases or decreases in the number of people with the disease, and identify possible methods to assist with reducing the impact of the disease.

This chapter will be primarily on the prevalence of mental, behavioural and neurobehavioural disorders (the disease) among older people with IDD (the population). However, as it is known that some disorders may present for the first time in older persons, in particular many neurobehavioural disorders, there will also be consideration of the factors that may affect incidence in this population. Solely for the purposes of this chapter, both autism and dementia are not considered as either mental or behavioural disorders, as there is separate and more detailed focus (in Chaps. 8 and 12 respectively) within this book on these important issues.

Prevalence of Mental, Behavioural and Neurobehavioural Disorders

What Is Prevalence?

Prevalence, as noted earlier, is the number of people who can be definitively identified as having a specific condition (e.g. disease, disorder, symptom). However, prevalence can be measured in a number of different ways. A 'snapshot' approach can be used, whereby the number of people with a disease at a specific point in time, such as 31 July, is determined, and this is called *point prevalence*. In contrast, *period prevalence* will measure the number of people who have the disease during a longer period of time, such as a month or a year. A third type of measurement, *lifetime prevalence*, examines the number of people who have experienced that disease at any time previously in their life. However, while lifetime prevalence has been commonly used within psychiatric epidemiology, concerns have been raised about its validity and recommendations have been made to cease using it [26]. With respect to prevalence, the focus in this chapter will be predominantly on point or period prevalence, as it is felt that these measures better establish current health issues facing older people with IDD.

What Is an Intellectual Disability and What Is 'Older'?

It is worth noting in advance that there are two pivotal concepts that may differ for readers around the world. These relate to the definitions used to determine whether a person has an IDD and also in relation to the commencement point for someone being considered 'older'.

There is no single accepted definition of precisely what factors allow for the diagnosis of an IDD [27]. There are a number of different clinical classification systems used in research and service delivery areas to define IDD, and these include the World Health Organisation's (WHO) International Classification of Diseases (ICD) [28] and International Classification of Functioning, Disability and Health (ICF) [29], the American Psychiatric Association (APA) Diagnostic and Statistical Manual (DSM) [30], and a definition by the American Association on Intellectual and Developmental Disabilities [31]. While these classification systems are in many ways conceptually quite consistent, the variance in usage between the models has resulted in data inconsistency with respect to accurately establishing the prevalence of IDD [32]. The clinical diagnosis of IDD has traditionally been based upon a measured Intelligence Quotient (IQ) for the individual, however recent years has seen a greater emphasis on also considering any limitations in adaptive behaviours in areas including socialisation, daily living skills and conceptualisation. A diagnosis of IDD for a person is then commonly sub-categorized in relation to its measured severity, including labels such as mild, moderate, severe and profound intellectual disability.

Similarly, there are numerous different, and sometimes conflicting, definitions pertaining to the ageing process that all people undergo. There is no universally accepted classification of what combination of symptoms comprise ageing or being 'older'. The common themes of most definitions relate to changes that occur progressively or over time that result in some loss of function for the individual. Determining precisely when ageing commences in a defined population, or when a specific individual is considered to be older, is not simple. Ageing is not an event that can be accurately predicted and takes place over an extended period of time and varies between every person [33]. As such, providing a clear definition of 'older' for an entire population sub-group, such as people with IDD, is problematic. While ageing should not be viewed as a singularity, many attempts to categorize or define it are usually framed in terms of a specific chronological age, such as 65 or 70. However, this approach has not resulted in a standard definition of ageing as there are no chronologically definitive and consistent physical, psychological or social phenomena associated with ageing that support the establishment of set ageing criteria in the same way that infancy or puberty can be so defined [34].

Researchers worldwide often use retirement or pension ages as a convenient marker in describing the onset of ageing [35], in spite of the fact there appears limited medical or social reasons for this decision. The United Nations has recognised this problem and nominated the use of chronological age as merely a crude mechanism for the identification of population groups and their respective needs [36]. However, various different classifications of ageing have continued to be proposed, with the United Nations generally using the age of 60 to define older people [37].

In the same way that IDD is sometimes sub-categorized into mild, moderate, severe and profound, there have been attempts to stratify the cohort of older people into smaller sub-sets. As one example, one ageing model defines a person aged 65–75 as 'young old', someone 76–85 as 'old old', and people over the age of 85 as the 'oldest old' [38]. Another methodology defines people between 65 and 80 who

are predominantly independent and socially active as ‘third age’ and people who are over 80 years old and requiring more support as ‘fourth age’ [35].

The lack of consistency regarding diagnosis of IDD, in conjunction with no universally accepted definition for ageing, means that there will be differences in the interpretations of research findings due to the variations in criteria used to derive any samples. To allow the reader to determine the relevance of results for their own particular situation, an attempt will be made to note throughout this chapter the specific location of studies (i.e. the city, state, territory or country) and what criteria (i.e. age brackets, severity of disability) were used within the research. This is not always possible, as not all research contains this demographic information.

Problems in Determining Prevalence

Historically, it has been difficult to accurately determine the prevalence of some health issues in older people with IDD. Prior to the 1990s, the majority of research or government reports that noted the prevalence of mental or behavioural disorders tended to be based on *convenience samples*. A convenience sample is one that is used because it is easily accessible and is therefore not necessarily representative of the wider population. For older people with IDD, convenience samples in this context were predominantly of composed of individuals referred through to specialist mental health services for support. This means that these samples are potentially biased, as they may miss individuals who were perceived, correctly or otherwise, to have less serious mental health issues that did not require specialist support, as well as people in areas where there were simply no services available for referral, such as rural locations. These convenience samples were often quite small and attempts to extrapolate their findings across the entire population of older people with IDD is inherently problematic. It is also important to recognise that there may be further data disparities depending upon the methodology used for collection. Personal interviews with an individual with IDD may not result in the same identification of issues as a personal interview with a parent or paid carer, online surveys, a review of medical records and so on.

In order to overcome some of these issues, many researchers have started to use large databases or registries from disability support organisations, health services and government records. Such data can be considered more representative of an entire community and allows for a retrospective analysis of all individuals at a given point in time. It can also include people living independently in the community with minimal support, as well as those referred to specific mental health services. While this approach is preferable to the convenience sampling method, it also has a number of limitations that limit the accurate estimates of prevalence. Registries are often still restricted to certain areas, such as one rural or urban location, and therefore the results may not be generalizable to other areas even within the same country. A solution to this problem would be to combine the findings from numerous different registries, but this approach then raises another issue that impacts on prevalence

estimates; how accurate and consistent is the diagnosis of mental or behavioural disorders across regions of one country, and internationally?

Diagnosis of a Mental Disorder in Older Persons with IDD

In order to determine the epidemiology of any disease, an accurate and consistent diagnosis is required. In the general community, mental disorders are primarily first diagnosed usually by a general practitioner, with additional support from a psychiatrist, gerontologist, psychologist or local mental health team. Mental disorders are not usually diagnosed through medical testing, such as a blood test or magnetic resonance imaging (MRI) scan of the brain. Instead, a doctor will usually discuss the presenting symptoms with the patient, and, after ruling out other possible causes of the symptoms, make a diagnosis based off the provided history. This diagnosis will usually be based upon specified criteria in either the *Diagnostic and Statistical Manual of Mental Disorders* (current version: DSM-V), as published by the American Psychiatric Association [30], or the *International Classification of Diseases* (current version: ICD-11) developed by the World Health Organisation [28].

Research since the 1960s has reported that, in general, people with IDD are more likely to be diagnosed with a mental disorder across their lifespan than the mainstream community [39, 40]. For older people with IDD, the process of diagnosis for mental disorders is largely the same as for the mainstream community, but there are a number of differences that are important to recognise and these are discussed in greater detail below. There has also been recognition of a number of specific mental disorders that require proactive monitoring in the population of older people with IDD. In particular, these include delirium, dementia, depression and chronic psychiatric disorders including anxiety and schizophrenia [41]. The symptoms of many psychiatric conditions in this cohort include impaired attention, a decrease in memory or language, or changes in cognition, motor skills and emotional state [41]. Specific mental disorders and their associated symptoms in older people with IDD are explored in greater depth later within this book (e.g. Depression Chap. 5; Autism Spectrum Disorders Chap. 8).

However, there are significant problems in accurately determining the prevalence of mental disorders, as the process of accurate diagnosis is complex. Assessing both the current and previous mental health of people with IDD presents difficulties for families, staff members and general practitioners, as they often do not have the necessary knowledge or information to accurately identify or recognise mental health issues in this cohort [42]. Even if the problem is accurately identified, the lack of available and appropriate mental health services is known [43], and the problem is magnified by the fact that health professionals are unsure of how or who to refer individuals on for further assessment [44]. This problem becomes even more complex, as psychiatrists believe they are generally "untrained and inexperienced" ([45]: p. 191) in supporting people with both IDD and an emerging or chronic mental disorder [15].

Many of the symptoms of a mental disorder can mirror behavioural disorders and it can be difficult for family members or support staff to distinguish between the two, or to determine whether there is a new psychiatric condition associated with ageing or if it is simply the progression of an existing behavioural disorder. It is therefore important that any change in functional skills or behaviour in an older person with IDD is evaluated carefully [46]. The symptoms of some psychiatric conditions can hide the development of other serious diseases and treatment can be unnecessarily delayed until a proper diagnosis takes place [41]. Further discussion of the assessment of behavioural and psychological disorders can be found in Chaps. 3 and 4.

When considering the use of registries and databases to establish prevalence estimates, the issue of accurate and consistent diagnosis becomes evident. If there is variation in how the presence of a disease is determined, this will result in discrepancy in the data. In particular, if the registry data is gathered across a variety of settings and with multiple different assessment criteria for decision making, the final reported data is potentially biased. This problem becomes evident in assessing mental disorders; there are often no universally accepted definitions for specific mental conditions, and practitioners will use different tools and measures, in combination with their own subjective judgement and clinical experiences, in order to make a diagnosis [47].

Diagnosis of a Behavioural or Neurobehavioural Disorder

While mental disorders are relatively well understood conceptually, albeit with the diagnostic difficulties as discussed above, behavioural disorders are less easy to define. Both DSM-V and ICD-11 refer to behavioural disorders, under a variety of different labels, but they are initially conceptualized in relation to the early life stages of infancy, childhood or adolescence. However, within the wider disability sector, issues associate with ‘challenging behaviours’, ‘problem behaviours’, ‘behaviours of concern’ or ‘complex behaviours’ are seen as occurring across the entire lifespan, and can disappear and re-emerge at various time points or in relation to changing life circumstances, stresses and physical health issues. The current frameworks for diagnosing behavioural disorders, and the distinction between an individual having a behavioural disorder as opposed to displaying behaviours that challenge convention, vary greatly between countries, and from an epidemiological perspective, it remains difficult to precisely define what constitutes a behavioural disorder in order to then determine the prevalence within older populations of people with IDD.

Within the adult population, behavioural disorders are generally diagnosed when an individual displays or performs repeated actions that fail to conform to societal expectations of age-appropriate conduct. As examples, such behaviours in adults with IDD may include physical or verbal aggression towards other people, animals or property, self-injury, impulsive or reactive deeds, or sexually deviant actions. In

this context, these behaviours may be perceived to be 'irrational' from the perspective of family, friends, carers or the wider community. This is not to say that the behaviour is irrational; instead it is often that the reasoning behind the actions is simply not immediately evident to anyone other than the individual. However, simply displaying such behaviours is not necessarily a sign of a behavioural disorder. For example, physical aggression is well known to be associated with undiagnosed pain, particularly individuals with severe or profound disability and communication impairments [48, 49]. Diagnosis of a behavioural disorder in this situation would be inappropriate, as the behaviour generally disappears once the cause of the pain is removed [50], but differentiation is highly complex and results in significant ambiguity in definitive diagnoses.

In contrast to general behavioural disorders, neurobehavioural disorders are considered more specific in that they occur in individuals who have either experienced a neurological event, such as a stroke or traumatic brain injury including ischemia or hypoxia, or have an emerging disease that affects their neurological function, such as Alzheimer's disease and other dementias, Parkinson's or multiple sclerosis. Behavioural disorders in people with IDD are usually diagnosed in childhood and then may recur across their entire lifespan. However, neurobehavioural disorders are different to behavioural disorders in that they arise following specific health events that are often associated with ageing. The manifestation of behavioural and neurobehavioural disorders may be quite similar, even if the origins may vary. Any older person is susceptible to such diseases, and may therefore experience neurobehavioural disorders as they age. It is known that the process of diagnosing neurobehavioural disorders in the mainstream community is poorly understood, and treatment and support for these disorders is either ignored or only cursorily considered [51]. However, for older individuals with IDD, the diagnosis process can be further complicated by the existence of pre-existing behavioural disorders and comorbid mental disorders.

Epidemiology of Mental or Behavioural Disorders in Older Persons with Lifelong IDD

While it is acknowledged that there are a large number of limitations in determining the epidemiology of mental or behavioural disorders in older people with IDD, there is a growing research quantum from around the world that estimates the occurrence of these issues. The following section is an attempt to synthesize the general findings of the research base; it is not designed as a systematic review or intended to encapsulate all the research in this area.

Again, it is worth noting that the varying definitions of both 'IDD' and 'older' will see reporting on potentially different samples. It is also important to recognise that, as the cohort of people with IDD has only recently started ageing with significant numbers, many studies will have aggregated data as the sample sizes would be

otherwise too small. This means that some research will combine together all levels of disability and all people aged over 60 (for example), while larger studies may have been able to stratify their sample into both different age cohorts (e.g. 60–65, 66–70, 71–75 and so on) and severity of IDD. Readers are encouraged to ensure that they are cognizant of these potential differences and to not attempt to compare findings across disparate samples.

The Pioneering Research

As discussed, both earlier within this chapter and then again in later sections of this book, ageing within the IDD population is a relatively new development associated with a rapidly increasing life expectancy during the twentieth Century [46]. One of the earliest research papers on the issue of ageing and disability was written in United States in the early 1960s by Dybwad, with the report detailing the need for improved planning and additional funding for individuals [52]. Further research over the following decade was spasmodic [53–55] commenting on the changing requirements of older people with a disability.

In the 1970s and 1980s, led by pioneers such as Janicki [56–59] in the United States, research into ageing with IDD was promoted and became more common, but the field was still very much in its infancy. Many of the studies were undertaken in the United States and Europe, and examined a variety of different aspects of ageing in this cohort including occurrence [60], predicted life-span [61], language training opportunities for older people [62], deinstitutionalisation [63], medical problems [64], decision making by ageing carers [65], and social care issues [66] amongst others.

While there was more research looking generally at ageing with IDD, there was minimal focus on mental or behavioural disorders amongst this cohort. Of this research, the majority was based around small clinical populations, with limited attempts to assess the epidemiology of disorders across wider areas. Corbett's 1979 research is recognised as one of the first reports on the prevalence of mental or behavioural disorders among older people with IDD [67]. Drawing on information gathered from carers of 110 individuals aged over 60 living in London, Corbett reported ten diagnoses of mental disorders (including schizophrenia, psychoses and neuroses) and 23 behaviour disorders.

While it was still spasmodic, the 1980s started to see more researchers examining the phenomenon of mental and/or behavioural disorders amongst older people with IDD. In a small comparison sample within a residential aged facility (nursing home) in the United States, Cotton and colleagues [68] noted that the prevalence of behavioural disorders was actually lower among people with lifelong IDD than their ageing peers without lifelong IDD. Day [69] reported on a retrospective analysis of the prevalence of mental and behavioural disorders amongst 357 hospital patients aged over 40 years in England. While there was an overall prevalence of mental disorders of approximately 30%, Day noted a consistent decline in the rate of

mental disorders as individuals grew older, with diagnoses of psychiatric illness falling from a high of 48.7% in the 40–49 year old group progressively to 30.3% (50–59 years), 25.2% (60–69 years) and 16.7% (70 years and above). This pattern was replicated for behavioural disorders, with a reduction in diagnoses from 29% in the youngest cohort to 4.2% in the oldest group. In a second English study, Day [70] noted a prevalence rate for psychiatric disorders of approximately 20% in a cohort of long-stay hospital residents aged 65 and over. Other small-scale United States or United Kingdom studies [71–73] noted the prevalence of mental disorders at similar rates for older people with IDD when compared to mainstream community peers. Others noted higher prevalence rates for older people with IDD; Menolascino [74] in the United States noted increasing prevalence of 30% in older people with IDD, while Iverson and Fox [75] reported prevalence rates of 35.9% in a United Sates sample of adults aged over 41 years.

One of the first national-level projects was undertaken by Andersen and colleagues [76] in 1987. They used 1977 nursing home data from across the United States and noted that 7% of residents aged over 54 years with an IDD also had a diagnosis of a co-morbid mental disorder. Jacobson and Harper [77] also used a United States national sampling approach to establish survey data regarding 379 people with IDD aged 55 and above. Their results differed from those reported by Day [69] in that mental disorders did not appear to decline with age. The rate of psychiatric diagnoses was 19.3% in the 55–59 year category, then went up to 22.7% in the 60–74 group, and finally decline slightly to 21.3% in those aged 75 and over. There were very high reports of 'behaviour problems' across all three age cohorts, with examples including *tantrums* (75.9%, 50.9% and 21.3% in the 55–59, 60–74 and 75 and greater groups respectively), *runs/wanders away* (37.3%, 20.8% and 21.3%) and *hyperactive* (27.7%, 53.2% and 21.3%) amongst others. It is also worth noting the report of *suicidal threats/attempts* occurring in 21.7% of 55–59 year olds; even now, the potential risk of suicide for people with IDD is still not widely recognised as being of concern [78].

An Increasing Knowledge Base

The data regarding mental health and behavioural disorders in older people with IDD that was gathered during the 1970s and 1980s was not particularly comprehensive and, as is evident from the previous section, was sometimes contradictory. For example, while there were studies that indicated the prevalence of mental disorders decreased with age [69, 79], some reported that it didn't change [77], and further others noted increases in the diagnosis of mental disorders as individuals got older [58, 80]. Nonetheless, this base provided a framework for future work to build on, and the following decades saw considerable growth in the number of larger-scale projects that focused on this area. However, in spite of this increasing knowledge, accurately determining prevalence of these disorders remained difficult, even within specific countries. As an example of the wide disparity in reporting, Campbell and

Malone [81] noted estimates of prevalence rates for mental disorders in people with IDD varied from a low of 14.3% to a high of 67.3%. Borthwick-Duffy [82] also commented on the vast discrepancy in prevalence rates, with a variation in estimates from below 10% to over 80%. However, much of this research was still focused on people with IDD across all age categories, with minimal consideration or stratification of the data specifically to consider the prevalence among older individuals.

Findings from a small project with 23 people aged over 65 without Down syndrome living in two regions of London was reported [83]. Of this cohort, 17 (74%) had psychiatric symptoms, although only 30% had a current or former diagnosis of a mental disorder. Behavioural disorders were also common, with 27% of the sample having more than 10 'maladaptive' behaviours, such as physical violence, property destruction or self-injurious behaviours. Some researchers have specifically examined whether age was related to the diagnosis of mental disorders, and they reported that there was no difference after adjustment for cognitive capacity and social skills [84]. A further study [85] of 105 individuals aged over 50 years with a range of level of disability reported that 21% of the participants would be diagnosed with a psychiatric condition.

In a study [86] of 124 individuals with IDD aged 60 years and over, prevalence of mood disorders (8.9%) and schizophrenia (6.5%) was reported. This was one of the earlier studies that started to report on specific diagnoses, rather than just a general heading of 'mental disorder'. A nation-wide study [87] reported on data gathered from 1581 Dutch individuals with IDD, 307 of whom who had Down syndrome, from 83 different group homes and 24 institutions. This study incorporated a wide range of ages, including 715 people aged 50 years and above. It also noted the severity of disability from mild to severe. This study defined 'psychological problems' as a category distinct to diagnosed mental disorders. No differences were noted in relation to the prevalence of mental disorders with respect to age, with prevalence rates varying between 23% and 29%. While the data showed a slight decline in psychological problems as individuals aged, this decline was not significant. Behavioural disorders showed a slight increase with age, but again this was not significant.

Cooper [88] undertook an analysis of the prevalence of psychiatric disorders among a cohort of 134 people with IDD aged over 65 years in the UK and compared them to a comparable group of individuals aged 20–64 years. Cooper found an overall higher rate of psychiatric disorder among the older cohort, however it is worth noting that much of this difference can possibly be attributed to not unexpected higher prevalence of dementia in the older cohort. Nonetheless, the prevalence of anxiety (9.0%) and depression (6.0%) were also both higher than the younger group (5.5% and 4.1%), while rates for schizophrenia, delusional disorders and behavioural disorders were similar across both cohorts. The prevalence of any psychiatric disorder, excluding dementia, in a combined sample of 602 people aged over 50 was 38.1%, with behavioural disorders estimated at 11.5%.

Evenhuis [89] undertook a prospective, rather than retrospective, longitudinal study over a period of 10 years between 1983 and 1993. A total of 70 participants aged 60 years and over with IDD in the Netherlands were tracked for a decade for

various health conditions. While mental disorders were not specifically reported, other researchers [90] have also followed a similar methodology to Evenhuis [89]. They completed a prospective cohort study over four time periods between 1990 and 1993 in order to determine the prevalence and incidence of a variety of health issues, including mental disorders, in a cohort of 1602 people with IDD in the Netherlands. Prevalence of affective disorders was found to be increasing in people over 50 years when compared to younger age cohorts, but there were no age related change to prevalence for other psychiatric disorders. Prevalence of affective disorders was noted at 6.8%, 8.3% and 5.9% for the age groups of 50–59 years, 60–69 years and >70 years respectively. The prevalence of all other psychiatric disorders was reported as 18.3%, 14.4% and 14.7%.

Findings from 134 people with IDD aged over 65 years in Leicestershire, which represented 93.7% of the known population that could be established through the Leicestershire Learning Disabilities Register, were reported [91]. In total, 63 (47%) of the sample had a psychiatric disorder which was not dementia. This prevalence rate was higher than reported previously [92] of a much larger cross-sectional analysis of 60,752 adults with IDD in the United States. Of this sample, 4878 were aged 60–74 years and 1248 were aged >74 years. Analysis showed 30% of people aged 60–74 had a psychiatric conditions, while this fell to 17% in the greater than 74 years cohort. Behavioural disorders, including emotional lability, aggression and passivity, were noted at high levels across all cohorts and did not vary with age. Emotional lability was noted at 36% and 35% in the 60–74 years and >74 years groups respectively.

Current Knowledge

It is now generally accepted that mental and behavioural disorders are more commonplace in older people with IDD when compared to mainstream peers without a lifelong IDD. While research in the area is still limited, the past decade has seen a focus on providing a more nuanced understanding of the prevalence and incidence of different types of disorders in this cohort, and how factors such as sex, severity of disability, co-morbidities and geographic location may impact on individual experience.

A recently published registry study [93] of people who were aged over 55 years in 2012 in Sweden examined the prevalence of mental and behavioural disorders over the previous decade (2002–2012) specifically in relation to the severity of disability. In general, they found that mental disorders were least common in people with severe/profound disability (54%), but there was minimal difference in prevalence between people with mild (67%) or moderate (66%) disability. However, the opposite was true for behavioural disorders, with the rate in the severe/profound (49%) and moderate groups (49%) being nearly double that of the mild group (25%). It is worth noting that while these prevalence rates appear higher than most other studies, the longer period for assessing prevalence must be considered.

Prevalence rates of 16.8% for depression and 14.1% for anxiety among a cohort of 693 people with IDD in the Netherlands aged over 55 years has been reported [94]. Sheehan and colleagues [95] reported on the incidence of both mental health and behavioural disorders in a cohort of United Kingdom residents with IDD. The incidence of severe mental disorder was reported at 32 per 10,000 person years, which is significantly higher than similar studies conducted on the mainstream population. When considering solely the cohorts of people aged 50–59 years, 60–69 years, 70–79 years and over 80 years, the incidence of behavioural disorders was estimated at 260 per 10,000 person years, 286 per 10,000 person years, 304 per 10,000 person years and 382 per 10,000 person years respectively. The rising incidence as people age is probably indicative of the increasing likelihood of a comorbid diagnosis of dementia, with little difference evident in the three age brackets between 18 and 49 (incidence rates of 218, 220 and 226 per 10,000 person years).

Scottish Census data [96] was used to examine the entire country's data on mental disorders, and to compare the prevalence of these disorders between individuals with and without IDD. Of this, 2455 people were identified as being over 65 years and having IDD, and 27.2% of this group also had a mental disorder. This was significantly higher than the prevalence of mental disorders in mainstream peers (4.5%). There were slightly more females (51.6%) than males (48.4%) in the cohort over 65 years who also reported a mental disorder, which is similar to some national level data that indicates mental disorders are more commonly diagnosed in females within the mainstream population [97, 98]. It is worth noting that increasing age from birth through to 64 years was associated with higher risks of mental disorder, however this effect was considered to cease from 65 years onwards.

A further study [99] examined the prevalence of problem behaviours in older Irish people with IDD. In a cohort of 683 individuals aged over 40, 53% were noted as displaying problem behaviours, including physical and verbal aggression, destruction and self-injury. Similar to previous reports [93], increases in the prevalence of problem behaviours were associated with the severity of disability, but also with the dual diagnosis of a mental disorder. Also looking at a cohort of Irish individuals with IDD aged over 40 years, other researchers [100] found 47.5% of the participants self-reported that they had been diagnosed at some point with an emotional, nervous or psychiatric condition. Slightly less than 20% of the sample had been diagnosed with depression; the prevalence of depression increased with age and was higher amongst females.

In 2019, Australian data based off a survey of 391 individuals with IDD aged over 60 years from both rural and urban areas was reported [101–103]. It noted prevalence of mental disorders at 35%. Depression or anxiety were the two most common mental disorders, with prevalence for each separately at 22%. Additional stratification by sex indicated that females experienced more mental disorders, and particularly depression and anxiety. Further mental conditions, such as schizophrenia, bipolar and other affective mood disorders, was reported at 3% for the entire sample. However, many of the participants had multiple mental disorders, with over 11% of the sample having two or more diagnosed mental disorders. There was minimal difference between sexes with respect to having multiple mental disorders, with

more men than women (13% versus 8%) in the under 65 year group reporting more than one mental disorder, but this trend then reversed for those over 65 years (8% of men and 10% of women). All of the mental disorders were noted to occur at a statistically higher rate when compared on the same survey measures with a control group of 920 people aged over 60 without lifelong IDD. Geographic location (i.e. urban versus rural) wasn't found to be a factor in the prevalence of mental disorders, but the availability of support services decreased dramatically with distance from a capital city. In a related paper, it was reported that Australian doctors had very low levels of pre- and in-service training in how to best support older people with IDD and health co-morbidities, which in turn was reflected in low levels of confidence in providing appropriate medical treatment, advice and assistance [104].

Conclusion

As discussed at the beginning of this chapter, the epidemiology of both mental and behavioural disorders in older people with IDD remains difficult to exactly quantify. The variations in assessment and diagnostic criteria, combined with no clear definition of what constitutes 'older', means that any dataset is open to interpretation, and attempts to combine data is problematic. With that in mind, and while acknowledging that there is still considerable discrepancy in the reported prevalence of mental disorders among older people with IDD, many studies find point prevalence rates that seem to fall roughly into the 20–30% range. There are certainly studies that note prevalence well above the upper figure, although some of these studies report lifetime prevalence rather than point prevalence, and equally there is also research that note lower prevalence. However, at this point, this range of 20–30% would appear to be a reasonable general estimate for point prevalence within the cohort of people with IDD aged over 50 years.

Of course, this figure is only a guide, and readers need to interpret findings with due consideration to their local situation and the specific population demographics. In particular, there appears to be differences based on sex, and also stratified age cohorts over 50 years in relation to both general mental or behavioural disorders, and specific conditions, such as depression. The prevalence of behavioural disorders is even more difficult to determine, as there is wide variation in definitions within the research quantum as to what constitutes a 'behavioural disorder'. Nonetheless, it clear that the prevalence of behavioural disorders is very high among people with intellectual disabilities of all ages, and that this prevalence continues and possibly even increases in older age.

There are a number of emerging trends evident in the more recent research that has started to better discriminate between disorders and various demographic factors, although it is worth noting they all require further investigation before any definitive conclusions can be drawn. Firstly, although people with IDD are more likely to have a co-morbid mental or behavioural disorder across their lifespan than mainstream peers, differences in the prevalence of mental disorders in particular

may reduce over the age of 65 years. However, mental and behavioural disorders appear to occur at a higher rate amongst older people with IDD when compared to younger people with IDD, although there is also some contradictory evidence for this premise.

Secondly, the prevalence of mental disorders tends to decrease in relation to the severity of IDD, but the reverse is true for behavioural disorders. Accurate diagnosis of mental disorders in people with severe or profound disability are problematic due to the high likelihood of the individual also having significant impairment to their receptive and expressive communication skills. This limits the capacity of assessors to correctly identify the presence of many mental disorders, such as psychoses, and therefore this data may reflect not actually reflect a decreased prevalence of such disorders but instead highlight flaws in the diagnostic process. Similarly, behavioural disorders are often difficult to distinguish from age-related diseases, such as dementias, and accurately determining the incidence of behavioural disorders in this group tends to be complicated by the subsequent or concurrent diagnosis of Alzheimer's disease or another type of dementia.

Thirdly, some research indicates a greater likelihood of mental disorder amongst the female population of older people with IDD. This finding is largely representative of the general community, where some mental disorders are more common in females, but there is the need for greater examination of this area, and particularly in relation to the varying prevalence of different types of mental disorders. Specifically, anxiety and depression have both been noted by a number of researchers as occurring more commonly in older females than older males, but it is unclear whether this also extends to other specific disorders.

We are gaining a greater understanding of the epidemiology of mental and behavioural disorders among older people with IDD, but the research base is still small, and a lack of standard criteria and diagnostic tools makes comparisons difficult. Further collaborative research is required to enhance our knowledge base, which will allow for earlier diagnosis and better treatment options to be provided for people with intellectual disabilities of all ages. While funding for such studies is difficult to obtain, additional large-scale multi-year prospective studies would allow for better identification of the prevalence and incidence of specific mental and behavioural disorders, and in doing so, facilitate our comprehension of how other key factors, such as ageing, sex and geographic location, may impact on both an individual and the wider community.

One final issue that is starting to emerge, but has not yet been subject to considerable examination, relates to the individuals who went through de-institutionalisation processes from the 1980s and onwards. Many of this cohort, who were successfully moved into more appropriate and supportive community-based accommodation while in their 30s and 40s, are now at an age associated with a high risk of being re-institutionalized; this time into residential aged care facilities. This transition back into congregate care models may be a trigger for the emergence, or re-emergence, of mental disorders arising from previous traumas, and is another appropriate focus of future research.

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Chapter 3
Psychopathology in Older Age

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Introduction

Recent decades have seen life expectancy increase for those with intellectual and developmental disabilities (IDD). Advances in medical and social care have improved longevity, however with this increased life span comes greater exposure to age related conditions [1–3]. The prevalence of health issues and multimorbidity increases with age in both the general and IDD, populations, those with IDD showing both a higher prevalence overall but also at a younger age [4, 5].

It is estimated that by 2050, 22% of the global population will be aged 60 or over, therefore the burden of care is expected to reach unprecedented levels [6]. The World Health Organisation reports prevalence figures for depression, dementia and anxiety in older adults of 7%, 5% and 3.8% respectively. Overall, approximately 15% of

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older adults have a diagnosis of a mental health disorder [6]. Direct comparisons between the general population and those with IDD are difficult given the differences in demographics, life experience and diagnostic issues, however studies suggest a higher prevalence of mental ill-health in older persons with IDD, with reference to the general population [7–9]. The higher prevalence of mental health disorders in those with IDD has been attributed to many factors including; genetics, life events and level of disability [10]. Standard assessment tools used in the general population are often not applicable or valid for use in an IDD setting. Assessing for mental ill-health can be more challenging in those with IDD. The development of specific guides have aided the diagnostician in distinguishing a person's disability from their psychopathology [10, 11]. In generic mental health services a person with IDD, may require extra supports in consideration of their particular needs [12]. The guidance from the National Institute for Health and Care Excellence (NICE) (Department of Health, UK) on Mental Health Problems in People with Learning Disabilities [13] sets out a baseline assessment tool for use in these situations. Bertelli and colleagues [14] considered the diagnostic issues encountered when assessing persons with IDD, for mental health disorders. In their review they identified key considerations; identification of symptoms, behavioural equivalences, diagnostic criteria, setting, source of information, screening and diagnostic tools. Ultimately, they recommended an individualised approach to diagnosis given the heterogeneity of the group but made no specific recommendations with regard to ageing.

Diagnosing mental health disorders in people with IDD, poses a challenge. Recognised clusters of symptoms, which when taken together in the general population suggests a diagnosis, may present differently in those with IDD [14]. Eliciting psychopathology is further complicated as many general assessment tools are of limited use in those with compromised cognitive function.

In this chapter we will investigate the psychopathology of common mental health disorders as seen in the context of older adults with IDD. In addition, the concept of psychological wellbeing will be discussed; this aspect of health rarely explored in IDD literature.

Mental Health Disorders

Anxiety Disorders

Anxiety disorders include disorders that share features of excessive fear and anxiety and related behavioural disturbances. Fear is the emotional response to real or perceived imminent threat. In DSM 5, anxiety is defined as the apprehensive anticipation of future danger or misfortune accompanied by a feeling of worry, distress and/or somatic symptoms of tension [15]. The focus of anticipated danger may be internal or external [16].

In the general population, anxiety is among the most prevalent mental health disorder [17]. The prevalence of anxiety disorders in English speaking adults is high

with 12 month estimates of 18.1% [18]. In the general population the conservative estimates for the prevalence of anxiety disorders in older people varies between 0.7% and 5.9% [19]. Individuals with IDD have a higher risk for anxiety disorders [20]. The longitudinal Intellectual Disability Supplement to the Irish Longitudinal Study on Ageing (IDS-TILDA) study from Ireland, using the Glasgow Anxiety Scale (GAS-LD) [21] reported that 15.1% of 291 older adult participants scored above the cut off for anxiety [9]. A large Dutch study assessed over 900 older adults with IDD for anxiety and depressive symptoms [22]. The authors reported that 16.3% of the study cohort scored positively for increased anxiety symptoms; this increase being significantly associated with female gender and mild/borderline disability.

People with IDD are diagnosed with the same diversity of anxiety disorders that have been documented in the general population [23, 24]. There are higher rates of anxiety disorders in the older IDD population than in the younger age groups [25, 26]. Depressive symptoms are frequently associated with anxiety making it challenging to determine which disorder is the primary diagnosis [27, 28]. Anxiety disorders with in IDD and the ageing IDD population can be difficult to diagnose due the numerous comorbidities such as mood disorders [29], other psychiatric disorders, behavioural presentation, physical illnesses and medical disorders.

Anxiety disorders are probably under-diagnosed in people with IDD [30] because of their limited ability to self-report anxiety, worry or fears [31]. The sources of information are usually carers, family and some of who may not be familiar with the individual and their behaviour, therefore leading to untreated anxiety. The diagnostic criteria for anxiety disorders in ICD-10 [32] and DSM-5 [15] are limited in people with IDD as both classifications rely heavily on the individual's subjective description of symptoms of anxiety. Therefore, the person would need to have adequate cognitive function and communication ability to describe their symptoms, both of which are probably compromised in people with IDD.

Making a diagnosis of anxiety disorders in this population is problematic, especially in those with moderate to severe disability. The literature in this population is limited. The Diagnostic Manual-Intellectual Disability [33] and the Diagnostic Criteria for Psychiatric Disorders for the Use with Adults with Learning Disabilities/Mental Retardation [11] were developed with proposed adaptations to both the DSM-5 and ICD-10 respectively to be used as guidelines/diagnostic criteria to facilitate making diagnoses of mental disorders in individuals with IDD. The use of an adapted diagnostic classification can increase the rates of diagnosing anxiety and other mental disorders in people with IDD [34].

Depression

Depression occurs more commonly in adults with IDD than other people without IDD [7, 9, 27]. Depression and anxiety may co-exist and/or perhaps share similar but overlapping developmental pathways. Depression is more challenging to

diagnose in individuals with communication difficulties and greater cognitive impairment due to the reliance of carer information [35]. Diagnosing depression in people in IDD may be complicated by an individual's medical history and side effects of medications that can obscure the clinical presentation [36, 37]. In DSM-5 [15] there are a few key developments:

1. Depressive disorders are now considered separately from bipolar and related disorders.
2. The additions of a new diagnostic category—disruptive mood dysregulation disorder.
3. When diagnosing major depressive episode it is no longer required to exclude bereavement

Two recent European studies reported data on depressive symptomatology in IDD cohorts. The IDS-TILDA study [9] reported that 10% of their study sample scored above the cut off of depressive symptoms using the Glasgow Depression Scale (GDS-ID) [38]. A higher prevalence of depressive symptomatology was presented by the Dutch Healthy Ageing and Intellectual Disabilities study group. They reported that 16.8% of their sample had increased depressive symptoms, this being associated with increasing age [22]. See Chap. 5 for a detailed review of depression in adults and ageing individuals with IDD.

Mania/Bipolar Disorder

First presentation of mania in later life in the general population is rare and usually part of a recurrent mood disorder that the individual has had previously in their life [39]. Following a literature review of the prevalence of psychiatric disorders in individuals with IDD, mania and bipolar disorders were rarely categorised [40]. A point prevalence of 2.5% in people with IDD has been reported [41] with a bipolar disorder being more likely to be diagnosed in persons with IDD, and more likely to be diagnosed in moderate to profound disability than those with mild disability [42]. However, a literature review on prevalence rates of mania and bipolar disorders in IDD suggest conflicting results, limited generalisability and lack of consensus.

Bipolar disorders may be missed in people with IDD due to the difference in their symptoms/clinical presentation to those in the general population. One reason is that the varying developmental status of persons with IDD may contribute to the differences in the clinical phenomenology observed. There are also numerous physical stressors such as delirium or the side effects of medication that can induce manic symptoms [43, 44]. A full physical assessment, including brain imaging, is required to rule out any underlying but modifiable cause.

An accurate assessment of psychopathology in IDD is challenging for various reasons, such as limited data and research in IDD and differences in clinical symptom presentation in IDD that is not captured in ICD-10 [32] or DSM-5 [15]. ICD-11 [45] is due to replace ICD-10 in 2022 when the complete manual will be published.

The introduction of a Bipolar II disorder classification will require the noted hypomania to be characterised by not only the presence of mood elation or irritability but also an increase in activity [46, 47].

Schizophrenia

Schizophrenia and psychotic disorders are a group of disorders characterised by common symptoms below [48]:

- Hallucinations
- Delusions
- Disorganised thinking (speech)
- Disorganised or abnormal motor behaviours
- Negative symptoms (deficits of normal emotional responses or thought process)

In the IDD population it is diagnostically important to distinguish between psychotic symptoms and the general presentation of IDD. Non affective psychotic disorders occur more commonly in adults with IDD than in other adults in spite of the diagnostic difficulties in IDD [11]. Limited verbal communication, varying presentation, aggressive behaviour, negative symptoms and limited or lack of insight can make diagnosing these groups of disorders in people with IDD challenging. In the older adults with IDD making a diagnosis of these disorders is further complicated by comorbid medical disorders i.e. delirium, dementia, infections and hearing and visual impairments.

A Scottish epidemiological study of adults with IDD [41] found the point prevalence of all psychotic disorders varied between 2.6% and 4.4% depending on the diagnostic method. When using the DSM-IV-TR diagnostic criteria for schizophrenia to make the diagnosis, the point prevalence was 3.4%. Other researchers [49] looked at two cohorts with IDD; aged 38–43 and aged 44–58 using ICD-9 and found 5.2% of the 44–58 age group with IDD developed schizophrenia compared to 4.5% of the 38–43 aged cohort.

The diagnosis of schizophrenia and psychotic disorders with regard to age has been noted in the literature. The main issue is of the validity of a diagnosis of schizophrenia in older adults, and the possibility of an alternative pathophysiological process, such as neurodegeneration, underlying the illness presentation [50]. Although the typical age of onset of schizophrenia is in early adulthood, a substantial minority of patients, 20% in some studies [51] have onset of first episode after the age of 40 years. Debate about the significance of this later onset from a neurobiological perspective continues and generates changes on diagnosis criteria across DSM editions. For instance, in the DSM-III, new onset in a patient over the age of 45 years disallowed a diagnosis of schizophrenia, which was repealed in the ensuing DSM-III-R where the diagnosis could be made with the addition of a “late-onset” specifier [50]. The DSM-IV [52] and DSM-5 [15] do not contain a separate diagnosis for late onset schizophrenia.

The DSM-5 states that "late-onset cases can meet the diagnostic criteria for schizophrenia but it is not yet clear whether this is the same condition as schizophrenia diagnosed prior to mid-life" [15]. It refers to late-onset as being after age 40 years, but somewhat confusingly contrasts late-onset with onset 'prior to mid-life' (i.e. before age 55 years), leaving a 15-year period of uncertainty.

The International Late-onset Schizophrenia Group [53] published an international consensus statement. The authors concluded that late-onset schizophrenia (onset after age 40 years) appeared to bear a reasonably close resemblance to schizophrenia of earlier onset, whereas a very late-onset group (onset after age 60 years) was better classified as having a schizophrenia-like psychosis based on a convergence of clinical, epidemiological, neuroimaging and neuropsychological data, although there was no consensus on the age cut-offs for this distinction. Howard's description of very late onset patients (after 60 years) stressed the high prevalence of sensory deficits especially long standing deafness, which is prevalent in the IDD population. Also when onset of psychosis is after age 60 years, formal thought disorder and negative symptoms are very rare but they are more likely to present with visual, tactile, olfactory hallucinations, persecutory delusions, third person, running commentary and accusatory and abusive auditory hallucinations.

The diagnosis of schizophrenia in adults with moderate and severe IDD continues to be problematic. The Diagnostic Criteria for Psychiatric Disorders for Use with Adults with Learning Disabilities/Mental Retardation (DC-LD) [11] states that for non- affective psychotic disorders it is not usually appropriate or valid to subclassify the disorders to the extent of ICD-10, as delusions and hallucinations are difficult to elicit in adults with IDD. In summary, a diagnosis of schizophrenia is made with a comprehensive assessment by an expert in the assessment and diagnosis of people with IDD and mental illness.

Dementia

Dementia is more common in people with Down syndrome (DS) than in the general population mainly due to the presence of dementia of Alzheimer's disease (DAT). A recent prospective longitudinal study reported that 97.4% of people with DS received a diagnosis of dementia [54]. Although there has been much research on the diagnosis and management of dementia in people with DS [55, 56], there has been less research on the psychopathology and the behavioural and psychological symptoms of dementia (BPSD) [57]. Dementia may mimic several other psychiatric disorders and the initial stages of the disease may present with depression, psychosis, behavioural changes or anxiety and it may be difficult to distinguish whether the illness represents a prodromal phase or a psychiatric or other illness [58]. Other researchers [59] have drawn attention to a frontal lobe presentation with apathy, loss of interest and change in personality presenting before cognitive decline.

Research on early symptoms of both cognitive and behavioural manifestations in a systematic review was recently published [58]. Whereas amnesic symptoms are

predominant in the general population, a more heterogenous presentation is noted in those with Down syndrome. In this cohort, the Behavioural and Psychological Symptoms of Dementia (BPSD) with particular difficulties in executive function appear early in the course of disease progression. Researchers [60] have looked at the association between BPSD and dementia in 224 adults over 45 years with Down syndrome and found associations between increased BPSD directed towards others and development of dementia as well as symptoms of sadness and anxiety. Changes such as restlessness aggression, repetitive speech and being uncooperative were reported in preclinical and early stages of DAT [61]. Emotional lability, lack of concern for others, stubbornness, disinhibition and impulsivity have been reported in people with DS prior to a diagnosis of dementia [62].

Several assessment procedures draw attention to differential diagnoses such as depression, grief and abuse as well as medical issues such as hypothyroidism, deafness, cataracts and sleep apnoea to name a few common disorders [63, 64]. For many people with DS, these illnesses may be comorbid, i.e., it is possible to have cataracts, hypothyroidism and dementia co-existing as a function of age. These lists, however, should not be prescriptively followed rather these conditions should be adequately treated before making a diagnosis of dementia. As dementia is a progressive illness, over time changes on cognitive tests and memory deterioration will advance even if mood and behaviour improve when adequately treated.

Particularly following a bereavement, the clinical picture of possible dementia can be an unclear one. Features of depression and anxiety can be present along with cognitive symptoms such as memory loss and poor attention span. Recent research [57] on BPSD and the development of a scale BPSD–DS (Behavioural and Psychological Symptoms of Dementia in Down syndrome) an informant interview administered to carers of 281 people with DS has shown an association with anxiety, depression, behavioural changes and the onset of dementia in people with DS. Since depression is common in people with DS a cautious approach to diagnosis is advised with treatment of the presenting issue and reassessment after treatment. Sleep wake cycle disorder and appetite changes may be seen as part of the prodromal phase of dementia and may exist by themselves or associated with other symptoms.

Due to the complicated neurochemistry of dementia and fluctuating levels of neurotransmitters there may be altered affective states (e.g., low mood, anxiety, irritability) and disrupted sleep-wake cycles as common features. There is often understandable anxiety around the diagnosis for the individual connected to having a life-changing diagnosis, loss of independence and increased reliance on carers for everyday tasks. The distress of the diagnosis and the implications for the individual and their family needs careful management and support as many people will be aware of friends and colleagues who have had the same illness.

Hallucinations and delusions may be a presenting feature of the illness and may be auditory, visual, tactile, olfactory and other [65]. Lewy Body variant should be considered particularly when associated with fluctuating cognitive symptoms, hallucinations and extrapyramidal features [66]. A recent study [57] did not find that hallucinations and delusions differentiated between people with DS with or without

dementia, however to describe a hallucination and delusion requires intact language ability and it may be difficult to elucidate these symptoms as dementia progresses. People with DS can be very sensitive to the side effects of antipsychotic medication, however the distress caused by the psychosis can be such that a balance has to be found between alleviating distress and side-effects of medication.

Behavioural changes that do not meet criteria for either a mood disorder or psychosis may also occur and may be difficult to treat due to the adverse side effects of medication. For instance, the increased risk of cerebrovascular accident with antipsychotics and the increased risk of falls and aspiration with any sedative medication. Many consensus guidelines do not advocate use outside stringent guidelines. Behaviours such as prolonged screaming, crying, self-injurious behaviour and verbal and physical aggression may not fit into a neat classification, but such symptoms indicate distress of either an emotional or physical nature. As with any indication of distress, infection, pain and discomfort, loss and loneliness and communication issues should be looked for, a behavioural analysis performed, and a full multidisciplinary approach to management adopted.

Dementia is a progressive degenerative disorder, psychiatric symptoms usually decrease and disappear in the late stages and there is a need for a palliative care approach at the end of life. In the vast majority of people with late stage dementia this will be a peaceful time, however for some people there can be emotional distress and anxiety and a collaborative approach with the palliative care team is advocated.

Positive Mental Health and Wellness

It has been noted that there is a higher prevalence of psychiatric conditions in older people with IDD. Whilst biological factors may provide an explanation for some mental illness, the reasons are diverse and psycho-social factors play an important and modifiable part [67]. Thus, for example, negative life experiences and rejection may be implicated in the development of mental illness [25, 68]. Furthermore, social isolation can lead to the absence of the social networks which may provide protective support [69]. This is in the face of significant adverse life events which can result in generalised symptoms of stress and depression [70]. These represent factors which may be addressed through the promotion of self-management and mentally healthy lifestyles.

People with IDD are more likely to experience more negative and multiple life events than those in the general population [71]. In Wave 3 of the Intellectual Disability Supplement to the Irish Longitudinal Study on Ageing (IDS-TILDA), 63% of respondents reported having experienced more than one such event in the previous 12 months, with nearly 20% reporting four or more [9]. The vast majority of these related to social changes (staff mobility, bereavements and new residents), and were associated with significant stress and mental ill health [68, 70].

The experience of stress and anxiety can influence an individual's vitality resulting in reduced energy. Whilst *vitality* is a pre-requisite for physical activity, the more encompassing *subjective vitality* may be a more holistic indicator of well-being, focusing on life meaning and energy for living. Indeed, it has been described as a phenomenological concept with a subjective 'sense of aliveness and energy' [72] and is suggested to have correlations with positive mental and bodily health [73]. Loss of such vitality often manifests as fatigue which, in turn, can result in reduced mobility, diminishing social contact, and impaired mental wellbeing [74]. Impaired vitality has been reported to be closely associated with depression in older people [75].

The Energy and Vitality Index (EVI) [76] was used to assess vitality in older people with an IDD [8]. It was noted that the scores from this cohort were similar to the results from a large scale study of the general population [77]. Ageing had little impact on the EVI scores with a slight decrease reported in older age groups.

Responses to psychiatric illness in the general population have increasingly become focused on individualised recovery-oriented approaches, which centre on the restoration of meaning in life, hope for the future and retrieval/reconstruction of self [78–80]. These may be achieved by re-establishing a locus of control (empowerment) in the person and thus supporting empowerment, in a supportive and connected context [80]. Such approaches have been somewhat radical requiring significant education and training of mental health practitioners, people with experience of mental health concerns and family members, and evidence has indicated that they are leading to positive change in people's lives [81]. This is particularly pertinent to people with IDD, many of whom have lived with the reality and stigma of mental illness and poor mental health throughout their lives and, for whom, access to recovery-oriented mental health services may have been limited [82]. The development of such approaches within IDD service provision may play an important part in addressing some part of the prevailing mental health problems that are so evident in the IDD population.

Conclusion

Successful ageing is a relatively new goal for individuals with IDD. Unfortunately, with increased longevity, more age-related morbidity has been reported. While the same mental health issues occur in those with IDD as in the general population; the presentation, associations and prevalence of these disorders differ, the most notable being the emergence of dementia in those with DS.

Diagnosticians should be aware of challenges which may present when engaging with people with IDD. Communication difficulties for the individual may interfere with their ability to verbalise symptoms, this results in placing more weight on biological features or collateral histories. Sensory loss can compound communication problems, and this may worsen with age. Education of carers to pick up on early signs of change such as altered adaptive function, increased irritability, sleep

disturbance or a change from baseline of the person they support should prompt further enquiry. Careful history taking which is mindful of the complex interplay of physical, environmental, behavioural and social factors will aid the clinician in reaching an accurate diagnosis and allow the formulation of a bespoke treatment plan.

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Chapter 4
Assessment of Psychiatric Disorders

Marco O. Bertelli and Annamaria Bianco

Background

In the last 20 years, the average life expectancy of people with intellectual and developmental disabilities (IDD) has increased considerably [1–3]. Since IDD represents a condition of high physical and mental vulnerability [4], the lengthening life expectancy has been associated with an increasing prevalence of physical health issues, mental health problems, and multi-morbidity related to aging, much higher than in the general population. In fact, in persons with IDD both the ageing process and the onset of mental disorders are reported to start earlier [5–7]. See Chap. 2.

This is particularly true for dementia, which appears to affect between 30% and 40% of persons with IDD and around 50–70% of those with Down syndrome (DS) older than 60 years [8]. In persons with non-DS IDD the average age of onset of dementia is around 10 years lower than the general population [9]. In DS, dementia in Alzheimer's disease affects at least one-third of individual in their forties [10, 11].

Psychiatric disorders are present in more than 40% of older adults with IDD, especially depression, anxiety, and psychotic disorders [5, 12]. In those with co-occurrence of autism spectrum disorder, the prevalence can further increase up to five times more than in people who present only IDD [13, 14], with a proportionate negative impact on individual functioning and quality of life as well as on family and the community [15].

Research indicates many different factors being associated with this high mental health vulnerability, including specific genetic syndromes, the extent of the central

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nervous system compromise (leading to associated neurological disorders and disabilities such as epilepsy, cerebral palsy, and sensorial impairment), life events, psychological factors (i.e. low skill for coping with stress, lack of environmental mastery), low level of health literacy, low compliance with hygiene rules, lifestyle and environmental conditions, health promotion/disease prevention practices, access to basic healthcare services, and reliance on other people for care.

The risk of psychiatric disorders in older people with IDD seems to be increased by exposure to more age-related life events than in younger adults. In particular, depressive and anxious symptoms were significantly heightened in individuals who reported more total and negative life events during the preceding year, also after controlling for a history of depression or anxiety disorder [1, 7]. Several surveys have reported positive correlations between lifetime exposure to life events and the development of psychiatric disorders, somatoform disorders and physical health issues, in persons with IDD [16–22].

However, in people with IDD mental disorders are difficult to diagnose, especially in old age. The first difficulties relate to clinical presentation, which is substantially different from that of the general population and which would, therefore, require the application of adapted diagnostic criteria as well as specific diagnostic procedures and tools, which are instead unknown to most clinicians. Differential diagnosis can be even more difficult to make, including that between dementia and some psychiatric disorders, such as depression or psychosis. Research findings on the co-occurrence of depression and dementia amongst older adults with DS are controversial precisely due to different interpretations of clinical presentations [23–25].

Peculiarities of Psychiatric Symptoms

In persons with IDD, the presentation of psychiatric disorders can considerably vary from that of the general population, especially for symptoms related to subjective experience in those with low or absent verbal communication skills, who may only be able to express themselves through changes in behaviour [4]. Symptoms can appear as both qualitative and quantitative changes of basic behaviours, the latter of which are indicated with the term “baseline exaggeration” [26–28]. Thus, symptomatology can be chaotic, intermittent, fluctuating, mixed, scarcely defined, extremely rigid, atypical, or masked [4, 29, 30]. Sometimes even the nuclear elements of some syndromes, such as suicidal ideation or delusion, may not be identifiable, especially if verbally mediated [31]. Some researchers have found that, concerning schizophrenia, the only first-rank symptom which can be detected with a good frequency through direct observation is auditory hallucination [32].

Scientists have proposed some explanatory models of this phenomenal complexity and difficulty to diagnose. One of the most common has been called “diagnostics overshadowing” [26], which consists in the tendency to attribute behavioural changes to the person’s disability rather than to one or more symptoms of an underlying psychiatric disorder and/or to dementia or another physical or mental problem.

Other explanatory models are the "intellectual distortion", that is the presence of alterations of the level of cognitive, communicative, physical and social functioning [33], the "developmental inappropriateness", that is the non-correspondence between the individual developmental level expected for chronological age and the level of effective individual development [34], and the "psychosocial masking", that is the peculiarities of social, cultural and environmental experiences [26]. Another aspect that strongly characterizes psychiatric symptomatology in people with IDD is "neurovegetative vulnerability": somatic symptoms, pain, changes in circadian rhythms, dystonias of the autonomic nervous system are frequently the main expression of many emotional dysfunctions [27].

Behavioural Equivalents

In people with IDD problem behaviours are common and represent one of the factors with the greatest negative impact on the implementation of rehabilitative interventions and on the quality of life of disabled persons and their families [35]. Some of the largest studies in this area found that about 25% of people with IDD have at least one problem behaviour, 5% show self-injury, and 10% physical outwardly directed aggression [36, 37]. When considered as a psychopathological category in persons with severe and profound IDD, problem behaviours account for 75% of all the diagnoses [6].

The term 'problem behaviour' refers to culturally abnormal behaviours of intensity, frequency, and duration such as to put at risk the safety of the person acting on them or others, or behaviour that severely limits or prevents access to the community's ordinary services. It is often difficult to determine whether problem behaviours are an expression of physical pain, organic problems, drugs, psychological, environmental, socio-relational factors, or whether they represent psychiatric symptoms or even a combination of some of these aspects. The same behaviour can be interpreted in a quite different way by the various professionals, even within the same multidisciplinary team, with consequences that are sometimes relevant for the interventions.

Several studies have shown a relationship between problem behaviours and psychiatric disorders [38–43] particularly evident in persons with lower levels of functioning [39]. Some problem behaviours have been identified as symptoms, or groups of symptoms, specific to some psychiatric disorders, named "behavioral equivalents" [44]; they are characteristic for onset, development, maintenance, and extinction, especially compared to other possible symptoms of a psychiatric disorder [44–47].

Not all researchers support the idea that problem behaviours can be considered as "behavioral equivalents" of an underlying psychiatric disorder. More they are indicators of underlying emotional stress [48, 49]. In one of the most recent researches in this area, problem behaviours have been found to be associated more with dysregulation of wide psychological dimensions than with specific

psychopathologic areas [50]. For detection and interpretation of psychopathological symptoms and "behavioral equivalents", dimensional symptomatologic models can have higher predictive validity than a categorical one. Further advantages can be given by a careful specification of problem behaviour and symptoms associated with them. For example, in persons with IDD and autistic features, self-oriented aggressive behaviours seem to occur more frequently during depressive episodes than during manic or hypomanic episodes. Within depressive episodes, verbal aggression seems to prevail in the clinical presentations characterised by the prominence of emotional symptoms (irritability, anxiety, and sadness). Physical aggression seems to be more common in individuals with more associated physical symptomatology (problems with energy, eating, sleeping, and stress) [51].

In persons with IDD, reference to behaviours as part of psychiatric psychopathology may represent relevant support for clinicians in the diagnostic processes and the treatment choice, especially during the old age, in which cognitive and communication skills often undergo further reduction.

Diagnostic Criteria

Research findings showed that the use of diagnostic criteria developed for the general population does not allow a correct diagnosis of psychiatric disorders in persons with IDD [45] and that adapted criteria should focus on observational-behavioural symptoms, especially in older people and in those with higher cognitive and communication impairment.

In the last 25 years, some adaptations have been produced, for both the Diagnostic and Statistical Manual of Mental Disorders (several versions) system and the International Classification of Diseases and Related Health Problems (several versions) by the Royal College of Psychiatrists (UK) and the National Association for Dual Diagnosis (USA). The former organisation has published an adaptation of the ICD-10; Diagnostic Criteria for Learning Disability (DC-LD) [52]. The latter organisation has adapted changes to the DSM-IV-TR and the DSM-5 and produced the Diagnostic Manual—Intellectual Disability (DM-ID) [53, 54]. As an example, the clinical guide of the DM-ID (version 2) states that depressed mood can be described by others in one or more of the following ways, that constitute a significant change from the baseline: sad or angry facial expression, crying, assaults, self-injurious behaviour, spitting, yelling, swearing disruptive or destructive behaviours [55].

Symptoms of mania or hypomania are more difficult to detect than depressive symptoms, and in some cases, the latter can mask the former, such as in the presence of irritability [56]. The differentiation can be supported by the identification of an increase of goal-directed activities, which are indicated by the clinical guide of the DM-ID (version 2) to present with engagement in activities in a "sped up manner", rarely sitting down, being up and down from seat a lot, pacing, walking rapidly, appearing "driven", racing around the room, and becoming very intrusive [57].

Another interesting example of behaviorally adapted diagnostic criteria can be provided in respect to visual, auditory, somatic, and multi-sensorial hallucinations, which are described in the DM-ID (version 2) and in some research reports in ways as the following: cover of eyes or ears, stare, nod, self-talk, make gestures as to remove something from the body, wear heavy, tight-fitting or multi-layered clothing, bandage ankles or wrists, move as if defending, fighting, or flirting [58, 59].

Although they need some improvements, these criteria have a valuable clinical utility, helping the clinician to better recognize and diagnose psychiatric disorders in persons with IDD and to better differentiate them from other psychiatric and neurological disorders [60].

Other Assessment Issues

In older people with IDD the assessment of psychiatric disorders should be appropriately modified and adjusted for the cognitive dysfunctions, language and communication limitations, sensory impairments, skill deficits, adaptations difficulties, and for physical disabilities often present. Another important issue is the reliability of the information source, including the persons with IDD, who may have poor verbal expression skills, tend to acquiescence and present cognitive distortion, that is difficulties in the ability for introspection, to define one's own experiences and to communicate the states of suffering or to recognize that they need help to identify the meaning of some changes in their psychic functioning [61].

Information from informants can be very helpful to the psychopathological assessment by adding historical information, clarity, and depth that might otherwise be missing. Conversely, some researchers have highlighted several limitations of the proxy perspective, which is often heterogeneous, contradictory, and with low inter-rater reliability. For example, family members are often struggling to answer questions about the presence of further disorder of mental functioning, and the indirect or atypical presentation of symptoms often limits the capability of identifying them for caregivers or other informants.

Several studies have shown that informant evaluation may vary depending on a number of factors, ranging from the cultural level and other personal characteristics to the specific nature of the relationship and involvement [62, 63]. Furthermore, the accuracy of the ratings depends strictly on the experience and insight of the informant, as well as on how well he/she knows the person being rated. The degree of agreement or disparity between ratings made through different informants is unclear [45]. The use of a comprehensive diagnostic system that includes the integration of different sources of information, such as the same individuals with IDD, carers, familiars, or other informants is highly recommended to measure any significant behavioural change that has occurred, to interpret it as symptom equivalent, and to increase the validity of the psychiatric assessment [45].

In addition to the sources of information, the setting of the evaluation also deserves special attention. The duration of the assessment should be short and

flexible, preferably in the context of the habitual life of the person with IDD, in order to reduce the impact of contextual variables on psychic and behavioural conditions. If this is not possible, a welcoming and silent place with limited sensory stimuli should be chosen. The evaluator should communicate in simple language, avoiding metaphors and idiomatic expressions. The question style should have many different answers and images and symbols are very useful for the communication between the evaluator and the person with IDD.

Assessment Tools

For all the above-mentioned issues, the application to people with IDD of the assessment tools developed for the general population also showed significant limits of validity and reliability. For example, screening tools and neuropsychological tests, such as the Mini-Mental State Examination [64, 65], determine a floor effect in most people with more severe disabilities and do not allow to identify a single cut-off threshold in mild and moderate forms [66]. Autoptic and neuroradiological studies have found changes in the central nervous system typical of Alzheimer's disease in almost all adults with DS over 45 years of age [67, 68], while the clinical manifestations do not seem so universal. The diagnostic difficulties could be precisely at the root of this discrepancy.

Researchers specialised in this field have worked partly on creating new tools, partly on translations and adaptations of questionnaires developed for the general population. In both cases, the tools have numerous limitations in applicability and effectiveness. The adjusted questionnaires also present sensitivity problems, due to the considerable atypical psychopathological symptoms of people with.

Currently, the most commonly used tools are general screening ones, which provide indications on all diagnoses compatible with the symptoms and behavioral equivalents detected in a person, but there are also tools which are specific for the psychopathological area. Instead, there are few standardized diagnostic interviews, which are long, expensive, and less useful for multidisciplinary collaboration. The tools must be chosen on the basis of the specific purpose, the characteristics of the person to be evaluated, the administration time, the training required, and the resource available [69].

Among standardized interviews addressed to people with good communication and introspective skills the Psychiatric Assessment Schedule for Adults with Developmental Disabilities (PAS-ADD) [70] was designed to provide all members of a care staff with a rapid psychopathological screening tool for adults with IDD and to collect information useful for setting the care plan. This instrument produces a diagnosis according to ICD-10. It showed reasonable reliability and validity [71, 72], and is also available in two other forms: the Mini PAS-ADD [73] and the PAS-ADD Checklist [74]. PAS-ADD boasts a high number of high-quality studies, is a reference tool in the United Kingdom and its use is also growing in other European countries [75–78]. However, some sensitivity problems have been identified, in

particular for disorders of the schizophrenic spectrum, reproducibility between different evaluators and consideration of anamnestic clinical information [4].

The Psychopathology Instrument for Mentally Retarded Adults (PIMRA) [79–81] is a structured interview, available both in a self- and other-report form, based on the DSM diagnostic criteria. It has been developed to screen for the presence of psychopathology in individuals with mild and moderate IDD. While showing increasingly frequent concordance difficulties between evaluators, PIMRA has proven to be particularly useful in the field of research, therapeutic planning, and evaluation of treatment outcomes. Furthermore, a revised version of the PIMRA has just been developed (PIMRA-II) and it seems to have good psychometric properties [82].

The Diagnostic Assessment for the Severely Handicapped (DASH-II) [83], which is the revised version of the DASH [84] is mainly based on the detection of key symptoms related to different syndromic groups, which can be defined by frequency, duration, and severity. The main limit of this scale is represented by the number and quality of the items, which are unbalanced for the various psychopathological dimensions, and present poor alignment with the criteria of the categories of the main diagnostic manuals [4].

The Psychopathology Checklists for Adults with Intellectual Disability (P-AID) is a battery of tools developed recently and allows to identify ten different psychiatric disorders and eight types of problematic behaviors according to DC-LD diagnoses. P-AID showed high values of internal consistency and inter-rater reliability, while sensitivity and specificity are still to be confirmed [85].

A comprehensive set of tools for all the different practical needs related to psychopathological assessment and monitoring, to be used by mental health professionals and the whole multidisciplinary team working with people with IDD is represented by the Systematic Psychopathological Assessment for persons with Intellectual and Developmental Disabilities (SPAIDD) [86]. It includes tools for every phase of the clinical intervention, such as general psychopathological screening, diagnostic categorical specification, dimensional diagnosis, and symptoms monitoring. The items of the SPAIDD system have been kept unchanged across all the battery tools, since they represent observable and behavioural aspects of cross-categorical symptoms. See Chap. 5 for further information.

In designing and implementing this system, authors tried to overcome also the other main limits of previous tools, such as the impossibility of being used for all the degrees of intellectual and communication impairment, the misalignment with the DSM or the ICD, the lack of some main symptoms or syndromes, the lack of chronological criteria, the long times of administration, and the scarce interdisciplinary usability [86]. The version for General screening (SPAIDD-G) is the first already validated and published tool of the SPAIDD system [4, 86]. It includes 56 items, which represent descriptions of the most frequent observable and behavioral aspects of all the symptoms that appear in the various DSM-5 diagnostic categories. These items were developed to be rated by a mental health professional through the information gathered by interviewing a family member of the person with IDD or another informant who has an awareness of changes in the behaviour of the people

for whom they care. The SPAIDD-G showed very good psychometric characteristics. To date, it is available only in Italian, although validations of the English, German and French translations are already underway.

Diagnosis and Differential Diagnosis of Dementia

Diagnosis of dementia is often difficult in people with preexisting cognitive deficits, and it is therefore important to make it by referring to valid specific diagnostic criteria and diagnostic tools. Important guidelines on this issue have been provided in the last decade by the British Psychological Society [87] and the National Task Group on Intellectual Disabilities and Dementia Practice [88], although there is currently no standard protocol for the assessment of dementia in persons with IDD.

Some of the early symptoms of dementia may be subtle or may present as an exacerbation of the existing behavioural traits or manifest differently in people with IDD than in the general population, as it has been described for psychiatric disorders. In most cases, these symptoms are noticed first by caregivers, as unusual changes in the individual's behaviour. Therefore, the use of informant-based instruments is recommended for screening or diagnostic purposes. Review of the literature [89, 90] in the last decade identified 79 instruments for direct evaluation and 35 for informant rating and provided detailed descriptions of selected neuropsychological assessments. Frequently used instruments in IDD research which have been thoroughly studied for their psychometric characteristics are; Dementia Scale for Down Syndrome (DSDS) [91], Dementia Questionnaire for people with Learning Disabilities (DLD) [92], and the Dementia Screening Questionnaire for Individuals with Intellectual Disabilities (DSQIID) [93].

The DSDS is an informant interview and includes 60 items organized into three categories, one for each of the stages in which the progress of the disorder is normally divided. The DSDS is designed for use with people with DS and standardized in a sample of adults the majority of whom had severe to profound disability, but may also be useful for people with IDD [94].

The DLD (formerly the Dementia Questionnaire for people with Mental Retardation—DMR) [95] has 50 items that have been divided up into the categories of 'short-term memory', 'long-term memory', 'spatial and temporal orientation' (making up the sum of cognitive scores; SCS), 'speech', 'practical skills', 'mood', 'activity and interest' and behavioural disturbance (making up the sum of social skills score; SOS). The cut-off scores for screening positive for dementia are different for individuals who have mild, moderate or severe disability respectively.

The DSQIID is another common screening tool, which is best used in clinical settings to deeply explore each item with a habitual caregiver in order to build up a valid clinical picture. It comprises 43 questions organised in three sections and should be used prospectively; however, certain items may show floor effect and total score may decrease as dementia progresses. The DSQIID has been included, with some adaptations, in the evaluation package of the National Task Group on

Intellectual Disabilities and Dementia Practices (NTG), called NTG-Early Detection Screen for Dementia (NTG-EDSD) [96], and recently translated and validated in various languages [97–100].

For people with good introspective and communication skills, there are also some adaptations of tools widely used in the general population, one of the most recent is the Cambridge Examination for Mental Disorders of Older People with Down's syndrome and Others with Intellectual Disabilities (CAMDEX-DS) [101]. The CAMDEX-DS consists of approximately 150 questions that ask about functional difficulties in different cognitive domains, whether such difficulties represent a deterioration in function, and the extent of the deterioration (slight or great) [102]. These tools can also support clinicians in the differential diagnosis between dementia and some psychiatric disorders, such as depression and psychosis, which are reported to be frequent co-morbid conditions and confounder of dementia in old persons with IDD [103, 104].

Most patients referred to as affected by depressive pseudo-dementia often show cognitive symptoms outside the range of dementia and a more rapid decline (over weeks to months rather than years). Depression does not significantly impair performance on cognitive testing if testers provide enough time and encouragement [105, 106]. Referral for complete neuropsychological testing can be helpful in clarifying the diagnosis in many cases. Treatment with antidepressants will significantly improve cognitive function in patients with pseudo-dementia, whereas persons with dementia may limit to partial improvements in overall functioning [107]. Recent research findings suggest that evaluation of facial recognition and left hippocampal volume may provide more reliable evidence for distinguishing pseudo-dementia from dementia in Alzheimer's disease [108].

Evidence on the relationship between psychosis and dementia in persons with IDD is lacking. Basing on data from the general population, psychotic symptoms are likely more common in prodromal and early dementia than previously indicated by factor analysis studies, although they are much more common in established dementia [109]. Psychotic features of dementia include hallucinations (usually visual), delusions, and delusional misidentifications. Delusional misidentification result from a combined decline in visual function and cognition and frequently manifest with strange suspicions or beliefs, such as family members being impostors, strangers living in one's own home, or with failure to recognize one's own reflection in a mirror. Hallucinations often do not upset patients and may be pleasant [110], contrary to what happens in the co-occurrence of schizophrenia, in which hallucinations usually have negative contents and determine severe problem behaviour. Symptoms of very late-onset schizophrenia show greater dissimilarity with psychotic symptoms associated with dementia with Lewy bodies than with those associated with Alzheimer's type, particularly more partition delusions and more auditory hallucinations of human voices. Processing speed and executive function seem to be comparably impaired among the three conditions. Patients with psychotic disorders at baseline such as schizophrenia seem to have a high risk of developing dementia over time, although finding are controversial [111].

Conclusion and Future Issues

Despite the evidence of need, adults and older people with IDD have a much lower rate of appropriate physical and mental healthcare (about a third of the population) and much larger difficulties in achieving services [112, 113]. The quality of mental health care is particularly poor and the rate of improvement is slow compared to that for the general population. Here the proportion of satisfied and unsatisfied care needs drops to a quarter of the norm [114, 115]. Already in early adulthood, the person with IDD and mental health problems is left without specialist reference. One of the main reasons for this serious situation seems to be the lack of experienced and properly trained specialists.

As mentioned in this chapter, people with IDD often have ways of communicating and displaying symptoms, especially symptoms of psychiatric disorders, which cannot be adequately understood by health professionals who have not received specific training. Graduate courses in medicine and specialization in psychiatry of national universities do not include the mental health issues of this population. The current attitude of neurologists, geriatricians, and psychiatrists towards IDD-specific problems is severely limited [116]. The need for training also concerns socio-health workers, educators, basic carers, family members, and other non-professional caregivers [117]. The current evaluation shortcomings are linked to even greater limits of socio-health systems and policies, determined in turn by long-lasting cultural issues, such as the prejudice that people with IDD cannot experience emotional suffering or the belief that rehabilitation interventions could have some efficacy only during childhood and adolescence. The idea that IDD is a life span condition is relatively recent. The first international document referring to it dates back to 2001 and the first European data on health problems are from 2008 [118].

The Edinburgh Principles, produced in 2001 by an international group of experts led by Wilkinson and Janicki [119], still represents a good reference for the care of the elderly person with IDD. Recently, the National Task Group (NTG) on IDD and Dementia Practices [120] and in the British Psychological Society and the Royal College of Psychiatrists [9] jointly produced consensus recommendations for the evaluation and management of dementia in adults with IDD. These recommendations follow a longitudinal perspective, forcing caregivers to take into account the individual's mental health history and to assess the symptomatological potential of behaviours, especially in terms of change from the baseline level. This perspective is not so obvious for providers of continuous assistance to people with IDD, often flattened into a sort of eternal present.

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Chapter 5
Neurocognitive Disorders Assessments

Tiziano Gomiero, Afia Ali, and Flavia H. Santos

Introduction

Neurocognitive disorders are core clinical deficits in cognitive functioning, which constitute a decay from a previously attained level of functioning. These conditions possess an acquired rather than developmental origin and imply different diagnosis which have in common identified brain diseases [1]. For instance, Huntington's, Parkinson's, and Alzheimer's disease, among several others. Consequently, each neurocognitive disorder will express specific neuropathological markers (morphological, electrophysiological, and biochemical abnormalities) and correspondent phenotypes, i.e., symptoms, stages, and age of onset. Even though some general manifestations will overlap regardless the disease [2].

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Table 5.1 DSM-5 indicators for neurocognitive disorders in the general population

Areas	Neurocognitive disorder	
	Mild	Major
Occupational functioning	Mild	Severe
Social functioning	None or mild	Moderate
Completing self-care activities	Preserved	Moderate to severe
Objective measures of cognitive function	1–2 standard deviation (SD) range (between the 3rd and 16th percentiles)	Below 2 SD or 3rd percentile
Completion of daily activities	Preserved	Moderate to severe

Neurocognitive Disorders

The DSM-5 [3], likewise ICD-11 [1], distinguishes mild neurocognitive disorder and major neurocognitive disorder in the spectrum of the functional and cognitive decline. These diagnoses exclude delirium and go beyond age-related changes[1]. The distinction between these two categories depends on quantitative and qualitative indicators. The key markers are functionality and independence that are preserved or slightly affected in the mild form. Besides, behavioural changes, if any, are less unsettling in this group. In the mild form pharmacological treatment and neuropsychological rehabilitation are available and aim to keep the individual autonomous. Currently, there is no cure for neurocognitive disorders. Cholinesterase inhibitors and memantine are used as cognitive enhancers, however, their effectiveness decreases as a function of the disease progression. Symptomatic treatments for behavioural and psychological symptoms have been used in the major forms, although nonpharmacologic methods are the first-line recommendation [4]. Experimental treatments are being developed.

Operationally, the diagnosis considers impairments in different areas of life, as listed on Table 5.1. The term cognition in this context includes complex attention, executive function, learning and memory, language, perceptual-motor, and social cognition [1, 3]. The main instruments available for neuropsychological assessment will be discussed further in this chapter.

Although not all individuals with mild form will worse gradually, advancement in cognitive deficits and functional losses are inevitably evidence of transition within the spectrum for the Major form. The Major neurocognitive disorder refers to what is widely known as dementia, this new term adopted by the DSM-5 [3] is an attempt to reduce social stigma. Dementia or "dementiae" in Latin means derangement of mind, madness, which does not exactly correspond to the clinical concept in question. In neuropsychiatry, dementia is currently understood as a progressive loss of cognitive ability and functional decline [5]. Ferri and colleagues [6] used the

[1] These conditions are not restricted to ageing, for instance, human immunodeficiency virus cause neurocognitive disorders in individuals at younger age (<50 years).

Delphi consensus method to determine dementia prevalence across the globe, they estimated that 42 million of individuals would have dementia by 2020, with two times more cases in developing countries.

Dementia in general population is well documented. By contrast, less is known about dementia in adults with intellectual and developmental disabilities (IDD). But there are a few epidemiological studies to date. A pioneer study by Janicki and Dalton [7] included 268 people with Down syndrome (DS) and 526 individuals with other forms of IDD. In people with IDD (except in person with DS), the presentation of dementia was likewise the general population, i.e., onset around age 67.2 years, diverse types of dementia (including nonspecific, vascular disease and Alzheimer's disease) and long-term duration of symptoms. This finding was consistent with the epidemiological study carried out in the United States using clinical diagnosis [8]. However, among individuals with DS the symptoms appeared earlier in average with 52.8 years and predominantly associated with Alzheimer's disease (75% of the cases).

Posterior epidemiological studies found that the incidence rates of dementia in people of IDD (except DS) were higher than the general population. For example, in a cohort of 222 individuals with IDD aged 60 years or older in the United Kingdom [9], 29 participants screened positive for dementia using ICD-10 and DSM-IV criteria, with the latter detecting more cases. Among individuals with dementia 66% individuals met criteria for Alzheimer's disease, the second most frequent cause was Lewy body dementia followed by frontotemporal dementia, and vascular dementia. Alzheimer's disease prevalence in this cohort was almost three times higher than in older adults in the general population [9].

The symptoms that clearly discriminated false positive in individuals with IDD (except DS) were deterioration in self-care ability, deterioration in instrumental activities of daily living, memory decline, development of incoherent thinking, difficulties with planning and thinking ahead, and newly developed perseveration [9]. In the follow up of this study carried out 2.9 years later, 21 (15.7%) out of the 134 adults with IDD aged 65 and older were diagnosed with dementia. The incidence was up to five times higher than in the general population using DSM-IV and ICD-10 criteria [10].

The most recent epidemiological study [11], investigated the prevalence of dementia in Japanese people with IDD among 493 out of the 900 users of support facilities. Diagnosis were made using ICD-10 and DSM-5 criteria plus expert physician examination. Thirty-four participants had DS and 459 other IDD, with a mean age of 46.57 (SD: 11.43 years; range: 19–83 years). No person with DS showed mild neurocognitive disorder, however, among them there was a higher rate of dementia (20.6%, seven cases). Regarding the other IDD, the prevalence was 2.2% (ten cases), higher than in a Swiss study [12]. Alzheimer's disease was the most prevalent cause, followed equally by vascular disease and Lewy body dementia. In DS and other IDD the presence of dementia increased as a function of intellectual deficits severity, although the average age was similar between mild, moderate, and severe intellectual disability. The authors also concluded that DSM-5 [3] was the most useful tool for diagnosing cognitive deficits.

Among the ten broader categories of dementia classified by the World Health Organisation (from 6D85.0 to 6D85Y; ICD-11 [1]), the 6D85.9 is the **Dementia due to DS**. The high occurrence of Alzheimer's disease in DS is well known and it increases as a function of age from the fourth decade onward [13, 14]. Major mechanisms related to Alzheimer's disease dementia in individuals with DS are discussed on Chap. 13.

Strydom and colleagues [13] contrasted findings from 17 (cross-sectional and longitudinal) studies and summarised the symptoms that are commonly related to Dementia due to DS. For instance, memory loss, speech slowness, disorientation, and functional decline. These changes course with neurological symptoms such as seizures (late onset of myoclonus epilepsy) and incontinence often associated with the progression of the disease. Dysfunctions in activities of daily living are pronounced in areas of personal hygiene, housekeeping, and dressing. A wide range of personality changes and maladaptive behaviours have been described, all of them leading to a social disengagement. In one side, irritability, aggression, or self-abusive behaviour. In the other side, general slowness, apathy, or loss of interest. In the late stages, individuals become unresponsive, unable to walk, speak, and eat (dysphagia). In the study by Margallo-Lana and colleagues [14] with 92 participants, 23 were deceased due to dementia with an average survival of 3.5 years from the diagnosis to death. Mortality rate is higher among individuals with IDD having dementia than in those without dementia [13].

The neuropsychological assessment is important for the diagnosis and follow up the neurocognitive disorders. It allows the person with IDD, caregivers and healthcare team to plan the best treatment alternatives respecting individual preferences, lifestyle and clinical care needs which will change according to the stages of dementia. In the following sections we will discuss the challenges of neuropsychological assessment in people with IDD.

Considerations on Neuropsychological Assessment

Although some persons maintain very high cognitive performance levels throughout life, the majority of older people will experience a decline in certain cognitive abilities. Similar to adults in the general population, ageing adults with IDD could become less capable of directing attention to and processing information necessary for tasks involving multiple cognitive operations [15, 16], which may parallel changes in physiological function. It is more difficult to evaluate such age-related cognitive changes in individuals with IDD because there are already considerable deficits in intellectual functioning (i.e., low intellectual quotient (IQ), delayed learning or impaired language development) and adaptive behaviour [17] across their life span. In addition, people with IDD are more likely to be exposed to anticholinergic medication, due to a high comorbidity of physical and mental health problems throughout the lifespan that increase with the ageing process [18–21]. More, consideration should be given to the negative outcomes of central

anticholinergics on cognitive, functional performance and behaviour in older adults with IDD [22, 23].

Early detection of dementia in adults with IDD has been limited by a lack of appropriate endpoints that accurately measure changes in cognitive and functional abilities [24–26]. Moreover, the use of direct neuropsychological tests in this population presents some difficulties: most of the screening tests used for the general population show floor effects in individuals with IDD because these instruments are based on the premise that tested individuals have an average IQ of 100.

Neuropsychological screening tests are usually influenced by schooling [27], which impacts on the ability of people with IDD to complete these measures due to their lower educational attainment [28]. By definition, individuals with IDD have cognitive deficits that pre-date the onset of dementia with a varied range of abilities and cognitive function (from profoundly impaired at one end to mild disability at the other). Therefore, it is not currently possible to determine a cut-off score for screening of dementia that can be applied to individuals with all degrees of severity of the intellectual functioning. It is difficult to make an early stage diagnosis of dementia in a person with IDD based on cognitive measures only [29]. It is important to consider that even in general ageing other aspects should be taken into account, for instance, changes in functionality and mental health symptoms [30].

Diagnostic criteria often states that a significant change from an individual's own baseline from a previous level of performance [31] is required for a diagnosis of dementia. An important part of the assessment is to establish this "baseline" in order to determine the highest level of functioning that the person with IDD previously had, and to determine which cognitive deficits may be longstanding, particularly in the presence of severe or profound disability, otherwise it may be more difficult to identify subtle changes in cognitive decline. Assessments over time are a key element in capturing the progression. One relevant question is the age that these follow up assessments should commence given that people with IDD are more likely to have early onset dementia. This is particularly the case for people with DS, where cognitive changes maybe observed in the fourth decade, or even earlier [25, 32].

A general cognitive assessment must examine a range of different domains such as attention, concentration, orientation, short- and long-term memories, praxis, language and executive function, but it is difficult to "optimise" the administration of cognitive tests due to factors such as emotional states, sensory deficits and medical status, then, only trained and qualified clinicians with experience working with people with IDD should conduct and interpret the test results [33, 34]. Nevertheless, the simple transpositions of neuropsychological tests to diagnose dementia in the general population to people with IDD should be avoided because it generates poorly specific or misleading results. This conduct has been inappropriately adopted in past years, for instance, the original version of the Mini-Mental State Examination [35] has generated questionable findings [36, 37] and general floor effects [38]. On the other hand, the use of specific instruments to assess changes in memory/cognitive functioning and to distinguish normal changes from early signs of pathology remains one of the most effective methods in discriminating dementia from

age-related cognitive decline [39]. In Box 5.1 we present a case study as an example of person-centred care focusing on cognitive and behavioural assessment.

Box 5.1 Case Study: An Italian Experience

The DAD Project. The "Down Alzheimer Dementia" project (see http://www.validazione.eu/dad/) is an ongoing prospective study, which aims to investigate the feasibility and effectiveness of person-centred dementia care and the related environmental and psychosocial interventions provided to a group of adults with IDD (including DS) and dementia.

The Service. "La Meridiana" is a temporary residential care setting, designed to proactively accommodate dementia-related symptoms in adults with IDD, funded in 2005 by a parent-based association in Trento, Italy. The DAD Project was almost entirely financed by *ANFFAS Trentino Onlus* including the architectural features (spatial layout, equipment, and furnishings). This daycentre provides individual grooming and routine domestic activities, also recreational, occupational, and music activities, motor rehabilitation sessions, or other physical activities, carried individually or in groups. External activities include shopping, walks, one-day trips, and weekly animal-assisted activities [40].

The assessment. A brief standard protocol to follow up neurocognitive changes was developed. The baseline screening used the NTG-EDSD_I (NTG National Task Group-Early Detection Screen for Dementia), adapted from the DSQIID [41, 42] and a repeated measure after about a year. The form can be completed in 15–60 min by family member, agency support worker, or health specialist that had been with the adult in the last 6 months. Check (www.aadmd.org/ntg/ screening) for further details. Significant deviation in the Real Change Index [43] is suggestive of deterioration. In this case, further instruments are applied such as the DMR [44, 45], the AFAST [22] and in the presence of a significant change in behaviour, they were evaluated through the AADS [46]. In the first year of this process, the DMR is repeated two or three times, if there are signs of a depressive state, with subjects with severe disability and whenever there is a change in therapy. For example, in 2017/2018, cognitive decline was identified in 22 out of the 156 subjects with IDD of various aetiologies (mean age 47 years ± 6.3) using the NTG-EDS-I. Among them eight (36%) persons had a depressive state that improved through changes in intervention plans and drug treatment. The staff—pedagogists, psychologists, general practitioners, psycho-geriatricians and nurses—examines the impact of specific stressor events and also all the medical conditions contained in the second sub score sheet. When a significant decline on DMR scores is observed, a geriatrician expert in people with IDD, in agreement with the general practitioner, proceed a clinical examination aiming to distinguish the type of dementia.

Post-Diagnose. The staff begins to support changes in people's life, meeting together the family, local social services and service operators to discuss the most adequate interventions available considering the person history and state and the social-cultural context. It includes contacting the associations of family members of people with dementia who are active in the territory for wider support to the family, whenever possible. The major challenges that have emerged in recent years are linked to the great difficulty of family caregivers in accepting a diagnosis of dementia in their loved one, neither individual/group training nor self-mutual help have proved effective support. In order to support people with IDD even in the most advanced stages of dementia when health and end-of-life issues may be more relevant, a prototype of a dedicated residence (hospice) was designed and will be built in the coming years.

Outcomes. In the last 15 years the project followed up approximately 70 people with DS within ageing services and all have shown signs of cognitive decline or dementia whereas it has not happened in people with IDD of other aetiology. Since 2013 the project expanded to other regions of Italy supporting more than 600 people with IDD.

Instruments for Measuring Dementia-Related Aspects in Adults with IDD

We have based our recommendations on the brief and practical indications stated in a recent report of the UK National Institute for Health and Care Excellence (NICE). This document suggests that the following steps for an assessment of dementia should be conducted in people with IDD and recommends the completion of a "baseline assessment of adaptive behaviour with all adults with DS" [51]:

Measure of Symptoms

These measures include an indirect examination that must be based on concern from an individual or knowledgeable informant, with instruments completed by caregivers of individuals with IDD, i.e., an informant-rated behaviour questionnaire. However, as it was stated above, an adjustment to the DSM-5 major neurocognitive disorders criterion for neuropsychological testing may be used, with the informant-based questionnaire to demonstrate decline.

Measure of Adaptive Function to Monitor Changes Over Time

A complete assessment of daily living skills can be conducted using the AAMD Adaptive Behaviour Scale [52] or the Vineland Adaptive Behaviour Scale [53]. However, it is also acceptable to use a simplified tool to monitor the changes in the daily living skills, e.g., the Alzheimer's Functional Assessment Tool (AFAST) [37]. The Daily Living Skills Questionnaire [54] was noted to be effective in early detection, showing changes indicative of dementia 3–4 years before diagnosis. The Activities of Daily Living Questionnaire [55] and The Barthel index [56] were better explained by disability level and comorbidity than dementia status, and therefore, these were the only informant measures that were considered to be unsuitable for use in dementia diagnostics for people with IDD.

This also applies to monitoring behaviours as people with IDD can manifest executive dysfunction and challenging behaviours superimposable with a pattern of dysfunctions known as the Behavioural and Psychological Symptoms of Dementia that are commonly observed during preclinical or early stages. These aspects may precede memory loss [32].

Measure of Cognitive Functioning

There are a variety of tools that have been used to test memory, attention and other aspects of cognitive functioning in adults with IDD, with standardized instruments that have been administered in clinical or research settings.

Table 5.2 describes some of the most widely used batteries. Overall, there are instruments specific for IDD and dementia of Alzheimer's Disease (AD); others are not specific for IDD or AD. However, there are specific instruments for IDD but not for AD and the opposite, specific for AD but not for IDD [68]. Not all the instruments have been studied for the purpose of dementia diagnostics in individuals with IDD. Like many authors of the specialised literature, among which we mention one of the first ones as Matthews [69], we suggest using only tools that have had, at least, a minimum psychometric calibration in this specific population.

Among the batteries indicated, for example the Arizona Cognitive Test Battery [ACTB [70]] was developed to assess a range of skills and has variable scores with low floor effects, good test re-test reliability and is suitable for a range of ages and contexts, and for a non-verbal population. However, this battery was validated in a sample of individuals with DS aged 7–38 and "*it would not be an appropriate outcome measure of cognitive function for clinical trials of dementia treatments without further modification and validation*" p. 1 [71].

Table 5.2 Main batteries for diagnosis of dementia in IDD

Instruments	Domains
CAMCOG-DS[a,b] (part of The Cambridge Examination for Mental Disorders of Older People with Downs Syndrome and Others with Intellectual Disabilities) CAMDEXDS [57]	Memory, orientation, language, attention, perception, praxis and calculation.
Cambridge Neuropsychological Test Automated Battery (CANTAB®) [58]	Visuospatial short-term memory, attention, rule Learning, set shifting and working memory.
DAMES[a] (Down's Syndrome Attention, Memory and Executive Function Scale) [59]	Attention; memory; executive functions.
Cognitive Assessment System (CAS) [44]	Planning, Attention, Simultaneous, and Successive Processes.
Neuropsychological assessment of dementia in adults with intellectual disability (NAID)[a,b] [60, 61]	Memory, language, picture and object naming and identification, actions on request orientation.
LonDownS adult cognitive assessment[a] [62]	General abilities, visuospatial and verbal memory, language skills, executive functions and motor coordination abilities.
Prudhoe Cognitive Function Test (PCFT)[a]—in a long and two Short Form [59, 63]	Memory, orientation, language, praxis and calculation.
Rapid Assessment for Developmental Disabilities (RAAD)[a] [64, 65]	Memory, language, executive functioning, motor performance, new learning.
Severe Impairment Battery [66] (SIB)[b]	Memory, orientation, language, attention, perception, construction, praxis and social interaction.
Test of Severe Impairment—Modified [33, 67] (TSI)[a]	Memory, language, conceptualisation, praxis and general knowledge.

[a]Designed specifically for use in individuals with IDD
[b]Designed specifically for evaluating dementia

In Box 5.2 we have answered common questions that might help family and professionals that have concerns about the assessment of cognitive functions in adults with IDD.

Box 5.2 Common Questions About the Dementia Assessment

What is best way to detect dementia: tests per functions or a battery?

The use of a battery, with good psychometric properties, is recommended, which should assess a range of skills. The battery must be applicable to a wide range of ages to allow easy comparison across various control populations and for the tracking of long-term change.

What age should follow-up of cognitive and behavioural changes start?

In adults with DS, it is recommended that a baseline assessment is carried out before the age of 40 and then prospective assessments should be carried

out regularly to identify early signs of cognitive and functional decline. In other people with IDD, assessments should be carried out when there is suspicion of cognitive decline or respecting the recommendations for the general population.

What is the ideal interval between dementia-related assessments?

The retest interval depends on individual contingencies but usually a period of 1 year is recommended between assessments.

Can we diagnose dementia with a single assessment?

A single point time assessment is not recommended [72], rather a longitudinal assessment is preferred. However, where there is a marked deterioration from previous levels of functioning then it is possible to diagnose dementia from a single assessment.

When should proxy assessments be carried out?

Direct testing of memory and sustained attention may be particularly useful measures to track decline in the preclinical/prodromal stages of AD in DS, whereas informant-measures may be useful in later stages of dementia or in people with more severe disability.

Why should we focus on early detection of dementia in adults with IDD?

Earlier age of diagnosis facilitates access to pharmacological treatment and behavioural interventions, which may enhance quality of life. Younger age of diagnosis and treatment with dementia medication are associated with longer survival in people with DS [73].

Can we use the same predictors from DS in other forms of IDD?

Executive function, disinhibition and apathy show a correlation with cognitive decline and Alzheimer's disease in adults with DS [68], but more research is needed to investigate the predictors of cognitive decline in other IDD samples.

Who should be involved in completing this comprehensive assessment?

A differential diagnosis requires multidisciplinary expertise complementing the neuropsychological assessment to ensure the delivery of person-centred care. It includes a psychiatric history, mental state examination, physical examination and appropriate clinical investigations (dementia screening blood tests and brain imaging).

Considerations About Dementia-Related Instruments

This chapter was focused on cognitive decline per se without scrutinize different types of dementia, then the neuropsychologist must consider specific instruments when targeting different diseases, for instance, Parkinson's disease in contrast with Alzheimer's disease.

In the last 20 years, several studies have used different tools and methods, for example 114 different instruments were extract by Zeilinger's review [40] (35 informant-based questionnaires; 79 direct cognitive tests and four batteries) and 44

instruments were reported in the Elliott-King and colleagues study [37] (11 informant-based questionnaires; 23 direct cognitive tests and ten batteries). There is still no international agreement upon a single neuropsychological test battery in adults with IDD [35, 37, 40, 47].

A systematic review by Paiva and colleagues [74] updated this list. Authors found out that the Dementia Questionnaire for Learning Difficulties (DLD) [48]—previously named Dementia Questionnaire for Persons with Mental Retardation (DMR) [49] was the most frequent instrument used in both cross-sectional and longitudinal studies for people with DS. Nevertheless, it was neither associated with changes in beta-amyloid precursor protein nor to The Severe Impairment Battery (SIB [66]) performance, which may indicate lack of sensitivity. As for other forms of IDD the DLD [48] and The Wolfenbütteler Dementia Test for Individuals with Intellectual Disabilities [50] have been used in longitudinal studies. Regarding cross-sectional studies the (AFAST [37]) along with (CAMCOG-DS [75] and DLD [48]) were effective in other forms of IDD. The TSI is appropriate to measure the progression of dementia in people with severe disability [76]. This review also detected behavioural measures such as Assessment for Adults with Developmental Disabilities (AADS [77]) and The Adaptive behaviour dementia questionnaire [78].

The most recent studies or reviews [38, 40, 74, 79–81] suggest that a battery composed of different instruments or the use of a multidisciplinary approach seems to be more successful in informing diagnosis and more effective in detecting dementia in people with IDD. Normally, the batteries contain a variety of instruments including both direct cognitive tests and informant reports, so they can be helpful and effective in discriminating between IDD dementia cases.

We have already highlighted that the longitudinal use of the chosen screening instrument is crucial in order to accurately inform diagnosis, as many studies have noted that the variability in cognition of individuals with IDD makes it almost impossible to recommend one specific instrument. Therefore, selecting appropriate tests for the individual is the key and they could be varied in length from 20 min (e.g. when using the SIB) [66], or up to 1–2 h (e.g. when using the CAS) [44]. This choice requires obtaining knowledge of the patient's history and making observations of their cognitive functioning as well as their emotional, motivational and daily functioning. It may be necessary to complete a shorter instrument with individuals with more severe IDD. Nonetheless, breaks should be offered to participants throughout testing, and it is always possible to split testing sessions into multiple shorter sessions. As for individuals with comorbid disabilities or who cannot be assessed through direct cognitive testing, proxy assessments should be completed. Informant tools like CSDS [81] may help to assess people with multiple disabilities or more profound cognitive deficits, but it is very important in this case to refer to an early or juvenile baseline assessment.

Following the NICE recommendation [51] every assessment must provide cluster scores per skills: motor, socialisation and communication, personal living, community living, and broad independence. A concise and practical guide on the role of some neuropsychological testing to support the dementia assessment can be found also in recent handbooks [82, 83].

Conclusion

Neurocognitive assessments in people with IDD can be a complex process and should be conducted by professionals with relevant expertise. The assessment requires a detailed assessment of the person's baseline functioning and subsequent assessments over time in order to identify a deterioration in cognitive functioning. Neurocognitive assessments should ideally include direct cognitive assessment of a range of cognitive domains or the use of an informant measure for people unable to complete direct testing. There is currently no single instrument that is recommended; the choice of instrument is dependent on individual factors such as the severity of IDD. In people with DS, baseline assessments before the age of 30 are recommended followed by regular longitudinal assessments. Neurocognitive assessments should be part of a multidisciplinary assessment that also includes a comprehensive psychiatric history, laboratorial examination, and physical investigations.

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Chapter 6
Depression in Ageing Adults

Vee P. Prasher, Victoria Lane, and Andreas Kitsios

Introduction

The occurrence of depression in ageing adults with intellectual and developmental disabilities (IDD) remains an under investigated area of research [1–3], especially when contrasted to our understanding of depressive symptomatology in the older general population [4, 5]. Depression is rarely recognised as a treatable illness, despite the significant detrimental impact it can have on quality of life [6] and all aspects of physical and mental health. It is therefore extremely important that the appropriate assessment, diagnosis and treatment of depression and indeed all other forms of mental illness is completed by multidisciplinary professionals working with vulnerable older adults with IDD.

This chapter provides an update highlighting important clinical and research findings relating to depressive symptomatology in the field of IDD, with a particular focus on ageing individuals. Individuals with IDD generally have a lower life expectancy that their counterparts in the general population (See Chap. 1), therefore the term 'ageing' is variably applied in the published literature from the age of 40 years [7]. Considerable heterogeneity exists in the methods and criteria used to diagnose study participants with depression. This chapter therefore includes all relevant research pertaining to adults with IDD with symptoms suggestive of a depressive illness, regardless of if these symptoms meet the specific diagnostic thresholds outlined in the Diagnostic Criteria for Psychiatric Disorders for Use with Adults with Learning Disability [8], the International Classification of Diseases [9] or the Diagnostic and Statistical Manual of Mental Disorders [10].

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Epidemiology

In the general population, depression is one of the most common psychiatric disorders affecting approximately 25% of women and 10% of men in their lifetime. In 2005, a nationwide survey in the UK on the prevalence of depression in 4269 individuals aged 65 years and over was completed using the self-administered 10 item Geriatric Depression Scale. Overall, 28% of women and 22% of men reported a score of three or more (indicating a probable diagnosis of depression). The gender disparity reduced when individuals aged over 85 years were assessed, with 43% of women and 40% of men reporting high numbers of depressive symptoms [11].

Overall, the risk of an affective disorder in adults with IDD appears to be increased when compared to the general population, with studies quoting a four to sixfold increase [12, 13]. Early data from 1993 on 798 individuals with IDD living in a residential setting identified that 4.8% had an operational related diagnosis (Diagnostic and Statistical Manual of Mental Disorders [10]) of current depressive disorder and that women were more likely to manifest dysthymia than men [14]. Preliminary data supports this finding that the gender distribution of depression in the IDD population mirrors that of the general population, with women having a higher risk [15, 16]. Mild depression may be more common in adults with mild/moderate severity of disability; in a sample of 151 adults assessed using the Beck Depression Inventory II, 39.1% of participants reported symptoms of depression. Of these, 43 were mild, 14 were moderate and two were severe [17]. In a Swedish sample of subjects with IDD, the relative risk of any mental disorder was 1.34 when compared to a reference sample, with 44% of subjects having a mental disorder according to Diagnostic and Statistical Manual of Mental Disorders-IV criteria and 11.5% of these having a mood disorder [18]. In a different sample of 142 adults with different degrees of IDD and no previous psychiatric diagnosis, 29% were found to have a previously undiagnosed psychiatric disorder, with depression being more prevalent in the milder subcategories of IDD (assessed using the Psychiatric Assessment Scale for Adults with Developmental Disability (PAS-ADD) checklist, mini PAS-ADD and PAS-ADD-10) and anxiety being more prevalent in severe disability (assessed using DASH-II) [19]. The finding that depression occurs more commonly in mild when compared to severe level of disability has been replicated elsewhere [14, 18, 20] and it has been proposed that psychiatric models of illness based on observations from the general population become less applicable as the severity of disability increases [20]. Despite the consistently higher prevalence rate of depressive disorders reported in the IDD population, a recent German cohort study (651 individuals with IDD living in a defined community) provides evidence that the incidence rate of depression in adults with IDD is similar to that of the general population (7.2%). The authors suggest two possible explanations for this discrepancy; either that depression in IDD subjects is more enduring or that this population is undertreated [21].

Regarding older individuals with IDD specifically, prevalence rates of mood disorders have been estimated to be between 7% and 27%. An early study by Patel

reviewed 105 mixed community and institutional subjects with IDD aged over 50 years using a semi-structured clinical interview (PAS-ADD) and identified that 11.4% had a psychiatric disorder (mostly depression and/or anxiety). Interestingly, many of these individuals were not known to mental health services, despite their care staff being aware of their difficulties [22]. An Irish study reviewed 753 individuals with IDD aged above 40 years using the Center for Epidemiological Studies-Depression (CES-D) scale and reported a case-level depressive illness in 11% and sub-threshold depressive symptoms in a further 27.1% [23]. A Dutch sample of 990 adults aged over 50 years with borderline to profound ID reported depressive symptoms in 16.8%, major depressive disorder in 7.6% and mixed anxiety and depression in 0.7%. No relationships were noted in this sample between gender, age or severity of disability [24]. An earlier study also reported no significant associations between depressive symptomatology and increasing age [25]. A large cross-sectional study screened 990 adults with IDD aged over 50 years and identified 17% had symptoms of depression [26]. A recent large Swedish cohort study utilised the National Patient Register (information on inpatient and outpatient care) to compare 7936 individuals with IDD aged above 55 years living in a specialist care setting to a matched sample from the general population. In the IDD cohort, 7% had an affective disorder compared to 4% of controls, with a calculated odds ratio of 1.74. It appeared that fewer psychiatric diagnoses were made as the age of individuals with IDD increased; in 2012 30% of those aged 55–59 had a diagnosis compared to 20% of those aged over 75, though the reasons for this were unclear [27].

There remains a paucity of data on the epidemiology of depressive symptomatology in the IDD population. In addition to this, considerable heterogeneity in the published literature makes it extremely difficult to compare groups directly to draw robust and reliable conclusions. Several methodological differences should be considered when reviewing and comparing the data;

- Broad inclusion of a number of affective presentations including; low mood, current/lifetime episode of major depressive disorder and bipolar affective disorder.
- Deficiencies in sample selection (for example, the use of a small sample size or sole recruitment of institutional subjects) resulting in poor external validity; with the study sample not accurately representing the wider population.
- The variable use of instruments with poor internal validity and reliability to detect psychopathology.
- The variable application of diagnostic criteria to confirm diagnoses. The study by Mantry and colleagues [28] demonstrates the potential impact of this; in a sample of 89 individuals with DS, the point prevalence of mental illness was found to be 23.7% when diagnosed clinically, 19.9% when diagnosed using the DC-LD, 11.3% when diagnosed using the ICD-10 and 10.8% when diagnosed using the DSM-IV criteria.
- Poor methodological control of variables including the severity of disability and other demographics such as age. Furthermore, the quality of matching of the control groups also varies.

With the above limitations in mind, a number of tentative observations can be made. When adults with IDD are compared to matched control populations, they appear to have a higher prevalence of depressive illnesses. It is possible that this discrepancy equalises with increasing age, as when results for the older IDD population (40 years and above) are compared to the older general population (65 years and above), prevalence rates appear to be approximately equal (IDD population, 7–27% versus general population, 22–28%). Rates of depression in adults with IDD appear to be higher in females and in those with a milder degree of disability.

Aetiology

No single factor has been identified as the cause of depression in subjects with IDD or in the general population. It is likely that combinations of biological, psychological and social factors are important (Fig. 6.1).

Genetic Factors

With regard to specific conditions that commonly present in individuals with IDD, studies outline specific results relating to persons with Down syndrome (DS) and Fragile X. It is unclear if these conditions truly have a higher prevalence of

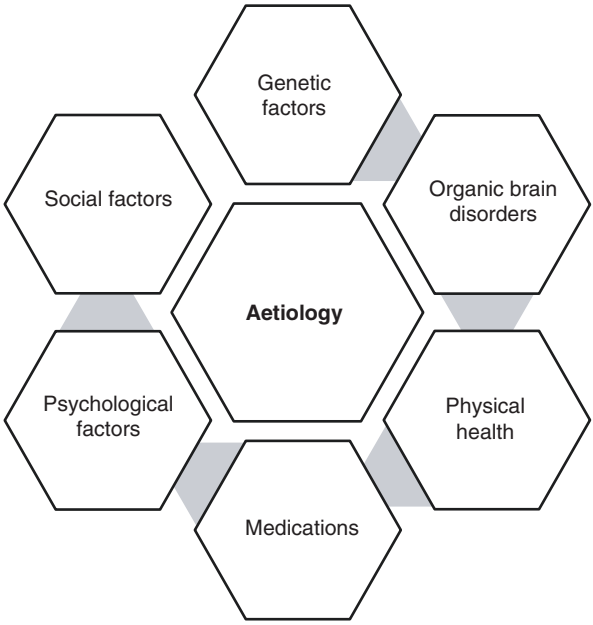

Fig. 6.1 Aetiological factors contributing to depressive illness in IDD population

depression than other IDD subcategories, or if they appear to, simply because there is a paucity of data in other areas.

In the DS population, one study reported a 2 year incidence rate of 5.2% for depression. Poor mental health in this study was specifically associated with urinary incontinence [28]. Another study reported that in a sample of 89 individuals with DS (mean age 42 years) the prevalence of mood disorders was 43%. This study highlighted the increasing life expectancy of the DS population and stressed the importance of improving our understanding of the challenges these individuals face as they age. The study sample included behavioural difficulties in 53%, epilepsy in 22%, osteoporosis in 40%, hypothyroidism in 53%, hearing impairment in 82% and severe cognitive impairment in 67% [29]. Walker commented that individuals with DS are subjected to a number of factors that may act to increase their risk of depressive illness, including; reduced hippocampal volumes, variations in levels and activity of neurotransmitters, deficits in working memory and language, a tendency towards certain attachment behaviours and an increased risk of somatic complaints [30].

High prevalence rates of depressive disorders were also seen in subjects with Fragile X; out of a sample of 47 individuals with Fragile-X associated tremor/ataxia syndrome (mean age 66 years), 65% met the DSM-IV-TR criteria for a mood disorder which was significantly higher than population-based controls [31]. A second study reporting on 46 women with the fragile X mental retardation 1 gene premutation, identified that 75% of participants had a lifetime history of either a depressive or an anxiety disorder, with 43% reporting both [32]. It has been hypothesised that mood disorders may form part of the clinical phenotype of fragile X [31] and that this effect may arise secondary to progressive mRNA toxicity in the limbic system [33].

Organic Brain Disorders

It might be expected that abnormalities in the structure and functioning of the brain might pre-dispose individuals to difficulties with mood, behaviour, personality and language. The positive association between depression and milder forms of disability has already been discussed. In addition to this, adults with both epilepsy and IDD had more than seven times the risk of developing a psychiatric condition, particularly a mood disorder when compared to individuals with IDD alone [34].

Physical Health

Chronic illness in the preceding 5 years is associated with an increased prevalence of depression in older adults with IDD, including congestive cardiac failure, ischaemic heart disease, cerebrovascular events, chronic obstructive pulmonary

disease, diabetes mellitus, malignancy [24] and visual impairment [23]. Many of these conditions may have a bidirectional influence on depressive symptomatology. For example, depression may contribute to an indolent lifestyle that increases the risk of metabolic syndrome and cardiovascular disease. Subsequent cardiovascular disease and associated polypharmacy may contribute to worsening of mood, which may in turn influence recovery and long term outcomes. These links are complex and not always immediately apparent; in a large study on 990 adults with IDD aged over 50 years assessing the link between psychopathology and physical illness, the only association to reach statistical significance was anxiety symptoms and diabetes [26].

Medications

Individuals with IDD have a higher rate of drug administration and polypharmacy [35]. Older adults with or without IDD are more susceptible to adverse drug reactions stemming from toxicity, side effects or drug interactions. Drugs of particular concern include steroids, beta-blockers, antihypertensive medications, antiarrhythmic drugs and anti-epileptic medications. As medications can have a significant impact on mental state, they should always be reviewed and excluded as the causative factor prior to a diagnosis of depression being made.

Psychological Factors

A number of psychological factors have been associated in the literature with a higher prevalence of depressive disorders, including loneliness [15, 23] and higher stress levels [15, 36]. A study assessing 43 adults with mild to moderate severity of disability using an adapted version of the social comparison scale identified that adults who reported low social attractiveness, low levels of group belonging and poor self-esteem, were more likely to suffer from depression [37]. Associations with downward social comparisons, poor self-esteem and negative automatic thoughts were replicated in a later study on 151 adults with mild to moderate disability [17]. Excessive reassurance seeking behaviour has also been associated with depression in adults with IDD and it has been suggested that this association is partially mediated by the resulting negative and rejecting reactions that subjects may experience from those around them [38]. A study comparing 47 subjects with IDD with depression to 47 subjects with IDD and without depression reported higher levels of stressful social interactions, a correlation towards a negative attribution style and more passive coping mechanisms in the depressed group [36].

Social Factors

In the general population, it is common for social factors and life events to act as precipitants for a depressive episode [39]. In the IDD population, poor social support has been associated with an increased risk of depression [17]. Life events, which might include a change in residence, relationship difficulties including bereavement, difficulties with authority, substance misuse, trauma and unemployment have also been consistently shown to have a positive correlation with depression [17, 21, 24, 40–44]. It has been identified that adults with IDD living in staffed accommodation may have increased exposure to life events compared to individuals living with natural or foster families [40]. Life events occurring in childhood [44] and those perceived to be negative [41] or traumatic [43] might have a more significant impact on mood. On the other hand, individuals who are able to complete more activities of daily living appear to be less likely to develop depression [24].

Presentation

The National Institute for Clinical Excellence (NICE) recommends the use of DSM-IV criteria to diagnose major depression. Subjects must exhibit at least one core symptom (persistent sadness or low mood and anhedonia) in addition to other symptoms (fatigue, feelings of worthlessness or guilt, suicidal thoughts or acts, reduced attention and concentration, psychomotor agitation or retardation, difficulties with sleep and changes in appetite). A total of five or more symptoms are required to make the diagnosis of depression (Fig. 6.2). Symptoms need to have been present for at least 2 weeks and in this time need to have caused significant distress or impairment. Increasingly it is being recognised that many individuals do not reach the diagnostic criteria for major depression, but they still live with

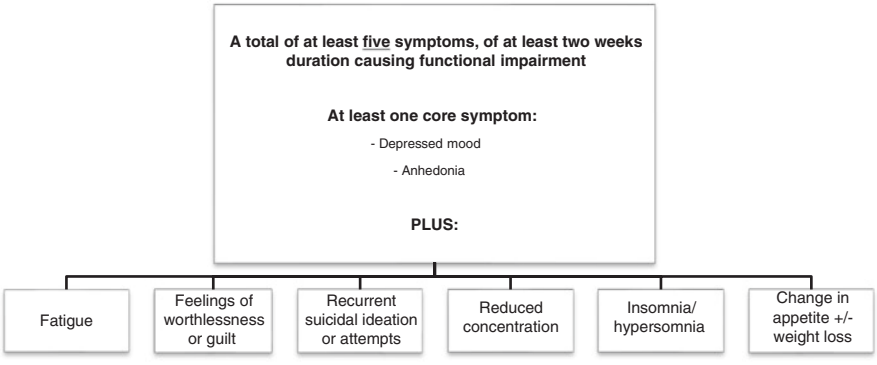

Fig. 6.2 Criteria for a DSM-IV diagnosis of major depressive disorder

disabling and distressing symptoms that impact upon their quality of life. This group are classified as having sub-threshold depressive symptoms (NICE Guideline).

In the general population, it is recognised that older people with depression often experience their symptoms differently. Somatic symptoms, such as sleep become less important diagnostically, as they often occur as a normal part of ageing. Where somatic symptoms do exist, it can be difficult to relate these directly to the depressive illness as the associations are often blurred by multiple physical co-morbidities and polypharmacy. Furthermore, cognitive impairment can commonly occur both as a result of depression and as an early sign of dementia, of which depression can be a preliminary sign [11] (Health survey for England 2005). The diagnostic challenges of differentiating depression from dementia are discussed in greater detail later in this chapter.

Our understanding of depressive presentations in individuals of all ages with IDD is less developed than for the general population. It appears that individuals with mild to moderate severity of disability often present in a similar way to the general population; symptoms may include low mood, a subjective feeling of sadness, crying, fatigue and self-criticism [17]. Following a retrospective review of 300 adults with IDD, the researchers proposed that three core symptoms: sad mood, crying and anhedonia should be used to differentiate and identify depression. This study suggested that many depressed adults with IDD do not meet diagnostic criteria for diagnosis. Possible reasons for this include atypical presentations and deficiencies in both self-reporting and in the observational skills of informants and assessors [45]. It has been proposed that with careful behavioural observations, a pervasive mood state can be distinguished from more rapidly changing emotions. Additionally, a reduction in reactions to positive stimuli over time might point towards a depressed mood in subjects who are less able to communicate their subjective inner experiences [46]. Further research is needed to determine whether impaired concentration, guilt, suicidal ideation and perceptual changes do not occur in individuals with IDD or are present but are not elicited.

Sleep difficulties including insomnia and hypersomnia commonly present in association with depression. A systematic review of sleep disorders in adults with IDD identified a prevalence rate of 8.5–34.1%, with 9.2% suffering from significant problems. Poor sleep was associated with challenging behaviour, breathing problems, visual problems, mental health difficulties and medication use (including drugs commonly prescribed in IDD psychiatry; psychotropic, anti-epileptic and anti-depressant medications) [47]. Sleep dysfunction has a complex bi-directional relationship with depression; it can occur as part of the depressive symptomatology, as a result of medication or it can occur independently but lead to a worsening of mental state and behaviour.

Another important area that requires discussion is the contentious role of challenging behaviours in the diagnosis of depression. It has previously been proposed that challenging behaviours, including self-injury and aggression should be considered as ‘depressive equivalents’ in the IDD population and should be used as evidence to support a diagnosis of depression, particularly in subjects with severe and profound disability [21, 48–53]. In a large study on 4069 participants, it was found

that difficulties with impulse control, mood regulation and perceptions of threat contributed to most episodes of aggressive behaviour [54]. In addition to challenging behaviour, one study also proposed that psychomotor changes and functional decline should point towards a diagnosis of a depressive disorder in adults with severe disability [55]. It is important to note that, publications report mixed findings on this subject; with several studies concluding that there is no evidence to support the association between challenging behaviours and depression [56–60]. Further research is required to examine this association in specific cohorts of the IDD population, specifically with regard to the role of potential confounding factors such as medication use and institutionalisation. As the published literature provides no clear consensus to guide practice, it would be sensible for professionals to screen for challenging behaviours during assessments and to consider the formulation for these on an individual case by case basis. Caution should be exercised when using challenging behaviours as evidence of a depressive illness, particularly when no other features are present.

Suicide

Fortunately, suicide in adults with IDD appears to be a rare occurrence. The reasons for this have not been fully established and may include; a lower cognitive and behavioural repertoire, limitations caused by physical disabilities and reduced access to lethal means. In a nationwide cohort sample of 2677 subjects followed up over 35 years (1962–1998), only ten cases were reported [61]. Risk factors for suicide in this population appear to include ageing, physical disabilities, psychiatric illness (especially if hospitalised), challenging care needs and social difficulties leading to loneliness [61, 62]. It should not be assumed that individuals with IDD will not attempt or commit suicide and professionals should therefore screen for risks, as they would for any other patient and intervene where appropriate.

Assessment and Diagnosis

A number of challenges may be encountered when diagnosing and treating individuals with IDD, including limitations in communication and atypical clinical presentations [45]. Interactions should be modified to the communication needs of each individual patient, with consideration of their severity of disability, their response style and their degree of capacity and ability to consent. Collateral information should be obtained from additional sources, including family members and carers [63] and where appropriate multidisciplinary team involvement should be facilitated. Assessments should be systematic and include; measurement and diagnosis of psychopathology according to specific diagnostic criteria (charts may be useful to monitor weight, sleep and behaviour), an assessment of risk and

identification of possible aetiological factors using a biopsychosocial framework and developmental information.

When assessing a patient with IDD for a primary depressive disorder, it is imperative that a number of other differential diagnoses are considered (Fig. 6.3). This is particularly important in the elderly, as depression may occur in conjunction with or secondary to a number of different conditions. A detailed history, physical examination and appropriate investigations should point towards potential causes. This will also highlight any psychological or social antecedents contributing to the clinical presentation that are extremely important to the development, presentation and course of depression, but are often poorly captured by the current diagnostic systems. The choice of investigations needs to be adapted to the individual case, but may include bloods tests (full blood count, renal function, liver function, thyroid function, blood glucose, calcium, magnesium, inflammatory markers, *human immunodeficiency virus* and syphilis serology), drug screening and neuroimaging if an intracranial cause is suspected. The drug history (including substance misuse) should also be considered including medication side effects, toxicity and

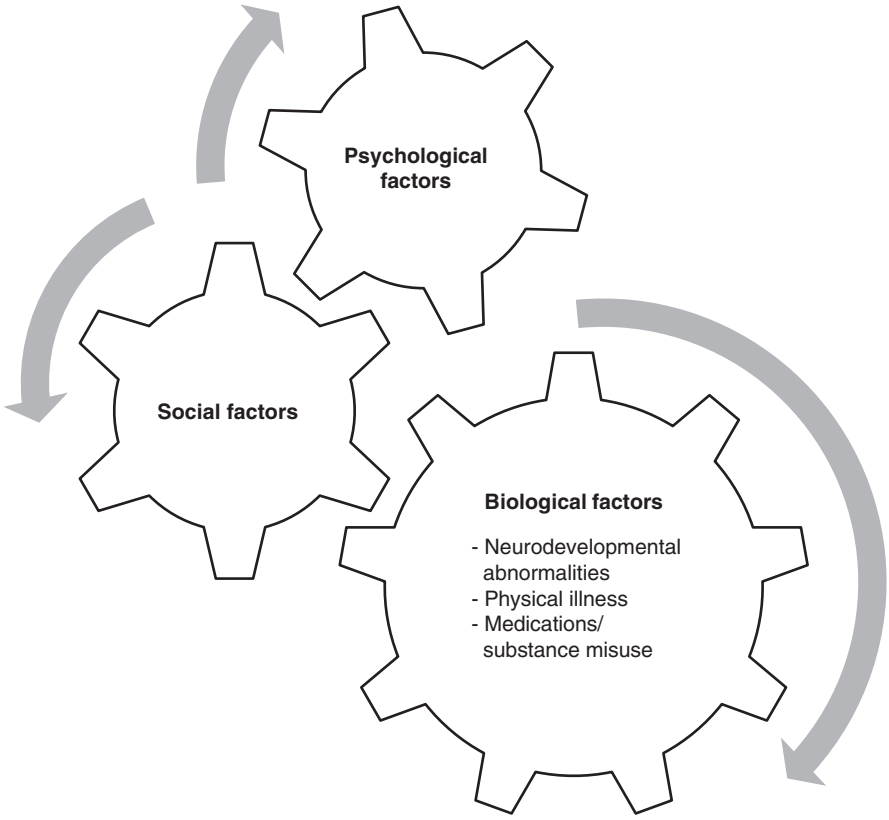

Fig. 6.3 Considerations for differential diagnosis of depressive symptomatology

interactions. The above list is not exhaustive and should be modified at the discretion of the reviewing clinician. It is reasonable to expect that mood changes occurring secondary to another cause are less likely to fulfil the diagnostic criteria for a depressive disorder than primary disease.

A number of other psychiatric disorders may present in a similar way to depression. In the mental state examination, additional features of psychopathology should be screened for including a history of suicidal ideation, elevated mood, anxiety symptoms, psychosis and cognitive impairment. As for the general population, individuals with IDD and bipolar affective disorder tend to present with elevated mood, irritability, increased verbalisation with pressured speech and inappropriate sexualised content, increased appetite and poor concentration [45].

Differentiating depression from dementia can be difficult [1, 64–66], not least because the conditions can co-occur, or can imitate each other; depression can manifest as cognitive impairment termed pseudo dementia and dementia can be associated with mood symptoms [67]. When symptoms do co-exist, caution is advised on underdiagnosing depression as treatment can lead to significant improvements in quality of life and functioning [68]. It can take time to clarify the diagnosis and clinicians could consider longitudinal assessment, including (1) a review of the subject's history in detail with careful inclusion of collateral information, (2) an evaluation of previous assessments of cognition, behaviour, mental health and functioning to detect a change (clear records and baseline assessments can assist this process), (3) monitoring the pattern and evolution of symptoms, (4) an assessment for differentials using targeted investigations, (5) combing all of this information to refine the diagnosis [69].

Psychometric Assessment Tools

A number of psychometric assessment tools exist, that can aid the clinical diagnosis of depression. These tools can improve the detection of psychiatric illness and monitor progression, reduce diagnostic disagreement, improve communication between healthcare providers, structure research findings to allow comparisons both within and between studies and facilitate service planning. Assessment tools may take account of the subject, in addition to their family and carers. Both modified and unmodified rating scales have been proposed for use in persons with IDD (Table 6.1), but their correlation to clinical diagnoses have not been fully established, particularly for older individuals. Such scales include the Beck Depression Inventory [70], the Zung Self-Rating Depression Scale [71], the Hamilton Rating Scale for Depression [72], the Mental Retardation Depression Scale [73] and the Self-Report Depression Questionnaire [74].

Although the use of these instruments enhances clinical assessment, particularly when used repeatedly as a means of monitoring treatment response, they should not be used as the sole means of making a diagnosis. They do not replace history taking, examination of both mental and physical state and appropriate investigations. It is

Table 6.1 Commonly used instruments to detect depression

Instrument	Comments
Psychiatric Assessment Scale for Adults with Developmental Disability (PAS-ADD)	Interview schedule for completion by trained interviewer of patient and informant
Diagnostic Assessment Scale for the Severely Handicapped (DASH)	Rating scale assessing the severity, frequency and duration of symptoms for 13 psychiatric disorders. Informant based. For severe/profound IDD
Reiss Screen for Maladaptive Behaviour	Rating scale for eight psychiatric disorders. Informant based. For mild/moderate IDD
The Psychopathology Inventory for Mentally Retarded Adults (PIMRA)	Scaled self-report screening questionnaire. Informant based. For mild-moderate IDD

also worth remembering that although studies have generally shown good internal validity for subject versus informant reports, the subjective experiences of the patient are extremely important when assessing for a mental illness and should not be overlooked simply because they can be more difficult to obtain. Generally it has been shown that family reports tend to agree more fully with subject reports, than those of staff informants [75]. Prior to the application of any given instrument, clinicians should always satisfy themselves as to the reliability, validity and practicalities of the test.

Treatment

As for all mental health conditions, the treatment of depression should involve a biopsychosocial approach. NICE recommends that individuals with persistent subthreshold depressive symptoms, mild depression or moderate depression should initially be offered a low intensity form of psychological therapy, which might include individual guided self-help cognitive behavioural therapy (CBT), computerised CBT or a structured group exercise program. The routine use of anti-depressants in those with subthreshold or mild depression is not recommended as the risk to benefit ratio is poor; a systematic review identified no difference in the treatment versus placebo arms in this group [76].

In persons with persistent/long-term symptoms (usually 2 years of more), or a current/previous episode of moderate or severe depression, an anti-depressant medication can be considered as part of the treatment strategy. NICE recommends that first line treatment should usually be a selective serotonin reuptake inhibitor (SSRI) and that this should be continued for at least 6 months following remission of the episode (Table 6.2). These medications have a more favourable side effect profile (reduced propensity for sedation, cardiotoxicity and anticholinergic side effects and a reduced risk of toxicity on overdose) and have been found to be as efficacious as other antidepressants. Medical therapy can lift mood and facilitate improved engagement with high-intensity CBT or interpersonal

Table 6.2 Examples of SSRI medications

SSRI	Minimum effective dose in general population (mg/day)
Citalopram	20
Escitalopram	10
Fluoxetine	20
Fluvoxamine	50
Paroxetine	20
Sertraline	50

therapy (IPT). Mindfulness-based cognitive therapy can be useful for subjects who are well but have a significant past history of depression, with three or more previous episodes.

Treatment for older individuals with depression follows the same principles as outlined above. Clinicians should be cautious when prescribing medications as there is an increased risk of toxicity, side effects and drug interactions, not least because older people tend to be prescribed more medications (particularly anticholinergics and those acting on the central nervous system) and the absorption, distribution, metabolism and elimination of drugs changes with age. It is therefore appropriate in this population to prescribe lower initial doses and reduce the rate of drug titration [77–79].

The NICE guideline for the management of depression outlines the evidence based guiding principles that all health care providers should following when treating depression. It is worth noting however that this guideline was created using data drawn from the general population. People ageing with IDD have unique patterns of morbidity and mortality that are different from the general population and as a result, aspects of this clinical guideline might be difficult to apply and may in fact contribute to the health inequalities already experienced by this vulnerable group [80]. A number of studies have explored specific aspects of treatment and these are summarised below:

Biological Management

With regard to medication, SSRIs are recommended as first line medications by NICE for depressive illnesses. It has been suggested that these drugs may be particularly efficacious in the IDD population, as they may work to correct some of the underlying serotonergic abnormalities that occur secondary to a range of developmental pathologies [81]. There is also evidence, that in addition to their role in treating depression, SSRIs and tricyclic antidepressants can improve problem behaviours [82].

Melatonin has been reported to be an effective treatment for sleep disorders associated with a number of psychiatric conditions, particularly for subjects with

co-morbid autism spectrum disorder. No information is given on the dose or titration schedule that should be used. No adverse effects were noted with its use [83].

For many of the reasons discussed in detail in this chapter, making a diagnosis of depression in individuals with IDD can be extremely challenging and as a result there is the potential for misdiagnosis. It has previously been proposed that in cases where the diagnosis is unclear, response to medication can be used as evidence to confirm a solid diagnosis. Some clinicians [84] provide a compelling argument that this approach should be avoided as it is divergent from what is known about psychopathology in general, the biochemistry of drugs and their complex interactions. Caution is recommended when using this argument in clinical practice and it is recommended where there is doubt about diagnosis, the differential and formulation should be revisited.

In addition to medication, other biological therapies with specific research findings relating to the IDD population include light therapy and Electroconvulsive Therapy (ECT). Light therapy is commonly used to treat seasonal affective disorder in the general population. It has been suggested that this treatment might be a useful adjunct in the management of depression in adults with IDD, not least because it is an inexpensive therapy that is unlikely to cause harm. Further research is needed to identify if light therapy confers any benefit for seasonal or non-seasonal depression in the IDD population [85, 86].

Despite the high prevalence of psychiatric illness in the IDD population, reports on the use of ECT remain scarce. Electroconvulsive therapy is currently indicated in the general population for the treatment of severe or refractory depression, catatonia and prolonged or severe mania. A review of the use of ECT in the IDD population identified a positive outcome in 79% of treated subjects and a complication rate of 13%, with no reports of cognitive decline. Approximately 70% of subjects were maintained on medication following treatment and 32% of subjects were reported to relapse. The authors recommend that ECT should be recognised as a valuable treatment and not viewed only as a last resort. The challenges of using ECT in the IDD population are recognised, these include; diagnostic uncertainty, complexities around capacity and consent, ethical and legal issues and a scarcity of data on the safety of this treatment [87].

Psychological Management

Psychological therapies for individuals with IDD are commonly used but their evidence base is not as robust as many of the medical therapies. Often these therapies are adapted to the individual's ability and motivation to engage with the intervention.

Cognitive Behavioural Therapy (CBT) is increasingly being used as part of the management strategy for individuals with IDD. It is centred on the concept that thoughts can impact on the way subjects feel (emotionally and physically) and behave. It involves modifications of distorted or dysfunctional thinking and aims to help subjects achieve a more realistic and adaptive perception of events. When using

CBT in IDD populations, it is important to recognise the challenges that these people face; not only do they have to manage the limitations and effects of cognitive distortions, they also have to cope with baseline cognitive deficits. Cognitive Behavioural Therapy can be used in individuals with IDD, but adaptations of the therapy are often necessary. Such adaptations include involving carers, simplifications of the delivery of therapy and simplifications of the model [88]. Often, therapists need to adopt a more direct approach, rather than the collaborative approach traditionally used in the general population [88]. Studies have shown that it is possible to deliver CBT [89], including computerised CBT [90] and staff-administered CBT [91] to individuals with mild to moderate severity of disability.

Additional therapies referred to in the literature that may be beneficial for persons with IDD with depression include group treatment programmes, behavioural activation interventions and guided self-help interventions. These approaches have been shown to improve depressive symptoms, boost positive feelings about the self and reduce the number of negative automatic thoughts for at least 3 months [92, 93]. A life story work program has also been demonstrated to be of benefit in an older IDD population, with regard to improving quality of life and enhancing social skills [94].

Uncertainties remain about the clinical and cost-effectiveness of psychological therapies for depression in the IDD population. Further research is needed to provide clarity on these issues and also to provide guidance on how to best modify therapy to make it accessible to subjects with lower levels of baseline cognitive functioning.

Social Management

An assessment for depression should always include a detailed social history, with consideration of contributing social factors. Combining treatment with a simple and comprehensive explanation of the illness, directed towards both the patient with IDD and their carer, can aid treatment co-operation and compliance. Research has also reported specific findings relating to the importance of physical activity, self-manipulatory behaviours and the use of technology in IDD population.

Physical exercise has been recommended as a treatment strategy for depressed adults, both with and without IDD, as it has been shown to improve physical wellbeing, mental health, self-confidence and social acceptance [95, 96]. A study investigating the changes exercise induces in electroencephalographic activity reported modifications in cortical activity in frontotemporal regions [96]. A number of challenges may need to be overcome before a physical exercise programme can be implemented, these might include; a person's reliance on family or carers to plan and engage in exercise, a requirement to travel and physical restrictions [97]. To meet these challenges, novel individualised approaches to treatment need to be adopted. An example is a controlled study that explored the impact of a mini tennis programme on the psychological well-being of 12 individuals with mild to

moderate severity of disability in semi-residential care setting. It was reported that visuo-manual co-ordination, anxiety symptoms and self-esteem improved in the treatment group and this approach has been advocated as an adjunctive treatment in rehabilitation programmes [98].

It has been proposed that self-manipulatory behaviours may provide a soothing function that facilitates emotional regulation in individuals with IDD [99]. The example discussed in the research is tonguing in DS. It was identified in a study of 40 persons with DS that depressed individuals that did not tongue, were more likely to manifest internalising and externalising behaviours, than those who did engage in tonguing. The relevance of this observation in adults with different aetiological backgrounds requires further review.

Many adults with IDD, particularly younger subjects enrich their quality of life through the recreational use of technologies, such as mobile phones and tablets. A number of opportunities to utilise technology to improve clinical practice are being realised. One study investigating mobile phone use in individuals with Fragile X found that many subjects were proficient in interacting with technology and that these tools could be used to develop skills and independence [100]. As our population ages, the role that technology plays in the management of psychopathology will becoming increasingly prominent and the challenges and opportunities that this presents should be met with enthusiasm by professionals working in IDD.

Conclusion

Limited research exists to help us consolidate our understanding of how to approach depression in older people with IDD. This chapter therefore incorporates other relevant learning from the general population, the older general population and the general IDD population (Table 6.3).

In ageing individuals with IDD, depression is a broad and heterozygous diagnosis that is often missed or misdiagnosed. Making a diagnosis of depression in this population is complicated by atypical presentations on a background of multiple co-morbidities, a poor history from the patient and/or the informant and limitations in completing both physical examinations and investigations. Additionally, it is often difficult to apply diagnostic manuals, as they were created for the general population.

Where diagnostic uncertainty exists, a multidisciplinary approach should be utilised to better understand the presentation and underlying condition. Evidence based practice should be utilised that takes into consideration other comorbidities, existing medications and the individual's social situation and personal preferences. Therapeutic interventions, especially the use of psychopharmacology should be considered carefully, as there is a risk that polypharmacy and drug interactions may negatively impact upon the individual.

Table 6.3 Summary of main findings

Summary	
Epidemiology	Depression is a common illness in ageing subjects with IDD, with an estimated prevalence rate of 7–27%.
Aetiology	Depression in this population is complex and unlikely to be due to a single factor. Possible contributory influences include; genetics; degree of organic brain abnormalities; physical health ailments; the interactions, side effects and toxicity of multiple medications; psychological factors and social factors.
Presentation	Depression in subjects with mild to moderate IDD may present in a similar way to the general population (see DSM-IV criteria). Frequently IDD subjects do not meet the criteria for a DSM-IV diagnosis; this may be due to atypical presentations, deficiencies in reporting/assessment or misdiagnosis. Elderly subjects may be more prone to presenting with cognitive difficulties and somatic complaints including sleep difficulties. It remains unclear if problem behaviours are indicative of underlying depression; the relevance of these symptoms should be considered on an individualized basis using a biopsychosocial framework.
Suicide	Suicide is rare in the DID population, but the risk should not be overlooked and a risk assessment should be completed for each patient at the time of review.
Assessment and diagnosis	Individuals with IDD can pose challenges for assessment and diagnosis, not least because of differences in forms of communication and atypical presentations. Possible differentials should be considered including biological (neurodevelopmental abnormalities, physical illness and medications), psychological and social difficulties. Psychometric tools can be useful in aiding this process.
Treatment	There are no specific guidelines relating to treating depression in ageing individuals with IDD; so NICE guidelines for treating depression should be adhered to where appropriate. Treatment can involve biological, psychological and social interventions. Any medications should be used cautiously, with consideration of the pharmacokinetic and pharmacokinetic interactions that commonly occur in the elderly. Generally, medications should be started at a low dose and titrated slowly, with careful monitoring.

Much of the uncertainty associated with the treatment of ageing individuals with IDD is rooted in their unique and atypical presentation. To rise to the challenge this presents, a carefully individualised, dynamic and responsive treatment plan should be developed to obtain the best outcomes.

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Chapter 7
Behavioural Manifestations of Medical Conditions

Jenniffer T. Herrera and Stephen Sulkes

Introduction

As adults with intellectual and developmental disabilities (IDD) age, they are subject to the same array of health challenges as older individuals without IDD. Many of these conditions are known to result in emotional disorders and behavioural symptoms in persons of all ages. Some are more commonly seen in people as they age, and others are more frequently seen in conjunction with IDD. This chapter provides an overview of some medical conditions that commonly present with behavioural manifestations in older individuals with IDD, and provides a framework for cost-effective medical evaluations when such conditions are being considered.

It has been recognised for decades that physical illness can present as psychiatric disease. In 1978, a study [1] reported on a series of 658 psychiatric outpatients in which there was an incidence of medical disorders productive of psychiatric symptoms in 9% of cases. Forty-six percent of these patients had not had their medical conditions previously recognised. Presenting psychiatric complaints were of

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depression, anxiety, confusion, and speech and memory disorders. In a meta-analysis of 21 studies dealing with the rate of diagnosed and undiagnosed physical illnesses in the psychiatric patient population, it was found that about half of the patients suffered from significant physical illness, with 58% being previously undiagnosed, and 27% of these physical illnesses producing symptoms related to the psychopathology of the patient [2]. Significant associations between anxiety disorder and cardiac disorders, hypertension, gastrointestinal problems, genitourinary problems, and migraine in a series of 262 patients have been reported [3], suggesting that individuals presenting with anxiety need to be carefully evaluated for comorbidity. The bidirectional relationship between psychiatric disorders and organic cardiovascular and endocrine illness is known [4]. Physicians at times have difficulty in differentiating mental disorders from physical illness due to overlap of symptoms.

Several studies have examined the relationship between physical and mental health in older adults with IDD [5–7]. Moss and Patel [6] compared health and functional ability of 101 people with IDD over 50 years of age. Twelve were patients identified as having dementia, and they were reported as having greater chronic physical health problems and subsequent disability. Subjects were reported as having reduced capacity to enjoy things, more irritable, and more prone to aggression. Torr and Davis [7] explored the mental health of older people with IDD, finding that there are more cases of dementia and physical health problems as compared to younger people with IDD, whereas the mental health problems are similar. Finally, when comparing older people with IDD (n = 7936 aged 55 or older) to an age and sex-matched cohort in the general population, researchers found that older people with IDD seem to be more likely to have psychiatric diagnoses in inpatient or outpatient specialist care than their peers in the general population [8]. Seventeen percent of those in the IDD cohort had at least one psychiatric diagnosis recorded, compared to 10% in the general population cohort.

A large cross-sectional study [9] examined the association between health status and behaviour disorders with increasing age in a cohort of 60,752 adults with IDD clustered into four age groupings (21–44, 45–57, 60–74, and >74). Age grouping data suggested an association between morbidity and increased likelihood of behaviour symptoms in all but the oldest age grouping. The magnitude of the association and trend varied by disease across age groupings compared to those found in healthier cohorts. About 25% of these adults with IDD had psychiatric diagnoses, not changing with age level. These results suggested that health status might increase the likelihood of persistent behavioural disturbances in older individuals with IDD, and that behavioural symptoms might be sentinels for potentially treatable medical conditions.

Many conditions associated with IDD have associated behavioural features that evolve from childhood onwards, e.g., self-injury in Lesch-Nyhan syndrome, or self-hugging and sleep problems in Smith-Magenis syndrome [10]. The following discussion will focus on behavioural manifestations of some conditions presenting initially in adult life in people with IDD. This chapter will exclude a discussion on dementia as this will be fully reviewed in Chap. 12.

Types of Specific Psychiatric and Behavioural Manifestations

Depression

The most common presentation of behavioural significance in the older individual with IDD is depression. As described elsewhere in this book (Chap. 5) typical manifestations of depression include withdrawal, sleep disturbance, diminished interest, feelings of worthlessness or guilt, change in activity level, change in sexual performance, change in attention, change in appetite, psychomotor agitation or slowing, preoccupation with death, and self-injurious or suicidal behaviour. In the elderly, rather than simply depressed mood, there are more likely to be somatic complaints, hypochondriasis, and delusions due to dementia. These manifestations, evident in people without IDD, are not the only features seen in people with cognitive limitations. While individuals with mild IDD may be reflected in the standard diagnostic criteria, with increasing disability, there are more signs of aggression, screaming, and self-injurious behaviour [11]. Before assuming that such behavioural outcomes are the result of depression, it is prudent to rule out underlying medical conditions.

Symptoms of depression can be manifestations of a myriad of medical conditions [12]. Pathology in virtually any organ system can cause pain or discomfort that might result in depressive symptoms in a person with limited communication skills or IDD. Similarly, chronic functional failure of virtually any organ system can present with fatigue, sleep, and appetite changes, and affective changes that can mimic depression. An excellent history is crucial in identifying associated health problems. This becomes even more critical when the ability of the individual to provide historical information is limited due to cognitive delays and impaired communication, when there are multiple caregivers and observers, and when old records must serve as the primary source of information.

Depression has been specifically associated with lacunar infarcts and stroke [13], Parkinson's disease [14, 15], and epileptic seizures [16, 17]. Late onset depression (onset beyond 60 years) can herald an increased risk of progression, especially to vascular dementia [18]. There is an association between depression and cardiovascular diseases like hypertension, coronary heart disease [19], respiratory illness like chronic obstructive pulmonary disease [20–22], sleep apnea [23–25], renal illness like chronic kidney disease [26, 27] and overactive bladder [28–30]. Folate and vitamin B12 deficiencies have been associated with increased risk of depression, though results of meta-analyses suggest that treatment does not decrease severity of depressive symptoms over a short period of time [31, 32]. Viral infections like human immunodeficiency virus and hepatitis C virus may manifest with depression due to changes in neuronal signaling by the inflammatory pathway [33]. Malignancies like leukemia, lymphoma, and pancreatic cancer may also function this way [34]. Other illnesses associated with depression include endocrine disorders like hypothyroidism, hyperthyroidism, hypoparathyroidism, hyperparathyroidism, hypoadrenocorticism, and hyperadrenocorticism. Diabetes

and depression occur together approximately twice as frequently as would be predicted by chance alone [35].

Depression may also be substance-induced, such as with methyldopa, benzodiazepines, steroids, anti-Parkinsonian drugs, beta-blockers, hormonal therapies like estrogens/progesterones, and chemotherapeutic agents like tamoxifen, vinblastine, and vincristine [12].

Sleep Disturbance

Sleep is a life activity in which disruptions are highly noticeable to the individual experiencing them and to the individual's caregivers for those living in a supervised environment. Sleep disruption is a common expression of a variety of medical concerns, and can manifest as inadequate night-time sleep or daytime drowsiness. Mood can affect sleep quality and, conversely, disrupted sleep can mimic mood disorders. A detailed sleep history, considering an individual's sleep and wake times (whether night- or daytime) and factors relating to sleep hygiene, is essential in identifying precipitating factors for sleep disturbance. Searching for the underlying cause of a new-onset sleep change is very valuable, helping to avoid the use of sedatives or other sleep aids that may complicate the individual's medication situation as well as impact daytime function.

The International Classification of Sleep Disorders-Third Edition published in 2014 identifies seven major categories that include insomnia disorders, sleep-related breathing disorders, central disorders of hypersomnolence, circadian rhythm sleep-wake disorders, sleep-related movement disorders, parasomnias, and other sleep disorders [36]. Individuals and caregivers might describe sleep disturbances in more functional terms, such as those utilized in a 2013 assessment of sleep problems in older adults with IDD [37]. In a cohort of 551 older adults with IDD and sleep problems, 24% had a sleep onset problem, 63% had a night waking problem, 21% had shortened sleep time, and 9% had an early waking problem. In a systematic review [38], sleep problems in individuals with IDD were found to be associated with the following factors: challenging behaviour, respiratory disease, visual impairment, psychiatric conditions, and use of psychotropic, antiepileptic and/or antidepressant medication.

Many older individuals suffer from conditions that, by affecting respiration, also impact sleep quality. Congestive heart failure [39], chronic obstructive pulmonary disease [40], and asthma [41] are just a few. Other medical conditions known to affect sleep include end-stage renal disease, gastrointestinal (celiac, irritable bowel, Crohn's disease) and chronic liver disease, and diabetes, which may all provoke or exacerbate periodic limb movement disorders [42–47].

Gastroesophageal reflux disease (GERD) is also strongly associated with sleep disturbances, and predicts poorer health-related quality of life [48]. Epilepsy and antiepileptic agents influence sleep, with more severe epilepsy provoking sleep disturbance or hypersomnolence [49]. Sleep disorders are more common among those

with traumatic brain injuries (TBI) than among the general population, with up to 68% of patients on rehabilitation units having sleep disturbances [50]. In TBI patients, common sleep disturbances included insomnia, poor sleep maintenance and sleep efficiency, delayed sleep onset, early morning awakenings, and nightmares; with more chronic TBIs, disturbances ranged from obstructive sleep apnea (OSA), post-traumatic hypersomnia, narcolepsy, and periodic limb movements [51].

Obstructive sleep apnea is also common in patients without TBI, and affects sleep by temporarily stopping or decreasing breathing repeatedly during sleep, resulting in fragmented, non-restorative sleep. Its prevalence increases in those older than 60 years old, and it is associated with cardiac arrhythmias, depression, congestive heart failure, stroke, hypertension, coronary artery disease, and diabetes [52]. Some genetic syndromes, like Trisomy 21 and Prader-Willi Syndrome, have an increased risk of OSA [53, 54]. Low levels of iron have been linked to restless legs and periodic limb movement disorder [55]. Abnormal circadian rhythm (melatonin onset timing) happen in individuals with blindness [56], but also in genetic syndromes like Smith-Magenis Syndrome [10]. Pain and mistreatment are also worth considering when there are long-standing or acute sleep disturbances. Psychiatric diagnoses like anxiety, depression, post-traumatic stress disorder, and mania must also be considered in this population, as should the medications for treating these [57].

Appetite and Weight Changes

A quality dietary history is essential in evaluating anyone who is failing to maintain weight or gaining excess weight. Information regarding vitamins, minerals, or other supplements should be included in addition to what the individual's typical food intake is like. Dietary history may be a simple 24-h recall of what the patient has consumed in the last 24 h, or detailed history including access to food, amounts eaten, frequencies, and methods of preparation of foods. Need for assistance versus the ability to self-feed should also be considered. It may be helpful to observe the patient eating.

Failure to maintain weight in older adults may be the result of physiologic decline associated with ageing or psychiatric illness, but may also be related to chronic lung disease; chronic renal insufficiency; chronic inflammation as in rheumatologic disease; chronic infections like tuberculosis, bronchiectasis, endocarditis; stroke; congestive heart failure; endocrine aetiology such as hyperthyroidism or diabetes causing malabsorption and end-organ damage; gastrointestinal aetiology provoking malabsorption and malnutrition, liver failure; or malignancy. Symptoms like dental pain, dry mouth, odynophagia, dysphagia, anorexia, early satiety, nausea, vomiting, diarrhea, constipation, bloating, and abdominal pain should be explored and put into the appropriate clinical context. A complete physical examination should include orthostatic blood pressure measurements; oral cavity examination to assess for dental abscess and caries, poorly fitting dentures, thrush; palpation for thyroid nodules;

and rectal exam for fecal impaction and occult blood in stool. With failure to thrive in older adults, the psychosocial context should also be considered, gathering history regarding social network and family support, and the possibility of mistreatment. Increased appetite and excess weight in adults are often associated with conditions like diabetes or hypothyroidism.

Maladaptive Behaviours: Self-Injury, Aggression and Agitation

Self-injury can be a behavioural manifestation of pain or discomfort anywhere, or it may reflect specific discomfort in the body area toward which the self-injury is directed (e.g. head banging in response to headache or dental pain). This type of behaviour can be seen in older people without IDD experiencing chronic health conditions like tinnitus, malignancy, diabetes, and chronic pain [58]. In a cohort of 25 patients with self-injurious behaviour (SIB), 28% had previously undiagnosed medical conditions that could be expected to cause pain or discomfort, and the majority of those experienced decreased SIB with treatment of their medical conditions [59]. Other researchers [60] found in a cohort of 198 patients with IDD admitted to a specialised inpatient psychiatric unit, most had been referred for aggressive, disruptive, and self-injurious behaviours. The most frequent medical comorbidity amongst these patients was constipation (60%); GERD was identified in 38%.

This being said, individuals with IDD disorders may also process pain differently. Where a typical individual's nociceptors and amplification of their central nervous system pain pathways work to de-incentivize harmful behaviour, individuals with IDD may initiate SIB to address irritating or painful sensation, and require exaggerated stimuli to engage the negative feedback loop [61]. Self-injury is also known to be associated with some genetic syndromes like Lesch-Nyhan Syndrome, Smith-Magenis syndrome, tuberous sclerosis complex, and Cornelia de Lange Syndrome [62].

Aggression is often a non-specific behavioural symptom, associated with agitation. Pain, again, is a major patient risk factor. Acute medical conditions, unmet needs, sensory changes, poor sleep hygiene, and male sex independently predict aggression [63]. Agitation can be secondary to delirium or the sequelae of TBI. Increasing intracranial pressure, due to malfunctioning ventriculo-peritoneal shunts and brain tumors, can also cause agitation. Metabolic disturbances such as hyperthyroidism, parathyroid abnormalities and hypoglycemia can cause agitation. Agitation can also be a side effect of many medications, or the manifestation of medication side effects like akathisia. Agitation can also be the result of ingestion/intoxication and pica. History surrounding aggression and agitation should qualify frequency of episodes, risk for

actual injury to self/others, and triggers. Physical examination should include thorough neurologic examination. As with the other behavioural manifestations described, aggression as a result of abuse and neglect must also be considered for these vulnerable patients.

Pain

Acute and chronic pain may precipitate behaviour change in people with IDD and functional communication limitation. Individuals with communication barriers manifest pain in different ways from other people. How to accurately identify pain in persons with IDD and associated communication difficulties remains problematic. No generally accepted assessment tool is yet available but checklists of nonverbal pain indicators, detailed carer interviews, direct observation are areas of ongoing research. Pain may be produced by psychological states and may also be a cause for psychological disturbance; treatment approaches need to be multipronged. An empiric trial of a non-sedating analgesic can help to differentiate behaviour due to pain when it is suspected.

Known Behavioural Associations with Specific Medical Conditions

Nutritional Factors

Nutritional issues are often anecdotally associated with behaviour problems. Since nutrition in older individuals occurs more often with identifiable disease and disruption of well-being, evaluation of nutritional factors in this population often identifies treatable conditions that might affect behavioural outcomes. As in pain syndromes, nutritional disorders can be both cause and effect of behavioural conditions. As can easily be imagined, individuals suffering from prolonged protein-calorie malnutrition will appear depressed, with decreased activity, increased sleep, withdrawal, irritability, inattention, and decreased self-directedness [64]. More subtle dietary deficiencies can also have behavioural impact. For example, scurvy (vitamin C deficiency) presents with asthenia, and mood disturbances [65, 66]. In contrast, vitamin A toxicity causes increased intracranial pressure and peripheral neuropathy and pain [67].

Skin

A number of skin diseases are associated with anxiety, depression, and sleep disorders. For example, one autoimmune skin condition associated with anxiety is alopecia areata [68]. Conditions causing itching like atopic dermatitis and chronic urticaria can be associated with behavioural symptoms beyond habitually self-injurious scratching [69]. Vitamin C deficiency (scurvy, as noted above), can present with an apparent folliculitis due to ingrown corkscrew hairs.

Eyes and Ears

Ophthalmologic conditions affecting behaviour can be grouped into those affecting functional vision and those associated with optic or periorbital discomfort. Rubbing of the eyes may be in response to discomfort (pain, itching, photophobia). Glaucoma can cause both eye pain and disrupted vision, and may result in both eye-directed and less specific behavioural manifestations [70]. Functional vision testing and more detailed eye examination may require the skills of an ophthalmologist. Similarly, otologic conditions can be grouped into those affecting hearing (either decreased hearing or undesired sound perception, like tinnitus), those associated with otologic discomfort, and those associated with acute or chronic vestibular disorders. Simple ear examination may reveal impacted cerumen that both affects hearing and causes pain, and functionally based audiologic assessment may reveal hearing loss that can affect behaviour [71].

Mouth and Throat

Weight loss and apparent depression may be the result of swallowing disorders or oral-pharyngeal discomfort. Dental examination for decay, infection, gum disease, and plaque build-up may identify sources of discomfort that, when treated, with result in decreased agitation and improved eating and sleep. Watching the individual eat can be extremely informative, demonstrating discomfort localizing to one side or the other, and showing problems with swallowing that could be painful or even frightening to the individual [72].

Cardiac and Respiratory Disorders

Cardiac and lung disorders, as mentioned above, have their effects through fatigue and respiratory distress. Cardiac or pulmonary failure can present both with symptoms of depression and sleep disturbance [73]. Pulmonary rehabilitation has been

reported to improve fear and depression and improve self-esteem in individuals without IDD [74]. Cardiac surgery can improve function, but cardiopulmonary bypass has been reported to have emotional and behavioural sequelae [75].

Gastrointestinal Disorders

Some sources of abdominal pain and nutritional disorders have been discussed above. In individuals with IDD, particularly those with adapted or limited diets or decreased physical mobility, problems worthy of consideration are gastroesophageal diseases (reflux and eosinophilic esophagitis), constipation (and other causes of intestinal obstruction). Abdominal pain can also occur due to celiac disease or other food sensitivities.

Gastroesophageal reflux disease (GERD) is well-established to have a negative impact on quality of life in people without disability, where associated symptoms reduce work productivity and activities of daily living. Quality of life measures improve after fundoplication. Although the behavioural symptoms attributable to GERD in individuals with IDD vary [76], when rumination, dental erosion, or hematemesis is present, GERD should be suspected. Rumination is a self-injurious form of GERD in which stomach contents are voluntarily regurgitated into the esophagus or mouth. Whether this is viewed as a self-simulating behaviour causing a medical problem or the converse, a combination of anti-reflux treatments, behavioural intervention, and psychotropic medication is often required to bring it under control [77].

Many people with IDD suffer from constipation, leading to a wide range of behavioural manifestations. Dietary differences, particularly low fluid and fiber intake, lack of mobility, and medications taken for other conditions (e.g., iron, psychotropic) slow gut transit time and combine to make this a particularly compelling problem [78]. Some get into a vicious cycle of stool withholding, which allows stools to pile up in the rectum, become dryer and harder, and then become harder and more painful to pass. Sometimes only the liquid stool from above the fecal impaction can pass, erroneously presenting as apparent diarrhea. Elderly individuals with and without IDD are notably prone to constipation, for the above reasons and also associated with obsessive-compulsive disorder, depression, and anxiety.

An often-overlooked source of abdominal pain in persons with IDD is the pancreas. Pancreatitis is an infrequent but well-known complication associated with the use of the anticonvulsant valproate [79], as well as with abdominal blunt trauma. When abdominal pain or vomiting is noted in a person with IDD, particularly if taking this medication, assessment for pancreatitis should be considered.

Celiac disease is an increasingly recognised cause of abdominal pain. This sensitivity to gluten, a protein found in wheat, barley, and rye, is associated with abdominal pain, malabsorption and associated nutritional deficiencies, bowel changes, and skin and behavioural changes [80]. It occurs in up to 10% of people with Down syndrome, and is also more common in people with Williams' syndrome.

Gynaecological

Cyclic behavioural changes in women may reflect hormonal cycling with menstruation or contraceptive use, especially in women with other neurological disorders [81].

Thyroid Disease

Thyroid disease is well known to be associated with Down syndrome, both with hypothyroidism and, less frequently, hyperthyroidism. The former can present as depression, cognitive slowing, and mood disturbance, while the latter can be associated with agitation and overactivity [82]. Notably, a presenting feature of hypothyroidism can be carpal tunnel syndrome, manifesting as hand flapping behaviour. An association has also been made between hypothyroidism and seasonal affective disorder.

Implications for Practicing Providers

Implicit in the above discussion is the essential role for history and physical examination. Primary care providers need to ask about behaviour concerns: as issues to address in themselves, as possible manifestations of known health conditions or as side effects of medications, and as clues to unrecognised medical conditions. It equally behooves behavioural health providers to get complete histories of previously identified health conditions and current medications to consider their possible impact on current behavioural symptoms. At minimum, providers should be aware of the Canadian Consensus Guidelines [83] with regard to behavioural health. As stated in those guidelines (extracted in Table 7.1),

> Before considering a psychiatric diagnosis, assess and address sequentially possible causes of problem behaviour, including physical (e.g., infection, constipation, pain), environmental (e.g. changed residence, reduced supports), and emotional factors (e.g., stress, trauma, grief) … [and] … Consult available information on behavioural phenotypes in adults with IDD due to specific syndromes.

History and physical examination, combined with judicious use of laboratory studies based on initial findings, will often uncover unrecognised associated medical problems. In addition to disease states, medication side effects can often include behavioural symptoms, and medication changes should be considered as initial interventions.

Table 7.1 Preventive care checklist for adults with developmental disabilities (DD): behavioural considerations and recommendations (adapted from Sullivan et al. [83])

Considerations	Recommendations
Problem behaviour, such as aggression and self-injury, is not a psychiatric disorder but might be a symptom of a health-related disorder or other circumstance (e.g., insufficient supports).	Before considering a psychiatric diagnosis, assess and address sequentially possible causes of problem behaviour, including physical (e.g., infection, constipation, pain, medication side effects), environmental (e.g. changed residence, reduced supports), and emotional factors (e.g., stress, trauma, grief).
Problem behaviours sometimes occur because environments do not meet the developmental needs of the individual.	Facilitate "enabling environments" to meet these unique developmental needs as they will likely diminish or eliminate problem behaviours.
Despite the absence of an evidence base, psychotropic medications are regularly used to manage problem behaviours among adults with DD. Antipsychotic drugs should no longer be regarded as an acceptable routine treatment of problem behaviours in adults with DD	Regularly audit the use of prescribed psychotropic medication, including those used as needed. Plan for a functional analysis and interdisciplinary understanding of problem behaviours, and consider reducing and stopping, at least on a trial basis, medications not prescribed for a specific psychiatric diagnosis.
Psychiatric disorders and emotional disturbances are substantially more common among adults with DD, but their manifestations might mistakenly be regarded as typical for people with DD (i.e., "diagnostic overshadowing"). Consequently, coexisting mental health disturbances might not be recognised or addressed appropriately.	When screening for psychiatric disorder or emotional disturbance, use tools developed for adults with DD according to their functioning level (e.g., Aberrant Behavior Checklist-Community (ABC-C) [84]; Psychiatric Assessment Schedule for Adults with DD (PAS-ADD)).
Increased risk of particular developmental, neurologic, or behavioural manifestations and emotional disturbances (i.e., "behavioural phenotypes") is associated with some DD syndromes.	Consult available information on behavioural phenotypes in adults with DD due to specific syndromes.
Establishing a diagnosis of a psychiatric disorder in adults with DD is often complex and difficult, as these disorders might be masked by atypical symptoms and signs. In general, mood, anxiety, and adjustment disorders are under-diagnosed and psychotic disorders are over-diagnosed in adults with DD.	When psychiatric disorder is suspected, seek interdisciplinary consultation from clinicians knowledgeable and experienced in DD.
Psychotic disorders are very difficult to diagnose when delusions and hallucinations cannot be expressed verbally. Developmentally appropriate fantasies and imaginary friends might be mistaken for delusional ideation, and self-conversation for hallucination.	Seek interdisciplinary input from specialists in psychiatry, psychology, and speech-language pathology with expertise in DD to help clarify diagnoses in patients with limited or unusual use of language.

(continued)

Table 7.1 (continued)

Considerations	Recommendations
Input and assistance from adults with DD and their caregivers are vital for a shared understanding of the basis of problem behaviours, emotional disturbances, and psychiatric disorders, and for efficiently developing and implementing treatment and intervention.	Establish a shared way of working with patients and caregivers. Seek input, agreement, and assistance in identifying target symptoms and behaviours that can be monitored. Use tools (e.g. sleep charts and antecedent-behaviour-consequence charts) to aid in assessing and monitoring behaviour and intervention outcomes.
Interventions other than medication are usually effective for preventing or alleviating problem behaviours.	To reduce stress and anxiety that can underlie some problem behaviours, emotional disturbances, and psychiatric disorders, consider such interventions as addressing sensory issues (e.g., under-arousal, over-arousal, hypersensitivity), environmental modification, education, and skill development, communication aids, psychological and behaviour therapies, and caregiver support. Cognitive-Behavioural Therapy can be effective in decreasing anger and treating anxiety and depression in adults with DD. Psychotherapy can sometimes be effective for emotional problems (e.g., related to grief, abuse, trauma) that might underlie aggression, anxiety, and other such states.
Psychotropic medications (e.g., antidepressants) are effective for robust diagnoses of psychiatric disorders in adults with DD as in the general population.	When psychiatric diagnosis is confirmed after comprehensive assessment, consider psychotropic medication along with other appropriate interventions
Psychotropic medications can, however, be problematic for adults with DD and should therefore be used judiciously. Patients might be taking multiple medications and can thus be at increased risk of adverse medication interactions. Some adults with DD might have atypical responses or side effects at low doses. Some cannot describe harmful or distressing effects of the medications they are taking.	"Start low, go slow" in initiating, increasing, or decreasing doses of medications. Monitor safety, side effects, and effectiveness through frequent communication with patients and caregivers. Review medication at least every 3 months, including diagnosis and appropriateness of treatment.
When unable to pinpoint a specific diagnosis, behaviours of concern might serve as index behaviours against which to conduct a trial of medication.	Having excluded physical, emotional, and environmental contributors to the behaviours of concern, a trial of medication appropriate to the patient's symptoms might be considered.
Antipsychotic medications are often inappropriately prescribed for adults with behaviour problems and DD. In the absence of a robust diagnosis of psychotic illness, antipsychotic medications should not be regarded as routine treatments of problem behaviours in adults with DD.	Do not use antipsychotic medication as a first-line treatment of problem behaviours without a confirmed robust diagnosis of schizophrenia or other psychotic disorder.

Table 7.1 (continued)

Considerations	Recommendations
Antipsychotic medications increase risk of metabolic syndrome and can cause other serious side effects (e.g., akathisia, cardiac conduction problems, swallowing difficulties, bowel dysfunction.)	Carefully monitor for side effects of antipsychotic medication, including metabolic syndrome. Educate patients and caregivers to incorporate a healthy diet and regular exercise into their lifestyle. Reassess the need for ongoing antipsychotic medications at regular intervals and consider dose reduction or discontinuation when appropriate.
Behavioural crises can arise that might need management in an emergency department.	When psychotropic medications are used to ensure safety during a behavioural crisis, ideally such use should be temporary (no longer than 72 h). Debrief with care providers to minimize the likelihood of recurrence, including review of crisis events and responses (e.g., medication, de-escalation measures) and identification of possible triggers and underlying causes. If the patient is at risk for recurrent behavioural crises, involve key stakeholders, including local emergency department staff, to develop a proactive, integrated emergency response plan.

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Chapter 8
Emotional and Behavioural Disturbances in Adults with Down Syndrome

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Introduction

The prevalence of mental health conditions at all ages in people with an intellectual and developmental disabilities (IDD) is much higher than the general population [1–6]. A review of epidemiological data that looked at the true co-occurrence (co-morbidity) and course of mental disorders in populations of children and adolescents with IDD revealed a three to fourfold increase in the prevalence of co-occurring mental disorders [7]. A recent study has shown the prevalence rates for co-occurring psychiatric symptoms or disorders ranges from 13.9% to 75.2% with much of the variation attributed to differences in the diagnostic criteria utilised and the specific samples examined in the studies [3].

People with IDD are at a higher risk of developing a mental disorder than the general population, partly due to their difficulties in communication, processing skills, cognitive functioning and social skills [8]. However, psychiatric disorders are often underdiagnosed in people with IDD especially for those with severe or profound IDD due to their difficulties in communication and diagnostic overshadowing. A recent cross-sectional study found a previously undiagnosed mental disorder in 29.6% of the sample with depressive and anxiety disorders being most prevalent [9]. Moreover, the available assessment tools for clinicians to use when screening for psychiatric disorders in people with mild or moderate IDD [10–12] and for people

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with severe or profound IDD are limited at best. More research is needed to develop instruments with better sensitivity and specificity for the IDD population [13, 14].

Down syndrome (DS) is the most common genetic and identifiable cause of IDD, accounting for around 15–20% of the intellectually disabled population [15]. A review of the cognitive and behavioural functioning of individuals with DS reveals a pattern of development that is characterized by unique strengths and weaknesses that emerge in stages throughout the progression of the lifespan [16]. Both children and adults with DS are commonly described as charming, affectionate, cheerful, and sociable with other observed personality assets that include kindness, humour and forgiveness [17]. Although DS children are prone to psychopathology, they are at a lower risk compared to other children with IDD [16]. The lower rates of behaviour problems in DS children may be attributed to their unique social development and an overuse of social behaviours [18]. The changes in emotional and behavioural functioning in adulthood were found to be associated with neurodegeneration and older DS adults are at higher risk for dementia in Alzheimer disease (DAD) [16].

Individuals with DS display a different pattern of behavioural and psychiatric disorders when compared to the general IDD population. The syndrome is unique with respect to its cognitive, behavioural, and psychiatric profiles [16, 18]. Some have proposed that people with DS exhibit a different behavioural phenotype compared to those with unspecified IDD [19–21] and to those with other syndromes [22, 23]. A unique cognitive phenotype for DS that is different from other individuals even with matched level of cognitive functioning has also recently been proposed [16].

A higher prevalence of specific psychiatric disorders occurs in people with DS compared to the general population and to people with other types of IDD [15, 24]. Certain mental disorders such as depression, anxiety disorders, dementia, and psychosis NOS are more common in people with DS compared to other types of IDD [24, 25]. Attention deficit hyperactivity disorder (ADHD), disruptive behaviours and obsessive-compulsive behaviours are common in the childhood and adolescent years in comparison to depression and anxiety disorder which are more common in DS adults [26]. The higher prevalence of attention deficit hyperactivity disorder in DS may also contribute to the occurrence of behaviour issues in childhood [27, 28].

People with DS experience different age-specific physical and mental health problems in comparison to people with IDD without DS [29]. The extent of multimorbidity in adults with DS, however, appears to be similar to that of adults without DS, but the prevalence of specific problems differed with such conditions as obesity, visual impairments, hearing impairments, and constipation being the most common [30]. Congenital heart disease, which is common in children with DS, remains a leading cause of death among DS children and adolescents [31, 32]. Hypothyroidism, osteoporosis, gastroesophageal reflux disease, pulmonary conditions, obstructive sleep apnoea, and heart septal defects are common in adults with DS whereas dementia, seizure disorder, and skin conditions are more common in older adults compared to younger adults with DS [33].

Survival of infants born with DS has considerably improved [34, 35], and overall life expectancy has increased [36]; yet people with DS still die earlier than people

with IDD without DS [35, 37]. Respiratory diseases are now the most common primary causes of death in adults with DS [37, 38] and reduced mobility, poor vision, and epilepsy are also associated with reduced survival in later years [32, 39]. Studies have consistently revealed that along with the increased life expectancy comes the increased risk and prevalence of Alzheimer disease [40–43]. A recent study has shown a mortality rate of around 70% in DS adults with dementia [44] and cognitive impairments in DS occur earlier than in the general population [45, 46]. For adults with DS and DAD, memory and executive functions are significantly affected [47, 48] whereas personality and behaviour changes are more common in people with DS who don't meet the DAD criteria [47]. It has been speculated that the functions of the frontal lobes may be compromised early in the course of the disease, which is evident by prominent personality and behaviour changes associated with executive dysfunction in an early presentation of Alzheimer disease in adults with DS suggesting these might represent preclinical signs of DAD [47, 49].

Adults with DS are known to age prematurely and present with chronic conditions commonly seen in the geriatric population with a high prevalence of comorbidities, age-related conditions, cognitive and functional deficits, and social problems which makes assessment and treatment very complex [50]. However it should be noted that a functional decline in an adult with DS should not automatically be assumed to be due to dementia, which is not inevitable in all adults with DS. A functional decline may be the result of a range of disorders, especially sensory and musculoskeletal impairments [32].

The literature has mostly focused on the areas of depression and dementia when addressing mental health issues in older adults with DS. There has been less attention to studying challenging behaviours and the presentation of psychiatric signs and symptoms associated with a reported change or decline in behaviour. This chapter highlights some of the challenging behaviours, the presenting psychiatric signs and symptoms, and the types of psychiatric disorders that can be found in people with DS. Eight case examples of adults with DS who were referred to an outpatient clinic for evaluation are presented to highlight some of the important behavioural and psychiatric issues that can present challenges to clinicians in arriving at a proper psychiatric diagnosis and a course of treatment.

Challenging and Maladaptive Behaviours

There is a lower incidence of challenging and maladaptive behaviours in the DS population compared to other individuals with IDD [17, 20, 51]. Aggression is one of the most common challenging behaviours displayed by people with IDD and can be exhibited in many ways. Recent studies have revealed that individuals with DS exhibit a lower incidence of aggressive and disruptive behaviours when compared to people with other genetic syndromes [16, 23, 52]. The distinctive DS behavioural phenotype can shape how DS children, adolescents and adults react to people and events around them. Children with DS can be susceptible to disruptive behaviour,

aggressiveness, attention problems, and conduct-oppositional disorders; however, the risk of psychopathology is lower than it is for others with IDD even though they do show more problems than typically developing children [53]. As the DS child matures, it appears that externalizing behaviours decline and internalizing symptoms (i.e. social withdrawal, anxiety and secretive behaviour) may develop during adolescence into adulthood [18, 52]. Additionally, both the frequency and type of challenging behaviour(s) when displayed can vary but appears to decline into adulthood [53, 54].

In a study comparing four genetic syndromes, temper tantrums and physical aggression were found to diminish in childhood and adolescence for those with Williams syndrome, DS and fragile X syndrome and in adulthood for those with Prader-Willi syndrome. Individuals with DS were also found to have the lowest rates of temper tantrums, however, verbal aggression showed no evidence of diminishing with age across any of the four syndromes [52]. For other maladaptive behaviours, some have described people with DS as "being excessively uncooperative" [55] or having a temperament characterized by obsessional slowness or stubbornness [56]. One report highlighted the distinction between the observed low rates of overt (physical) aggression and higher rates of low-level aggressive behaviours (i.e. non-compliance) in children and adolescents with DS [53]. Compulsive-like behaviours (i.e. ritualistic habits) were reported to be more evident in DS children than in other children with IDD and were thought to be a component of the DS behavioural phenotype [57, 58].

In a review of an outpatient clinic sample of 206 DS adults ages 20–71 who were referred for evaluation, the primary challenging behaviours of physical aggression, disruptive behaviours, inappropriate sexual behaviours and self-injury were reported to be lower in DS adults above age 40 [54]. Noteworthy, non-compliance behaviours (e.g. refusing to follow directions) were the foremost challenging behaviour in more than half the study sample regardless of age and suggest that this behaviour was also likely to be part of the DS behavioural phenotype.

Quite the opposite to the often described friendly and social DS stereotype, a small percentage of the DS population have co-morbid autism spectrum disorder (ASD), which can present a distinctive pattern of challenging behaviours not usually seen in DS individuals [16, 18, 59, 60]. Children and young adults with the dual diagnosis of DS and ASD show a greater delay in developing language and adaptive behaviour skills and display a distinctive set of aberrant behaviours marked by characteristic odd/bizarre stereotypic behaviour, anxiety, and social withdrawal [59, 61]. Additionally, when the co-morbid diagnosis of ASD is present the majority are more often severely cognitively impaired with severe to profound IQ levels and show greater delays in language and adaptive behaviour, whereas DS individuals without ASD more often have moderate to severe IQ levels [16, 18, 60]. In a recent study of individuals diagnosed with DS and ASD, and those with only ASD the findings revealed more stereotyped behaviour, repetitive language, overactivity and

self-injury were exhibited than by those with only DS. Additionally, those with both DS and ASD and those with only DS were described as less withdrawn from their surroundings than those with only ASD indicating a different behavioural and cognitive profile for individuals with both conditions [62].

It has been well established that older adults with DS are at a higher risk for developing dementia (predominately Alzheimer type) with the average age of diagnosis in the mid-50s [32]. A diagnosis of dementia in people with DS was a predictive indicator for increased maladaptive behaviour [63] and can include mood changes and depression, daytime wandering, restlessness, resistiveness, aggressive behaviours, sleeps disturbances, incontinence, amotivation, slowness and social disengagement [39, 55, 64, 65]. These behavioural problems are also a common feature of individuals suffering from dementia in the general population, especially those in the latter stages of the illness. The difficulties in dealing with such challenging behaviours in the general population rather than cognitive decline were found to be the main reasons why carers seek placement in nursing homes [66]. This is less of an issue for people with IDD, as many of them as older adults are in supervised group home or residential settings or receive supportive home services where the level of care and oversight is greater than any care provided by only one or two carers in a person's private home.

To summarize, the prevalence rates for severe challenging or maladaptive behaviours in people with DS are low and do not constitute part of the DS behavioural phenotype [17]. The most commonly described problematic issues in the DS population include non-compliance, stubbornness and compulsive-like behaviours with only low-levels of non-physical aggression. However, the incidence of behaviour problems does change and increase in frequency and occurrence for some DS individuals when there is a comorbid diagnosis of ASD [62, 67] or dementia [68]. A substantial proportion of people with DS with questionable dementia present with increased anxiety, sleep disturbances, apathy and depressive symptoms, suggesting that these changes may be early warning signals of dementia [68].

Typically, young adults with DS prior to age 40 display a positive profile of functional skills with few problematic behaviours than their non-DS counterparts [69]. This may explain why a higher proportion of people with DS live outside of institutions and function well in the community without the presence of few or any challenging behaviours. After age 40, however, DS adults were found to have a higher risk for health, functional decline and cognitive problems as they age [69, 70]. It is therefore important when studying the incidence of challenging or maladaptive behaviours to take into account the ages of the cohort being studied since both the frequency and types of challenging behaviours can vary and differ across the lifespan. It is also essential for clinicians to be aware that the typical behavioural phenotype seen in the DS population can be different when a co-morbid diagnosis such as ASD is made since different treatment approaches may be necessary to address the presenting problems.

Common and Uncommon Psychiatric Signs and Symptoms

A presentation of depression in people with IDD has marked diversity and may present with the typical symptoms such as low mood, irritability, disturbed sleep, lack of interest, social and withdrawal [71, 72], as well as atypical symptoms such as aggression and self-harming behaviour especially in people with profound and severe IDD [73]. For adults with DS, depression tends to present commonly with symptoms like low mood, crying, withdrawal, disinterest, mutism, psychomotor retardation, and decreased appetite [17, 74, 75]. A decline in adaptive functioning with anxiety and agitation are significantly associated with depression in adults with DS whereas cognitive symptoms such as loss of concentration, disturbance of memory and suicidal ideation are less common [75].

Previous reports indicated that anxiety disorders are common in the IDD population ranging from 14% [76] to 31% [77] and commonly present with symptoms such as increased polydipsia, hyperventilation, irritability, anger, sweating, self-injury, and avoidant behaviour [78]. Early reports found anxiety disorders to be less common in people with DS [79, 80]; however for those presenting with symptoms of anxiety, older DS adults were more likely to present with an anxiety disorder than younger DS individuals [24].

Psychotic symptoms of hallucinations and delusions were generally found to be low in DS individuals and remain an infrequent phenomenon in this population [2, 54, 81, 82]. Nevertheless, some studies have reported auditory hallucinations in some individuals with DS and depression [83] or delusions and hallucinations in older DS adults with possible or definite dementia [84]. An earlier report showed that patients with DS and dementia had a higher prevalence of mood disturbance, restlessness, disrupted sleep, uncooperativeness and auditory hallucinations [55].

Assessing psychiatric signs and symptoms in people with DS present challenges to clinicians, especially for individuals with severe disabilities due to practical difficulties of eliciting abnormalities in thought process and thought content [85]. A common behavioural feature observed in many people with DS was talking to self, which may represent another aspect of the DS behavioural phenotype. The presence of self-talk was reported to be a common occurrence in many people with DS and was seen as an adaptive behaviour rather than a sign of psychopathology [86–88]. Self-talk behaviour is typically directed to oneself but can also involve a real or imaginary person or object and mostly occurs in the form of a monologue or a commentary. It can serve as a method of self-regulation, decision-making, and working through emotional problems [86]. If misinterpreted as psychotic or hallucinatory behaviour (e.g. responding to voices) self-talk may result in a diagnosis of psychosis being made [86, 88, 89]. In assessing self-talk, it is important for clinicians to observe an individual with DS for any change in the frequency and quality of their self-talk (e.g. an increase or decrease in the volume of speech, expressions of fear or anger) as such a change in one's self-talk rather than merely the presence or self-talk may indicate psychopathology [25, 54].

Compulsive-like behaviours may also be a part of the behavioural repertoire of individuals with DS [57]. Routinized and compulsive-like behaviour (RCB) was significantly higher in younger people with DS when compared to typically developing children and adults with behaviour problems [90]. Behaviour rituals as a psychiatric sign/symptom can manifest as compulsive hoarding of objects or food, clothing obsessions, and bathroom rituals which may be driven by anxiety or stress as seen in obsessive compulsive disorder [54]; it may also present as obsessional slowness an extreme form of obsessive-compulsive behaviour [56]. Recent studies reported that restrictive behaviour and restrictive interests (RBRI) can serve the purpose of reducing anxiety or fear in some DS individuals [91–93]. For older DS adults with functional decline, the hoarding of objects is a common behaviour in many people with Alzheimer disease [66] and may be an adaptive way to minimise forgetting where personal effects are in the environment.

It has been reported that some DS adolescents and adults experience unexplained deterioration characterized by regression with impairing symptoms across motor, speech, behaviour, mood, and daily living skills consistent with catatonia [94, 95]. A recent study suggested that the presence of catatonia, a neuropsychiatric disorder, may be relatively common and represent an unrecognised cause of a reversible decline in adolescents with DS [96]. Motor disturbances (e.g. freezing, grimacing, abnormal movements, etc.) was a prominent and impairing feature in four reported DS cases and could not be explained on the basis of a primary affective illness. The diagnosis of catatonia was supported by the fact that each individual responded favourably when established anti-catatonic treatment along with benzodiazepine and Electroconvulsive treatment was instituted [96]. The DSM-5 now includes the diagnostic category of "unspecified catatonia" when no discernible underlying neuropsychiatric or medical diagnosis is evident.

There have been few studies reporting the presence of tics/involuntary movements (exclusive of tremors or myoclonus) in people with DS. In a sample of 206 adults with DS, 12% were observed to exhibit vocal or motor tics as part of their behavioural repertoire [54]. Two earlier reports described a smaller number of DS cases diagnosed with Tourette (syndrome) disorder [97, 98]. It has been suggested that the use of neuroleptic medications can precipitate observed tics or abnormal movements and may cause tardive or atypical Tourette disorder [98]. Medication-induced tics can also resolve on cessation of medication [99]; in other instances the use of neuroleptics can also block tics or influence their frequency of occurrence [54]. The presence of tic-like behaviours may be under-reported or go unrecognised in the DS population and requires further investigation to determine if they are a true psychiatric sign indicative of Tourette disorder, chronic tic disorder or a medication side effect.

As a psychiatric sign self-injurious behaviour is a low occurrence in the DS population however it was found to be associated with significantly higher levels of autistic behaviour in some individuals with DS [100]. Suicidal ideations and attempts are infrequent among people with ID but completed suicides and potentially fatal attempts have been reported. In the DS population, suicide or self-talk

about harming oneself is extremely rare. Only one study was found that reported two cases of suicide attempts by people with DS, both with a major depressive episode and a potentially lethal suicide attempt [101]. In an extensive population survey on suicide attempts, individuals with DS had significantly fewer incidences of suicidal behaviour compared to the IDD control group without DS [102].

Psychiatric Diagnoses

People with DS have a lower prevalence of psychiatric disorders than people without DS, even when their IDD level was taken into account [103]. The reported presence of mental health disorders significantly increases with age in people with DS [24]. A considerable proportion of older adults with DS present with mental health and behavioural problems, including attention deficit hyperactivity disorder (ADHD), depression and DAD [58]. The early onset and high prevalence of Alzheimer disease in DS adults has been relatively well studied and overshadows other age-related disorders that result in significant morbidity, functional decline and early mortality [32].

The prevalence of depression is higher compared to other psychiatric disorders in adults with DS [2, 17, 104, 105]. In looking at the behavioural phenotype, depression along with dementia and hypothyroidism can distinguish the adult DS population from age-matched or developmentally matched control groups [19]. Both depression and dementia can present as a dual diagnosis, however it is often difficult to separate the two since depression is also common in the early stages of dementia. Depressive disorders are also mistaken for dementia when a DS adult presents with functional and/or cognitive decline, however the symptoms of depression can be reversed with appropriate treatment with antidepressant medications [32, 106]. The use of antidepressants was also found to be associated with delayed dementia-onset and may increase longevity in DS adults [107]. Another factor to take into account is that there is a high incidence of subclinical and transient hypothyroidism [36, 108] in people with DS, which can produce signs and symptoms of depression and coexist as a second illness in depressed patients. However, a successful reversal of depression can occur following treatment and normalisation of the thyroid dysfunction [109].

The prevalence of schizophrenia is higher in people with IDD compared to the general population. However, in comparison to other people with IDD, people with DS were not reported to be particularly susceptible to schizophrenia or psychosis [2, 6, 24, 54]. One recent study, however, suggested that the prevalence rate of psychosis, not otherwise specified (NOS) or depression with psychotic features might be higher than previously reported [25]. Since hallucinations and delusions were found to be an infrequent phenomenon in DS but talking to self (self-talk behaviour) has been reported to be a common occurrence in the DS population, it is important to evaluate the individual to determine if their self-talk behaviour may be indicative of hallucinatory or psychotic behaviour [86].

The incidence of anxiety-type disorders can occur across the lifespan in the DS population [54] but does not appear to differ from the IDD population [25]. Past reports indicated the prevalence of obsessive compulsive disorder (OCD) within the DS population to be low with estimates from 0.8% [80] to 4.5% [81] with ordering and tidiness being the most commonly presenting behaviours [110]. In another report 18% of a DS study group presented with anxiety and approximately 50% of these had repetitive, compulsive-like behaviours, although none met the criteria for OCD [90]. As noted earlier, compulsive-like behaviours appear to be a part of the behavioural repertoire of individuals with DS although a formal diagnosis of OCD is often not made. Currently there are no current large-scale studies in the literature for any firm conclusions about the incidence of OCD in the DS population.

The incidence of impulse control disorders in the DS population was also found to be lower than in the IDD population; however, when impulse control problems are present they were reported to be more common in males, and in adolescents versus young DS adults [25]. A low incidence of stereotypic movement disorders and tic disorders (i.e. Tourette disorder, chronic tic disorder) were reported for people with DS [54] and two cases examples are presented in the next section for review.

Bipolar disorder is less common in people with DS compared to IDD or general population [25], however few studies have investigated the incidence of bipolar disorder or mania in the DS population [111, 112]. One past report suggested a genetic explanation, the presence of an extra copy of chromosome 21, which confers protection against bipolar disorder [113]. In a clinic sample of 206 DS adults ages 20–71, less than 6% received a diagnosis of bipolar disorder, depressed phase [54]. There is some evidence of mania episodes being more common in DS males following a rapid cycling pattern [111] and one report described a case that developed a manic episode in later life [112]. Further studies are necessary to better understand this phenomenon and how it may be expressed in people with DS.

The current prevalence rates of ASD among children with DS were estimated to be between 6 and 10% [59, 62, 114]. There is also a higher prevalence for both ADHD [27, 28] and ASD [36, 115] to occur in DS children and adolescents. As noted above, with a diagnosis of ASD, the types of challenging and/or maladaptive behaviours displayed will often be different and more problematic than in those DS individuals without ASD. A recent study on the prevalence of ADHD and obstructive sleep apnoea syndrome (OSAS) in DS has also shown not only the high prevalence of 34% for ADHD, but also 42% for ASD, suggesting that screening should be introduced in the early school years [27].

Illustrative Case Examples

Six cases of adults with DS who were evaluated and followed by the first two authors in a community diagnostic and treatment clinic are presented below and were part of the original chapter published in 2003. Two additional cases have been added for this revised edition to highlight other overshadowing psychiatric conditions.

Decline in Functioning Misdiagnosed as Dementia

MM was a 40-year-old female with mild disabilities who lived in a community residence. Referral was made for a second opinion to rule out dementia. Prior to evaluation, a diagnosis of DAD was made and donepezil was prescribed based on the reports of withdrawal, an inability to focus on tasks and a decline in adaptive functioning.

The reported regression in MM's behaviour occurred within a 10-month period. The presenting complaints included an increase in anxiety, irritability and avoidance behaviours, insomnia, weight loss, withdrawal, non- compliance, loss of interest in self-care, becoming confused and forgetful, and talking to herself. A number of life events occurred prior to MM's decline in behaviour. In 1993 the death of her father, with whom she had been living with, resulted in a series of changes at her day program and living situation over 4 years until her eventual placement in a group home at age 38.

Cognitive testing revealed the absence of memory impairment, agnosia or apraxia. A mental status exam found depressed mood and constricted effect with some evidence of psychotic features (e.g. talking to objects, fabricating stories). A diagnosis of major depression, single episode with possible psychotic features, and anxiety disorder NOS (not otherwise specified) was made. No clinical findings to support a diagnosis of dementia were evident. Olanzapine and sertraline were prescribed and an improvement in MM's behaviour and condition was reported on the 6-month follow-up. Two subsequent follow-ups revealed MM to be active and doing well with major depression in remission.

Delusional Disorder and Onset of Dementia

DL was a 50-year-old male with mild disabilities who lived in a community residence. Referral was made due to the onset of paranoid ideation and possible auditory hallucinations. Other presenting complaints included periods of agitation, suspiciousness, mental confusion, memory deficits, clothing compulsions and an altered time concept. After the death of his aunt (his only remaining relative), DL fabricated the delusion that he was married and had a wife and an older son.

Initial evaluation found cognitive impairment, confabulation, poor insight and paranoid ideation. A diagnosis of delusional disorder, mixed type was initially given; the diagnosis of dementia was added on 6-month follow-up when a further decline in memory and general functioning was observed. Continued follow-up over a 5-year period noted a gradual loss of cognitive and adaptive skills in all areas. As mental confusion and disorientation increased, delusions and paranoid ideations decreased and were no longer expressed by age 54. Late onset seizures developed at age 54 and a loss of ambulation resulted in DL being relocated to another community residence that provided increased care and supervision. A loss of all self-care

skills and total dependence in all areas by age 55 led to his eventual placement in a nursing home where DL died within a year at age 56.

Major Depression, Single Episode with Selective Mutism

SC was a 28-year-old female with moderate disabilities who lived at home with her parents. Referral was made due to a significant regression in behaviour, which occurred over a 1-year period. The presenting complaints included selective mutism, a lack of energy, non-compliance, social avoidance, insomnia, changes in appetite and weight, lability of mood and behaviour rituals (e.g. excessive hand washing, tooth brushing). The reported changes in behaviour occurred after the birth of her married sister's baby. This event caused a change in focus of the family's attention from SC, especially by her parents, and directed to the new baby in the family. Behaviourally, SC closed herself off from her surroundings choosing not to speak; she insisted on sleeping in her parent's bedroom at night and displayed a fear and general lack of interest in previously enjoyable activities.

Initial evaluation found SC anxious and mildly depressed with flat affect. Speech was minimal and nearly inaudible, with frequent throat clearing and complaints that her throat hurt. When not engaged she was detached and withdrawn. Some evidence of auditory hallucinations was suspected; though on subsequent follow-ups this was found to be self- talk. Memory was intact and although functioning had declined, there were no signs of dementia. The start of fluoxetine resulted in improvement with an increase in expressive language and energy levels within a few months and a gradual return in functioning after 1 year. Follow-up was maintained over a 17-month period with good results.

Tourette Disorder

RC was a 36-year-old male with severe disabilities who lived at home with his family. Referral was made for his obsessive, compulsive and other maladaptive behaviours. The presenting complaints included checking/touching compulsions, hesitation in executing motor actions (e.g. descending a staircase), insomnia, and weight loss due to restricted dietary preferences (he ate only pureed foods). Evaluation of RC revealed depressed mood, short attention span, disruptive behaviour rituals and the presence of vocal and motor tics. Memory was intact with no signs of psychosis. The initial diagnosis of anxiety disorder with obsessive-compulsive traits was made. A trial of paroxetine was ineffective in altering RC's behaviour. On follow-up, a diagnosis of Tourette's disorder was made after it was clear that motor and vocal tics were part of RC's daily repertoire, which waxed or waned along with ritualistic behaviour depending on his level of discomfort in different settings. Treatment with clonidine was effective in reducing the intensity and

duration of his behaviour rituals and tics. The clinical impression was that the compulsive and repetitive behaviours displayed were complex tics and not a function of obsessive-compulsive disorder. The positive response from the use of clonidine appeared to support this impression. Follow-up was maintained over a 5-year period with continued good results.

Anxiety Disorder with Stereotypic Movement Disorder and Onset of Dementia

AB was a 48-year-old female with mild disabilities who lived in a community group home when she was first referred for evaluation. The initial complaints were an increase in her behaviour rituals of impulsive stealing and hoarding of objects. By history, AB was reported to take and hoard objects (e.g. towels, cups, straws and audiotapes) in her bedroom. Mild self-injurious skin picking behaviour to her hands and arms was also reported, which appeared motivated by anxiety. Our assessment found AB to be mildly anxious and in a state of psychomotor excitation. Memory was intact; insight and judgment were fair. A diagnosis of anxiety disorder (not otherwise specified) and stereotypic movement disorder was made. Follow-up at 6-month intervals over a 6-year period revealed some reduction in stealing, hoarding and skin picking behaviours with behavioural programs and the introduction of buspirone and later paroxetine.

At age 55 years, reports of forgetfulness in daily routines and getting lost in the community were first reported by group home staff, and updated cognitive testing revealed a decline in her memory-recall scores from baseline. Increasing difficulties in memory, recall and general orientation on follow-up led to a diagnosis of DAD at age 56. Stealing and hoarding of objects decreased in frequency; however, skin-picking behaviour continued to be displayed. AB's long term hearing deficits also contributed to her difficulties. The addition of donepezil and replacing risperidone with aripiprazole had been helpful. On our last follow-up at age 62, AB's condition remained marked by disorientation, severe memory impairment, anomia and dependence on staff for all levels of care. At age 64, AB died while still residing in the same group home.

Major Depression with Catatonic Features; Obsessive-Compulsive Disorder

CM was a 31-year-old male with moderate disabilities who lived at home with his family. Referral was made due to a significant decline in his behaviour and adaptive functioning. Presenting complaints occurred within a 12-month period and included extreme lethargy, apathy and extreme hesitation in approaching objects and

initiating simple motor actions (e.g. standing up, feeding and dressing self). There was also an increase in previous existing behavioural rituals that included touching/checking compulsions and obsessive behaviour (e.g. counting). A history of self-talk had been present with no evidence of psychotic features. Medical treatment for varicose veins, dermatitis and hyperthyroidism, and a number of social losses and environmental life events occurred prior to the change in behaviour. These events appeared to have precipitated the change in behaviour. A suspected diagnosis of dementia and Parkinsonism was made prior to his being seen in our clinic; however the parents were seeking a second opinion.

Initial evaluation noted depressed mood, behaviour rituals and social withdrawal. There was a marked rigidity in posture and movements connected with his compulsive behaviour, anxiety and ambivalence. The diagnoses of major depression, single episode with catatonic features and OCD were made. Treatment initially with fluoxetine and then with fluvoxamine and risperidone resulted in stabilisation of mood, a return in general functioning and a gradual decrease in behaviour rituals and obsessions. Follow-up and treatment for OCD was maintained over 2 years and his parents reported CM to be doing well with fluvoxamine and clonazepam.

Undiagnosed Autism

LB is a 20-year old male with translocation DS and moderate disabilities who was referred because of escalating aggressive behaviours, especially towards his father. He was brought to a neurologist who prescribed escitalopram for anxiety but LB refused to swallow pills so his mother chose not to give it after a few days. Review of history during the evaluation revealed that LB's father had surgery recently and needed to convalesce at home. LB reportedly does not want his father home when he arrives from school at around 3 p.m. He has a set routine of eating "his bagel with cream cheese," and was not used to the father being around at this time of the day. He wants his father to leave the house and when his father had a medical appointment and was not home at 3 p.m. on a particular day, LB did not exhibit any behaviour problems in the home. His family reported that LB is inflexible, rigid and demands that things are done only his way. He has difficulty with transitions or changes in his routine and gets agitated when his routines are not followed. He typically does not do well in social situations or gatherings (opposite of most DS individuals who are very sociable) and does not want to be around other people and gets extremely anxious.

During the clinic evaluation LB had very poor eye contact and looked away from the examiners. He did not like being asked questions and would only look at his mother when he wanted to say something. While it was very easy for the clinicians in our clinic to identify and diagnose LB with ASD, this was not recognised prior until he was evaluated at age 20 years.

The diagnosis of ASD was based on his clinical symptoms and presentation, which include stereotypies, need for sameness (routine driven) and lack of

relatedness as well as impaired means of communication, which is more typical of individuals with Autism. The differential diagnosis of OCD was considered but LB had no predominant compulsive behaviours. Two clinical psychologists, who were experts in Autism, confirmed the diagnosis as well. Behaviour interventions and a medication trial of quetiapine to address mild anxiety were recommended. Future follow-ups have been scheduled to see how LB will respond to behavioural and medication interventions as he was only seen recently for the first time.

This case illustrated the tendency of clinicians to focus on the obvious presentation of DS while missing the more prominent diagnosis of ASD.

Bipolar Disorder, Type II with Depression as Initial Presentation

MI is a now a 34-year-old male with mild disabilities who was initially referred to a community outpatient clinic at age 19 years because of crying spells, depressed mood, lost of interest in previous pleasurable activities (i.e. going to Broadway shows), sleep changes, decreased focusing, impaired attention and mild irritability. He also exhibited psychotic-like symptoms of looking into the mirror and talking to himself. As per mother's report, before he got "depressed" MI was described as a happy, bright and confident boy who dreamed of attending college, having a family and driving to the city to attend Broadway shows.

However, MI's mood and behaviour changed after he was told in school "he was different from other children and could not attend college classes." It was implied that he had 'mental retardation' and could not proceed to a higher level in school along with his peers. After he heard this, he began to get isolative, no longer wanted to attend school and became nervous around other people. In 3 months he became very depressed and exhibited symptoms stated above. He was started on fluoxetine 20 mg and risperidone 1.0 mg with minimal relief of depressive symptoms. Fluoxetine was titrated up to 30 and then to 40 mg after about 3 months but MI became hypomanic. He did not sleep, was up all night and had pressured speech. He also became very irritable and mood was labile. Fluoxetine was brought down to 30 mg and risperidone was discontinued. MI also began to have distinct panic attacks and was afraid to go out. Alprazolam 0.5 mg as needed was prescribed and clonidine 0.1 mg was added after to help with sleep. Symptoms remained however and MI remained amotivated, anxious and showed no interest in any activity. Later, the diagnosis was changed to Bipolar Disorder, Type II from Major Depression with Panic Attacks. Lamotrigine (a mood stabilizer that is effective for Bipolar depression), was started at 25 mg and added to fluoxetine and clonidine. MI's mood slightly improved and he became less anxious and started to go to Broadway shows again. Lamotrigine was slowly increased to 100 mg and MI became mood stable. Clonidine was stopped as it affected his blood pressure and he became hypotensive.

MI remained mood and behaviour stable with fluoxetine 20 and lamotrigine 100 mg daily and continues to take both medications to this day. MI has now been followed in this clinic for more than 14 years and has able to attend a "special College" for people with DS, attend a summer camp in a different state and was able to find a job and work in a local supermarket. He is also participating more in local community activities.

Although not so common in the DS population, this case illustrates Bipolar Disorder, Type II with Depression and Panic Attacks as the initial presentation. Activation with higher doses of an SSRI (fluoxetine), and mood stabilisation with lamotrigine support this diagnosis.

Conclusion

A number of research studies confirm the higher prevalence of mental health conditions in older persons with IDD as compared to the general population; particularly in those persons with DS [75]. The older DS population being very much susceptible to Alzheimer disease dementia [32] but less so to challenging and maladaptive behaviours [17]. The extent of multimorbidity in older adults with DS, such as obesity, visual impairments, hearing impairments, epilepsy, thyroid dysfunction and constipation being important conditions alongside mental health issues does further impact on the treatment and provision of care for this vulnerable population. A number of psychological disorders present in younger adults with DS e.g. ASD, ADHD, anxiety disorders, may be less of concern for older adults with DS.

Undoubtedly over the next decade many advances in the health and social care of older persons with DS will happen. Genetics will play a major role in our understanding of Alzheimer disease and anti-dementia drugs along with the development of therapeutic inteventions which will help treat and prevent this debilitating illness, and improvements in social care will ensure better community support. Older adults with DS in good physical and mental health surviving into their 70s will then not be unusual.

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Chapter 9
Mental Health in an Ageing Population with Autism Spectrum Disorder

Leona Oakes

Introduction

Understanding ageing in individuals with autism spectrum disorder (ASD) is a field of needed growth across all areas of concern, and especially in regards to mental health. Several challenges complicate this understanding, from the research and clinical emphasis on early childhood and transition age to changing diagnostic criteria that creates significant differences in functioning levels across age cohorts, this field of ASD research is still just emerging [1–4]. Calls have been made across research and clinical professionals to increase our knowledge and practice base for working with adults with ASD, particularly in regards to understanding life span trajectories, ongoing assessment of autism symptoms and mental health concerns, and treatment [5, 6]. Considering the high impact of mental health on quality of life, understanding the mental health concerns in this population is paramount to providing optimal, long-term care [7]. As the prevalence rate of ASD continues to increase, so will this special population continue to age and need evidence-based, specialised care to meet their ongoing needs.

Most individuals with ASD will experience some psychiatric symptoms at some point in their life. Mental health concerns in children with autism are well documented, with high co-morbidity of attention-deficit/hyperactivity disorder (ADHD), oppositional disorder, mood disorders and anxiety disorders [8] starting with school age children and continuing through adolescence and young adulthood. Emerging studies have begun to examine comorbidity and incidence rates of mental health diagnoses in adults with ASD but have limited applicability to current older

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individuals with ASD and limitations in their generalizability to future cohorts as they age. Many studies focus on cross-sectional comparisons of subgroups of individuals with ASD that cannot be generalized to individuals with higher or lower intellectual abilities or language levels. Other studies lump adults with ASD into one large group with little concentration on elders and how their presentation may differ from younger and middle aged adults. In reflection of the information available, this chapter will first present what we know about mental health in adulthood overall, examine work that has looked at mental health trajectory over the lifespan, and present the limited findings on mental health concerns in the oldest age group. Available tools for the assessment and treatment of mental health concerns in elders with ASD will be viewed as well as clinical implications and guidelines.

Comorbidity Studies of Adults

A recent review of autism and ageing found consistent reports of adults with ASD experiencing mental health challenges, particularly in regards to depression, anxiety, aggression, hyperactivity, obsessive compulsive disorder, oppositional conduct and self-injurious behaviour [2]. One of the largest studies examining the health of individuals with ASD, reviewed the medical records of an integrated healthcare delivery organisation in an effort to capture a community-based sample. Through the medical record, they identified 1507 adults with ASD (ages 18–65+), 15% of which were also diagnosed with intellectual disabilities. Within this sample, 54% of individuals with ASD were diagnosed with a psychiatric condition, the most common of which was an anxiety disorder. Individuals with ASD were also five times more likely to have attempted suicide and significantly less likely to have an alcohol- or drug-use disorder [9]. Data taken alongside a larger epidemiological study used the Psychiatric Assessment Schedule for Adults with Developmental Disability (PAS-ADD) interview to evaluate individuals originally diagnosed with ASD in childhood. In a sample of 129 adults with childhood IQs of 70 or greater, 57% met criteria for at least one current psychiatric disorder and 69% for at least one disorder in their lifetime. The most common diagnoses were anxiety disorders [10]. Another set of studies specifically explored the risk and protective factors of depression and anxiety as the most common co-occurring mental health diagnoses in individuals with ASD. One such study sent adolescents and adults with ASD common mental health questionnaires and identified high rates of depression with 49% of scores on the Patient Health Questionnaire in the clinical range for depression and 36% of respondents reporting recent suicidal ideation [11]. A similar study identified rates of about 38% each for anxiety and depression with high overlap of individuals that reported high levels of both [12].

While these studies attempted to get a range of abilities within their groups of individuals with ASD, their reliance on self-report questionnaires biases their results

towards individuals with ASD with at least borderline IQ. Other studies purposely exclude individuals with ASD and intellectual and developmental disabilities (IDD) in an attempt to examine the unique characteristics of ASD that may specifically impact mental health. In one such study, adults with ASD and average IQ were identified to also have common comorbidities, including mood disorders (~53%), anxiety disorders (~50%) and ADHD (43%) with high psychosocial impairment despite average cognitive abilities [13]. Another study of adults with ASD and average cognitive skills used a combination of self- and informant-report to evaluate anxiety and depression specifically. Within this sample, mental health concerns were reported at high rates and with a range of severity: 28% mild to moderate, 23% severe, and 5% very severe across symptoms and behaviours associated with depression, anxiety, and self-injurious behaviours. Severity of mental health concerns was weakly correlated with social functioning and ASD symptom severity as evaluated by the Autism Diagnostic Interview-Revised (ADI-R) [14, 15]. Following, a study focused on individuals with ASD in the community split between those with a professional diagnosis and those self-diagnosed (both groups confirmed through high Autism Quotient scores [16]) and therefore assumed to be "high functioning", found that both groups had similarly elevated rates of anxiety and depression compared to the general population [17]. While these rates seem high considering the higher cognitive level of the participants, it is important to note that these studies relied on recruiting participants from clinics and service providers rather than a community sample, so they have an inherent recruitment bias.

While we expect individuals on the spectrum with widely varying IQ to present differently in regards to clinical symptoms, several aspects of their presentation of mental health concerns are shared. Consistent with research findings presented above, particular difficulty with adjustment was echoed in a study comparing adults with ASD and Intellectual Disability (ID) to those with just ID alone with about 50% of individuals age 14–57 years with ASD and ID having severe adjustment problems compared to 20% of individuals with ID only and the greatest group differences were found in rates of anxiety [18]. Similarly, adults with ASD and ID that were referred to mental health settings were four times more likely than their peers with just ID to exhibit challenging behaviours [19]. However, other studies have found that adults with ASD and ID are *not* at greater risk for mental health concerns compared to individuals with ID alone. Instead, results suggested that severity of intellectual disability, adaptive behaviour skills, and social skills, play a much greater role in comorbidity. However, nearly all authors of these studies warn that the generalizability of these findings are limited due to small sample size within groups across studies and difficulties in the accurate assessment of psychopathology in this population [20]. A large study of over 1000 individuals with ASD and ID found that compared to their peers with only ID, individuals with ASD were less likely to have depression but more likely to have an anxiety disorder and to have challenging behaviours [21]. While a few promising studies about the mental health individuals with ASD and ID have been published, this is still a much neglected

population [22]. Furthermore, concentration on this end of the spectrum may better inform current adult populations as they age but as children more recently diagnosed with widening criteria age and benefit from early intervention and broader school services, it is possible that these finding may quickly become out of date.

Another lens through which to consider comorbidity of mental health concerns in ASD is through an understanding of gender differences. Studies have examined gender difference in psychiatric comorbidity of adults with ASD, as they are common in the neurotypical population [23] and because autism diagnosis has its own gender biases. One study examined gender differences through medical record review of men and women with ASD and ID that were referred to a specialist mental health service. In regards to differences, comorbid personality disorders and schizophrenia were more common in males while dementia was more common in females. Overall, the majority of individuals across both groups failed to meet criteria for a specific psychiatric disorder despite presenting with enough impairment to seek and receive professional intervention [24]. Another, larger study found some conflicting results when examining sex-difference, finding that women with ASD were significantly more likely to be diagnosed with anxiety, bipolar disorder, dementia, depression, psychotic spectrum disorders and to have attempted suicide than men with ASD [9]. Other studies have found that although self-report of depression is higher in women with ASD than in men, reports of suicidal ideation are equal [11]. These conflicting results suggest much more research needs to be done in this area.

One final viewpoint from which to explore mental health in adulthood overall is to examine points across the lifespan. A research team in Australia has examined the trajectory of behaviour and emotional problems in ASD by following a cohort of individuals over 18 years with 5 study time points spread across that follow up period. Overall, this cohort demonstrated high rates of comorbid behavioural and emotional problems that declined over time as measured by the Developmental Behaviour Checklist. A little over 61% of individuals demonstrated significant decreases in symptoms while about 16% experienced an increase. Individuals with more severe intellectual disability were described as having more behavioural and emotional problems [25]. Another study performed a retrospective review of individuals with ASD aged 30 and older and compared those over 50 to those younger than 50. Between groups, there was a trend for more residential supervision and more physical aggression in those younger than 50 years of age. Within individuals, there was a significant decrease in the prevalence of challenging behaviour over time [26]. This trajectory focus may become more important over time as we attempt to make conclusions relevant to the currently ageing and to generations to come.

Overall, research on mental health comorbidity and trajectory of adults with ASD across the spectrum of cognitive ability is a slowly growing field. While promising studies have been produced there is still little consensus regarding specific numbers and comparisons between individuals with ASD with and without ID and between men and women on the spectrum.

Focus on Older Adults with ASD

A handful of studies specifically investigate mental health concerns in elders with ASD. Most of their results are similar to the trends identified in studies including the full adult span but some examine specific factors of special concern to ageing populations.

A recent Swedish study of adults with ASD aged 55 years and older found that 60% of those adults had been in contact with some kind of mental health care and 50% had at least one mental health diagnosis. Individuals without comorbid intellectual disability were more likely to have a co-occurring psychiatric disorder than were those with ID [27]. Another study attempted to eliminate diagnostic bias by screening older adults (age 50 or older) with ID for autism spectrum disorder at the time of the study in the hopes that this study would better inform expectations for more currently diagnosed individuals as they age. Identified seniors with ASD were then separated into groups of older adults with ASD and ID and those with just ID and compared. That comparison revealed that senior adults with ASD and ID did not differ from their peers with ID in regards to behaviour problems, psychiatric disorders, or quality of life. Rather, the greatest predictor of psychiatric disorders and low quality of life was decreased adaptive skills.

Additionally, compared to younger adults with ASD and ID, older adults with ASD and ID had fewer behaviour problems and psychiatric diagnoses [28]. A review paper of 13 studies of autism and ageing found that overall adults with ASD 50 years and older have lower rates of psychiatric diagnoses than young adults with ASD, regardless of intellectual ability [3]. In another study comparing older adults (ages 61–88 years) with indications of broader autism phenotype, those that scored higher also reported higher depression and anxiety compared to peers with lower symptomology [29]. These results suggest that while older adults with ASD still present with higher rates of comorbid mental health concerns than their neurotypical peers, they are not so different than others their age with broader intellectual and developmental disabilities.

Following, research has just begun to explore whether or not adults with ASD will follow typical ageing trajectories associated with neurotypical ageing that are associated with greater social isolation and depression or whether some cognitive functions will be spared because they function differently [4]. As a group, adults with ASD have several age-related risk factors for the development of new or the worsening of pre-existing mental health concerns. For example, older adults with ASD may be more acutely affected by loss and bereavement for parents and siblings as they often rely more on these family members for support and caretaking [3]. They are also more likely to have chronic physical health issues that negatively impact their quality of life [30]. Following, many seniors with ASD have the added implications of ageing common to others with intellectual and developemntal disorders, including limited opportunities for socialisation, stigma, and lack of fulfilling occupation and recreation, which also create risk factors for emotional distress [3]. A meta-analysis examined quality of life of adults with ASD across the

lifespan and conducted a smaller study to specifically focus on the elderly with ASD. They found that as a group, individuals with ASD have significantly lower quality of life compared to same age peers. There was no effect of age across the entire age range as evaluated through the meta-analysis nor in the follow up study that focused specifically on those in the oldest age group (53–83 years) [31]. Another study included seniors with broader symptomology of autism and found that those with more ASD symptoms had more executive functioning problems and lower social support compared to their peers, two significant risk factors for age-related issues [29].

Even in consideration of these promising studies, some argue there is still too little known about ASD in adulthood to make conclusive statements about comorbidity rates across the whole adult age range, and especially in older adulthood [1]. Studies tend to be small and provide inconsistent and sometimes conflicting results with highly variable psychiatric comorbidity depending on recruitment strategy, setting, and assessments used. The most consistent statement appears to be that adults with ASD have considerably poorer mental health than their peers. An analysis looking specifically at anxiety and depression estimated that in adults with ASD, current prevalence of an anxiety disorder is 27% with a lifetime prevalence of 42%. Depression was similar with a current prevalence of 23% and lifetime prevalence of 37%. Conclusions could not be made about these rates in older adulthood due to lack of research [22].

Assessment of Mental Health in Adults with ASD

One of the challenges to understanding mental health concerns in older adults with ASD is in identifying and using valid assessment instruments for this population. Most mental health assessments are designed for use with typically developing adults and even those created for use with individuals with intellectual/developmental disabilities are subject to bias. A full review of assessments available and used to evaluate psychiatric disorders in adults with ASD identified a significant lack of tools with robust evidence of appropriate use in this population and advises a multimodal approach with several assessments that allows for judgement of how ASD symptoms impact the presentation of mental health concerns [32].

Anxiety presents as a particularly difficult concern to address due to diagnostic overshadowing of common symptoms between ASD and anxiety, ASD-specific presentation of anxiety symptoms, overlap of symptoms between anxiety and other common comorbidities, and the need to rely on informants rather than self-report of internalizing symptoms [33]. One study comparing different assessment methods found that rather than self-report of physiological arousal symptoms associated with anxiety, questions about general adjustment problems were much more indicative of significant levels of anxiety in adults with ASD and multimodal assessment was the most reliable method with which to collect that information

[33]. Following, in an evaluation with adults with ASD who had IQ scores of 70 or above in childhood, participants often rated their mental health concerns more severely that did their identified informants and diagnostic criteria were met less frequently when measured by questionnaire compared to by interview. The only exception to this was in symptoms of obsessive compulsive disorder, which participants often rated highly on questionnaires but, when asked to describe their obsessions and compulsions, were found to fit best under the repetitive and restrictive behaviours characteristic of ASD [15]. These findings were tempered by a recent review that found that rates of anxiety disorder were similar as assessed by questionnaire or interview with the continued exception of obsessive-compulsive disorder, which was also over-indicated when evaluated by questionnaire [22]. It is important to note that most of these studies only included individuals with ASD and borderline IQ or higher so their results may not be generalizable to the broader spectrum.

In response to assessment limitations, the Autism Spectrum Disorders-Comorbidity for Adults (ASD-CA) assessment was created to better understand the co-occurrence of psychiatric concerns in adults with ASD regardless of their cognitive abilities [34]. This assessment is rather new and is still building its evidence base. Several studies have also identified the Psychopathology Assessment Schedule for Adults with Developmental Disabilities (PAS-ADD) to be promising as it has been found to be sensitive to both ASD symptoms and to other psychiatric symptoms exhibited by individuals with ASD [24, 35].

Treatment of Mental Health in Adults and Ongoing Implications

The vast majority of articles reporting on rates of mental health concerns in adults with ASD make little mention of treatment approaches and the few that do mention access to some kind of treatment or focus on medication management. A review of the medical records of individuals with ASD and ID receiving services at a specialty mental health clinic found that 70% of those individuals were prescribed at least one psychotropic medication with men more likely to be prescribed a combination of medication and women more likely to receive a sedative [24]. A community-based study found that 59% of adults with ASD were on at least one psychotropic medication [10]. A similar study based in Australia found that individuals with ASD were prescribed psychotropic medication significantly more often than neurotypical peers, even when controlling for a co-occurring disorder and about 14% of people with ASD were prescribed medication without any comorbid diagnosis [36]. These results are echoed in research specifically with individuals with ASD and ID that compared them to peers with ID alone. Their findings also identified that individuals with ASD were significantly more likely to be prescribed medication for mood disorder, psychosis, anxiety and challenging behaviour [21].

While there are no systematic reviews of therapeutic approaches to mental health concerns for adults or elders with ASD, we can extrapolate from the available literature and current clinical practice. Overall, cognitive behavioural therapy (CBT) is the most empirically supported treatment of anxiety and depression in individuals with ASD [37, 38]. Standard manuals for CBT can be used as written or slightly modified for use with individuals with ASD and average cognitive abilities and individuals with ASD and ID may benefit from CBT modified specifically for those with cognitive differences [39]. Regardless of treatment approach, early and consistent management of mental health concerns, including appropriate monitoring of medication [3] with careful reduction of medications used to control disruptive behaviours as these reduce with age is vital.

Additionally, given the repeated finding that low adaptive skills are associated with higher mental health symptom severity, special attention must also be paid to the learning of everyday skills across environment for individuals on the spectrum of all ages. A focus on increasing adaptive skills early on and continued into lifelong learning and development may be protective against mental health concerns [28]. Lifelong learning is highly recommended across several areas [3], especially as it relates to increasing independence and improving relationships [7].

Lastly, early and regularly updated future planning for loss of family caregivers and end of life decision making [3] and awareness of poor quality of life risk and protect factors is crucial to the unique needs of an ageing population and especially applicable to elders with ASD.

Conclusion

There remains an ongoing need for future developments in our understanding of ageing in individuals with autism spectrum disorder (ASD). Especially in regards to associated mental health issues which can have a high impact on quality of life in individuals who already have a significantly lower quality of life compared to same age peers [6]. Even expected life events such as loss and bereavement for parents may affect older adults with ASD more acutely than their peers [3].

Adults with ASD are reported to have high co-morbidity of attention-deficit/hyperactivity disorder (ADHD), challenging behaviour, mood disorders and anxiety disorders [8] and subsequently more likely to be prescribed medication [21]. Some evidence suggesting that women with ASD are significantly more likely to be diagnosed with anxiety, bipolar disorder, dementia, depression, psychotic spectrum disorders and to have attempted suicide than men with ASD [9]. As the population of older adults with ASD continues to increase, so will the need for evidence-based, specialised care to meet their ongoing needs which along with greater awareness of mental health concerns will improve the chances of providing optimal, long-term care [7]. This previously under researched group are beginning to be the focus of important clinical and scientific advances.

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Chapter 10
Bereavement, Grief Reactions and End of Life

Suzanne Guerin, Damien O'Riordan, Geraldine Boland, and Philip Charles Dodd

Introduction

Our understanding of grief and bereavement as it is experienced by people with intellectual and developmental disabilities (IDD) has, for the most part, developed substantially in the last 20 years. Our understanding has grown from the early opinion, which suggested that individuals with IDD lacked the cognitive capacity to experience grief following the death of a loved one, to a developing literature on the experience of end of life care for people with IDD. In addition, given that complicated grief, also known as traumatic grief or prolonged grief, is a current issue of debate in the medical and psychological literature, it is reassuring to see this topic being considered in the context of IDD. It is in this context that the issue of complicated grief as experienced by adults with IDD is included in this chapter.

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The existing literature to date has tended to focus on adults with IDD, with very little literature considering either children or older adults in particular. Given the focus of this text on older adults with IDD, this chapter attempts to capture current understanding of the experience of grief—both typical and complicated grief—among adults with IDD, considering the nature of grief and bereavement among this group, the factors that appear to influence this experience, as well as the existing knowledge about how best to support people with IDD during this time. However, one key point for the reader is the focus of the body of this chapter on adults with IDD, with extrapolation to the experience of older adults. In reviewing the existing literature in this area, there has been a growing interest in the experience of palliative and end of life care among people with IDD and so, we have included a short section on this issue.

A Note on Definition of Key Terms

Before considering the body of knowledge on this topic, it is important to reflect on key definitions, given that there is some variation in the language in this area. It is interesting to note that there is little explicit discussion of the key concepts of grief and bereavement in the IDD literature, though many articles will refer to common definitions and models from the general population [1–6]. For the purpose of this chapter, we adopt the definitions used by Reynolds and colleagues [7], whereby grief is presented as feelings of intense sorrow, grieving is the process of feeling this sorrow and bereavement is the grief experienced by the loss of a loved one. We would also note the consideration of the mourning process as one that supports the transition to the new reality without the person who has died [8] and will often involve engagement in public and private rituals [9]. It is interesting to note that in a number of papers, these experiences are associated with attachment [10] and indeed separation distress and the breaking of bonds [11, 12]. While these feelings can result from losses other than death, this chapter focuses on the feelings following the death of a loved one or significant person in the life of a person with an IDD. Given the focus on older adults with IDD, it is accepted that this group is likely to have experienced significant index losses, including parental death, which are considered in the context of associated losses such as the family home and existing care arrangements.

A key aspect of IDD related to the understanding of grief and bereavement is the concept of death, and its related components including irreversibility, non-functionality and universality [13]. While the early literature on IDD questioned if people with IDD had a sufficient concept of death that would allow them to experience grief [14], several empirical studies have clearly established the evidence of concept of death among people with IDD and the ability of people with IDD without a full concept of death to show understanding of emotions that follow bereavement [15, 16]. Therefore, while a full understanding of the concept of death may not be achieved by all people with IDD, this does not necessarily minimize or prevent the experience of the emotional aspects of grief.

Considering broader aspects of grief, there are several that have been discussed in the IDD literature. The first is considered as a separate section later in this chapter, that is the issue of complicated grief (our preferred term). However, the concepts of anticipatory and disenfranchised grief are also considered in the context of IDD. Authors have considered these issues in the context of IDD, with disenfranchised grief [2, 3] representing the extent to which loss among people with IDD may not be recognised by those around them, thus reducing the likelihood that they receive needed supports [17–19]. Weise and colleagues [20] suggest that historic practices of preventing participation in bereavement rituals by people with IDD can increase the risk of disenfranchised grief. Less frequently considered, though an important concept for IDD is anticipatory grief. Elliot and Dale [21] note that an expected or anticipated death may allow for some preparatory work to be done, but that this can also influence the experience of anticipatory grief [22], while other researchers consider the fear that can come from the death of a peer [12].

Finally, the issues of loss and grief have also been considered in the context of palliative care for people with IDD. The provision of supports towards and during the end of life are discussed in many texts [23, 24]. However, within the IDD literature there is an interesting body of work that focuses on the person with IDD in the context of the experience of palliative care services or the experiences of peers with IDD (including both self and staff reports), and indeed their experience of the period before their own death. This is one of the contexts in which anticipatory grief and the extent to which fear of their own impeding death may influence this type of grief is considered [25].

Understanding the Grief Reactions of People with IDD

There is a significant body of literature that directly and indirectly (e.g. studies on interventions), though self and proxy reports, describe the grief reactions experienced by people with IDD [26–28]. Indeed, later publications have extended knowledge of grief reactions from those with mild and moderate IDD, to include those with severe and profound IDD [29, 30]. Reviewing the broad content of this literature, it is possible to frame reactions as physical/behavioural, emotional, and cognitive, with a key message being the extent to which many of the grief reactions experienced by the IDD population are common in the general population also.

Physical and behavioural reactions include shock, physical illness, poor appetite and weight loss (associate with refusal to eat or drink), and disrupted sleep. Behavioural reactions include both the display of behaviours that challenge (including self-injury [9]), but also refusal or inability to engage with educational work and social activities.

Emotional reactions frequently include sadness and loneliness, fear and confusion, anger and worry [31]. McEvoy and Smith [32] report a study of family perceptions that highlight many of these emotions in relatives with IDD. Multiple studies have reported that these emotional responses can include mental health difficulties [33].

Cognitive reactions can include avoiding thoughts or conversations about the person who has died, and dreams of the person who has died [34]. More significant reactions include visual and auditory hallucinations (in the context of complicated grief), as well as general cognitive decline [35].

These reactions represent a constellation of responses that are, in many ways, typical of the general population. However, the literature, in particular case studies [36, 37] highlight the complexity of the reactions to grief displayed by individuals with IDD. The impact of these reactions on both paid and family carers is also evident in studies which highlight the process of supporting people with IDD as a challenging task for carers [10].

Across the literature several contextual factors are associated with the above detailed grief reactions, relating to the individual and their immediate care (service and family) context. One issue considered is involvement in bereavement rituals [8, 32], including the importance of activities designed to promote positive memories of the person who has died [38]. However, it is interesting to note that a correlation between complicated grief reactions and involvement in bereavement rituals has been reported [39], suggesting that involvement without preparation may be associated with more distressing grief reactions. Another key factor are the views and attitudes of carers towards the individual's bereavement experience [26], which also highlights the potential for training and education for these groups [7].

Support and Intervention for Bereaved Individuals with IDD

A key strength of the literature on the issue of grief and bereavement in IDD, is the focus on practical supports and interventions for this population. While the literature is still somewhat exploratory in its assessment of these interventions [37], there is no shortage of discussion of how best to support individuals with IDD at this time, with many studies suggesting that these interventions show promise. The supports discussed fall broadly into several categories, including education/training regarding bereavement for both people with IDD and carers (including formal and informal), participation in key rituals as intervention, and therapeutic interventions based on a number of models of therapy. We will consider some of the literature highlighting the strengths and limitations of these approaches in this section.

Educational programmes for people with IDD are discussed both as individual and group-based programmes. Read and colleagues [40] describe a group-based educational workshop, which they describe as more structured than standard bereavement supports and drawing on more creative activities. They note the advantages of the workshop approach including contributing to normalizing loss and providing inbuilt peer support. The importance of the provision of early support and education for families on supporting individuals with IDD has been reported [32]. This may involve including funeral directors with experience in this area, who can support the provision of information for families [8].

Studies have also highlighted the impact of training for professional staff, particularly on confidence in supporting adults with IDD who have been bereaved [7, 41]. One particular area of research has been supporting staff to break bad news to

people with IDD, whether in relation to the death (or expected death) of a loved one, or their own death. Tuffrey-Wijne and colleagues [42–45] have considered the factors that can influence the process of breaking bad news, the need for training for staff in this area and the development of guidelines for disclosure. Given the focus on education, combined workshops on bereavement for staff, carers and people with IDD have been proposed [46].

While noted as a factor that impacts on grief reactions above, ritual participation and in particular, supported ritual participation can be an important support for people with IDD. This is interesting given the historical context in which people with IDD might have been excluded from these rituals out of a concern for their ability to understand the rituals. While some may suggest that this is an outdated perspective, frustration at families' decisions not to involve the person with IDD in rituals and the concerns for the family's ability to cope with supporting the person with IDD have been reported [10]. Funeral attendance, supported remembering and engagement with anniversaries can be beneficial [20, 25, 47]. Young and Garrard [19] highlighted the benefits of techniques that support remembering the deceased person; in this study a memory box was utilized, which was found to support connections and understanding of the loss among adults with IDD.

The final area of intervention discussed included therapeutic intervention including models of bereavement counselling. Again, studies have considered both individual and group based therapeutic interventions. In an example of individual therapeutic support, Summers and Witts [48] described an approach that combined both psycho-educational and psychodynamic approaches with a bereaved woman with IDD. They suggest that this intervention was associated with a decrease in intensity of behaviours that challenge and improved understanding of the death. Considering other modalities, cognitive therapies have been reported to be effective for anxiety associated with the death of a loved one [34]. Group-based psychotherapy interventions supporting the grieving process for people with IDD can be valuable [49].

A key conclusion from the research on interventions in this area is that there is a range of options for services supporting adults with IDD in advance of or following a significant bereavement. Though the research represents perhaps lower levels of evidence than would definitively guide decisions around effective support, there is no doubt that there are many promising and appropriate options available to support people with IDD though the period of bereavement.

Understanding the Experience of End of Life Among Adults with IDD

The issue of the experience and indeed care of people with IDD at the end of their life is important in the context of grief and bereavement. As noted above, people with IDD may be at increased risk of disenfranchised grief and may also experience anticipatory grief, both very much related to an understanding of one's own impending death.

Several helpful review papers exist in the published research, which capture some of the key information available on this issue. Tuffrey-Wijne and colleagues [50] present their review in the context of increased longevity as well as increasing rates of diagnosis of life limiting illnesses among adults with IDD, also noting the increase in interest in the issue of palliative and end of life care for this group since a previous review in 2003. A more recent review highlights the extent of the development of this body of research, with consideration of the specific issues of decision making at the end of life [51].

A number of issues are underlined in the literature on this topic. One issue is that of communication, with communication difficulties among adults with IDD identified as a barrier to the provision of care at the end of life and recommendations including the use of clear language [52]. This can extend to the experience of people with IDD not being aware of a terminal diagnosis or challenges associated with the timing and frequency of relevant discussions [51]. Related to the issue of communication, the ability of adults with IDD to report their experience of pain and for professionals to assess and manage this pain appropriately is also highlighted as a challenge. People with IDD may communicate pain and distress though the presentation of behaviours that challenge [45]. The issue of decision making is also to be considered, with studies reflecting struggles with the issue of continued treatment, the capacity of people with IDD to make decisions to end treatment and the role of third parties in this process [51]. Perhaps unsurprisingly people with more significant disabilities are more likely to be excluded from the decision-making process [53]. Linked to this is the small body of literature on do not resuscitate orders and euthanasia [54–56], issues that present challenges in the typical population, and added layers of emotion and complexity in the IDD population.

Perhaps unsurprisingly, a substantial body of literature focuses on the need for and provision of supports and services for people with IDD as they approach the end of their life [57, 58]. Highlighting the shared and unique insights among groups of staff working in IDD and palliative care settings along with issues of confidence among both groups in addressing the intersection of these two areas [59]. It is interesting to note that the growing support for the provision of palliative care in non-specialist settings [60] would fit nicely in this context, though clearly there are training needs among both groups. There remains the potential challenge of family perspectives in the provision of care, including conflict regarding wishes and decision making [59].

What is impressive about this body of literature is the inclusion of multiple perspectives, despite the potential sensitivity of this issue. Studies explore the views and experiences of individuals with IDD [61, 62], paid carers [57, 63] and medical/clinical professionals [53]. However, there is less focus on the specific views of family members [64]. These different perspectives are important, as they allow for a clearer understanding of the needs of adults with IDD approaching the end of their lives and the how professionals and family members can support this process.

From one area that has grown in terms of research interest, we move to another that has shown similar growth in interest in more recent times, perhaps in the context of increased discussion of the concept in the typically developing literature [65].

Complicated Grief in People with IDD

There has been a growth of interest in the research of complicated grief in this population, which reflects the increased focus on this issue in the general population. Unsurprisingly, research in this area is much less developed, though it has been suggested that people with IDD might be at a greater risk of complicated grief [39]. Within the general population there are challenges and uncertainty surrounding the definition of complicated grief. The upcoming ICD 11 for example, refers to prolonged grief disorder whereas DSM-5 refers to persistent complex bereavement disorder [66].

There are numerous additional challenges when carrying out research with people with IDD, which makes defining and diagnosing complicated grief in this group even more challenging [26, 67]. Cognitive deficits, varying levels of understanding death, communication difficulties, along with the heterogenicity of this group have been cited as challenges leading to deficits in our understanding of this important clinical presentation [68, 69]. Despite these challenges, there are similarities but also noticeable differences between complicated grief in the people with IDD when compared to the general population.

Regarding terminology, prolonged grief disorder is the diagnostic term most often used in the general population, whereas complicated grief is most used when referring to this form of grief in people with IDD [68]. Importantly, there is evidence that complicated grief is more common in people with IDD compared to the general population. Dodd and Guerin [68] found that one third of bereaved people with IDD experienced ten or more clinically apparent symptoms of complicated grief following the bereavement of a parent. There is approximately a 10% prevalence of prolonged grief disorder in the general population [65]. People with IDD are at a higher risk of developing traumatic grief (a term sometimes used to describe complicated grief) [70].

Signs and Symptoms of Complicated Grief in People with IDD

Studies have considered the usual features of complicated grief in people with IDD. Firstly, grief symptoms are delayed and prolonged beyond those experienced in the general population [33]. Traumatic grief symptoms such as disbelief and bitterness over the loss are present in people with IDD post bereavement. Separation distress symptoms such as yearning for the deceased, being unable to think about anything other than the deceased, and distrust of others are all also present in people with IDD post bereavement. Interestingly, separation distress symptoms appear to occur more frequently than traumatic grief-type symptoms in this population. The prominence of separation distress in people with IDD may be in keeping with the proposition developed by Bowlby [5] that grief is an extension of a general response to separation where an attachment has been broken [39]. People with IDD may be more vulnerable to attachment difficulties.

Developing accurate screening and diagnostic tools are crucial to allow for better understanding, diagnosis, and treatment of this condition. Carer reported scales such as the complicated grief questionnaire for people with IDD (CGQ-ID) measures the maladaptive symptoms of loss and has been shown to accurately distinguish bereaved and non-bereaved people with IDD [69]. Moreover, a self-report version of the CGQ-ID has been recently developed and similarly appears promising as a means of diagnosing cases of complicated grief [71]. It is likely a combination of self-report and proxy report scales which will prove to be pivotal in diagnosing complicated grief going forward, but further research is required.

Risk and Protective Factors

People with IDD have numerous risk factors that increase the likelihood of developing complicated grief including environmental factors. The importance of secondary losses, such as loss of the family home and having to move in with paid carers have repeatedly been highlighted in the literature [67, 71, 72]. Moreover, if hidden losses go unrecognised the original grief is compounded, increasing the risk of complicated grief further [73]. Disenfranchised grief, discussed earlier as grief that is not acknowledged and deficits in the supporting environment, including low staff confidence, have both been proposed as risk factors for more complicated grief reactions [73].

Risk factors, relating specifically to IDD such as communication difficulties [68, 73], cognitive difficulties [26, 68, 73], difficulties understanding the finality of death [70, 74], and masking of grief [72] have all been highlighted. Indeed, people with IDD may lack the internal resources to manage death related emotions as well as an inability to obtain external support systems when needed, thus increasing the risk further [19].

The relationship between the person with IDD and the deceased loved one, including the degree of dependency on the deceased, may influence the likelihood of developing complicated grief [73]. Insecure attachment and the associated absence of a secure psychological identity may increase the probability of poor grief outcomes and complicated grief [72]. As mentioned previously, Dodd and colleagues [39] also found that bereavement ritual involvement, was associated with increased risk of complicated grief symptoms. They proposed that ritual involvement, may actually exacerbate separation distress symptoms, especially if the people with IDD have limited previous exposure to ritual involvement or has limited prior understanding of what is involved in the bereavement ritual. Contextualizing this result by acknowledging how meaningful ritual involvement can be for the individual. A bereavement history should be taken along with bereavement ritual education prior to deciding on the appropriate level of bereavement involvement.

The literature highlights potential protective factors against CG include providing supports during and after the death for the people with IDD and for the person's family [18], providing skills training for staff [18], reducing secondary losses [26],

and providing death education for people with IDD and families [19]. Finally, the people with IDD should have death education prior to deciding on the level of bereavement involvement [39].

Treatment Approaches

Reflecting the points made in relation to supports for typical grief reactions among people with IDD, the lack of evidence-based treatment options for complicated grief in people with IDD is also noted [26, 68]. It is proposed that we must first improve our understanding of grief responses and be able to accurately diagnose complicated grief in people with IDD to then allow for the development of effective treatment approaches [68, 71, 74].

The research that is available highlights the importance of optimizing environmental factors. The use of a ten question, bereavement needs assessment tool (BNAT) tool to clarify the specific needs (practical, emotional, and social supports) of the people with IDD post bereavement has been proposed [73]. Similarly, the use of targeted supportive treatment which is an individualized treatment that focuses on environmental factors including responding to secondary losses has been suggested [26]. It is hoped that such environmental interventions will not only reduce the frequency of complicated grief but also improve the impact and outcome of complicated grief in this population.

Again, reflecting the supports for typically presenting grief, numerous therapeutic approaches have also been proposed for use in the context of complicated grief. One approach is relational psychotherapy whereby unconscious processes at play within relationships, such as the relationship between paid carer and client, are considered [67]. Through training, staff may better understand the dynamics within their supportive relationships enabling them to provide insightful and skilled care, thus reducing the frequency and impact of complicated grief. Other techniques include the use of goal setting and communication focused counseling [26]or the use of a systems-based approach including a model for a time limited intervention consisting of both bereavement group and individualized treatment [75]. The effectiveness of any psychological intervention for complicated grief in people with IDD has yet to be experimentally proven.

The importance of carer training in the context of complicated grief, as a prerequisite to cases being identified, referred to appropriate services, and treated properly has been highlighted numerous times in the available literature [18, 76]. Appropriate staff training can help staff provide more tailored support systems, and ideally help people with IDDs work through or bypass traumatic grief reactions [70].

Despite the growing body of literature on complicated grief reactions in people with IDD, there is a clear need to develop complicated grief treatment approaches and the effectiveness of these need to be experimentally tested. A structured treatment should be developed and trialed in people with IDD who have CG. This could be achieved by adapting a recognised treatment approach from the general

population such as the complicated grief therapy approach [77] which is currently underway by some of the authors on this chapter.

Recommendations from Published Literature

Across the body of literature that is available on grief and bereavement in IDD, many recommendations have been made. Many of these have related to recommended interventions for people with IDD, as well as education and training for families and staff (as discussed above), though as was noted earlier the research falls short of providing any definitive evidence on the impact of the different options discussed.

Related to appropriate support for people with IDD who have been bereaved, one common recommendation highlights the importance of assisted mourning for people with IDD following a bereavement. A central element of assisted mourning is involvement in culturally appropriate bereavement rituals, with multiple authors overtime stressing this issue [11, 18, 78]. Assisted mourning also encompasses elements of pre-bereavement death education using accessible information, involvement in post bereavement planning and support with the transitions that may follow the loss, for example when a parent's death requires new accommodation arrangements [79]. It remains important for person-centred planning to take place in supporting the experience and expression of grief and loss after the death of a loved one [80]. In the context of death education, there remains a lack of research evidence on the issue of preparation for one's own mortality [81] and on the rights of people with IDD to understand they are dying [25].

The growth of literature on palliative care for people with IDD highlights the increasing rates of receipt of end of life care by this group [53], and with it comes the need to support people with IDD to understand this phase of their lives. Researchers have highlighted the importance of supporting a dignified death in a place of one's choice for people with IDD and highlighted the need for advanced care planning with this group [20, 23, 82]. As with general issues of grief and bereavement, there remains a need for training of professionals and support for family members in the context of end of life care for people with IDD [50, 51, 59].

Another recommendation relates to the need for effective assessment strategies to understand more fully the experience of grief among this population and to identify individuals with IDD in need of perhaps more specialist supports. These would involve the need for independent assessments of concept of death to establish understanding [11] and the importance of comprehensive bereavement assessment [83]. Our own work on the assessment of complicated grief in people with IDD highlighted the ability of staff to identify symptoms in adults with IDD [39, 69], and our current project is showing initial evidence that self-report of these symptom can be supported among people with IDD [71]. Perhaps unsurprisingly there are multiple recommendations for further research on the topic of grief and bereavement in IDD.

Conclusion

Reflecting on the content of this chapter, two points are immediately clear. There is a significant body of literature on grief and bereavement in IDD, which includes empirical consideration of the grief reactions experienced by adults with IDD, preliminary evidence of a range of supports to assist adults with IDD in negotiating the process of bereavement, and a body of recommendations for practice in this area. Also impressive is the growing recognition of the needs of people with IDD as the end of their own lives approach. However in contrast, and important to the focus of this text, there is little explicit consideration of the experience of older adults with IDD, though we know that this group is more likely that younger adults to experience multiple bereavements of family members and other loved ones, including peers. We do note that the research on end of life is more likely to include older adults. Nevertheless, given that the research discussed here has shown that the methods are available to explore individuals' experiences, there is a clear need to explore the lived experiences of grief and bereavement among this group.

A final concluding point though, is reserved for the issue of complicated grief, which also shows increased focus, perhaps reflecting increased interest among the typical population. Despite this growth, complicated grief remains ill-defined in the IDD population. The evidence that is available suggests that grief is not only delayed and prolonged in IDD, but there is a prominence of separation distress symptoms such as yearning for the deceased. Accurate screening and diagnostic tools are crucial to allow for better understanding, diagnosis, and treatment of complicated grief among people with IDD. There is also a lack of evidence-based treatment options in this area. Standardized bereavement assessments along with practical, environment and social supports aimed at reducing secondary losses are likely important components to any effective intervention. It is hoped that work being completed by the authors of this chapter to adapt Katherine Shear's complicated grief therapy model from the general population for an IDD population will result in an IDD specific and effective psychological treatment approach for supporting people with IDD experiencing more complicated forms of grief.

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Part II
Treatments and Interventions

Chapter 11
Psychological Interventions and Psychotherapy

Germain Weber and Alina Streicher

Introduction

In this chapter the term 'psychotherapy' is used to mean classical psychotherapeutic methods, reaching from psychoanalysis, humanistic therapies to behaviour therapy and cognitive therapy. However, the term 'psychological interventions' is used as a generic term for various psychologically founded interventions, including classical psychotherapies, as well as general psychological counselling and specific psychological interventions aiming at support for defined groups.

Psychological interventions and psychotherapy in older people with intellectual and developmental disabilities (IDD) are topics largely neglected both in systematic research and general practice. On the one hand this can be explained by the fact that psychotherapists and clinical psychologists show in general a reserve toward therapeutic interventions for older people, and on the other hand, very often psychotherapeutic techniques are thought not to be indicated for people with IDD, as these people might lack language capabilities required for verbally-focused psychotherapies, or their capacities for insight are thought to be reduced.

Availability and access to psychotherapy and other psychological interventions remains very limited for adult people with IDD with those 50 and over and those with profound and multiple disabilities experiencing the greatest barriers of access to psychological interventions [1]. The figures from a 14-year longitudinal study are straightforward in this respect: From those meeting the criteria for a mental health

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disorder only 10% received any form of mental health treatment [2]. Regarding psychological interventions and psychotherapy this provision disparity might be linked to the fact that most people with IDD do not have the necessary financial resources for paying for psychotherapy.

However, for children and younger adults with intellectual disabilities there is a growing body of literature with respect to psychotherapy [3]. Furthermore, disability therapists have adapted and discovered concepts like *handicapped smile* and *secondary handicap as a defence against trauma* for the clinical work in the population with intellectual disabilities [4].

Finally, prevalence figures for mental health needs and mental health disorders are up to four times higher for younger adults with IDD as compared to their peers without disabilities, and these numbers are reported invariant while people with IDD are growing older. For the overall adult population with IDD—age range 16–83 years—figures reported for a clinical diagnosis of mental-ill health of any type are 40.9% [5]. With increasing age, behaviour problems like aggression and self-mutilation show dropping prevalence figures, however, mental disorders like depression, anxiety and sleep disorders might increase in frequency, with a prevalence of psychiatric multi-morbidity of the population 50+ varying between 40% and 71% [6]. In addition, new mental health needs related primarily to declines in cognitive functions and frequently related to neurodegenerative processes and other conditions, provide a challenge for psychological interventions with older adults with IDD. A good overview for the state of research in mental health and IDD in general was recently published [3]. In addition, the different interactions between psychotherapeutic and psychopharmacological interventions, including the potential for facilitative or inhibitory effects of one treatment modality to the other, should be considered in cases of dually diagnosed individuals [7]. Further, pharmacotherapy used in severe, potentially harmful behavioural syndromes or in the more biologically driven mental disorders has to be tailored to age-related vulnerability [8].

This chapter offers an overview of the research and reported experiences related to psychological interventions in the adult population with IDD, and especially with respect to the older segment of this population. In addition, emerging models of psychological interventions for older people with IDD will be outlined, and psychological interventions as successfully developed for the general older population or people with Alzheimer's disease will be reflected on to look at their appropriateness and usefulness toward an application in elderly persons with IDD. To start, the chapter introduces two distinctive markers specific to psychotherapy in IDD and old age. Though respectable advances can be observed in psychological intervention and psychotherapy research for children, youth and younger adults with IDD during the last decade with an emerging and positive evidence base for treatment effects [9–11], extensive literature research has shown in this period however an absence of psychological intervention research with respect to the older segment of the population with IDD.

Characteristics Affecting Psychotherapeutic Interventions

Focusing on psychotherapy in older people with IDD requires the consideration of main characteristics for psychotherapy and psychological interventions in the population of IDD, and characteristics of therapists, as well as main characteristics for psychological interventions related to old age. These specifications should be considered within a general framework for psychotherapy and psychological interventions for older people with IDD. Standards for psychotherapeutic interventions for various therapeutic orientations can easily be derived from a general geronto-psychotherapeutic model. IDD and old age can, in many ways, be an issue for and during psychological interventions. Major issues linked to IDD have been addressed earlier [12]. More recently, Beail and Jahoda [13] offer a broad overview on those elements to reflect and to consider for direct psychological interventions in people with IDD or when working relationally with adults with IDD [14]. Overall, creative adaptions of techniques and settings, as well as openness towards non-conventional communication channels create unique opportunities and possibilities for psychotherapy in the IDD population [11]. Finally, Whitehouse and colleagues [15] offered a comparative review of cognitive–behavioural and psychodynamic findings to adapting individual psychotherapies for adults with IDD.

Limitations in Communication

A limited basis for communication, typical for people with IDD, will first be addressed. Communications competencies depend on multiple factors, such as the person's intellectual-cognitive level, the verbal and non-verbal expressiveness, and the general developmental level of the person. The etiological background of the disability might be considered for additional understanding of specific communicative features. In general, the therapist should use plain syntax when addressing his or her client. In adult and especially older people with IDD, initiating communication can take longer (i.e. several sessions might be needed for developing a relationship between therapist and client for assuring a solid therapeutic basis). Further, non-verbal communication signs, like quality of eye contacts as well as gaze expression, have to be prominently considered. When explaining something or inquiring, it is important to make use of simple examples that are understandable and meaningful to the client. The therapist is advised not only to make sure and to inquire about the client's understanding within the session but also to refer in later sessions to options, intentions and objectives formerly discussed or defined.

Lack of Goal-Orientation

The ‘rehearsal’ strategy points to a second characteristic common in people with IDD, which is the lack of goal-orientation. Many adults with IDD and especially older adults with IDD show difficulties in remembering the reason why they are meeting the therapist, or the reason remains unclear to them from session to session. This very often calls for a directive approach by the psychologist, in order to get the adult re-informed about the course and the objectives of the therapy plan, when addressing the client according to his or her perceptive and intellectual grasp. In addition, it will be important to point to the rules of the therapy, stick to them, and make sure the client ‘plays the game’. Aggressive behaviour as well as behaviour of extreme attachment, which are both reported in older people with IDD, can best be handled when addressed directly [16]. In such cases, the therapist would clearly point to the limits and inappropriateness of such behaviour, though recognizing the client’s current emotional need, and elaborate with the client on alternative behaviour.

Lack of Self-Initiating Communication

A more directive approach might also be adopted to face the lack of self- initiative in communication reported to be especially common in older adults with IDD. The therapist might encourage such adults to think about questions or evoke their attention by stimulating some relevant events. Once the client raises questions it is imperative to respond to these immediately by answering in a clear and simple way. One can proceed on the assumption that older people with IDD have little experience with this kind of very personal communication, engaging however with ongoing therapy in this mode. As the therapy advances, adult clients can more frequently be asked about their coping with specific situations and problems. This helps to strengthen the client’s competences in problem solving (e.g. his/her coping strategies in stressful events and situations with high emotional loads).

Alternative Approaches

Adaptations in Therapeutic Techniques

Often the therapy is at risk of getting paralyzed. Rigidity in the therapy can be overcome by adaptations of therapeutic techniques. A greater flexibility in techniques, methods and settings, without changing the objectives of the therapy, is recommended in such a situation. However, it should be considered whether the selected methods and techniques are conforming both to the mental and chronological age of

the adult. Length and frequency of sessions are largely determined by the adult's endurance and attention. In general, psychotherapeutic sessions for people with IDD are of shorter duration and are more compactly scheduled as compared to sessions with adults without IDD. For older people with IDD sessions of 20–25 min might be enough while meeting twice a week.

Use of Third-Party Informants

In contrast to most persons without IDD, adults with IDD frequently rely on information of third parties. As people grow older, the need for information from third parties—such as family members, staff or other caregivers—might increase, due to memory losses associated with Alzheimer's dementia or other cognitive dysfunctions. A rule of thumb is not to assign an informant in order to achieve the therapy objective. However, if an informant is required in order to keep the therapy process going, it should be assured that the confidential and trustful relation between client and therapist is not violated. As a matter of fact, therapies conducted in an isolated form (i.e. without indirect contacts to a client's environment) have been shown often to be not effective [15].

Disability as a Reality in Therapy

Independent of the clinical symptoms that might have led the older individual with IDD to enter psychotherapy, for most of the older adults their disabilities remain an issue. A number will raise this issue during psychotherapeutic intervention. For instance, some researchers [15] recommend that the adult first develops an awareness of his or her disability, thus making the understanding of the disability a prerequisite for a therapeutic intervention. An awareness of the disability and concurrent stigma experience might especially apply for people with limited needs of support (such as mild disability) [17]. This virtually requires special guidance and counselling related to the issue of disability. In the case of older adults with IDD this guidance often might be a reflection across the individual's lifespan and his or her experiences as a person with a disability. In this case, counselling should not only focus on areas of personal weakness linked to disability but should also point strongly to the personal strengths developed during the life span and relate to the disability. Besides raising issues of disability, older people with mild to moderate disability might experience further difficulties with advancing age. Besides age-related morbidity and co-morbidity they might express concerns over anticipating additional experiences of social rejection and non-acceptance as they grow older. For the psychologist and therapist, it is good to remember the high sensibility of people with IDD toward social non-acceptance as reported in literature [18]. Finally, challenging behaviour of clients with high difficulties to verbally express their

views can be proxy behaviour struggling to establish or maintain positive social identity is in not a few cases can impair their mental health.

Therapists' Attitudes

Distorted attitudes either toward disabilities and/or toward old age might be an additional reason for undesirable effects during therapy. A therapist's personal attitudes and views often influence the relation between client and therapist, and hence can affect the course of the therapy. A typical distortion in a therapist's attitudes might be his or her overprotective or paternalistic views. Additional sources for dissonances in the relation between client and therapist might be a therapist's state of general discomfort while working with the client. Though signs of dissonance might only be present in very subtle forms, clients are generally highly sensitive in capturing these. Therapists should avoid over-estimating the clinical symptoms, and are recommended to focus more on progresses, even the smallest. This contributes to the establishment, the strengthening or the re-establishment of behaviour and attitudes promoting relative independence and self-determination in old age. The therapist, in general, being younger than the older patient with IDD, patronizing attitudes might play a crucial role, especially for an older client with IDD, then losing motivation and abandoning therapy.

Finally, issues like therapists' attitudes or disability as a reality in therapy are major events during a psychotherapy, with only recently upcoming contributions in psychotherapy process research in the field of IDD [19].

Therapists' Orientation

For psychotherapeutic interventions with people with IDD to become a success, McGee and Menolascino [20] have emphasized the importance of moving from personal disconnectedness to human engagement. Especially when conducting behavioural interventions, a sensitive orientation toward the client that emerges from attachment theory should be maintained during the process. This is particularly important when dealing with adults with autistic symptoms, and older persons with IDD. Promoting and maintaining a mutual relationship that is based on the principle of empathy is crucial for the intervention. In general, the recommended approach is human engagement, with the goal of establishing attachment, also referred to as the gentle teaching program. Gentle teaching, though having its origins when addressing emotional problems in the younger population, asks for being unconditional, meaning that our attitude toward a person is not depending on his or her behaviours [21]. This approach is independent of the client's age. Thus, regardless to what the person is addressing or doing, the therapist will keep his promises and will give him the help and protection he needs. While opting for this orientation,

the adult is taught that human presence signifies safety, consistency and positive interaction or reward.

Motivational Issues in Old Age

In addition to the characteristics mentioned for IDD, older people with IDD, just like their age-peers without IDD, are often indifferent or even reluctant toward psychotherapeutic interventions, though they may show symptoms which according to experts definitely require treatment. However, some adults might not be ready for a 'traditional' psychological intervention if they are not fully aware of their mental health state or if they are not motivated for psychological treatment (that is, if they have negative expectations with respect to the effects of treatment on their lives or just reject or quit therapy on the basis that it is not worthwhile), or they experience therapy to be too exhausting and hard for their age. This motivational gap is well documented for the general older population, showing an ongoing motivational decline as people are growing older [22]. Designing strategies for enhancing motivation toward psychological interventions in old age is a major challenge for improving quality of life in life's last stage. However, successful motivational strategies might need age-adjusted adaptations in 'traditional' therapeutic techniques as well tuning the contents of psychological support toward those subjective needs that the individual expresses in old age. For example, instead of approaching an anxiety disorder with the traditional classical behavioural or cognitive-behavioural setting, an older person might be highly motivated for reviewing his or her most meaningful episodes of life in a systematic way. This intervention alone, with higher face validity for the client, might be a more appropriate approach for raising the adult's readiness to engage in a therapy. Deducing a general framework for psychotherapy in old age, independent of the psychotherapy 'school', might help raise the motivation for psychological support especially in older people with IDD.

A General Model for Psychotherapy in Old Age

The founders of classical psychotherapies largely concentrated their attention on young to middle-aged adults, later focusing also on childhood and youth. The theoretical models and intervention techniques often neglected older people, with some schools (e.g. classical psycho-analysis) directly stating the therapy not to be appropriate for older people. Behaviour therapy and cognitive therapy though extensively developed in an evidence-based way for applications with people with intellectual and developmental disabilities since the early years of the 1990s, were nevertheless mainly aimed at children, adolescents and young adults [23]. In general, the purpose of psychotherapy is modifying or removing existing symptoms and promoting personality growth. With most schools of psychotherapy assuming the roots for major

psychological processes to be in the development of early childhood, and in the family history, or that behaviour is largely based on learning processes and can be environmentally controlled, the schools in their early years did not, in general, focus on older people. Rather, old age was seen as a period with declining and ending development, with learning processes becoming more and more irrelevant while growing older. This view is best expressed in some of the early ageing theories, nowadays often criticized within the ageism discourse, with theoretical models more recently popping up, reflecting both strengths and challenges typical for the late developmental phase.

Emerging Geronto-Psychotherapeutic Model

A first psychological developmental model considering old age was proposed by Erikson [24]. His eight-phased model relates from infancy to old age. In old age, developmental processes are assumed to be competing between two contrasting outcomes—integrity versus despair. Erikson's model suggested major developmental stages after youth, as outlined in his later work [25]. Erikson's eight developmental stage model enjoys continuing consideration in psychological research, with specific attention to vital involvement in old age [26].

Cohort-Based, Context, Maturity, Specific Challenge (CCMSC) Model

In modern gerontology there is a broad agreement for research on life-span approaches. Only recently a framework based on a life-span approach has been proposed in psychotherapy by Knight [27]. Knight's CCMSC model offers a framework for adapting psychotherapy to the work with older people. The model is based on methods, concepts and recent findings in psychology and on research outcomes from the gerontology life-span approach. The model is designed to be trans-theoretical, i.e. it allows implications for various theories and schools of psychotherapy.

The model outlines four 'factors':

1. The cohort factor considers, besides an individual's cognitive performance, verbal fluency and his education career, the person's normative course of life and life-experiences from a social-historical point of view
2. Context effects refer mainly to current environmental characteristics such as age-adapted accommodations, residential facilities for senior citizens and spare time

options for older persons, as well as to general health and long-time care provisions and to old-age specific acts like regulations related to social or health insurance
3. Maturity, deals with gathering information on the person's cognitive and emotional complexity, his or her post-formal reasoning, including the areas of expertise and competencies, including experiences in family life and the person's accumulated inter-personal competencies
4. Dealing with specific challenges, includes areas such as chronic diseases, disabilities, and grieving while experiencing deaths of relatives and friends, as well as the person dealing with their own end-of-life, including preparation for death

Up to now the CCMSC model has generated old age specific therapeutic principles for various schools of psychotherapy, like behaviour therapy [28], cognitive-behavioural interventions [29], systemic family therapy [30] and psychodynamic therapy [31, 32]. Finally, a CCMSC for older people with IDD would consider specifically on trauma during the life-course of people with IDD.

Model Adaptions for Older People with IDD

The model offers a common basis for defining requirements related to change and adaptation in old age. However, applying the CCMSC model in the field of IDD requires specific complementation, which will contribute to a meta-theoretical framework in guiding an integrated psychotherapy approach for the ageing population [33].

Maturity Effects

Referring to the element of maturity it seems that many psychological variables show a high level of continuity and stability during adulthood of people with IDD (e.g. personality, crystallized intelligence), whereas other areas are declining in their functioning similarly to the general population (e.g. fluid intelligence, capacity of working memory, audition, and vision [34]). However, opportunities for extensive vocational and family experiences are in general limited during the adulthood of people with IDD. Accumulated expertise and competences may just be limited in the same way, thus giving a clear disadvantage for older people with IDD for effective coping in their later years. Thus, in old age, people with IDD might show similar difficulties when coping with a depression as in younger age.

Cohort Effects

Cohort effects might be different to those of the general population. Many people of 50 years and older at the beginning have a record of living many of their adult years either in institutions or in their family of origin. The personal identity might be more imprinted by these circumstances than by typical identity cohort effects like marriage, fatherhood and motherhood in the general older population. Requirements for a therapist will be—besides the understanding of verbal codes typical for specific cohorts—the understanding of differences between cohorts, and acceptance of the adults' accounts related to important events in their lives. Cohort effects are important for understanding onset, development and maintenance of a depression.

Environmental Effects

For older people with IDD the effects of the circumstances and environments they are living in as older people (i.e. the effect of context) might not be that important, as for older-age peers in the general population. People with IDD in general live in supported and assisted accommodation settings of various community-based degrees for most of their lives. However, when living in larger old-age residential facilities, adults might have been imprinted by institutional characteristics. Permanent or ongoing confrontation with their peers' chronic diseases or multiple disabilities might play a role for triggering condition, such as depression. After moving to new accommodations, syndromes related to relocation in old age and behavioural and/or mood adaptation might be present for more than a year.

Co-morbidity Effects

Depression might be a common co-morbidity when there are major health problems, or long-term hospitalisation is required, or if the adult suffers from chronic diseases. Older people with IDD, who may develop dementia, might show increases in behaviour disorders or abnormal behavioural signs like aggressiveness, and delusions or hallucinations, and if relocated to nursing homes these signs might increase. In general, consideration of the clinical record is needed during the first assessment, as for older people with IDD it might be that high blood pressure, diseases of the cardiac system or diabetes can go along with co-morbidities of mental disorders [35]. In general, in old age various reasons may play a role when the emotional balance for people with IDD gets disturbed (e.g. chronic disease, personal loss, neurodegenerative processes, additional disabilities, age-related attitudes, attitudes related to the original or primary disability).

Considerations for Psychological Interventions

When determining and selecting the therapeutic approach, the cognitive functioning of the older person must be considered first. For some methods good functioning of short-term memory is needed, as information has to be integrated during the session. When opting for a group therapy, the group should be homogeneous with respect to the level of cognitive functioning and communicative skills, as more capable adults might feel they are not being taken seriously. Further, digital and smart communication technologies can open up new possibilities and better access to psychological therapy for those people with IDD who are more and more familiar with this kind of technology [36].

Institutional-Based Therapists

When working as a therapist in a residential facility for older people with IDD (for example, residential homes or nursing homes), a certain flexibility within the professional's role should be the norm. In many cases, the professional might be obliged to act on behalf of his/her client's interest without forgetting the needs of staff supporting the adult. Cognitive and behavioural modification techniques have been reported to be quite effective within residential settings [9]. However, the success of such interventions is highly dependent on staff compliance to the therapeutic process. Hence, discussing and designing a plan with activities agreeable for the older person and the staff should be the first step before starting a therapy. Next might be preparing staff with respect to the therapy process, thus assuring coherence between professional acts of staff and therapist. Next would include recording a baseline, expressed in the frequency of agreeable and non-agreeable activities, as well as of behaviour problems. This will be followed by the determination of priorities for areas of change. These areas might in addition be processed by functional analysis, thus getting first information on possible contingencies related to target behaviour. Finally, a realistic plan for change has to be developed, including a concept for evaluating the intended therapy goal. These steps largely follow a traditional behavioural intervention strategy.

Combining Therapeutic Approaches

It has been reported that using behaviourally-based therapies mixed with so-called 'simulated presence therapy', and/or 'individualized music therapy', with older people in general resulted in higher ratings for efficacy [37]. During individualized music therapy, the client actively listens to his preferred pieces of music. In simulated presence therapy, the client listens to prepared audio sequences offering him

or her selected segments of his memory which are known for their calming and comforting effects on the individual, thus raising the client's participation for stimulating activities and allowing him or her an uplift in affect. It is thus assumed that combined and integrated therapies might show similar effects when applied to select older people with IDD, as some are reported to have high affinities to rhythm and music [38]. Within the general population of people with dementia music therapy as well as chair yoga have demonstrated positive effects on participants mood and social skills [39], with chair yoga having in addition a positive effect on reducing the risk for falls in this population [40].

Behavioural Interventions and Specific Disorders

As reviewed in chapter "Bereavement and Grief Reactions and End of Life", therapies and interventions based on behavioural concepts including cognitive behaviour therapy and adapted dialectic behaviour therapy have been tested and applied to a large variety of disorders found in adults; however, these have been applied mostly to younger adults with IDD. Conditions cited in the literature include anxiety and mood disorders, schizophrenia, stress disorders and sleep disorders. For people with severe and profound IDD, interventions are reported for stereotypic behaviour, problem behaviour, self-destructive behaviour, aggressive and destructive behaviour and challenging behaviour as well fits of rage or tantrums [41–44]. Behaviour therapies either use positive reinforcement techniques for stimulating desirable social behaviour, or/and aim at reducing problem behaviour by referring to aversive behaviour modification techniques. Since the mid-1980s, the emphasis has clearly shifted to positive reinforcement techniques, primarily using proactive and ecological strategies to prevent behaviour problems [45]. Further cognitive behaviour therapy has largely been tested in the IDD population with verbal communication with therapists' manuals available [46].

Behaviour modification procedures can be grouped in several types of differential reinforcement techniques, with the differential reinforcement of other behaviour (DRO) being one of the most popular strategies for decreasing maladaptive and aberrant behaviour and improving more socially acceptable behaviour. As part of this, functional assessment is a central part of the management of behaviour problems, as it aims at determining the multiple conditions that maintain aberrant behaviour [47].

Functional Communication Training

Results, generated through functional assessment, indicate self-injurious behaviour, for instance, to be an aberrant form of communication. Based on these findings, functional communication training has been developed and tested in many IDD subgroups including those with severe IDD since the 1980s. Results indicate that FCT is

effective in decreasing the level and frequency of problem behaviour and increasing the level of appropriate alternative communication response. However, effectiveness varies according to factors as disability type and age, with higher effectiveness in people with autism spectrum disorder and IDD as people with IDD [48, 49].

Anger Management Training

Through anger management training introduced by Benson [50], adults with IDD are taught better management of their tantrums and aggressive behaviours. This training refers to—besides techniques such as self-instruction—relaxation techniques, identification of emotions and problem-solving strategies. Well-designed studies revealed relaxation techniques and self-monitoring to be highly effective strategies for anger control [51, 52]. Behaviourally-based interventions, such as cognitive behavioural treatment and positive behaviour support are known to be highly effective, with outcome effectiveness rates varying between 40% and 75% among various behaviour problems [53, 54].

Severe Communication Disabilities

When therapeutically approaching adults with IDD who are non-verbal or have severe communication impairments, behavioural techniques (as described above) are highly popular. Further, a therapist might first screen for other modes of communication of the adult, adapting the communication to the individual's full communication capabilities before starting therapy. Known as Augmentative and Alternative Communication (AAC), this approach screens for any residual speech or vocalisations, gestures, signs and areas for aided communication [55]. This means that AAC techniques act as a gate to communicate with people with severe to profound IDD (people requiring pervasive support), thus enabling additional opportunities for psychological interventions [56]. Communication with gestures and other natural modes is known to be helpful for interacting and building relationships, with the effectiveness of AAC having been underpinned by meta-analysis [57]. Additional alternative therapies, such as Snoezelen or multi-sensory stimulation therapy and active therapy, have been employed with people with profound IDD, showing increases in positive communication and decreased agitation levels [58, 59].

Pre-therapy

A further asset in psychological interventions is Prouty's pre-therapy. Derived from the person-centred approach, and originally developed for interventions with schizophrenic patients, it has been applied for treating emotional disorders in

non-verbal adults with IDD or with severe contact disturbances, thus making the client accessible to more regular kinds of therapy [60]. Essentially, the therapist sets up contact by repeating gestures, vocalisations and body postures of the adult, thus offering reflections. Pre-therapy aims at restoring or developing an adult's contact functions (that is, either his contact to reality, his affective contact or his communicative contact). The effects of reflection on contact functions can be observed and measured. Changes in an adult's behaviour are interpreted in terms of increases in interpersonal communication. Referring on the one hand to techniques related to AAC, while on the other hand following the principles of client-centred therapies, pre-therapy is shown to be a highly promising approach for building a therapeutic communicative basis in adults with IDD showing mute behaviour [61].

Interventions Related to Specific Situations in Old Age

Techniques which appear to be effective in older adults include relaxation training, featuring pleasant events, behaviour modification based on reinforcement plans, and cognitive restructuring, as well as emotional processing based on expressing and naming emotion [33]. Whereas emotional processing is recommended for improving the general emotional well-being, the other techniques are applied to specific states. Most of the techniques reported in this section require a minimum of verbal communication skills, present in general in people with mild to moderate IDD.

Relaxation Techniques

Relaxation training, especially progressive muscle relaxation is indicated for anxiety symptoms related to chronic diseases and related anxiety symptoms to other therapeutic interventions [62] Instructions for muscle relaxation have to be adapted for those muscle groups that might be involved in the adult's present clinical pain record. A script for old-age adapted relaxation training can be found in other literature [27]. In addition, relaxation techniques can be used as additional treatment for memory impairment and controlling pain.

Behavioural and Cognitive Reshaping

Behavioural reinforcement or cognitive restructuring procedures are recommended in older age, especially for improving the emotional and affective state following diseases or loss of functional competences [63]. In cases of disease and additional disability, older people with IDD, just as peers in the general ageing population,

show depressive reactions and depressions. These symptoms diminish in frequency and gravity as these people participate in pleasant and/or meaningful activities. During the therapeutic process, areas of activities meaningful to the person prior to the disease or functional loss should be explored, and adaptations for activities to be maintained or replacement activities outlined and defined with the adult. This would be followed by a stepwise implementation plan. For a good understanding of depression in situations of specific challenge, a therapist needs, in addition to his/her knowledge about various therapeutic techniques, a sound knowledge about the adult's condition and the relation between the condition and his or her age.

Grief Therapy

Grief therapy is reviewed in detail in chapter "Successful and Positive Ageing". It aims to offer emotional support to adults after the loss of close family members and friends. Though mourning after death of a loved one is a normal reaction, people with IDD often show prolonged mourning reactions. Mourning is characterized by depressive and anxiety symptoms and/or by a mixed affective state combined with behaviour problems, with major depressions occurring occasionally. After loss of several close persons within a short time, mourning reactions are shown to be especially intense. Older persons with IDD who continued to live with their parents are generally confronted with a change of residence in the case of death of their parents.

This extra life-event brings an additional load to the mourning process and has to be considered during grief therapy. Adults and older people with IDD may over years show repeated depressive reactions related to the death of a loved one [64]. Such reactions point either to the fact that the mourning process has not been successfully completed and/or to a lack of adequate coping mechanisms in cases of loss of a close person. Besides expressing emotions, grief therapy also focuses on developing an understanding for viewing the loss in a longer perspective, and on offering support and active encouragement for the development of perspectives for life without the loved person.

Life-Review Therapy

In general, gerontological life-review therapies have been applied since the early 1960s [65]. Life-review therapy is offered in either a structured or an unstructured way, and life-review therapy is referred to when applied in the context of a mental disorder [66]. In the general population of older adults, life-review therapy has been shown to be effective in the treatment of depressions, and of special effectiveness when dealing with post-traumatic stress disorders. Reasons for this effectiveness are thought to be related to the fact that the traumatic event often overshadows other positive life-events, thus preventing a balanced review for an older person. Memories

related to a traumatic event often lack consistency and appear in a fragmentary form. This indicates incomplete coping processes. From trauma-based memory research it is known that pending affairs are better remembered then settled matters or other autobiographical events. In addition, traumatic events are often not following the way autobiographic memory traces are processed and structured. Life-review or reminiscence work as a therapy aids in more successful coping and adjusts autobiographic memory, and thus contributes to a differentiated self-assessment of a person's life [67].

Psychotherapeutic Interventions for Persons with Alzheimer's Disease

People with IDD and especially people with Down syndrome while growing older, have a higher risk than their age peers of being affected by Alzheimer's disease. This process goes along with changes in emotional functioning [68], among others. In the early phase, psychological interventions aim at stabilizing the person's decline of independence in various areas. A structured daily activity plan might be required, and the use of external memory supports might help the person to better perform activities of everyday life. Further, the expression of emotions related to the changes the person is experiencing might help to balance the affective state.

As the neurodegenerative process is progressing, emotion-centred therapies, such as validation therapy according to Naomi Feil, enjoy widespread application. As one of the core techniques in validation therapy, highly confused verbal production or behaviour is understood as emotional communication and messages, with the factual content being given no major relevance. The fact that the messages of many patients with Alzheimer's disease refer to their parents is interpreted as a special need for affection and attachment. The gap between the propagation of validation therapy and empirical evaluation is reducing [69]. However, in the field of intellectual disabilities validation therapy might receive growing attention as professionals see more reasons for emotional-centred assistance, as compared with ameliorating cognitive functioning, with the knowledge that there is no remedy for stopping cognitive decline in patients with dementia [70].

Supporting Mental Health

Maintaining mental health in balance is one of the key issues for a high quality of life in older age. Supporting quality of life in older age for people with intellectual disabilities might start years before the individual gets in his or her fifties. Factors such as former lifestyle or former episodes with mental health problems are known to affect a person's quality of life in later years. However, for people with

intellectual disabilities a preparation on ageing issues might beneficially contribute for their later years.

Person-Centred Planning for Later Life

As people with IDD in general show low degrees of self-initiated anticipatory analysis related to their future, or rarely reflect on upcoming events and their effects on their life, a systematic discussion of situations to be expected and an examination of the personal wishes and goals might contribute to entering older age with good prospects from a mental health perspective. Increasing adults' awareness of retirement issues and of later life is the aim of programs as outlined in the Person-Centred Later Life Planning Project [71] based on Person-Centred Planning methodology (PCP) [72, 73].PCP, strongly based within the social model of disability, is a set of approaches designed to assist a person to plan his or her life and supports. It aims at empowering people with IDD, supports them in their choice making and promotes growing relationship. PCP encourages being responsive to the needs of persons with IDD rather than prescriptive in the types of services offered. PCP is accepted as evidence-based practice throughout the world.

The Person-Centred Later Life Program offers training in goal planning, choice and decision-making, and it aims at increasing awareness of options for sound health and wellness. Further, issues such as leisure activities, work after retirement, living arrangements, accommodation and friendships can be discussed in a systematic way in this training designed for groups. A study on the program's effectiveness showed increased knowledge of later-life options and increased participation for individuals who were living at home [74, 75]. In addition, the program includes training components for staff and families on later-life planning issues.

Finally, healthy ageing, as promoted by WHO global ageing agenda is reflected with regard to people with IDD's later life, with the perceptions of self-advocates and their families towards their later life being the centre of attention [76–78].

Retirement Programs

Though often there are few policies or supports that permit people with IDD to officially retire from their 'work', people with IDD do retire from those programs that they used to be in over many years. Preparing people with IDD for retirement and offering adequate retirement programs is a central issue [79]. During the transition to retirement people with IDD are at risk for decreased social integration, with decline in self-esteem and a reduced range of activities. Changes in these factors might have an impact on the mental health status of ageing adults.

Recreational Leisure Programs

Recreational leisure programs adapted to old age might offer older individuals with IDD additional protective effects for their mental health while ageing. Such programs generally aim to increase community inclusion, while providing support for those interests and activities that are meaningful to the individual. Recreational programs vary from group programs to more individualized forms, including the person's informal network [80, 81].

Reminiscence Projects

Reminiscence projects focus on "remembering memorable events and experiences of long ago". Some older people with IDD show interest or have the need to share those moments and events of their life, which have been meaningful to them. Reviewing with an individual, his or her life might help the person develop integrity and well-balanced attitudes with respect to his past life and experiences thus contributing to his or her ageing successfully. The importance of this approach for the field of IDD has been emphasized [82]. Life-review can contribute to maintaining the well-being of the person, as opposed to re-establishing mental health when life-review is used as therapy in mental disorders. For effectively conducting a life-review program or a biography project, information and biographic material related to the person's life is highly useful. Unfortunately, often staff members have little information about an older person's earlier life episodes as well as life events meaningful to them. Thus, for future use, in particular with applications to successfully reconstructing autobiographic memory traces, adults with IDD and their families should be encouraged to keep a biographic or autobiographic record, including happy and less-happy moments and valued and less-valued episodes of their lives. The record might include notes, letters, cards, photos, short videos and personal items, thus offering material outreaching the information of traditional medical or institutional records [83]. The availability of such a biographic record might be a useful resource for maintaining an individual's wellbeing in old age and allow staff to better understand the life trajectories of those persons with IDD they are assisting in old age.

Carers' Emotional Well-Being

Caring for an older person with IDD, especially when that person suffers in addition from dementia, can be a high burden for the staff members and other carers. This topic is reviewed in depth in chapter "Legal, Ethical and Due Process Issues". The carer's burden (perceived or real) often results in depression, anxiety, irritation and

frustration. Changes in carers' emotional well-being might be accompanied with various effects on their environment, including the quality of life of the older person with IDD. Therefore, special techniques should be used by frontline staff and carers to prevent distortions in their own emotional well-being. Techniques that staff might use include collecting information of the process specific to the disease, enhancing one's capabilities to cope with stress (i.e. making appropriate use of relaxation techniques, getting emotional support in supervision sessions, and allowing oneself more frequently agreeable events [84]). Further it will be of need to target at the actual needs of family carers as they themselves are ageing while continuing to take care of their family member with IDD in the community [85]. Considering the special needs of the carer may contribute positively to the emotional wellbeing of the person for whom care is provided. Finally, when caring for older people with IDD and dementia, professionals not only have to adjust their support in terms of identifying and responding to the needs of the persons they support but also through collaborative team working within and across services [86].

Conclusion

Psychological interventions and/or psychotherapy offer a wide variety of techniques with promising use in ageing and older people with IDD and carers. However, while reviewing recent developments from the field of geronto-psychotherapy, and considering advances of psychotherapy in IDD, a clear need for combined research between these two areas is evident. Substantial advances in research might require a general model for psychological interventions in people with IDD. The observations of a tendency for convergence between the techniques of different schools of psychotherapy when applied in the field of IDD [12], as well as the growing agreement on a common basic attitude for such interventions, might be a suitable starting point for shaping such a theoretical model [9]. Indeed, besides major contributions from behaviour and cognitive therapy, psychodynamic approaches of psychotherapy show since the 2000s an increasing number of reports focusing on therapies with adults with IDD, with a spectrum of problems that can be addressed [9, 10, 23, 31]. The convergence hypotheses between therapeutic techniques of different schools find support when observing psychoanalytic and humanistic approaches to integrate more directive techniques in their settings, so getting very close to or even directly referring to behaviour techniques [87]. In addition, recent advances from frameworks in geronto-psychotherapy for the general population, including an emotional-developmental perspective, might be considered to specify psychological interventions for older people with IDD. Subsequently well-designed evaluation studies would follow, as there is particularly a need for randomised controlled trials (RCTs), and a need for better specification of treatments (e.g. manualised protocols), and better outcome measures. Finally, clinical trials need to make use of improved reporting standards larger samples, and multi-center studies are recommended for addressing the challenges of RCT studies in this population.

Concluding, there is evidence that psychotherapy with adult and older persons with intellectual disabilities is at least moderately effective [10]. Further, there is evidence that a range of therapeutic interventions is effective, with behavioural, humanistic as well as psychodynamic techniques showing good outcomes in the population with mild to moderate IDD, and with cognitive-behaviour therapy (CBT) showing specifically good outcomes for both anger and depression [36]. In general, adults and older people with IDD and concurrent mental health problems appear to benefit from psychological interventions. In the end, future efforts focusing on the development of mental health promotion and mental health literacy programs appear promising for empowering this highly vulnerable population.

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Chapter 12
Role of Psychotropic Medications in Older Adults

Elizabeth Jane Evans and Julian Norman Trollor

Introduction

Psychotropic medications are those which affect the nervous system, with a resulting impact on cognition, behaviours or emotions. Under traditional classification systems, such drugs have been subcategorised based on their therapeutic action: stimulants, antidepressants, hypnotics/sedatives, antipsychotics, mood stabilisers, anti-anxiety agents, movement disorder medications, anticonvulsants, anti-emetics and anti-nauseants, and others, which includes anti-dementia drugs [1]. Some drugs have multiple therapeutic effects [2, 3], and the current nomenclature for psychotropics includes many inconsistencies [4]. Therefore, more recently, an alternative classification has been proposed, based on the neuropsychopharmacological action of each drug [2, 5]. This neuroscience-based nomenclature imparts to the prescribing clinician a rationale for a particular medication [5].

When they are used appropriately, psychotropic medications offer substantial benefit to people affected by mental disorders. However, when used inappropriately psychotropics can induce serious side effects, and the risk of these may be amplified in older people [6]. Adults with intellectual and developmental disabilities (IDD) are known to be at higher risk of undiagnosed health conditions, multimorbidity and polypharmacy. Therefore, older adults with IDD fall at the intersection of two groups at higher risk of adverse events. Furthermore, psychotropic medications themselves impose an additional risk for morbidity [7].

In recent years, structured protocols have been developed to redress inappropriate prescribing to the general population aged over 65 [8, 9]. However, despite

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successfully reducing medication use, they have not been shown to improve clinical outcomes or be cost-effective [10]. However, instruments to detect inappropriate medications in older adults are not readily applicable to people with IDD, who have different patterns of both multi-morbidity and medication use [11, 12]. Moreover, a different age threshold for such medication review may be needed for people with IDD, who show increased risk of frailty at younger ages [13, 14]. An individual approach is therefore required [15], and for people with IDD and substantial polypharmacy, medication reviews and rationalisation may offer potential benefits.

Clinicians working with elders with IDD therefore require a thorough understanding of the risks and benefits of psychotropic medications, clarity regarding when to recommend their use, and skills in prescribing and monitoring their safety and efficacy in this population. Here, we discuss the additional complexities to consider when making decisions regarding psychotropic medications for adults with IDD, before outlining principles good practice for psychotropic use in this group. We will not discuss anti-dementia drugs here, as this issue is covered in chapter "Role of Anti-dementia Medication".

Considerations When Prescribing to Older Adults with IDD

Prescribing to older adults with IDD is made more complex by a range of additional and interacting risk factors experienced by people in this group.

Multimorbidity and Premature Frailty

People with IDD of all ages experience mental health problems at a rate that is higher than the general population [16, 17], and these may escalate with advancing age [18]. Adults with IDD also experience age-related health disorders at a higher rate and early age than those without IDD. This includes seizure disorders [19] the dementias [20–22] and falls [23].

The high rates of medical and psychiatric comorbidities in older adults with IDD includes both diagnosed and undiagnosed medical conditions [24–26], with substantial multi-morbidity [27]. Adults with IDD also show higher rates of premature frailty [13, 14, 28]. Frailty can involve both physical, psychological, and social elements [29, 30]. Physical changes in frail individuals are underpinned by altered physiology, which reduces the body's capacity to maintain or regain homeostasis [31]. Frailty can result in decreased mass and strength, reduced appetite, increased risks of falls and osteoporosis, and a high vulnerability to health conditions following even minor stressors [32, 33]. In older adults with IDD, frailty is associated with increased medication use [34]; long-term care admission [35]; and mortality [36]. These factors likely increase the risk of adverse drug reactions for older people with

IDD, and underscore the importance of thorough assessment and of prescribing recommendations that consider multimorbidity and frailty.

Atypical Patterns of Medication Use

Adults with IDD are prescribed psychotropic medications at high rate, and polypharmacy is common [37–45]. Use of antipsychotics is particularly common in people with IDD [46–48]. Of relevance to older adults with IDD is the high anticholinergic burden [49] which exposes the person to an increased risk of adverse drug reactions [12]. Some increase in medication use is to be expected, given that this group also experiences higher rates of mental illness and seizure disorders. However, adults with IDD are frequently prescribed psychotropic drugs in the absence of a definitive indication or diagnosis [46, 50], and without adequate monitoring or review [44, 51]. These inappropriate practices increase risk of adverse drug reactions, may complicate subsequent assessment and are best avoided, especially in the elderly.

Specific data on prescribing in elders with IDD also suggests an unusual profile of medication use. Older people with IDD have been found to take a greater number of both medications and supplements than older people without IDD [52]; including more psychotropic medications (see also 11 for a review, 46, 53, 54). They show high rates of polypharmacy [12], including psychotropics [55]. By contrast, they may be less likely to receive preventative medications [54, 56] or pain management [12, 54]. Such data highlight health gaps for older people with IDD, particularly in health screening, and access to preventative healthcare and to tailored mental health care.

A Less Substantial Evidence-Base Is Available to Inform Prescribing

People with IDD are often excluded from research [11, 57]. Many clinical trials also systematically exclude older people [58]. This means that for many drugs there is no established efficacy and safety profile in adults with IDD, particularly older adults. As de Leon [59] points out, randomised controlled trials are unlikely to ever be funded for smaller subgroups, and furthermore, systematic reviews based on scant available research are likely to favour older medications over newer ones.

In recent years, specific practice guidelines have been developed to guide treatment of mental disorders and management of behaviours of concern in people with IDD [15, 60, 61]. However, these guidelines contain little or no information specific to older adults with IDD [47]. For some specific drugs or drug classes, practice guidelines for people with IDD may reference information relating to

older age [59, 62], but the available evidence is scant and drawn from studies of the elderly without IDD.

In this context, the best available evidence about when and how to use many psychotropic medications will be that available for the general population [63], and clinicians must acknowledge that the risks of a particular drug or class of drugs cannot be fully known for this group. Through consultation with the person and their carers, they must carefully weigh and communicate the potential benefits against the potential risks for the patient's health, and impact of uncontrolled symptoms on wellbeing.

Additional Challenges for Assessing Both Symptoms and Response to Treatment

There are also difficulties inherent in assessing both symptoms and response to treatment in adults of any age with IDD, including reduced verbal skills and capacity to introspect regarding symptoms [15, 64], and an increased reliance on carer reports [65]. An increased likelihood of atypical presentation of mental disorders also complicates both the initial assessment and symptom monitoring.

Contextual Issues in Supporting Elders with IDD

Several contextual factors create additional complexity. People with IDD, including elders, have reduced access to appropriate health care, with lower utilisation of some preventative care [66–68], including preventative medications [54, 56]. This means that detection of potential baseline treatment risks, and of side effects during treatment, may be less likely to occur.

Access to mental health care for those with more severe disability requires carers to recognise symptoms and liaise with healthcare providers, but many carers are ill-equipped for this task. Staff in either aged or disability care receive minimal training in mental disorders, and staff in either sector receive little or no training in the other sector. Carers require a good knowledge of a person to support the identification of symptoms, and both the beneficial and detrimental effects of treatment. However, the care context is often characterised by changes in carers, which jeopardises this process. Furthermore, older adults with IDD may enter residential care following a series of losses, including loss of a family carer, independence and community contacts. Issues of adjustment and grief may compound assessment of mental health, and staff in the new care context may lack sufficient knowledge of the person to inform approaches to treatment.

Such contextual complexities potentially expose elders with IDD to poorly tailored prescribing and adverse events. For this reason, clinicians must adopt a cautious approach to prescribing to older adults with IDD. Below we outline principles guiding psychotropic use, and their application to older adults with IDD.

Principles for Prescribing Psychotropics, and Their Application to Older People with IDD

Prescribing psychotropic medications to older people with IDD should be underpinned by the same principles as those which apply to any patient. Any approach to mental health care for people with IDD should be rights-based, inclusive, and person-centred. Therapies are appropriate when they are recovery-oriented, strengths-based, and promote independence [63, 69].

A Comprehensive Assessment Informs Responsible Prescribing

The decision to use psychotropic medication should always be informed by a comprehensive biopsychosocial assessment. Elucidating the symptoms, their duration, severity, and impact is only one part of the assessment. The broader context in which they are appearing is of equal importance: the person's physical health; their psychosocial wellbeing; risk profile; and their current medications. The individual's prior responses to current, past and, if relevant, proposed medications should also be noted [63, 70]. The additional physiological, psychological, and psychosocial risk factors faced by older adults with IDD necessitate a deliberate and methodical approach to exploring each of these areas.

As for any patient, the first step of assessment is always to systematically check for undiagnosed physical and then psychiatric comorbidities [71]. A thorough multi-system physical examination is essential. Structured tools such as the Comprehensive Health Assessment Protocol (CHAP; 72) can be effective for screening unidentified health problems and sensory issues, and promoting uptake of preventative healthcare, even when used at a single point in time [72, 73].

It is also important to systematically screen for frailty, even in middle-aged adults with IDD. Validated tools such as the Frailty Marker [74] may require some modification for those with IDD [28]. Using a frailty index that permits selecting those markers most relevant to people with IDD is another promising approach [14].

Particular physical and psychiatric comorbidities associated with specific genetic causes of IDD should be considered. For example, people with Down syndrome have a high rate of Alzheimer's disease [75]; those with Williams syndrome are at high risk of hypertension, diabetes and vascular disease, as well as sensorineural hearing loss [76]; and the obesity associated with Prader-Willi syndrome impacts a person as they age, including through cardiovascular diseases and diabetes [77].

Review all existing medications, checking both the dosage and the way they are taken. This should include non-psychotropic drugs also, to check for potential interactions [10]. Ask regarding herbal medicines and supplements: older people with IDD may use supplements at an even higher rate than the general population [52]. Routine bloods relevant to establishing or monitoring disease status should be performed. These also provide a baseline against which to determine the impact of

psychotropic medications on blood and biochemical parameters [70]. Additional investigations should be performed as clinically indicated.

Assessing anticholinergic load is especially important for older people, since high load increases the risk of cognitive impairment, in the context of vulnerability to dementia [22, 78, 79]. Structured tools may assist to assess anticholinergic burden [80]. However, thresholds for concern need to be lowered for those with specific physiological vulnerabilities, including elders with IDD, in whom physiological reserve may be reduced, and frailty may be present.

Alcohol intake [15] and illicit drug use [81] should be considered. Illicit drug use or abuse of prescription medication can occur in people with IDD, most commonly in those with mild IDD or those living independently. As is the case in people without IDD, it is often aimed at self-medicating distress resulting from negative life-experiences [82]. Substance use screening should therefore be considered as a routine part of clinical assessment. However, common terminology for substances might not be understood by people with IDD; slang terms and pictures can aid communication [81].

As part of a comprehensive psychosocial assessment, assess what supports (if any) the person requires to manage their medications. The support needed will depend on both the mental capacity of the person with IDD and the complexity of their medication schedule. Both things can change with age: small decrements in cognitive function can have a larger functional impact, and concurrently, age may bring with it an increased number of health conditions and hence a more complex medication regimen. Graded support matching the person's needs may involve arranging the supplying pharmacist to create a tailored blister-pack, or teaching the person to use medication reminders [47], through to arranging community nurse visits support administration of medication where this is available. For those living with family or in supported accommodation, it is helpful to establish who is an appropriate person to be responsible for administering the medication [47], and for monitoring side effects, and liaising with others involved in the person's care [63].

Responsible Prescribing Considers Up-to-Date Evidence

Any selected mental health treatment should be informed by the best available evidence, and this is particularly important for psychotropics, which risk causing harm. In the absence of evidence regarding elders with IDD, consider the risk profile of all potential medications, and match this to what is known about the patient's risk factors [11, 47, 63]. Evaluation of risk is especially important for antipsychotics, which carry a high risk of metabolic side effects [47]. Psychotropic drugs are the highest contributor to anticholinergic burden in older adults with IDD [49]. The extrapyramidal side effects of antipsychotics are common in people with IDD [83, 84] and can be long-term and severe, reducing a person's quality of life, particularly as they age.

A Holistic Plan Considers All Elements of the Assessment, and All Available Treatment Modalities

Where it is used, psychotropic medication should form just one part of a multimodal approach to addressing the older person's physical and mental health and promoting psychosocial wellbeing [63, 69, 70]. Talking therapies such as cognitive behaviour therapy can be successfully modified for older people with IDD using the information gleaned from a comprehensive assessment [15]. For those with more severe disability or limited communication, environmental and behavioural approaches should be trialed first. In some cases, psychotropics are an appropriate adjunct to such therapies, and they may also be needed where non-pharmacological treatments fail, and symptoms have a severe impact.

Consider also the way selected therapies benefit or compromise the person's overall wellbeing, especially for elders with IDD with serious physical health problems or multimorbidity. For older adults with IDD, pay attention to cardiometabolic and falls risks. If prescribing psychotropics which potentially increase cardiometabolic risk it is appropriate to pre-emptively assist the person and their carers to develop a plan to improve the person's overall cardiometabolic health. Obesity, poor diet and lack of exercise are common lifestyle-related risk factors for adults with IDD [85]; these compound metabolic side effects of psychotropic drugs. A falls and balance assessment may be warranted prior to commencement or increase of psychotropic medication in an older person with IDD who has a history of falls or who is at risk. Allied health professionals such as exercise physiologists, dieticians and physiotherapists can play an important role assessing and mitigating risks.

Establishing Goals Within a Shared Care Context

Goals of care should be established through discussion with the older person with IDD and their carers. This includes communicating a rationale for any medication, and regular and ongoing review. For people with reduced verbal communication, use of visual aids can be helpful.

Shared care with primary health providers forms the basis of good healthcare for older individuals with IDD. In establishing a shared care arrangement, establish responsibilities of all parties and ensure they are clear to all [15]. With regards psychotropic medication, this should include identifying who is responsible for administering the medication if the person is unable to manage this themselves, and who is monitoring for side effects. Carers may require explicit instructions on how to monitor these [47]. It is also sensible to discuss who is responsible for communicating medication changes with other members of the support team [70], particularly where support networks are complex [47].

Adopt a Conservative Approach to Psychotropic Medication Use in Older Adults with IDD

Older age warrants a conservative approach to medication use. Older age is associated with increased polypharmacy due to multimorbidity [86]. Ageing results in changes in the metabolism and clearance of drugs [87, 88]. As a result, older people are at higher risk of both adverse drug reactions and drug-drug interactions [86]. Guidelines therefore recommend adjusting medication choice and dose according to the age and frailty status of elderly individuals, and adjusting further as the person ages [88].

It has been suggested that most adults with IDD can commence medication at the same dose as for the general population, in order to balance the risk of side effects against the risks of under treatment [60]. However, if a person shows signs of premature ageing, it is prudent to use starting doses for a group that matches their physiological age over their chronological age, giving careful consideration of any markers of premature frailty. For elders with IDD, beginning at low initial doses with slow titration and careful monitoring of responses is appropriate [47].

Monitor and Review Over Time

Most importantly, regularly review the benefits and side effects of the medication, as well as other relevant psychosocial factors. Structured tools such as the Matson Evaluation of Drug Side Effects (MEDS; [89]) or the Modified Monitoring of Side Effects scale (MOSES; [90]) can assist clinicians to measure side effects. This will inform decisions to reduce or cease the medication. The same principles outlined above for prescription should also guide deprescription of medication. A multidisciplinary approach which includes a pharmacists' insights has been shown to be helpful for reviewing and reducing medications in adults with IDD [12]. Physical health parameters should also be monitored regularly, particularly where drugs are known to have adverse cardiometabolic effects. Comprehensive annual health assessments should continue as a priority and take on increasing importance as the person ages and in the context of psychotropic medication prescription.

Deprescribing Psychotropic Medications to Older Adults with IDD

For some individuals, psychotropic medications can be successfully reduced without worsening of symptoms [91, 92]. However, ceasing medication is not without risk: a recent systematic review identified a subgroup of people for whom discontinuation resulted in worsening behaviour [83]. The reasons for

deprescribing failure are varied and may include staff or carer characteristics, or an unrecognised withdrawal reaction [83, 93, 94]. It is also possible that for some, an increase in challenging behaviours following medication reduction will be only temporary [95]. Hence, an individual approach to the reduction of antipsychotic use is required [15, 61].

Antipsychotics Are Not an Acceptable First-Line Treatment for Challenging Behaviour

Special mention must be made regarding the use of antipsychotics for challenging behaviour, particularly in the context of ageing and dementia in people with intellectual disability. Studies have found that antipsychotics are prescribed to adults with IDD at far higher rates than the general population and often in the absence of a severe mental illness diagnosis [46, 50, 53, 56, 96, 97]. Antipsychotics are also overprescribed amongst older adults with dementia in the general population [98], though guidelines and interventions to redress this mean the trend is reducing in some locations [99, 100]. Calls to reduce the inappropriate prescription of psychotropics and particularly antipsychotics for people with IDD are longstanding [101–103] but have gained greater prominence in recent years [83].

Managing challenging behaviours should aim to uncover the underlying cause [70] and thereafter treat it—this may include medication as an adjunct to other therapies [61]. Where no medical reason is identified, the first-line treatment should be behavioural and environmental approaches [47, 63]. Medication is only appropriate where the behaviours are high in severity and frequency; pose a severe risk to the person, others, or their placement [61, 70]; and where it can be documented that an appropriate behavioural or environmental intervention has been attempted and failed [59].

The Intersection of Psychotropics and Complementary and Alternate Medicines (CAMs)

In recent years, developed countries have seen an increase in the use of complementary and alternate medicines (CAMS), with close to one third of middle aged and older adults using CAMS including herbal medicines [104, 105]. Supplement use may be more prevalent in older people with IDD compared with those without IDD [52]. Guidelines on healthcare topics often overlook CAMS [106], as do protocols redressing polypharmacy in the elderly [107].

CAMS are often self-prescribed and patients frequently do not disclose their use to physicians [108]. As some herbal medicines can have a psychotropic effect, or can interact with prescription psychotropics, it is important for prescribers to

specifically ask regarding CAMS, and to educate patients and carers regarding potential interactions with their prescribed medicines.

Medicinal Cannabis

Cannabinoids, particularly cannabidiol and *tetrahydrocannabinol*, have been suggested to hold potential as a therapeutic agent for a wide-range of conditions, including epilepsy, anxiety, sleep, behavioural symptoms of dementia, chronic pain, nausea, particularly in late-stage diseases such as AIDS or cancer, and psychosis [109, 110]. More recently, the potential use of medicinal marijuana as a potential treatment for severe behavioural problems associated with neurodevelopmental disorders such as autism has been proposed [111], though the lack of quality evidence in this regard has also been highlighted [111, 112].

Overall, research on medicinal cannabis limited, and the 2019 National Institute for Health and Care Excellence guidelines echoed other calls for further research to understand its benefit and safety for most suggested therapeutic applications [113, 114]. Much of the existing research has excluded older adults [115]. A small number of studies have investigated the safety of cannabis in certain older adults, with promising results (e.g. 115, 116). However, there appears to be high inter and intra-variation between individuals in its pharmacokinetic profile [116, 117], with adverse events more frequent than for some more traditional therapies [118]. Specific research regarding the use of cannabinoids in older people with IDD has not been undertaken at the time of writing.

Given that this is an emerging area of medicine, it is suggested that clinicians who are considering prescribing medicinal cannabis to older adults with IDD appraise themselves of up-to-date guidelines and evidence. It is important to consider a person's prior history of use or abuse of this or other drugs, and their liver, renal or cardiovascular health [113]. Lucas [117] recommend a cautious 'start low and go slow' approach for prescribing cannabinoids to any population group. In those with IDD, who are already at higher risk of adverse effects, this would be essential.

Conclusion

Psychotropic medications have a role in managing mental disorders in some older people with IDD, but they should be prescribed in the context of a multimodal approach to therapy. For older adults with IDD, an increased number of medical conditions, high risk of frailty, and an unusual medication profile, can make prescribing more complex, as can the reduced evidence base available to inform the use of psychotropic medications in this group. Both prescribing and deprescribing

psychotropics to this group therefore requires an individualised approach which considers the information gathered from a thorough biopsychosocial assessment, and which establishes goals of care with the older person and their carers. Commencing medication at low doses with slow titration and systematic monitoring for side effects can help to mitigate the increased risk of adverse events in elders with IDD. Appropriate prescribing for older people with IDD is complex and requires diligence on the part of the clinician, but ultimately it is underpinned by the same principles which inform responsible prescribing to any person.

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Chapter 13
Role of Anti-dementia Medication

Vee P. Prasher, Kasim Qureshi, and Sharmin Fatema

Introduction

The treatment of dementia poses formidable challenges to all involved. The impact of cognitive decline can have profound effects from the level of the individual to society at large. Within this context, the presence of intellectual disabilities complicates both recognition of dementia and its management. This chapter will primarily focus on the role of anti-dementia medication within the context of the most striking relationship between dementia and intellectual and developmental disabilities (IDD)—linked conditions of Down syndrome (DS) and Alzheimer's disease.

The realisation that people with DS are prone to early dementia has been noted since at least the late nineteenth century. Clinicians of the time speculated that a type of early "precipitated senility" [1] was apparent in adults with DS and it was speculated that this may represent a form of premature ageing. Building upon this foundational observation, modern DS research has proved instrumental in our current understanding of Alzheimer's disease across populations. As we will discuss latterly, this has unfortunately not translated into parity of research involvement for people with DS compared to the general population.

The relationship between DS and Alzheimer's disease led to the hypothesis that some part of chromosome 21 (triplicated in DS) may contain the key genetic features that predispose individuals to developing dementia [2, 3]. What subsequently followed was the discovery that beta amyloid proteins appears to mediate the development of Alzheimer's disease and that the amyloid precursor protein (APP) gene plays in key role both in people with DS and in the general population with familial

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Alzheimer's disease [4, 5]. Contemporary research reflects the realisation that links between Alzheimer's disease and DS are myriad [6, 7], and this has significant implications for pharmacological treatment.

The major societal impact of dementia in Alzheimer's disease has driven diverse research approaches around the world. The most common age-related dementia globally, Alzheimer's disease is heterogeneous and characterised by a wide range of cognitive deficits and other neuropsychiatric symptoms [8]. Although the detection of early symptoms of dementia in Alzheimer's disease may at times be difficult in individuals with intellectual and developmental disabilities (IDD), the clinical picture seen is not too dissimilar to that for the general population [7, 9]. Insidious progression of memory impairment, personality change, dysfunction in language and motor skills, onset seizures and behavioural abnormalities, marked loss of self-care skills and weight is typical. Variability in presentation does exist, but the clinical pattern broadly reflects dysfunction in a core temporal-parieto-frontal network [10]. Personality or behavioural disturbances have been reported to possibly be a more prominent early feature than forgetfulness [11]. Death usually occurs within 5–10 years of onset. The disease having a devastating impact not only on the individuals themselves but also on carers and the State.

If we exclude individuals with DS the prevalence of dementia is no higher in other types of IDD than in the general populace [12]. The onset of dementia in Alzheimer's disease has been reported in individuals with DS as young as aged 30 years, with a dramatic increase in prevalence rates over the subsequent decades [11, 13]. Increasing age in adults with DS, as per the general population, markedly increases the risk of Alzheimer's disease, as illustrated in Table 13.1 but persons with DS tend to be affected approximately 30 years earlier [11]. However there is considerable variation in estimates depending on the population sampled. The impact of Alzheimer's disease on people with DS is highly significant for prognosis: in those aged over 60, the presence of Alzheimer's disease is associated with a mortality rate of 44% compared to only 11% in individuals with DS alone [14] (Table 13.1).

Neuropathological Alzheimer's disease is characterised by the formation of amyloid plaques and neurofibrillary tangles, loss of cortical brain matter, synaptic and neuronal loss and presence of inflammatory changes [15, 16]. Neurochemically there is a deficit in a number of cerebral neurotransmitters, such as acetylcholine,

Table 13.1 Approximate prevalence of dementia in people with DS by age

Age of individuals with Down syndrome	Estimated prevalence of Alzheimer's dementia (%)
30–39	3
40–49	10
50–59	40
>60	>40

Adapted from Holland and colleagues [11]

neuroadrenaline, and serotonin [17]. Findings over the last few decades would suggest that the principal chemical deficiency in Alzheimer's disease is that of degeneration of cholinergic neurones, neocortical deficits in choline acetyltransferase, reduced choline uptake and in acetylcholine release. This "cholinergic hypothesis of Alzheimer's disease" has been the main thrust of drug development in Alzheimer's disease [18]. The aim being to enhance selective cholinergic transmission in the brain by increasing the supply of choline, stimulating cholinergic receptors or by reducing acetylcholine metabolism (by inhibiting cholinesterase action). Interest remains in the hypothesis that glutamate mediated neurotoxicity is involved in the pathogenesis of Alzheimer's disease [19, 20]. In this hypothesis glutamate receptors (N-methyl-D-aspartate [NMDA]) are overactive and may interact with beta-amyloid or tau protein metabolism resulting in the characteristic changes of Alzheimer's disease.

Pharmacological Treatments for Alzheimer's Disease

Although serendipitous discoveries can still play a role in drug development, devising new pharmacological treatments is predominantly informed by an understanding of disease pathology. Drug treatments for Alzheimer's disease are no exception. In the 1980s major developments in Alzheimer's disease research included the discovery that abnormal protein structures (beta amyloid plaques [21] and tau tangles [22]) were associated with the disease. Furthermore the dysfunction in the brains of people with Alzheimer's disease was principally linked to reduced activity in acetylcholine brain systems [23]. Given the importance of acetylcholine, it was speculated that if the breakdown of acetylcholine could be reduced, communication between neurons (and hence cognitive function in Alzheimer's disease may be improved). This swiftly led to first clinical trial of a treatment for cognitive decline in Alzheimer's disease.

Several decades have passed since the first ant-dementia drug tacrine was used [24]. More recently the "second generation" of cholinesterase inhibitors donepezil, rivastigmine, and galantamine, have made a significant impact on the clinical management of Alzheimer's disease [25, 26]. In the UK the National Institute for Clinical Excellence [27] and in the US the American Academy of Neurology [28] concluded that anti-dementia drugs have a significant benefit in patients with Alzheimer's disease and that these agents should be made available. However, both reports limited their recommendations to the general population. This article reviews recent advances in the drug treatments (donepezil, rivastigmine, galantamine and memantine) for dementia in Alzheimer's disease in adults with DS. Donepezil and galantamine are selective inhibitors of acetylcholinesterase (AChE), whilst rivastigmine is a dual inhibitor of AChE and butyrylcholinesterase (BChE). Memantine is a non-competitive antagonist of N-methyl-D-aspartate receptor, i.e., the NMDA receptor (Table 13.2).

Table 13.2 Main pharmacological treatments for cognitive symptoms of Dementia in Alzheimer's Disease

Medication	Major mechanisms	Licensed indication (UK)
Donepezil	Selective reversible inhibitor of Acetylcholinesterase	Mild-moderate dementia in AD
Galantamine	Selective reversible inhibitor of Acetylcholinesterase	Mild-moderately severe dementia in AD
Rivastigmine	Reversible non-competitive inhibitor of Acetylcholinesterase and Butyrylcholinesterase	Mild-moderate dementia in AD (also licensed for dementia in Parkinson's Disease)
Memantine	Non-competitive NMDA antagonist	Moderate-severe dementia in AD

Pharmacological Properties

The pharmacological properties of donepezil, rivastigmine, galantamine and memantine are given in Table 13.3. All are given orally in tablet/capsule form, with donepezil and memantine also as orodispersible tablets. All can now be administered to individuals with poor compliance or swallowing problems in liquid form. The transdermal patch formulation of rivastigmine is useful but advice does need to be given to carers on how to administer the patch and in particularly to remove the previous' day's patch before applying the new patch. All prescribed once or twice a day at maintenance dosage. Good clinical practice would suggest that all of the anti-dementia drugs should be started initially at a sub-therapeutic dosage and gradually increased. In the learning disabled population with greater caution and greater monitoring than that for the general population. All of the cholinesterase inhibitors are licensed for use in mild to moderate dementia in Alzheimer's disease; memantine is the only drug licensed in the UK for the treatment of moderately-severe to severe dementia in Alzheimer's disease. Rivastigmine reaches its maximum concentration and is eliminated the quickest of the four drugs Table 13.4. Memantine the slowest, with an elimination half-life of up to 4 days. Donepezil and rivastigmine are metabolised by the liver and memantine and galantamine excreted by the kidneys.

Although there are differences in the pharmacokinetics and formulation there are still considerable pharmacological similarities between all of the four anti-dementia drugs. They all affect the central cholinergic system either directly as AChE inhibitors or indirectly by acting on related pathways (glutamate receptors). The AChE inhibitors are licensed for mild to moderate dementia in Alzheimer's disease. The anti-dementia drugs should be initiated with a similar degree of caution. Treatment should be withdrawn if tolerance or compliance is poor, if the patient's condition continues to deteriorate at a rate after 3–6 months, or if little benefit has been determined during this period.

Table 13.3 Summary of pharmacological parameters of anti-dementia drugs

Drug	Chemical class	Action	Type of inhibition	Route of administration	Frequency of administration	Dosage/day	Indications
Donepezil	Piperidine	AChE inhibitor	Rapidly reversible	Oral tablet Oral solution Orodispersible tablet	Once a day	5–10 mg	Mild-mod AD
Rivastigimine	Carbamate	AChE and BChE inhibitor	Pseudo-reversible	Oral capsule Oral solution Transdermal patch	Twice a day Patch once a day	6–12 mg	Mild-mod AD
Galantamine	Phenanthrene alkaloid	AChE inhibitor	Rapidly reversible	Oral tablet Oral solution Modified-release capsule	Tablet twice a day MR capsules once a day	8–24 mg	Mild-moderately severe AD
Memantine	Glutamatergic modulator	NMDA antagonist	N/A	Oral tablet Oral solution Orodispersible tablet	Twice a day	10–20 mg	Mod severe-severe AD

Table 13.4 Summary of pharmacological parameters of anti-dementia drugs

Drug	Time to reach max conc (h)	Elimination half-life (h)	Protein binding (%)	Total body clearance[a] (L/H/kg)	Time to steady state (days)	Excretion
Donepezil	3–5	50–70	96	0.13	14–22	Hepatic
Rivastigimine	0.5–2.0	0.6–2.0	43	N/A		Hepatic
Galantamine	1.2	5–7	<20	N/A	2	Hepatic and renal
Memantine	3–8	60–100	45	N/A	11	Renal

[a]Drug clearance from plasma

Therapeutic Efficacy

For the general population it has now been established that patients with dementia in Alzheimer's disease do benefit clinically both in the short and long-term [25, 26, 29, 30] with anti-dementia therapy. In one of the earliest studies Mohs and colleagues [31] in a study of 431 Alzheimer's disease patients randomised to donepezil 10 mg or placebo for 54 weeks found that the former group maintained their function for 72% longer, and were less likely to decline over the year compared to placebo by approximately 40%. Treatment with donepezil delaying decline by approximately 5 months. Subsequent studies have shown that with active treatment with AChE medication benefit is seen in global functioning, cognitive abilities, neuropsychiatric symptoms, behavioural problems, in day-to-day skills and in the reduction in carer stress [32, 33] There is less scientific literature on the role of memantine in the treatment of dementia in Alzheimer's disease, either used on its own or in combination with an AChE, but positive findings are reported [34–38]. There are, however, some on-going debates as to whether these modest improvements consistently represent a clinically significant and cost-effective change [39–42]. Previous analyses have suggested that long-term AChE inhibitor treatment may be cost-neutral at best [40, 43] when longer timescales are considered. As such, both on grounds of clinical risk and cost, it is both important and challenging to determine when to withdraw dementia medication when its effectiveness is unclear.

For persons with IDD and dementia the evidence of benefit is unclear due to the paucity of drug trials. Four Cochrane systematic reviews of dementia medication in DS pre 2008 were undertaken in 2009 [44–47]. The reviews were not able to identify studies of sufficient methodological quality that could be synthesised to make reliable and valid recommendations. Subsequently, Livingstone and colleagues [48] conducted a more recent systematic review and meta-analysis of studies evaluating dementia medication in DS. This utilised broader inclusion criteria and incorporated nine clinical trials three of which investigated cholesterol or dietary interventions [49–51]. The included trials of AChE inhibitors or NMDA antagonists are illustrated in Fig. 13.1. Importantly, no cognitive or behavioural benefit was identified within the meta-analysis of randomised controlled trials for any treatment. One

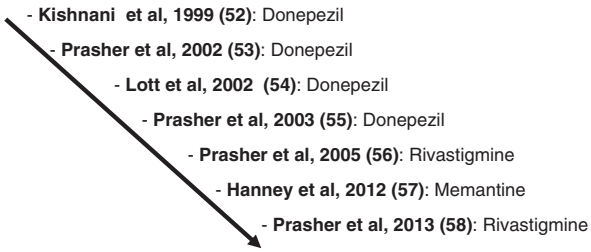

Fig. 13.1 Timeline of studies of dementia medication in Down syndrome

trial of donepezil identified a modest possible benefit in global functioning in for people with DS and Alzheimer's disease, however the evidence underlying this conclusion was deemed to be of low quality. As such, the scientific knowledge of the role of anti-dementia drugs in IDD was mostly limited to small scale studies.

Kishnani and colleagues [52] published findings of four adults with DS who were treated with up to 10 mg donepezil for between 26 and 68 weeks. Two younger individuals aged 24 and 27 years were not demented but two older persons aged 38 and 64 met DSM-IV criteria for dementia. The report was an open trial of donepezil has subject to considerable sources of error. On the one objective test used Vineland Adaptive Behavior Scales there was improvement in scores for the non-demented individuals but little change for the demented persons.

Prasher and colleagues [53] published findings from a 24-week, double-blind placebo-controlled trial of donepezil in 30 patients with DS and dementia in Alzheimer's disease. The Dementia Scale for Mentally Retarded Persons (DMR) was used as the primary outcome measure with secondary outcome measures for cognition, neuropsychiatric features and adaptive behaviour also used. The DMR can be used to give a global impression. The donepezil group had non-statistically significant reduction in deterioration in global functioning, in cognitive skills and in adaptive behaviour. The active group scored worse on the presence of neuropsychiatric symptom profile, which was explained by the authors as being a reflection of drug induced adverse effects being detected by the questionnaire. No life-threatening events occurred during the study period. The authors concluded that donepezil is probably efficious in the treatment of dementia in Alzheimer's disease in adults with DS.

Lott and colleagues [54] reported results form an open-label study of donepezil to treat dementia in six DS patients. Treatment was for between 83 and 182 days with dosage up to 10 mg. Findings for the active group were compared to six matched historical control subjects. Dementia was assessed before treatment and after an average time interval of 5 months using the Down Syndrome Dementia Scale (DSDS). A significant improvement in dementia scores was seen for the treated group, although the authors highlighted a number of drawbacks with the study.

Prasher and colleagues [55] went on to report in an open-label study the evaluation of the long-term (104 weeks) safety and efficacy of donepezil in the treatment of dementia in Alzheimer's disease. The 25 patients in this study had previously

completed the 24 week randomised double-blind placebo-controlled trial [53]. Patients were assessed in this study with the same measures as in the 24-week study. The primary outcome measure was the DMR. The mean total DMR score showed initial improvement from baseline for the donepezil group with subsequent deterioration in both the treated and untreated groups over the study period. At 104 weeks the deterioration in global functioning and adaptive behaviour was statistically significantly less for the treated demented DS subjects. This study demonstrated that donepezil was beneficial in the treatment of dementia in Alzheimer's disease in the DS population for up to 2 years.

The largest and most methodologically rigous study was published in 2012 by Hanney and colleagues [56]; a randomised, double-blind placebo-controlled trial of memantine for dementia in adults with DS over 52 weeks. Eighty eight DS individuals with and without dementia received memantine and 85 DS individuals received placebo. Both groups declined in cognition and function but rates did not differ between significantly the two groups. The researchers concluded memantine was not an effective treatment for dementia in the DS population.

To date there has only been two studies specifically reporting on the role of rivastigmine to treat dementia in older adults with DS [57, 58]. In the 2005 study Prasher and colleagues [57] found a non-statistically significant benefit of oral rivastigmine to treat dementia compared to placebo. In the 2013, study Prasher and colleagues [58] reported findings of 10 persons with DS and dementia treated with rivastigmine transdermal patches was compared with 13 non-demented subjects and with 17 DS persons with dementia previously treated with oral rivastigmine. Over a 6 month period. Patients treated with rivastigmine (oral or patches) had borderline significantly less decline in cognitive and global functioning as compared to the untreated group. However, rivastigmine patches were found to have better patient compliance and less significant side-effects.

Eady and colleagues [59] recently reported on a naturalistic longitudinal follow-up study of 310 adults with DS diagnosed with dementia in Alzheimer's disease. Median survival time for those treated with anti-dementia medication was significantly greater than for those not on prescribed medication.

There remains, and will always do so, difficulties in undertaking drug trials in the IDD population which are of the same standard as those undertaken in the general population. This particularly applies to studies of older adults with dementia in Alzheimer's disease. Problems of small sample size, non-blindness of carers and raters, inclusion criteria for dementia in Alzheimer's disease, reliability of measures used and type of statistical analysis used are at present inherent sources of error. However, from the limited information available from studies of dementia in Alzheimer's disease in the DS population and the inferences from findings from the general population, it is reasonable to conclude that AChE inhibitors can both in the short-term and long-term be efficacious in the treatment of dementia in Alzheimer's disease in older adults with DS. In keeping with results for the general population there is a reduction in the deterioration of cognitive skills, neuropsychiatric symptoms and in adaptive skills. No information is available regarding the possible beneficial impact on carers.

The use of all four anti-dementia drugs in older adults with DS have been reviewed [60]. They are being used in the clinical setting and further ongoing

studies on the efficacy of these drugs to treat dementia in Alzheimer's disease in the DS population, along with studies of other medications, such as antioxidants to treat cognitive decline in adults with DS [61, 62] are very much required. From the available information it would appear that patients with IDD and with dementia in Alzheimer's disease are likely to benefit from anti-dementia therapy. The importance of methodological rigour in clinical trials and the need for large multi-centre studies involving people with IDD remains necessary.

Tolerability

Side-Effects

All of the anti-dementia drugs discussed are generally well tolerated, and most of the adverse events that may occur are mild and transient. In the case of the AChE inhibitors these are related to the cholinergic system. The rate of adverse events of are often similar to placebo [53, 63] and are dose-related. The commonly reported adverse events are listed in Table 13.5. In drug trials approximately 5% of individuals withdraw from studies because of adverse events. Many of the side-effects listed

Table 13.5 Commonly occurring side-effects of the anti-dementia drugs

Adverse event	Donepezil	Rivastigmine	Galantamine	Memantine
Nausea	+	+	+	
Diarrhoea	+	+	+	+
Insomnia	+	+	+	+
Fatigue	+	+	+	+
Vomiting	+	+	+	+
Muscle cramps	+		+	
Anorexia	+	+	+	
Headache	+	+	+	+
Dizziness	+	+	+	+
Syncope	+	+	+	
Urinary incontinence				
Psychiatric disturbances	+	+	+	+
Pruritus				
Weight loss	+	+	+	
Abdominal pain		+		
Drowsiness		+	+	+
Hallucinations		+	+	+
Cardiac changes			+	+
Aggression	+	+	+	+
Agitation	+	+		
Common cold	+			
Seizure	+			

in Table 13.3 are relatively minor, transient and can be stopped by reducing the dose of medication used.

In the studies involving adults with DS, Kishnani and colleagues [52] found that none of the four study individuals experienced any serious adverse effect. Transient agitation and loose stools were the only side-effects noted [53, 55]. The researchers found fatigue (44%), diarrhoea (38%), insomnia (25%), nausea (25%), dizziness (19%) and anorexia (19%) as the commonest treatment-emergent features. Hemingway-Eltomey and Lerner [64] reported three cases of DS patients with dementia who developed adverse effects of agitation, aggression, urinary incontinence and worsening memory loss whilst being treated with donepezil.

Though the medications have a common therapeutic aim, there are some differences in their side-effect profiles. These treatments are associated with some common (and usually transient) side effects—for AChE inhibitors these are nausea, vomiting and diarrhoea, whereas memantine is more commonly associated with dizziness, headache and constipation [65, 66]. There are important, albeit less common, adverse effects which warrant close monitoring. For AChE inhibitors, there is an increased risk of worsening symptoms in individuals with bradycardia, urinary obstruction and chronic obstructive pulmonary disease. An important consideration for memantine is that recently a significant increase the risk of visual hallucinations has been reported [67].

Clinicians should be aware of the more serious side-effects, e.g., reduced heart rate (which can be significant in an individual with DS who may already have a low resting heart rate), stomach ulcer with bleeding, seizures, and depression. Starting at a low dose and slower titration of dosage can reduce the frequency of side-effects, particularly in an ageing population.

Contra-Indications

There are a number of medical conditions where the use of the anti-dementia drugs is contra-indicated (Table 13.6). These conditions are generally similar for all four of the anti-dementia drugs. They include asthma, sick-sinus syndrome, supraventricular

Table 13.6 Conditions were anti-dementia therapy should be used with caution

Drug	Condition
Donepezil	Sick sinus syndrome, supraventricular conduction abnormalities, history of peptic ulcers, asthma, chronic obstructive airway disease, hepatic impairment
Rivastigmine	Renal impairment, hepatic impairment, sick sinus syndrome, supraventricular conduction abnormalities, history of peptic ulcers, asthma, chronic obstructive airway disease
Galantamine	Sick sinus syndrome, supraventricular conduction abnormalities, history of peptic ulcers, asthma, chronic obstructive airway disease, hepatic impairment, urinary obstruction
Memantine	Renal impairment. Caution in patients with epilepsy, cardiovascular disorders

conduction abnormalities, history of peptic ulcer, chronic airway disease, anaesthesia, hepatic and renal impairment. Older adults with DS are more vulnerable than their general population counter-parts to presenting with concurrent medical conditions and a detailed review of their physical health status is necessary prior to prescribing the drugs for dementia in Alzheimer's disease.

Drug Interactions

The formulation of medication can be a particularly pertinent factor when ensuring safety and concordance for patients with IDD. All four medications can be administered as an oral solution (as well as tablets) and donepezil and memantine as orodispersible tablets; a significant advantage when individuals have impaired swallowing. Rivastigmine is also available as a transdermal patch, the only dementia medication which can be administered via a non-oral route in the UK [68].

Rivastigmine and galantamine are rapidly cleared from the systemic circulation (half-life 1–2 h and 5–7 h respectively) and therefore are at lower risk of significant interaction with other drugs. They have low plasma protein binding and are eliminated by the kidneys. As such there is also a lower risk of long-term accumulation [69, 70]. In contrast, donepezil has a level of high protein binding (which is a particular concern in overdose) and a longer half-life (50–70 h). Memantine has the longest elimination half-life (60–100 h). These factors are relevant when assessing the risk of serious adverse events such as bradycardia [71]. The more common antidementia drug interactions involve share metabolic pathways in the liver. In the IDD population, this may commonly include medications such as anticonvulsants and antidepressants. Later in life, people both with and without IDD are at risk of side effects from polypharmacy and the potential benefits of dementia medication need to be evaluated against the individual risk factors that include other medication and comorbidities.

There is limited adverse effect data from AChE inhibitors and memantine in patients with IDD. Common themes from small studies reflect the same transient, dose-dependent side-effects described above. There is insufficient data to establish whether the absolute rates of these effects are equivalent to the general populace. Certain specific considerations are important when monitoring medication for patients with IDD—specifically the potential for AChE inhibitors to produce bradycardia particularly in DS patients who are prone to abnormalities of heart rate variability [72], and also the theoretical potential for memantine to reduce seizure threshold in this population with an increased risk of epilepsy.

There are some indications (based on adverse event reporting in North America [66] that rivastigmine may be associated with a greater risk of mortality due to cardiac effects than other AChE inhibitors. This may be attributable to high doses associated with administration errors when transdermal patches are used, and clinical guidelines which advise use of rivastigmine in more complex cases such as individuals who have not responded to donepezil or galantamine [66]. As such it is not

clear whether a higher rate of adverse effects are attributable to rivastigmine's pharmacological properties, but in patients with IDD, the higher likelihood of medical comorbidity warrants an increased level of caution for all involved in clinical decision-making. As with much pharmacotherapy for patients with IDD, a prudent approach in the face of uncertainty is to commence at low doses, increase gradually and monitor closely for serious adverse events.

Memantine differs from the AChE inhibitors above, in that (while it has some anticholinergic effects) it predominately acts on glutamatergic systems and its distinct mechanism also serves to improve neuronal signalling in Alzheimer's disease. Memantine also differs in its clinical usage, in that it is prescribed in cases where individuals have not tolerated AChE inhibitors, or those for whom dementia has progressed to a severe stage. AChE inhibitors are thought to act on the (predominantly neocortical) cholinergic neurones by interfering with the breakdown of acetylcholine, thereby increasing the available supply of the neurotransmitter. In contrast, NMDA antagonists (of which memantine is the only widely approved medication) is thought to reduce the neurotoxic effects of overactive glutamate receptor expression. It has been proposed that memantine protects against the damaging inflammatory effect of beta-amyloid build-up and this enables increased neuroplasticity and hence cognition [72].

Other Issues

Due to the considerable absence of evidence-based research findings for the IDD population, there remain several unresolved issues. Do adults with IDD and dementia other than persons with DS also benefit from anti-dementia therapy? Concerns remain regarding the maximum dose and exact dosage schedule to be used. Is efficacy dose-related? There remains considerable clinical uncertainty regarding when should anti-dementia drugs be stopped. Medication should be withdrawn if significant adverse effects occur, compliance is poor or a significant contra-indication occurs. NICE recommend that a repeat assessment should take place 2–4 months after the maintenance dose has been reached and only if there has been no decrease in assessment scores (Mini-mental State Examination for the general population) together with improvements in behaviour and/or functioning.

Other issues are, could AChE inhibitors be beneficial to young individuals with DS who present with Young Adults with Disintegrative Syndrome (YADS) syndrome [73] or those that have the neuropathological changes of Alzheimer's disease but as yet not presented with clinical dementia? As for the general population, are there particular predictors of response, are there any significant differences between the drugs, can an AChE inhibitor be used in combination with a NMDA antagonist and can the drugs be used for other forms of dementia, other than dementia in Alzheimer's disease?

The health economics of the prescribing of anti-dementia therapy has been researched in the general population [37, 74–76]. Limited information is available

for people with IDD. Yearly healthcare costs for providing care for adults with dementia are a significant proportion of the State budget. Such costs are usually related to provision of residential/nursing home care. It is argued; therefore, maintaining a person with dementia in Alzheimer's disease in their family home by using drugs which delay severe deterioration of dementia in Alzheimer's disease or improve functional abilities would significantly impact on healthcare costs. Many older adults with DS are often prior to the onset of dementia in Alzheimer's disease already living in residential-type accommodation and cared for by paid carers. The economic benefit will, therefore, be markedly less as compared to those for the general population. Nevertheless further health economic analyses evaluating the cost benefits of all the anti-dementia drugs for adults with IDD is recommended. Further, considerable emotional and financial stress is put upon family carers.

Future Directions

The precise manner in which beta amyloid relates to clinical symptoms in DAD is still being elucidated, however the importance of this pathological process has given rise to novel drugs targeting amyloid more directly. Though at the early stages, novel drug treatments which reduce amyloid toxicity have demonstrated feasibility and tolerability in patients with DS [77]. This may yield the potential to directly reverse fundamental pathological processes in Alzheimer's disease; however the duration of the pilot study was 4 weeks, and perhaps unsurprisingly, clinical changes were not observed during this initial phase.

Approaches successful in other neurodegenerative conditions may hold promise for patients with DS and Alzheimer's disease. Neuromodulation techniques can be used to directly and indirectly alter the electrical activity of the central nervous system. One such family of approaches—transcranial electrical stimulation—appears well-suited for translation to clinical settings due to its non-invasive nature, low potential for side effects such as seizures, and relatively low cost [78]. These represent significant advantages in comparison with other non-invasive approaches such as transcranial *magnetic* stimulation. Transcranial direct-current stimulation (tDCS) applied to the temporoparietal regions has been shown to enhance recognition memory in patients with Alzheimer's disease who do not have IDD (albeit transiently). As this technology is refined and adapted to produce more robust and long-lasting effects, its low cost, benign side-effect profile and non-invasive nature may render it well-suited to use in settings for persons with IDD.

Arguably one of the most significant paradigm shifts in dementia research in recent years concerns not new technology, nor pharmacological agents, but rather a more rigorous system of classification. A critical mass of converging evidence across clinical, pathological and translational studies has revealed that a very significant proportion of individuals previously diagnosed with Alzheimer's disease should not be classified as such. Nelson and colleagues [79] report the recent conclusions of a consensus working group on the recently-proposed diagnostic

category *Limbic-predominant age-related TDP-43 encephalopathy* (LATE). LATE dementia may encompass as many as a third of people diagnosed with dementia in Alzheimer's disease, but describes a distinct (and highly prevalent) disease entity with its own pattern of neuropathological change, radiological features and neuropsychological profile. The implications of this for Alzheimer's disease research are vast. The remaining proportion of individuals with 'pure' Alzheimer's disease may exhibit less heterogeneity, thereby facilitating more targeted research and treatment. Because LATE is associated with a distinct proteinopathy and other pathological changes, those who remain diagnosed with dementia in Alzheimer's disease may represent a group for whom effective disease-modifying treatments can be developed for the first time.

Conclusion

The reported efficacy of the use anti-dementia drugs (donepezil, rivastigmine, galantamine and memantine) in IDD is limited and remains ambiguous. However the absence of positive evidence should not be mistaken for evidence of absence. As with other neuropsychiatric conditions, the dearth of research involving people with IDD risks limiting access to the best treatments. Furthermore, the detection of subtle cognitive improvement is as fraught in individuals with IDD as the recognition of subtle cognitive decline. It is reasonable in many cases to extrapolate positive findings from the general population, and yet there is evidence that dementia medication is prescribed for people with IDD at rates far lower than for the wider populace [80]. It is unclear whether this reflects appropriate caution due to risks such as bradyarrhythmia, or the lack of access to appropriate treatment that people with IDD routinely face.

With growing research evidence and growing clinical experience, it is likely that the above drugs will also become first-line treatment for dementia in Alzheimer's disease in older adults with IDD without or without DS. Whether adults with DS can tolerate higher doses remains uncertain but the initiation of drug therapy at low dose with gradual titration will reduce adverse effects and lead to greater compliance.

There are many other drug therapies which are being developed and many become alternatives or supplements to the present medications. These include metal chelators (e.g. clioquinol), nonsteroidal anti-inflammatory drugs (e.g. indomethacin) antioxidants (e.g. vitamin E), hormones (e.g. oestradiol), herbs (e.g. Gingo biloba), and vitamins (e.g. folic acid). Irrespective of the type of drug therapy it remains important that drug therapy is used as part of a wider management plan with carer support, psychological and behavioural treatment and the assessment of physical health status also components of care.

In the field of IDD the decision when to stop medication can be difficult, e.g. anticonvulsant medication. This particular applies to the use of anti-dementia therapy. There is limited information available regarding the natural progression of dementia in Alzheimer's disease in adults with DS. For clinicians the ongoing

prescribing of a drug that modifies deterioration over a short period of time continues to be a dilemma. Ideally, an objective measure is required. Several potential markers, such as red blood cell cholinesterase inhibition, cerebrospinal fluid monoamine or beta-amyloid measurement and platelet amyloid precursor protein, are being investigated but as yet have not been established.

There remain several areas of further research. How do the different drugs compare? Can they be used in combination? Do any significantly improve the quality of life? Is there a cost-benefit to the State? Are they any outcome predictors (e.g. apolipoprotein status)? Do people with IDD but without DS respond differently?

The development of drug therapy for dementia in Alzheimer's disease, a devastating medical illness, is a major advance in medical care. Adults with DS should be allowed access to the same types of treatments as the general population but with caution and modification in the management plan as appropriate. At present, it is recommended that treatment should follow, in principle, NICE guidelines with initiation of therapy and responsibility for monitoring of the patient by a specialist.

The field of dementia research is in a transitional period in which the understanding of the disease process is advancing, but it is not clear from which direction therapeutic advances will come. Refining the classification of other amnestic syndromes which were conflated with Alzheimer's disease (such as LATE dementia) has great potential to allow a more precise development of treatments for "pure" Alzheimer's dementia. As understanding moves beyond plaques and tangles, toward other relevant factors such as epigenetic, neuroinflammation and many other mechanisms, there is an opportunity to ensure people with IDD are included in research and accessing advanced treatments at an early stage. The IDD population is vulnerable to exclusion throughout life, perhaps even more so with the onset of dementia. As such, ensuring early access to appropriate interventions, may serve to partially redress the inequalities that affect many aspects of care for people with IDD.

It is outside of the scope of this chapter to discuss the vital importance of treating comorbidities in dementia, in particular depression, which can have a major impact on people with and without IDD who experience cognitive decline. Similarly, there are pharmacological treatments for behavioural and psychological symptoms in dementia which warrant detailed consideration in their own right. Furthermore it is important to highlight, that while this chapter discusses the role of medication, this is not to imply that pharmacological treatment has primacy in the management of dementia. Addressing psychological and environmental issues can often hold greater potential to positively impact individuals with IDD and cognitive decline.

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Chapter 14
Legal, Ethical and Due Process Issues

Christine D. Cea, Celia B. Fisher, Alexa G. McKnight, and Philip W. Davidson

Introduction

Since the middle of the last century, we have known that the world population is graying. Data from a recent United Nations Population Fund report [1], shown in Fig. 14.1, indicates that by the year 2050, 52% of the world's population will be 60 years of age or older and that octogenarians are the fastest-growing segment of the population. There are no comparable statistics for people with intellectual and developmental disabilities (IDD). Two factors provide indirect evidence, at least in states with developed economies, that these trends should be no different for people

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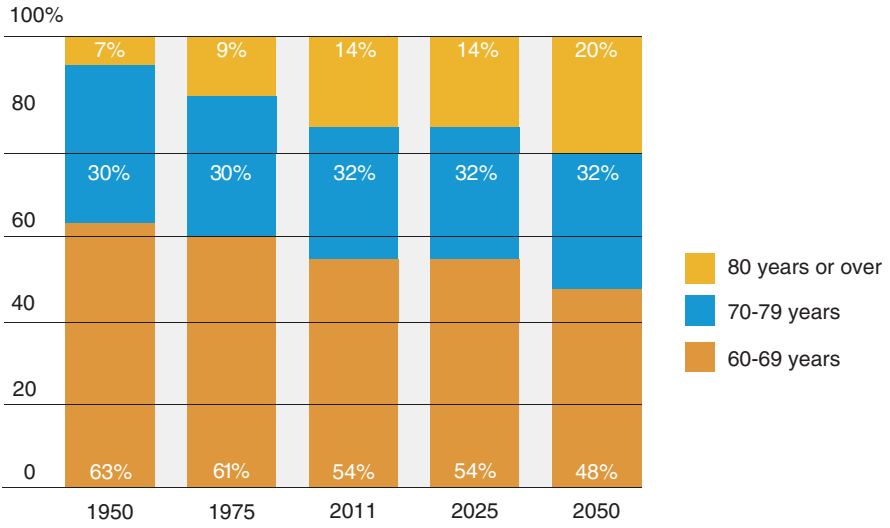

Fig. 14.1 Distribution of population aged 60 years or over by broad age group: World, 1950–2050. (Source: UNDESA, *World Population Ageing: Profiles of Ageing 2011* (Geneva, 2011), CD-ROM)

with and without disabilities: improved health care leading to lower infant mortality rates and better health outcomes in older age; and the shift from institutional care to community living [2]. It is also not known whether or not similar shifts in population statistics are occurring amongst people with IDD who have mental health needs. We do know from cross-sectional studies that older persons with IDD have more mental health needs [3, 4] and take more medications for nervous system disorders [5] than younger IDD cohorts; that some people with IDD are at higher risk for dementia than similar age groups of people without IDD [6] and that recent increases in prevalence of Autism Spectrum Disorders will increase prevalence rates of behavioural disorders among affected individuals as children [7] and has already led to older cohorts of people with IDD requiring mental health services [8].

Psychoactive medicines have been used for centuries to address challenging psychiatric symptoms and behavioural disorders in persons with IDD [9]. Much of this practice has occurred in the context of symptom relief or control, with little regard for addressing the underlying psychiatric or behavioural disorders themselves. Frequently, the description of these symptoms emanated from sources other than those of traditional diagnostics, including poorly documented reports from caregivers.

In more than a few cases, the symptoms for which relief was sought may have been an adaptive response by the individual with IDD to life stressors, limited adaptive skills, or inappropriate, non-enriched or restrictive environmental factors or contingencies that might have been prevented by adjustments in care-giving practices. While estimates suggest that approximately 30–50% of persons with IDD are

prescribed psychoactive medications, the actual prevalence of psychiatric disorders in this population is about 3% [9]. The over use of these psychoactive medications is attributed to the faulty belief that challenging behaviours in persons with IDD are associated with psychotic disorders [9–11].

These historical and contemporary practices often set the stage for misuse of psychoactive medicines, ineffective treatment for psychiatric disorder, unnecessary and potentially permanent and even lethal side effects, poly-pharmacy, and a reduction in the individual's functional capabilities. Misapplications of pharmacological treatments may often lead to violations of ethical practice standards issued by professional, governmental, and other regulatory organisations, such as the Federal Medicaid program, the American Psychiatric Association and the American Psychological Association [12]. Such violations include compromising the rights of individuals to appropriate treatment and to pre-treatment informed consent.

There are many reasons for the misuse of psychopharmacologic interventions for persons with IDD. These include well-intentioned attempts to treat psychiatric disorders that have been difficult to diagnose or poorly understood in persons with disabilities. But the list also includes the deliberate use of medicine as a substitute for alternate habilitative settings or practices that might be more expensive or more difficult to provide or for which family members or other caregivers may have insufficient skills, understanding, or training [12]. These conditions have frequently been associated with institutional settings. However, there is a very high risk that they will persist in residential settings as institutions continue to close, and as personnel shortages, unsatisfactory reimbursement mechanisms, lack of health care coverage, and lack of professional experience with IDD in the mental health system increase [12].

Ageing persons with IDD are among the most vulnerable to misuse of psychotropic medications. Older persons with IDD continue to manifest psychiatric disorders that began earlier in their lives. With increasing age, there is an increasing likelihood of changes in metabolic responses to pharmacotherapy, carrying with it a higher risk of medication side effects such as reduced liver and kidney function, confusion, blurred vision and dizziness leading to greater risks of falling and injury [13–17]. There is also high likelihood of age-related functional decline, including symptoms of depression, aggression, agitation or sleep disturbances. These symptoms, coupled with preexisting chronic medical and behavioural disorders, can be mistaken for new psychiatric morbidity, leading to the introduction of new and unnecessary medications [14, 15, 18, 19]. Equally as problematic are attributions by untrained staff in group residences caring for ageing persons with IDD to attribute changes in cognition and behaviour to symptoms of the normal ageing process, and thus delay needed treatment for potentially serious medical and mental health disorders before they worsen and require more aggressive treatment with even higher risks [20]. Taken together, these concerns are heightened by the fact that there are now a substantial number of persons with IDD living in group residences who are ageing [21].

Ethical Considerations

For some time, there has been a growing concern among both scientists and practitioners regarding the ethics of psychopharmacological treatment. Much of the focus has been on research and practice with children and adults with mental disorders [22–25]. Special issues emerge when considering psychopharmacological treatment of persons with IDD. These ethical crossroads can be classified into categories that may not be mutually exclusive. This chapter is designed to raise several ethical, legal, and due process issues in clinical practices involving adults with IDD. We begin with a conceptual discussion of each of these issues and follow with illustrative case studies.

Informed Consent, Proxy Consent, and Right to Refuse Treatment

Psychopharmacological treatment for adults with IDD raises ethical questions regarding patient capacity to give informed, rational, and voluntary consent to standard and experimental treatments. The serious and often permanent side effects of many of these medications raises complex ethical challenges for practitioners, scientists, and family members attempting to balance the obligation to respect the right of adults with intellectual impairments to make autonomous decisions with the need to protect them from poorly informed or incompetent decisions that place their welfare in jeopardy [26].

The moral foundation of informed consent rests upon three principles: Respect for Personhood, Beneficence, and Justice [27]. Respect for personhood acknowledges the right to self-governance and the obligation to protect persons with diminished autonomy from harm. Beneficence reflects a moral and ethical responsibility to maximize potential treatment and research benefits and minimize potential harm and suffering. Justice requires that all members of society share equally in the allocation of services and treatments, their risks and benefits, and that one particular group not bear the burden of risks for other groups. The significance of these principles in practice is evident in the notorious medical experiments conducted at the Willowbrook State School in mid-1950s, which violated each of these moral principles. Clinical investigators wanting to develop a vaccine for infectious hepatitis obtained parental consent under manipulative circumstances and proceeded to intentionally infect children with developmental disabilities as young as 3-years of age residing at the institution with strains of the virus to observe its progression [28]. Moral obligations of beneficence were denied the children who were infected with many later contracting the disease. The principle of respect was violated for parents, who believed they were acting in the child's best interest (as was their legal responsibility) but based their consent on misleading incentives offered by the investigators. Furthermore, justice was ignored, as the children confined to the

institution unfairly carried the burden of risk for a society who later benefited from the findings of these experiments.

Standards for Professional Practice and Research

Professional guidelines and federal regulations draw upon these principles in the creation of ethical standards for medical treatment and biomedical research. These guidelines address several issues related to informed consent and the right to refuse treatment [29]. The Doctrine of Informed Consent (the ethico-clinical principals that protect patient autonomy) has its roots in tort law addressing battery, the act of touching another person without permission [24]. This doctrine requires the provider of care (physician, allied health professional, mental health professional) to not only ascertain the patient's voluntary consent to be treated, but also assure that the decision is made following the provision of adequate information by a person legally competent to make the decision.

Existing case law spells out the definition of these requirements [30] including use of the most current medical or psychological diagnostic criteria in competency assessments of persons with IDD [31]. While informed consent is viewed by law as a contract, the law permits a provider to administer treatment without consent in an emergency, when the person with IDD has waived the right to informed consent, when a legal caretaker cannot be located, or when seeking consent might worsen the condition to be treated [24]. Under the Doctrine of Informed Consent, providers of treatments are legally required to present sufficient information to enable patients to make informed decisions about whether to accept or refuse treatment recommendations or research participation. This information must include: (1) a description of the expected benefits of the treatment; (2) the reasonable risks or adverse reactions; (3) steps to ameliorate discomfort; (4) alternative treatments; (5) and the consequences of no treatment at all. To ensure that valid and informed decisions are made, this information must be presented in a way that is understandable to the person. When consent is sought for a patient or prospective research participant with IDD, practitioners and scientists must tailor disclosure information to the differing levels of comprehension and language ability of the individual and jargon must be avoided [32–34]. The manner in which the information is presented must take into consideration other disabling or age-related conditions such as visual or hearing impairments. For persons with visual impairments, for example, the consent form should be written in large print.

The Doctrine of Informed Consent also requires that consent for treatment is given voluntarily, free from coercion, persuasion, manipulation, and undue influence [35]. Non-voluntary consent decisions violate individual rights of autonomy. The cognitive deficits of persons with IDD, compounded by emotional or personality disorders, restrictions of their physical and social environments, lack of experience and opportunity in making autonomous decisions, and reluctance to refuse requests from others in authority, makes persons with IDD uniquely susceptible to

pressure or coercion to accept treatment [33, 34, 36]. To ensure voluntariness and minimize coercion in research settings, it is recommended that participants with IDD be reminded throughout the study of their right to discontinue participation at any time without penalty [36]. In addition, care providers and clinical investigators must be sensitive to the dynamic of coercion in any given situation and fit their informed consent process to the strengths and vulnerabilities of the person with IDD to understand consent information and the potential exploitative or coercive factors within the context of the consent by modifying their approach to accommodate both [29, 37–39]. Incentives offered for participation must also be selected carefully so as to avoid coercion. Cea and Fisher [33] suggest that prior to offering compensation, clinical investigators consult with the individual, family members, or residential staff to determine the level and type of payment that would be consistent with the person's everyday opportunities.

Almost five decades ago, the President's Commission for the Study of Ethical Problems in Medical and Biomedical Research [40] urged practitioners to avoid determining the incapacity of a decision-maker based on his or her status as having intellectual and developmental disabilities. Assigning capacity to consent simply based on a diagnosis of IDD creates a threshold that can restrict participation or inadequately protect participant rights to decline participation [29, 41]. Since that time, policies have shifted from a protectionist stance of denying persons the right to consent or refuse treatment based on presumptions of incompetency, to placing emphasis on the assessment of decision-making capacity. The capacity requirement of the Doctrine of Informed Consent rests on the ability of the decision-maker to receive, understand, and use disclosure information to make informed and voluntary choices. While adults are legally presumed to have the ability to make competent decisions unless it is determined otherwise, persons with diminished capacity may not always have the ability to do so [42]. In a study of the *MacArthur Competence Assessment Tool for Treatment* [43], Cea and Fisher [32] found that factual understanding of treatment information by persons with IDD is not only linked to intellectual capacity, but also to the degree to which the person has had previous experience with the treatment, the type and manner in which information is given, and to the extent the practitioner educates the person about aspects of the treatment and his or her options.

Irrespective of varying levels of cognitive ability, impaired decisional capacity of persons with IDD may be further influenced by the presence of psychiatric disorders that can be permanent, cyclical, or fluctuate over time. Consequently, the issue of informed consent for treatment or research participation becomes even more complex when it involves persons with dual diagnosis [32, 33]. Due to the large number of individuals with IDD and co-morbid psychiatric disorders, assessment of the emotional state of the person prior to seeking consent is essential to understanding his or her capacity for competent decision making [44, 45]. For older adults with IDD, consideration must also be given to any age-related factors such as dementia or sensory impairments that can negatively impact their decision-making ability, as well as the influence of these disorders on the emotional well-being of the individual.

Many factors associated with IDD may disrupt contractual obligations under the Doctrine of Informed Consent. For example, competence to consent is often not present in young children or in persons with severe IDD. Thus, depending upon the unique characteristics of each patient or prospective research participant, ensuring a fair consent outcome may require the use of surrogate (proxy) or guardian consent [33, 38, 45]. By US law, surrogate decision-makers or guardians appointed by the courts may give consent to treatment or research participation for a person with impaired decision-making ability. Family members and friends appear to struggle the most with their dual commitment to promote autonomy while also keeping their loved one safe [39]. Surrogate decision-makers are obligated to make decisions using substitute judgment based upon their understanding of what the person with the IDD would have decided if he or she were capable of doing so. If preferences are not known, the surrogate makes decisions in the best interest of the individual. Adding to the complexity of this issue are the substantial differences among proxies' in terms of experience, knowledge, training, and background, which often lead to variation in healthcare decision-making. The dependence on proxies and their variation in decision-making contributes to the vulnerability of individuals with IDD [46]. Surrogate decision-makers may give proxy consent to routine medical treatments without involving the courts if the person with intellectual and developmental disability agrees to be treated [33, 34, 44, 45]. In most states, however, if the person with IDD refuses to be treated or if proposed treatments involve "forcible medication with antipsychotic drugs, electroconvulsive therapy, sterilisation, and abortion" (p49) [34] surrogates cannot give proxy consent. A guardian has the legal authority to make best interest decisions either of the person (all daily decisions) or of property (assets and income alone) of the individual with IDD. It is cautioned, however, that individuals acting on behalf of persons with IDD may not always have the individual's best interest in mind [47, 48]. Care taking responsibilities, economic pressure, and other factors may create conflict between the person with IDD and their guardian's interests. For this reason, proxy consent procedures are not considered ethically justifiable for individuals with the ability to communicate a choice, unless the person with IDD agrees that proxy oversight or assistance is a desirable means of protecting his or her interests, his or her assent to participate is sought, and his or her dissent over-rides proxy opinion [37, 38, 49, 50]. Additionally, as this population ages, there is a diminishing likelihood that parents and siblings will be available to assist as proxy decision makers. Exacerbated by the deinstitutionalisation movement increasing the number of individuals moving into community-based systems, nonfamily caregivers are increasingly called upon to assist with healthcare decision-making [9, 46, 49]. In this context, directors of these community based residential facilities often become default healthcare decisions makers for the individuals residing there, and are empowered to consent to a myriad of healthcare treatment decisions in their place [46]. It is estimated that as the number of individuals residing in community housing increases, the role of proxy decision makers will continue to expand.

Decisions regarding the ability of persons with IDD to provide valid consent must take into consideration the individual's vulnerabilities in relation to the

context in which his or her consent is being sought. For example, in restrictive residential settings that offer little chance for independent decision-making, persons with IDD may simply acquiesce to please and not disappoint staff, thus limiting their opportunities to develop decision-making skills that would support self-determination [33]. Viewing consent capacity as a product of both person and context, shifts ethical inquiry away from focusing exclusively on the intellectual and developmental disability of the individual, and to assessment of aspects of the consent setting that may be creating or exacerbating consent vulnerability and to considerations of how the setting can be modified to produce a consent process that best reflects and protects the individual's hopes, values, concerns, and welfare [29, 37].

By utilizing a relational framework [37, 50], care providers and clinical investigators can create a goodness-of-fit between the person and the consent context and thus maximize opportunities for patients or prospective research participants to provide informed, rational and voluntary consent. When such efforts are insufficient to ensure consent competence of persons with IDD, a relational framework also maximizes opportunities for surrogate decision-making that reflect the wishes and concerns of the patient or prospective research participant.

Case Study 1 The following hypothetical case study utilizes the relational approach to address some of the complex questions and ethical challenges faced by care providers and research investigators in balancing the rights to autonomy of persons with IDD while protecting them from harm.

T.J., a 40-year-old man with mild IDD, has been starting fights in the community residence in which he lives. Standard medications and behavioural treatments for aggressive disorders have not helped T.J., and he may have to move to the developmental centre if his behaviours cannot be controlled. T.J. is eligible for research at a nearby hospital testing a new drug for aggressive behaviour. Fearful about being treated by doctors he does not know, he refuses to participate. His supervisor and the treatment team at the residence think that despite T.J.'s concerns, he should be enrolled in the study because it may be his last chance to stay in the residence where feels happy and safe (adapted from Fisher [38]).

T.J.'s case raises some important questions. Prior to a determination of whether T.J.'s decision not to participate in the study is a valid one, the other possible causes of his behaviour need to be explored and ruled out. T.J. should be examined by a medical doctor for the presence of physical problems that may be causing him to act out, and a psychiatric evaluation needs to be conducted to rule out any underlying causes for his behavioural symptoms. If the latter was found to be the case, treatment with medication specifically designed to alleviate T.J.'s psychiatric symptoms, rather than merely mollifying his behaviour, would prevent subjecting him to experimental treatments that may not target his psychiatric symptoms or alleviate his aggression.

Having eliminated these influences on T.J.'s behaviour, an examination of the context in which the behaviour is occurring is necessary. The supervisor and treatment team need to assess the residential environment for antecedents to T.J.'s behaviour and explore whether any modifications can be made at the residence before attempting to enroll him in a study to test an experimental drug with unknown potential. Taking into consideration T.J.'s cognitive limitations, were efforts made to explain all of the issues to him in a manner that he would understand? Did the residential staff discuss their concerns with T.J. about the fact that he might have to leave the residence and help him think about the positive and negative aspects of his decision so that his choice can be made based on facts instead of fears? Were efforts made to lessen T.J.'s fear of new doctors by arranging to have a doctor from the research team visit him at the residence or have T.J. visit the hospital?

In attempting to determine whether T.J. has the capacity to make this particular decision, his right to autonomous decision-making must be weighed in relation to protection of his welfare. If T.J. is presumed competent, the residential supervisor and treatment team would have to respect T.J.'s right to not participate in the research study since the ultimate decision of weighing risks and benefits is his to make. To over-ride his decision-making authority could have harmful practical or personal consequences. T.J.'s legal capacity to consent in other situations may be investigated or his hard-won confidence in his own ability to make decisions might be jeopardized causing repercussions that may be just as dangerous to T.J.'s welfare as the possibility of his leaving the residence [38].

If T.J. is found not competent to make this decision, the residential team might seek proxy consent from a family member or petition the courts to appoint a guardian to make decisions on T.J.'s behalf. Neither the supervisor of the residence nor the treatment team is authorized to make this decision for T.J. Doing so would be a violation of his due process rights. Even if a surrogate decision maker is appointed and T.J. is ultimately enrolled in the study, his assent must be sought. Forcing an individual to participate in treatment or experimental research of unknown risk when he or she is unwilling to do so is ethically unjust and technically illegal [51]. Thus, regardless of T.J.'s capacity for decision-making in this instance, his right to refuse to be enrolled in the study must be respected.

This case study illustrates the importance of viewing consent capacity in the context in which it is sought and engaging adults with questionable capacity as partners in creating respectful and compassionate consent procedures [38]. All persons with IDD are unique. Consent procedures should be based upon an understanding of each prospective patient's or research participant's special characteristics, their consent strengths and weaknesses, life experiences, and practical concerns. Such understanding can be achieved through ongoing dialogue with patients, family members, legal advocates, and practitioners to ensure that consent procedures reflect an ethic of respect and care [38, 50].

Drugs Used for Non-approved Applications with Adults with Intellectual and Developmental Disabilities

Mental health professionals face special ethical challenges in working with children, adolescents, and adults with IDD. The cognitive and emotional characteristics of these individuals, as well as their limited social power and relative lack of legal status make them particularly vulnerable to treatment risks [26, 45]. When prescribing psychoactive drugs to treat mental health disorders, care providers and clinical investigators have a professional obligation to ensure the welfare and safety of vulnerable individuals and minimize potential harm and suffering, by carefully weighing and continuously monitoring risks against benefits for the treatments they provide. Researchers [52] have suggested that an additional responsibility of health professionals is to make sure information about prescribed medications is available and understandable to people with IDD, who are unlikely to seek information about their medications. Frequently, information is given to care takers, who know how to administer the medications, but do not know why they have been prescribed or the adverse side effects [52, 53]. In some cases, patients with IDD may find difficulty verbally communicating their reactions to the medication. As a result, care must be taken to monitor the effects of the drug objectively by defining, observing, and recording behavioural information that accurately reflects the medication benefits and side effects [14, 52, 53].

The pharmacopoeia of psychoactive drugs has grown dramatically over the past 25 years. Drugs are now available for selectively influencing specific neurotransmitter systems and for treating syndromes for which only psychotherapeutic or behavioural treatments were previously available. Unfortunately, few of these medications have been tested specifically for use with people with dual diagnosis of IDD and mental disorders. Very few of these trials have included efficacy or risk assessments specific to older individuals with IDD, because having an intellectual and developmental disability is often an exclusionary criteria for participation in randomised clinical trials. Recruitment challenges, obtaining informed consent, communication difficulties, and an increased sensitivity to medication risks that result from the presence of organic dysfunction have all be cited as reasons for not including persons with IDD or dual diagnosis in drug treatment studies [15].

Some medications, such as Tegretol, Valproate, and Tofranil, may have been approved for treatment of one condition (e.g., seizures or depression) but have been shown in clinical reports to be serendipitously effective in addressing other symptoms (e.g., elevated mood or behavioural dyscontrol). Yet, there are only limited controlled randomised clinical trials of these so-called non-standard uses of medications and these applications lack the U.S. Food and Drug Administration (FDA) approval.

In the mid-1990s, rule changes broadened the number of alternative uses of these medications to generate evidence that drugs approved only for adults may be beneficial for applications in pediatric populations [54]. These provisions for the use of *off-label* drugs do not address the concern that pharmaceutical manufacturers may

simply extrapolate information on pharmacokinetics on adults without IDD for application to adults with dual diagnosis. Doing so fails to recognise significant age-related and intellectual-status differences in many biological parameters that might affect the performance of a drug [15, 55]. Adults with dual diagnosis would benefit from more sound empirical research specifically investigating the effectiveness of treatments tailored to their needs. Yet, there remains a lack of consensus guidelines on issues such as study design, subject enrollment and recruitment, and long-term outcome and risk assessments across these different populations [15, 23].

The non-standard use of a medication, either in terms of target symptom or dosage, has been a significant addition to the intervention spectrum for individuals with IDD. At the same time, conducting randomised clinical trials or other outcome studies involving drugs and behaviour is complicated by difficulties in arguing that the drug's efficacy may justify its non-standard use [9, 15, 48] or that neurological and biological differences between disabled and non-disabled persons, or between one person with a disability and another, are so complex that isolating specific drug-behaviour associations may be impossible. The following two case studies illustrate ethical issues that can arise from the use of non-approved applications of psychopharmacological interventions for persons with IDD in research and treatment situations.

Case Study 2 In the 1990s, Lithium was a new medication for treatment of affective disorders and was still in the early testing stages for different populations. The purpose of this study was to test the efficacy of lithium as a treatment for aggressive and self-injurious behaviours in older adults with IDD (adapted from Fisher and colleagues [22]). A 4-month, parallel group (lithium vs. placebo control), double blind investigation of older adults with IDD living in a residential treatment centre was planned. While the effect of lithium on aggression had been documented in younger populations with typical mental development, there was little empirical support for its efficacy when prescribed for older adults with IDD. Although there was an absence of rigorous scientific validation, as is common in many treatment settings, staff utilized lithium to treat the aggressive and self-injurious behaviours of some of their residents. As a consequence, staff were reluctant to allow individuals with IDD to stop taking the lithium, based on their unsupported belief that it was an effective treatment. Despite the resistance of hospital staff, the investigators decided to use a placebo control group in the study. The Institutional Review Board concurred with this decision recognizing that the staff's belief that research demonstrating the pharmacological agent's efficacy in younger populations without IDD was not a sufficient substitute for direct validation research of older IDD populations. Case Study 2 raises a number of important issues:

Ethical Justification for Using Randomised Placebo Controlled Trials. The first ethical dilemma faced by the investigators was whether to use randomised placebo trials to evaluate the efficacy of a treatment that was clinically established, but not scientifically tested. It is not ethically responsible to deprive research participants of a medication whose benefits have been well documented for that population [56, 57]. On the other hand, continuing to use an empirically unsupported treatment can

also be harmful. When no effective treatments exist, and there is an honest disagreement within the research community about a medication's effectiveness, the use of placebo controls are considered to pose few risks and offer the potential of direct benefit to both treatment and control groups, with the potential of the latter group being prescribed the medication once its effect was demonstrated [29].

Withholding Collateral Treatments. The "Gold Standard for Clinical Trials" requires that internal validity requires that the effects of an experimental drug be studied in the absence of all other psychotropic medications. This presented a second ethical challenge to the researchers, since patients with IDD in the lithium or control condition might be expected to exhibit increased behavioural difficulties when collateral treatments (e.g. Serentil, often used to treat schizophrenia) were withheld. The investigators were concerned that this could expose patients, their peers, and hospital staff to increased risk of self-injurious or assaultive behaviours. Additionally, the investigators questioned the external validity of a study that withheld collateral pharmacological treatment, since it is common in clinical practice to use lithium in concert with other medications (i.e. a tranquilizer). The investigators decided that a clinically and ethically justified approach would be a two-phase design with lithium alone in the first phase, followed by lithium plus collateral medication in the second phase. The clinical staff supported this decision [22].

Monitoring and Protecting Participant Welfare. Meetings were held with residential staff to discuss potential risks of the study that would determine inclusionary and exclusionary criteria of participants and of monitoring procedures. Since the investigators were using a double-blind methodology, procedures had to be developed that would allow the blind to be broken when a participant had a negative reaction to his or her experimental condition [56].The investigators wanted to develop a procedure that would both protect participants from potential experimentally induced harm and avoid breaking a blind precipitously and thus endanger the scientific integrity of the study. One way to accomplish this when a negative reaction was suspected was to compare a patient's current behaviour to his or her baseline behaviour measured prior to the initiation of the clinical trials. If the patient's current behaviour was commensurate with baseline behaviours, in consultation with staff and parents, the blind need not be broken [22]. It was decided therefore that baseline data be collected for each participant and his or her behaviour continually monitored to ensure that it did not fall below baseline levels. A second mechanism developed for protecting participant welfare and the scientific integrity of the study was to avoid potential investigator and staff bias by appointing an independent clinician with no final decision regarding a patient's continued participation [58].

Dissemination of Research Findings. Data on participant reactions to treatment or control conditions has the potential to uncover information that can directly benefit research participants [12]. With regard to the study under discussion, an

older person with IDD's response to lithium or placebo trials and his or her reaction to the withdrawal of collateral medicines can provide valuable information for future treatment decisions. Often when a psychopharmacological study is completed, there is little communication between investigators and hospital staff of information that could directly benefit the treatment of individual research participants. In order to prevent this from happening, at the end of the first and second phases of the study, the investigators prepared a summary letter to the surrogate decision makers or guardians and (with their permission) to the hospital staff [59]. The letter documented the duration and nature of the medication or non-medication condition in which the participant had taken part and described the participant's observed behaviours at baseline, during, and at the end of the experimental period. Where appropriate, treatment based on a participant's reactions to the experimental conditions was recommended [22].

Case Study 3 M.A., a 61-year-old woman with severe IDD living in a supervised community residence, had a life-long history of concomitant over-activity and screaming. Her behavioural problems had been resistant to treatment with numerous psychotropic drug trials and behavioural interventions. Unrelated to her behavioural disorders, M.A. developed bilateral corneal abrasions. The ophthalmologic intervention involved bilateral placement of antibiotic ointment followed by covering each eye for 72 h. Staff was instructed to prevent removal of the patches or any rubbing of her eyes to avoid corneal infections that could result in permanent visual impairment.

The family and the treatment team agreed that M.A. could not be prevented from removing the patches without some type of restrictive intervention. Three choices were posed: (1) maintain M.A.'s least restrictive living conditions with no intervention and subject her to the risk of permanent ophthalmic damage; (2) introduce a behavioural intervention to restrict her eye-touching behaviour, risking failure of the intervention and escalation of her baseline behaviours; or (3) administer medication to sedate her.

The family and the team decided that the risk of M.A. incurring self-induced blindness was too great to not intervene or to use a behavioural intervention with an uncertain outcome. Instead, they chose to sedate her for the 72-h period during which the ophthalmic treatment was in progress. Following approval of the treatment plan by her agency's Human Rights Committee, M.A. was given a benzodiazepine sedative-hypnotic four times per day in a dosage sufficient to sedate her. Staff awakened her every 6–8 h for toileting and to assure that she maintained hydration and food intake. No behavioural outbursts or eye-touching behaviour occurred during the 72-h treatment period. After 72 h, the benzodiazepine was discontinued with no residual side effects. M.A. regained full visual function within several hours following removal of the patches.

This case illustrates one of a very few examples of conditions under which chemical restraint may be considered ethical. The decision-making process utilized in M.A.'s case follows Sprague's (1994) model, which considers ethical treatment to be the treatment that is most humane, most effective, and least risky to the person's health and well-being. In M.A.'s case all three of these conditions were met. The temporary application of a psychopharmacological agent prescribed in a dosage to sedate M.A. was beneficial in allowing her corneal abrasions to heal and in avoiding the potential of further damage that may have resulted in blindness. Thus, the benefits of the treatment far out-weighted the risks. Sprague [48] also recommends that the deliberative process regarding the use of psychotropic medications involve the family, the individual for whom the intervention is planned (if feasible), and the treatment team staff to assure that all options are fully considered. In addition, a tertiary review process of the treatment decision by an impartial body should be conducted. This review body may from time to time serve in a mediation role when all parties to the treatment decision cannot agree on an option.

Analagous to Sprague's [48] latter recommendation, 9 years ago the U.S. Food and Drug Administration (FDA) formed an advisory panel to review the longstanding use of electric shock treatment intended to modify the aggressive and self-injurious behaviour of children and adults with disabilities at the Judge Rotenberg Center (JRC) in Canton, Massachusetts. After hearing testimony from disability rights groups, parents, and students at the JRC, many who condemned the practice as unethical, causing severe physical and emotional pain with questionable results, in 2014 the FDA determined the treatment unsafe and called for an immediate ban of its use. However, at this writing, the FDA has yet to take steps to formally finalize the ban, and as a result, a considerable number of students at the centre continue to receive painful electric shock treatment [60, 61]. The FDA's inaction in this case is in direct conflict with Sprague's (1994) definition of ethical treatment as one that is most humane, most effective, and one that carries the least amount of risk. The JRC has continued to fight any effort to ban the controversial practice. Meanwhile the FDA has proposed banning all electrical stimulation devices for the treatment of self-injurious behaviour and aggressive behaviour in individuals with IDD. Many professional associations, including IASSIDD, have issued strong statements against its use [62].

Diagnostic-Based Pharmacological Treatment

In the field of IDD, most decisions concerning the use of psychoactive pharmacological treatment revolve around behavioural symptomatology. Within that framework, behaviours that threaten physical harm typically receive the greatest attention. In the absence of sufficient empirical data, psychiatrists providing care to persons with IDD often treat aggressive behaviours in persons with IDD in the same way, regardless of collateral diagnosis with psychosis or other disorders [9, 11]. Some have argued that this approach may overlook a bona fide psychiatric

diagnosis more often than not and perhaps lead to selection of the incorrect medication to treat behavioural symptoms, or to trial-and-error pharmacological management—a strategy that has led to ineffective and inappropriate poly-pharmacy in the past [9, 15, 55].

The alternative to treating symptomatology is to treat based upon diagnosis. Sturmey [63] argues that diagnosis-based treatment assumes that either (1) maladaptive behaviours stem from psychiatric disorders; or (2) that maladaptive behaviours are an expression of some underlying disorder. Different psychiatric disorders respond differently to different classes of medication, while often giving rise to similar behaviours. Specific medications should be used to treat specific disorders as often as possible, rather than used as a substitute to mollify generalized behaviours [9, 14].

Sturmey [63] describes a three-stage clinical decision-making model to evaluate the diagnostic basis of behaviour disorder. Stage 1 involves differentiating a true behaviour disorder from developmentally normal behaviours perceived by the caregiver as abnormal. Stage 2 differentiates transient behavioural changes from true disorders. Stage 3 follows a tradition differential diagnostic process to characterize the underlying cause of the behavioural symptoms.

Not every person with a behavioural disorder will be diagnosable, and some individuals may require many visits, extensive observations, and series of laboratory examinations to proceed through the differential diagnosis. Such individuals and their advocates and treatment team staff will invariably present compelling cases for pharmacotherapy based on a best guess diagnosis after only a few visits. Such treatments may even be desirable on a temporary basis if they can stabilize the individual or his or her environment by creating the opportunity to apply behavioural interventions. They should not, however, substitute for pursuit of a differential diagnosis.

Treatment involving application of a psychoactive medication based upon a best-guess cause of behavioural symptoms is often the only option for some individuals. It is complicated by the frequent lack of correspondence between the expected effects of a psychoactive medication based upon data from typical patients versus the effects of that drug on persons with IDD. Hence, the best-guess about a diagnosis may lead to the choice of a medication to which the individual has an idiosyncratic and unfruitful response. The inevitable trial-and-error medicating of behavioural symptoms may actually be the best approach in certain cases. But the process must be carefully controlled and monitored.

Supported Decision-Making

Recent international trends propose a paradigm shift from plenary guardianship protections to a less restrictive option that would allow people with IDD the ability to maintain their autonomy in decision-making situations. Language to support the supported decision-making (SDM) approach is found in Article 12 of the United

Nations Convention on the Rights of People with Disabilities [64] that upholds the right of persons with IDD to legal capacity in all aspects of life, and by holding States Parties responsible for developing appropriate measures that allow persons with IDD access to the supports they need to exercise their legal capacity [65, 66]. The underlying premise of SDM is that everyone, irrespective of level of intellectual ability, has the legal right to participate in decision making and to express their preferences using trusted supporters to help explain issues, explore options, and support their preferences [67, 68].

Although SDM under Article 12 has gained global attention, and a wide spectrum of possible models have been suggested, its more formal recognition is less prevalent [47, 65, 68–71]. Proponents of SDM propose that it can serve to protect autonomy and enhance self-determination in persons with IDD by providing them greater opportunities for choice making [47, 65, 66]. Others suggest that until the concept of SDM is precisely defined, questions regarding how the process would function and whether it would lead to self-determination cannot be empirically determined [70, 72]. Bigby and colleagues [66, 68] argue that regardless of the formal acknowledgement of decision-making support for persons with IDD from a legal perspective, it is equally important to garner empirical evidence on what works to ensure that the values and preferences of the person are at the centre of the decision-making process.

Drawing on a program of empirical research in Australia, Bigby and colleagues [66, 68] are the first to systematically address the complexities of supporting persons with cognitive disabilities in decision making contexts. They developed and piloted the Support for Decision-Making Practice Framework that provides capacity building tools and resources to enable supporters to understand and act on the will and preferences of the individual they are assisting, including his or her cognitive and communicative limitations, the context for the decision, and the level of support needed. Should it be determined that SDM not be appropriate for a specific individual, Bigby and colleagues [66, 68] suggest that information gathered using the framework can serve to inform surrogate decision-making by court appointed legal guardians. A larger program evaluating the efficacy of the framework is ongoing to determine its applicability across the diverse contexts in which support for decision-maker occurs [66].

Further Direction

As ethical values and attitudes regarding the rights of adults with IDD in decision-making situations continue to evolve, tensions remain on how best to support their autonomy, while ensuring their well-being from the consequences of decisions that are not adequately made. Intrinsic to the person with IDD are inherent characteristics that can influence the quality of their decision-making ability, including limited levels of cognitive ability, communication difficulties, restrictive environments, vulnerability to manipulation, and a willingness to please others [33, 34, 36, 70].

Some of these tensions will be realized regardless of the decision-making approach taken. Whether the SDM process alone can address the needs of older adults with IDD, with or without mental health issues, or whether it can be formally integrated into existing guardianship policies will need to be considered. As we grapple with how to best reduce the risks and enhance decision-making opportunities for persons with IDD, their perspectives are essential. Giving voice to persons with IDD will help ensure that treatment and research reflect their preferences and respect their rights and welfare.

Conclusion

When prescribing psychoactive medications to persons with IDD, practitioners and clinical investigators are obligated to ensure their welfare and safety and minimize harm by continually weighing and monitoring risks against benefits for medications prescribed. As much as possible, these medications should be used in the treatment of diagnostically based psychiatric disorders and not merely to relieve behavioural and other symptoms for which no clear cause is evident.

The use of psychopharmacological medications in treating older adults with IDD also raises complex ethical questions regarding their capacity to give informed, rational, and voluntary consent to standard and experimental treatments. Regardless of level of intellectual ability, the decisional capacity of these individuals may be further impaired by the same psychiatric disorder that the medication is intended to treat and by age related factors such as dementia or sensory impairments. All persons with IDD are unique. Individual capabilities as well as situational circumstances must be weighed and their implications to informed consent clearly understood.

If it is determined that an individual with IDD is unable to make treatment decisions for him or herself, consent should be sought from a surrogate decision-maker or guardian who makes the decision in accordance with the preferences of the individual or in his or her best interest. It is important to note that with increasing age and enhanced medical care, there is a likelihood that relatives may not be available to assist in these decisions and alternative means of obtaining the preferred or best interest decisions for the ageing person with IDD will need to be explored.

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Chapter 15
Psychosocial Concerns Among Ageing Family Caregivers

Philip McCallion and Lisa A. Ferretti

Introduction

Despite the funding and policy emphasis in most countries on out of home care, the most prevalent care of adults with intellectual and developmental disabilities (IDD) has been family caregiving, with many of those caregivers themselves ageing [1, 2]. United States datasets suggest that over 70% of individuals with IDD live with their families [2], further studies suggest that at least 20% live with siblings [3], and many more receive supports from siblings regardless of where they live [4].

Family caregiving in the family home, at all ages, has been reported to enhance quality of life for a person with IDD [5]. It has also been found to ensure, at least for some, greater likelihood that desires for community living, participation and integration are realized [6], with benefits for both the person cared for and the caregiver [7–9]. People live in communities as a result where they are known, with community resources they routinely access, and where the family is likely to have established linkages; for all ageing adults characteristics likely to support ageing in place and successful ageing [10].

The literature on caregiving has tended to talk about "primary caregivers" and for people with IDD these have been identified as primarily female and as parents but with a growing group of siblings [3, 11, 12]. The conceptualisation of primary misses that caregiving is more complex; that there are male caregivers too, grandparents, siblings, nieces, nephews and other relatives and friends play key supportive even if more limited roles, and that caregiving success is often about the caregiver feeling supported, not just the individual being cared for.

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Caregiving Challenges

As society changes so too does caregiving, caregiver availability and caregiving supporters' willingness and ability to assist. There are worldwide trends for increased participation of women in the out of home workforce, increases in single parent families, fewer children in individual families, continued movement from rural to urban communities, increases in families in poverty and greater geographic dispersal of smaller family networks [12, 13]. These demographic challenges are impacting the availability of primary and replacement caregivers, but perhaps are of most concern for the availability of supporters that primary caregivers rely upon. Equally, the shrinking pool of caregivers and caregiver supports is further challenged by changes for primary caregivers themselves. As they age many older caregivers are also assuming additional caregiving for

- spouses,
- adult children perhaps with mental health or substance abuse issues
- and/or of grandchildren where the parent generation in not available or is incapable of providing care [14].

Potential and actual sibling supporters may also be acquiring additional caregiving responsibilities [12]. Concerns and stress for caregivers may be less about a caregiving situation they know well and where they have experienced success and more about what is changing about themselves and the support networks around them [15].

Theories Guiding Responses to Caregiving Challenges

Theories of stress and coping, at the individual level and at the family level, have been borrowed from the general ageing and caregiving literatures [16] to understand the psychosocial needs and challenges of family caregivers of persons with IDD.

In the transactional theory of stress and coping (TTSC), Lazarus [17] summarized, "The way people evaluate what is happening with respect to their well-being, and the way they cope with it influences whether psychological stress will result, and its intensity" (p. 6). An important addition to his approach to stress (in this case caregiving stress) was that stress also affects the well-being of close others.

There are also family-level stress and coping constructs such as Hill's [18] ABC-X model. McCubbin and Patterson [19, 20] have taken the model further exploring why some family caregivers adapt and thrive through stress while others disintegrate in similar stress situations.

In TTSC, when a person feels that the stressful demands of caregiving outweigh their resources or ability to cope they will be overwhelmed; i.e., the person's

perception and interpretation of the stressful event may actually be more critical than the event itself [17, 21]. This is called the "primary appraisal," where the individual appraises the threat's significance; if it is seen as harmful or challenging. In a secondary appraisal they determine if the stress is controllable and if they have the coping resources (inner strength or will power; support from family, peers and professionals) to address it. Coping strategies then can be [22] problem-focused (when they believe they can manage or solve the problem), emotion-based (when they must cope but do not have the tools and support) or meaning-focused (where beliefs, and values—e.g., god's will—help provide positive meaning in the stressful situation that then motivates and sustains coping and well-being) [23].

Interventions using stress and coping ideas tend to emphasis changing appraisals of the situations and valuing coping strategies that work. These interventions may be delivered to individuals but are more often delivered in small groups of individuals facing similar caregiving challenges as the interactions with others experiencing many of the same stresses offers examples of different types of coping as well as peer validation of the stresses experienced and confirmation of the value in the moment of the strategies chosen. The groups may also be a safe place to consider, try out and get feedback on trying new appraisals of situations and for accepting that approaches chosen are not working.

Family stress and coping theories focus on indicators of positive family strategies and traits such as the extent of emotional bonding among family members, the ability among family members as circumstances change to alter role relationships and relationship rules in response to the stress of increasing or changing care needs or reduced caregiving ability as a caregiver ages. In the related ABC-X model—

- *A* stands for a stressor, i.e., an event requiring a response;
- *B* stands for resources or coping assets and strategies which may be sufficient for some stresses and be overwhelmed by others;
- *C* stands for the family's interpretation of the event, a challenge to be overcome or a catastrophe to be endured;
- *X*, stands for the crisis, or stressful event.

The ABC-X may also be the basis for individual counseling but group interventions may involve other family members working on their collective stress approaches or considering new strategies and the engagement of alternate coping assets (family assets not previously engaged; new strategies with existing assets or more openness to formal services). In some cases and for similar reasons as in TTSC a group intervention may include peer family caregivers.

TTSC and ABC-X are long established approaches to supporting caregivers but the types of issues considered and the coping work undertaken in individual and group meetings has been changing because of changing family roles, cultural issues, opportunities to access ageing services, transition challenges and the need for long-term planning.

Changing Family Roles

Siblings have always played a role in the care of their family members with IDD [24]. Although Burke and colleagues [25] report that female siblings are more likely to assume higher levels of caregiving when parents are no longer able., Zendell, in a national survey sample found a much fuller range of caregiving roles for siblings regardless of gender [4]. In addition, there are changing gender roles among parents. Traditionally, women have been the primary caregivers for persons with IDD. In more traditional gender role times, fathers worked to support the family, while mothers stayed at home. In the ageing years although initially research reports were that that these patterns continued, with fathers unlikely to take on additional responsibilities [26], these gender patterns are now changing. Yet, gender role stereotypes persist and, for example, some primary caregiving fathers report feeling isolated and judged by assumptions that caregiving is a mother's role [2, 15, 27].

What does emerge in these situations is that it is likely that older adults with IDD being cared for at home are more likely to have been brought up in one earner homes and are ageing in homes with limited savings and limited pension income adding to the stress situation for some families. In countries where there are carer allowances or where caregivers can receive payment from personal services budgets, those payments rarely make up this financial shortfall. Un-researched is the impact of reductions in income at retirement, or in previously two parent households of the loss of the primary earner, and/or of the person who drove a car if both parents did not drive. Assigned gender roles is not the only challenge.

Cultural Issues

While persons of all cultures, ethnicities and races have IDD, it has been documented in the past that families of some cultural groups are less likely to receive services from public disabilities agencies [28]. Mainstream disabilities agencies were found to poorly accommodate the needs of diverse families [28]. Equally, where services were provided, a lack of openness to family structures and ways of caring that are different from those of agency workers have not always been respected, and there has not been recognition that different values and structures may mean that different services are needed and desired [27, 28]. In particular, the emphasis of self-determination and the needs of the person with IDD being separate from the family reflect policy and historic desires to move from institution and family to independent community lives guided by the values of western white societies and histories of service scandals. Other cultural and ethnic groups were not part of this history and often but not exclusively have values that place greater value on family connectedness and family caregiving [29]. Lack of cultural sensitivity and competence may add to the isolation and challenges faced by caregiving families.

Ageing Caregiver Services

To the extent that there were services, care of persons with IDD and supports for their caregivers have remained largely within the purview of disabilities service providers. In general, there is at least ambivalence about support for family caregivers. Some countries do provide carer or caregiver allowances and there have been case finding efforts, in others, particularly to identify families unknown to the system where if an ageing caregiver is suddenly unable to provide care, there is likely to be a crisis. However, there has been both subtle and explicit policy and service beliefs that the provision of too "high" a level of service/support may lead to families relinquishing care creating an unprepared for burden on public services [27, 29].

In the U.S. National Family Caregiver programs, funded by the Administration on Aging/Administration on Community Living have included family caregivers of persons with IDD as a targeted population for Area Agencies on Aging. Services and supports are now provided but the ideas of not too "high" a level remain. There is also a recognition on the ageing services side that support of these caregivers, particularly if they too are ageing is part of the mandate for ageing agencies; there is still ambivalence for the disability services side that "disability" funds should be used for caregivers who themselves do not meet criteria for disability.

New solutions are needed that focus on the caregiving family unit, offer genuine integration between ageing focused and disability focused service providers, address intersystem barriers and genuinely recognise the value to society of families who continue to provide care [15].

There are examples of change. A greater emphasis in disability funding on home and community based services, use of consumer/participant directed services and the provision of personal budgets for purchasing services have led to

- some family members receiving wages for care given,
- the ability to support environmental modifications that benefit all family members
- giving both persons with IDD and family caregivers as proxies greater decision-making power over what is an appropriate expense.

Nevertheless, there remain concerns about such decision-making [30] with fears expressed concerning financial exploitation, improper use of funds and lack of oversight to ensure that services are adequate. This despite there being no evidence of additional risk for those in such programs [31].

Transition Challenges

In the past when persons with IDD were not expected to live into old age, parents would most usually outlive their offspring and offer a lifetime of care. Instead, ageing parents increasingly cope with their own health problems and eventual death [32]. This changed demographic reality has raised concerns about who will care but

has not always been accompanied by changing views of families and family caregiving. Responses are more likely of the one-size-fits-all type which usually means movement of the individual with IDD to an out-of-home placement and/or provision of more formal services in the home. For many situations, this is likely to be the best choice.

This is a crisis viewpoint seeing caregiving as doomed to fail. The priority has been to assist families with transitions and to have formal arrangements in place for the adult when his or her parent becomes incapacitated and/or dies. This philosophy of care mandate largely persists even as available out of home placements decline [12] and continues to be a major thrust of interventions. There is little recognition of the caregiving strength of family members even as they cope with their own diminishing health [24].

The ideas posited by Folkman, Lazarus, McCubbin and others that appraisal of a caregiving stress and differences among caregivers in terms of their coping resources suggest a more case-by-case even person-centred approach to supports for caregivers. Perhaps, for example, there should be less judgement that the mutual emotional and financial support workers often encounter in caregiving homes are not in the best interest of the individual with IDD [15].

Long-Term Planning for Families and Adults with Intellectual/Developmental Disabilities

Previously it was not uncommon for parents to be averse to creating detailed future care plans for their adult children with IDD [3]. That aversion often reflected stress, concerns for safety, and anxiety including regarding speaking of future living arrangements with other family members [15]. Nevertheless, four types of long-term plans have emerged as interventions: explicit succession plans transferring the responsibilities of overseeing care of the adult with IDD to an appointed person; implicit succession plans; financial plans; and residential plans. As legal frameworks around consent and adulthood have changed the issue of planning for guardianship has risen in importance. While family member or state guardianship used to be the norm for people with IDD, it is now used more specifically to protect individual rights using full, partial, or temporary guardianship, such as for help in managing finances or need for medical care. Planning for guardianship is often discussed in the context of future healthcare planning.

An individual with IDD who cannot express an understanding of a complicated medical issue has barriers to demonstrating consent. This necessitates some arrangement of surrogate medical decision-making, depending on an individual's ability to understand and make complicated medical choices. Although guardianship can help protect individuals with IDD, such as in allowing them to access medical care, it may also limit self-determination. Other complicated issues that individuals and parents might want to plan for include advance directives, such as a living will [27].

There is a need going forward that in helping families to understand the future of their caregiving and the person they care for, it may no longer be about movement to an out-of-home setting. Instead interventions are needed that target maintaining the person in their existing home; accessing services for both the individual with IDD and the caregiver; involvement of other family members, neighbors, and friends through "circles of support" [33], rather than as the person care will be transferred to; and the purchasing of services from a range of providers [3, 34]. Equally, the lauded and needed support of self-determination for the individual with IDD and support of caregivers with the stresses of caregiving, concern for the future and management of the caregiver's own health and mortality concerns all calls for new and different services. A strengths perspective, health promotion programming, work on greater community inclusion, the valuing of mutual support, and counselling to address disappointments, concerns and fears for both are yet to be available and widespread [35].

Engaging Caregivers in Interventions

As mentioned, there is a tendency for interventions to be either individual or group delivered. Services such as case management offered by public services or private agencies are often presented as interventions but are experienced by families as controlling and inadequate [24]. More recent person-centred planning strategies are often seen as more inclusive but the focus is upon the person with IDD, families may not always perceive themselves as treated as important and their needs are rarely included as a focus. Consumer or participant directed services and personal budget approaches also maintain a focus on the person with IDD but the ability of the primary caregiver to function when needed as a proxy decision-maker and greater consideration of household needs and possible payments for family members do mean a greater sense of involvement. A critical divide is that disability focused services do not see the welfare of the caregiver as their primary concern. For older caregivers this role may be played by ageing services but this sector is less well-resourced and services are rarely mandated. Limited respite services, information and referral and counseling is available and in some jurisdictions person-centred counseling is being initiated to deal more globally with long term care needs of older adults including caregivers. Access to individual counseling for caregivers tends to be privately paid, and cognitive behaviour strategies consistent with TTSC and ABC-X framework are most prevalent. There is little information on extensiveness of availability of such counseling or on uptake. However, the need to fund personally must necessarily limit availability and use. There is more evidence of use of group interventions and these primarily are family counseling, future planning groups and meetings, and peer and professional support groups. More recently there has been growth in online groups and communities.

Family counseling tends to take two forms. The first is a family meeting facilitated by a case manager or other health professional to address an immediate or

urgent challenge such as the primary caregiver going into hospital and care alternatives being immediately needed. The purpose is short-term and a time and place where immediate decisions will be made. It is unlikely that longer term planning will occur as this may not be seen by the case manager as her/his role and because discussing longer term issues may be too disruptive or polarizing and distract from the short-term need. Longer-term issues may be discussed in subsequent meetings if both the family and the facilitator are willing. Success will require a willingness by family members including the family caregiver to participate. The person with IDD should also participate if possible but not all family members may be ready for this involvement. Effective processes often require preparatory work and education including of the person with IDD, meeting with smaller groups and/or individuals to understand influential issues and histories and a framework to organize the meetings.

Futures planning is one framework around which to organize such meetings. Across a range of studies a number of domains of concern have emerged: housing, legal planning, identification of next primary caregiver(s), financial planning, day-to-day care, medical management, end of life care and transportation [12, 36–38]. A number of guides have emerged (see for example https://arcwi.org/content/uploads/sites/17/2017/12/WI-Future-Planning-Training-Binder.pdf and https://www.mentalhealth.org.uk/learning-disabilities/our-work/family-friends-community/thinking-ahead) that may be the basis for organizing a series of meetings with identified caregivers. Significant results both in terms of advancing future plans and in reducing stress for caregivers have also been reported [38–40]. However, numbers of participants in the reported studies are small and it has not been established that the programs can be widely replicated or that there are funding streams to support implementation.

A major challenge for all interventions is the extent to which the needs and experiences of caregivers of persons with IDD are unique and whether those caregivers may be accommodated within other caregiver group interventions. In a review of the caregiving intervention literature Toseland and colleagues [41] found that there is a broad variety of types of caregiver support groups and feeling that “my” issues are being addressed are critical to attendance and success. It is therefore likely that caregivers of persons with IDD will want to attend groups focused on their issues rather than general caregiving groups. One area where there are interesting emerging findings are health promotion groups that include caregivers and most recently include the person with IDD being cared for. Here, improved health has been found for both caregivers [42] and the person with IDD [35].

The absence of funding streams also means it is difficult to maintain professionally led groups. Here the overall findings are that both peer led and professionally led groups are successful; peer led groups do a somewhat better job of offering support and professionally led groups tend to offer more education [41]. Most of the research on futures planning in particular and on caregiver groups in general for caregivers of persons with IDD has been with professionally led groups. Findings by Heller and Caldwell [39] support the value of peer-led futures planning groups.

A criticism of the research to date is that there is still a tendency in the futures planning and other intervention literature to continue to approach caregiving solely/primarily from the perspective of supporting the person with IDD. Again the literature on caregiver support in general has a greater focus upon addressing the physical, mental health and support needs of the caregiver, and exploring resilience, coping and self-efficacy [41]. Unanswered throughout the literature is how to address the reality that those who would most benefit from support, counseling and intervention are the most stressed caregivers, i.e., those without the time, transportation, respite and alternate caregiver needed to be able to attend the meeting.

It is for this reason that the recent emergence of listservs, social media pages and online groups is of importance. The most visible is SIBNET https://www.sibling-support.org/connect-with-others-sibs/meeting_other_sibs_online/sibnet a virtual community designed to support siblings to respond to lifelong and ever-changing relationship needs with their sibling with special needs. There are also less developed and resourced virtual links among caregivers of unique forms of disability, for example caregivers of persons with Down syndrome and dementia. As these groups have evolved they now offer online support groups, workshops and training and are not just sources of information, as important as that is. These newer forms of intervention offer the greatest opportunity to reach the hard to reach and stressed caregiver. Attention will be needed however, to ensure that culturally diverse and less financially resourced caregivers are also included.

Conclusion

Nations across the world have always been dependent upon care by families to serve the greatest number of people with IDD and as longevity increases that includes ageing people with IDD. Challenges for continuing caregiving are being increased by the ageing and health concerns both for the caregiver and for the person with IDD as well as by growing financial and care challenges, and changes in size, dispersal and economic activity of caregivers and their family networks. Interventions even when successful, are reaching few caregivers, tend to be both under-resourced and professional rather than peer-led and are at best ambivalent about meeting the caregiver's needs. Health promotion may offer new opportunities to address caregiver psychosocial and other concerns.

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Chapter 16
Role of Assistive Technology

Gregory William McGrew

Introduction

Throughout this book, it is made clear that the number of people considered to have intellectual and developmental disabilities (IDD) is on the rise; especially the ageing population (over 55 years of age). It is also clear that many efforts are beginning to be underway to address the varied needs of this older IDD population; involving a broad range of human services and the structures, government policies, and in the institutions that support them. Technology is playing an ever more significant role in how individuals conduct their lives. It's role in addressing ageing health and wellness needs in the general population is increasing dramatically [1]. New and more powerful medical devices for prevention, diagnosis, and treatment of ailments and diseases are developed and produced in growing numbers [2]. The same can be said for technologies devoted to how we communicate with others. Smartphones, while having plateaued somewhat in computing power, are still evolving to provide more convenient and intuitive ways to stay in touch with friends, family, and caregivers. Even more so that in the 2020 Covid-19 pandemic.

Technology may prove in the coming years to enhance the independence and quality of life of persons with IDD, particularly in the areas of health care and social engagement. This chapter will initially focus on the types of needs and barriers faced by older adults with IDD. Subsequently some of the assistive technology (AT) products and devices developed to address these needs will be reviewed as well as providing additional resources for these products. This chapter will not list and

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detail such products as there is an abundance of readily accessible information online. This chapter will be devoted to describing their potential role in the lives of older persons with IDD, how they can facilitate greater independence for the user, and how they can provide greater confidence for caregivers that their friends or family members are safe, secure, and are having their health related needs met. The significant challenges associated with applying technological aids and solutions to the needs of ageing individuals with IDD will be discussed.

The end of the chapter will list and characterize key precepts that should be followed when looking for technology products and devices for persons with IDD, particularly those ageing with dementia and age-related problems.This includes a discussion of a supportive infrastructure that should be in place to:

1. ensure the proper 'fit' of products and systems to the needs and capabilities of the user(s), and
2. address adjustments and repairs as needed for whatever products and systems are being used.

Needs of Older Persons with IDD

The general population have a reasonable concept of the types of physical, sensory, and to a lesser extent, cognitive declines associated with ageing. This is particularly true of people like the author, who are approaching 'seniority' and starting to recognise incremental reductions in functional acuity in these areas. Other chapters (chapters "Psychopathology in Older Age", "Assessment of Psychiatric Disorders", "Neurocognitive Disorders Assessments", "Depression in Ageing Adults" and "Behavioural Manifestations of Medical Conditions") in this book will highlight the mental health diagnoses associated with such declines, and the physiology behind them. Beyond referring to medical diagnoses for context, this chapter will focus discussions on the physical, sensory, and cognitive nature of functional decline (including that posed by dementia) as they impact the lives of the elderly who experience them. Table 16.1 links these disabilities with associate functional limitations.

How can technology 'pick up the slack' for functional declines? The sensory and cognitive declines to which we refer tend to limit the amount and quality of information an individual receives. For example an older person with IDD and dementia does not always recognise his carer when they come to visit him. Is there technology that could recognise them, and tell him who they are?

More generally, are there technologies that could supplement the limited or unreliable information received? Could this additional, supplemental information help to mitigate the effects of such declines on their lives?

1. If someone has reduced vision, is their technology that can sense what would be in their visual field (and even beyond) and communicate information the user needs to engage/avoid/react to what is around them and where it is located?
2. If they are hearing impaired, is there technology that can alert them when the telephone rings, when someone is at their front door, or when their dinner is done in the oven or microwave?

Table 16.1 Disabilities and their influence on function in daily life

Ageing related disabilities, and their influence on daily life	
Cognitive	
Disability	*Functional manifestations*
• Reduced mental acuity	• Increases reaction times
• Difficulties in short term memory	• Increases risk of forgetting/missing appointments • Increases risk of forgetting/missing medications • Increases risk ignoring situations requiring attention/action
• Difficulty in interpreting information	• Takes longer to comprehend meanings • Impairs the ability to accurately appraise situations • Leads to confusion, leading to inaction • Leads to inappropriate or unsafe actions
• Prone to distraction	• Impairs the capacity to focus attention • Impairs the ability to concentrate
Physical	
• Reduced muscle strength • Reduced muscle coordination • Increased reaction time	• Increases risk of falls • Increases risk of dropping objects • Increases risk of injury through muscle/tendon/ligament strain • Reduces capacity for independent mobility
Sensory	
• Hearing • Vision • Tactile sensitivity • Proprioception • Olfactory sensitivity • Taste	• Reduces environmental awareness, leading to: – Missed communications – Missed opportunities – Unsafe actions or inactions – Inappropriate responses to environmental stimuli – Lack of response to environmental stimuli

3. If their prospective memory is affected and worsening, might be there technology that will alert them that the oven has been left on or time to get ready for bed?

Currently, there are products on the market capable of providing such assistance, and more and better ones being developed as you read this. For the three scenarios listed above, here are examples to technology products that address respectively these issues.

1. The WeWALK smart cane [https://wewalk.io/en/]
2. Braci PRO [https://www.youtube.com/watch?v=rpVKTN1y0oA]
3. Wallflower Stove Monitor [https://wallflower.com/]

The nature and variety of the disability characteristics in Table 16.1 need to be taken into account by product designers and manufacturers if they aim to provide products that can be used successfully. Certainly not all older adults with IDD are dealing with significant memory loss, impaired vision, or major muscle weakness, many are struggling with these and other physical, sensory, and cognitive impairments as they try to maintain their functional independence. Products designed for this segment of the population need to provide users with accessible controls and interfaces, facilitating their successful use by individuals with such disabilities.

Technologies Available

As discussed earlier in the chapter, resources for finding technology products, systems, and strategies for enhancing the quality of life of older persons in the general population are abundant and reasonably simple to find on the internet. If you perform a Google search with the specific phrase "AT for seniors" you get over 40,000 hits, with first 100 or so being ads for and articles on products and devices to help older individuals maintain their independence. There is an ever-growing variety of aids and devices designed specifically to help individuals maintain their independence with respect to various activities. Many, if not most of these products address needs associated with the loss of physical or sensory abilities, and are part a category of AT devices designed to help individuals "age in place", i.e., stay in one's home, instead of going to an assisted living or a nursing home. The functional focus of these products is to facilitate ability to independently carry out various daily living tasks they may not otherwise be able to perform due to physical and/or sensory decline. Used in the home, these would include products like stair lifts [Bruno.com], tilting toilet seats [EZACCESS.com], adaptive kitchenware [Rehabmart.com], adaptive landline phones [Independentliving.com], and many other types of functional aids and devices [Amazon.com].

However, products designed to address functional limitations associated with cognitive decline, while less abundant, are increasing in number at an accelerated pace. This is due in part to the evolution of the digital technologies on which these products are based. Increases in the variety, power, speed, and affordability of sensors and digital processors have made it feasible to create devices that identify and respond to needs of ageing individuals who, for example, have forgotten to turn off an appliance, or missed taking a medication, or could not recognise a family member by sight. These technical advances, as well as the accelerated growth in the market (number of older adults who may benefit from these products), have all combined to stimulate the fast- paced growth of product development in this area. In the Forbes online magazineof February 2018, Paul Irving calls the ageing population "a dynamic emerging market", and says they have more financial resources to spend on maintaining health and independence.

These types of AT products and systems will be discussed, but it is important to highlight first an underlying precept which is strongly tied to the eventual success of new and emerging ATs. That is, the input from actual users and stakeholders, particularly of persons with IDD into these products. Not only in their design, but earlier in the development process when they are actually conceived as how they will be of benefit when used.

One example illustrating early user involvement in product design regards medication reminders. Medication compliance is one of the most common concerns of older adults and their caregivers [3]. Given the large number of older adults needing help with medication compliance, developers have come up with a broad variety of products to address this need, based on their assumption that memory decline due to ageing is the significant contributor to medication non-compliance. Most of these

products are designed to remind individuals to take their medications at specific times during the day, and in some cases, they will dispense these medications at the time prescribed. Many of the reminder/notification products are in the form of apps for smartphones which carers or high functioning adults can use. They provide an option for setting notifications or alarms throughout the day cueing users and caregivers to take a particular medication at the appropriate time. Some offer the user the ability to record their own voice (or someone they choose) to provide the audio notification of what, when, and how much of a specific medication needs to be taken.

Beyond smartphone apps, a number of products in this category actually store and dispense the medications at prescribed times. The additional features of storing and dispensing one's medications can be beneficial to the user by consolidating all medications they take into one container, and by providing not only notification but actually provide access to the right medication—and the right one only—at the correct time. While these devices offer more features than smartphone apps, they can be more complicated to set up properly, making it more difficult for users and caregivers to ensure that the right medications are dispensed at the correct time each day. As you can surmise, there are a plethora of products on the market devoted to this one focus: ensuring an individual knows what medications to take at what time.

Given the large number of products available to deal with medication non-compliance, one might assume this problem is well managed and soon to be conquered. However, statistics on the current state of medication compliance for older adults tell a different story. While these products do help a large number of individuals with impaired cognition issues recognise what to take and when to take it, non-compliance continues to be a significant problem. Why is this? Studies show that medication non-compliance has much more to do with choice. While many individuals still do not take their medications as prescribed, it is less often because they forget to take them, but more often because they **choose not** to take them [4]. This is a significant issue in the IDD population.

Although medication reminders (e.g. smartphone apps) do not address the underlying cause of non-compliance, often many devices are difficult to set up and/or to use properly, particularly for many older adults. With respect to the design of these products, the user interface for persons with IDD—the device's buttons, knobs, switches and screens the user manipulates to operate the device—is often too complex, difficult to see, lacks feedback, and/or is foreign to the user's previous experience, such that the device is unusable for them. The frequent lack of literacy in people with IDD is also a barrier. The design flaws highlight the importance for developers to gain a reliable, relevant understanding of the population they are targeting with their products. This can only be achieved by engaging potential users in assessment of designs and prototypes, preferably throughout the development process, to ensure the product is usable and satisfying for such users when it carries out its prescribed functions.

However, what options may be on the horizon for the ageing IDD population looking for technological solutions to age-related barriers they are experiencing? Assistive technology has not been widely tested in older persons with IDD, thus examples of good practice are not readily available. That said, the functional

manifestations of cognitive impairment associated with dementia in the general population are often similar to those experienced by those in persons with IDD and dementia, then it can be addressed in some cases by the same technologies. Below is how AT can benefit an older person in the general population:

Jim is 73 years of age. Aside from his small dachshund, Marshall, he lives alone in a suburban condominium complex. His condominium has two bedrooms. It also has an attached garage. He owns a small car and drives occasionally to shop for groceries or to a recreation centre nearby where he has a fitness membership. He moves a bit slowly due to some arthritis but is independently mobile without a walker or cane. Jim has some cognitive disabilities associated with age, and possibly some mild dementia. For him, these conditions manifest in occasional memory lapses. He may not recall something he needs to do, or to remember, and/or he may remember something inaccurately, mistaking one thing (person, task, time, item, etc.) for another.

Jim takes several medications for his arthritis and a heart arrhythmia. And he tries to watch his diet fairly closely to avoid certain foods. Jim eats out occasionally, but usually prepares his meals at home. When fixing hot meals, he often uses his microwave oven, but occasional cooks something on the stove or heats something in the oven. After dinner, he sometimes likes to watch a movie or TV show that he can stream through Amazon. He also likes to touch base with his daughter on the east coast and see how her new job is working out. He has some medications he takes each night. Finally, he likes to make sure the exterior doors are locked before he goes to bed.

Several apps on Jim's phone help him with various daily living needs. To help him remember to take his medications, Jim has an app on his smartphone that notifies him when to take which pills and how many (Medsafe Medication Reminder app). Another phone application tells him if he has left his garage door open. It allows Jim to shut the garage using his phone, if he has a phone signal, from wherever he is (Tailwind iQ3 Smart Garage Door Opener). At night when Jim goes to bed, he gets a notification on his phone that his front door is not locked. He locks it from his phone (Yale Assure Lock SL). Helping Jim maintain a supply of healthy food items, he also has an app he uses when he goes grocery shopping that works with his "smart" refrigerator (Samsung Family Hub Smart refrigerator). The refrigerator tells the app what items are running low so Jim can purchase them. He uses another app at the store that tells him what items may be appropriate—or inappropriate—for his dietary needs. And lastly, he has "smart" programmable dog food feeder with an app that tells him whether there is food out, and allows him to dispense food to his dog, anytime from anywhere (WESTLINK 7L feeder).

There are a growing number of "smart" devices available for the home that can be controlled through the internet via a 'hub'. For a smart home system, the hub is the brain of the system that takes in the information from various products, appliances, and sensors around the house, as well as requests and instructions from others wirelessly connected to the hub. This would include caregivers and family members. Jim can use an Amazon Echo hub that allows him to speak to "Alexa" to handle many of his needs. He can give verbal commands to these various devices

through Alexa when he wants to control his television, control room temperature, open window blinds, turn on lights in various rooms, lock or unlock his front door, or contact a friend or caregiver to say that he needs assistance. And Amazon Echo is not the only system that can be used to do this. Apple and Microsoft each have their own systems for voice control of devices through the internet.

Emerging Technologies

There is significant research and development being carried out, not just in the United States but in Europe and Asia as well, on new and emerging AT applications to enhance quality of life for ageing persons with IDD. Two of these technologies and their applications in smart home development are briefly discussed here.

Sensor-based artificial intelligence (AI) and augmented reality (AR) both offer significant potential to enhance quality of life and independence. AI refers to decision making by computer-controlled devices based on information they receive from sensors in their environment. Driverless cars incorporate AI to steer the car away from obstacles, or stop it completely if appropriate. Sensors in the frame of the car sense the presence and location of an obstacle and tell the car's computer to activate some software that makes the car avoid the obstacle. In a smart home with AI, sensors may be located in the home to discern when someone enters or exits a room. This information may then be used by a smart home hub (described earlier) to tell the lights in that room to turn on or off. Another sensor in the kitchen may let the hub know if someone left a burner on the stove after a pot was removed. The hub may then alert the resident that is the case, or it may just turn the stove burner off, depending on how it is programmed. There are a wide variety of sensors available for sensing things like heat, light, motion, vibration, etc. With a controlling computer programmed to perform certain tasks based on something a sensor picks up, you can start to imagine the variety of assistive actions that can be performed in a home that might help someone with IDD.

Augmented reality occurs when a user of a mobile device or wearable device (like smart glasses) sees not only an actual real-time image of his or her surrounding through the mobile or wearable device, but also can receive supplemental images, text, and voice through the mobile device as well. The images or text are superimposed on the real surroundings image. These supplemental images can be provided through a computer by another individual, or by sensors in the mobile device or the surroundings. There is currently research being carried out that is looking at using AR as a tool for providing people with cognitive disabilities supplemental information and guidance as they engage in activities of daily living or work-related activities.

Both AI and AR tools, infrastructure and applications are being developed at an accelerated pace, there application to barriers faced by people with disabilities will be included in their evolutionary journey. As this scenario suggests, there are a number of technology devices and products that may help ageing individuals with IDD

remain independent in their home. However, let us look at what also must happen for these devices and their users to reach this potential.

Challenges Associated with Applying Technological Aids and Solutions

There are an ever-increasing number of technology products designed to enhance the independence, safety, and quality of life. However, there remain many unanswered questions about the efficacy of many of these products, and about the effectiveness of the strategies on which their functions are based. This is, in part, associated with the significant challenges developers face in designing, producing, and in selling such products for this demographic at a financially sustainable level. These challenges conspire to create engineering and business landscapes for product developers that are ever changing and unpredictable. Consider the following.

Over the past 40 years the accelerated evolution of digital technologies and their impact on our daily lives has eclipsed the level of technological change that occurred over the preceding 400 years. Roughly 40 years ago we experienced the arrival of the first personal computer. Now, over 80% of Americans own a powerful handheld computer called the smartphone [5]. The consumer technology landscape that the current generation of 65–75 year olds have experienced and matured in is very different than that of people over 75 years. Not long ago the assumption was that individuals with IDD as a group had little experience with digital technology, and were therefore resistant to adopting technological aids and devices of which they had little understanding, and were ill equipped to learn how to use. Now, a growing IDD population, particularly younger persons can contribute with experience—sometimes, extensive experience—with digital technologies. While this is obviously a positive situation for younger persons, it makes it more difficult for designers to produce a product that works effectively for the older IDD population as a whole given the broad variety of technical experiences represented in such a large diverse group of individuals.

The most significant challenge for product developers in designing technologies for older persons who have IDD is the diversity of needs inherent in the heterogeneity of this population, independent of age. That diversity manifests in not only the nature, severity and number of people's disabilities [6] but the cultural and life experiences people bring to the table [7]. Older individuals with IDD can have multiple disabilities, or just cognitive issues. The demographic has a broad variety of wants and needs. And they have a broad variety of skills and experiences. The challenge in providing effective, usable AT to the IDD population is in how to address diversity of needs as also seen in the general population [8].

While there are tricky defiances for product designers coming up with effective products, there are plenty of difficult challenges facing those responsible for seeing that the product or system works influenced by multiple stakeholders (caregivers,

healthcare providers, family members, etc.), each with their own priorities. The paid caregiver's role of an older person with IDD and what they think is important regarding that person's independence may be very different than what a family member may consider important.

Studies have pointed out a challenge often not considered when introducing and applying new technical solutions to functional limitations. Older persons with IDD, particularly those with severe cognitive disabilities find themselves undergoing significant upheavals around their living situation. They are often uprooted from their homes to unfamiliar surroundings, which can significantly increase the effects and impacts of even minor memory loss, as well as disrupt the effectiveness of formerly successful AT interventions. Even for the general population a situation where ATs and smart systems worked effectively at home can change significantly in a new location [9]. The AT may not be applicable in the new home as configured in the former residence, it may not address new functional needs at the new home, or simply may not work at all due to constraint on internet access.

Another problem is the dynamic nature of dementia onset and its cognitive manifestations. The decline in functions for an ageing person with IDD and dementia is typically progressive, which can cause ATs become ineffectual over time, but step changes in function, both for and against, can also occur that in certain situations can render AT solutions either ineffective or just in the way.

It should also be noted that there is little in the way of dependable research-based guidance available to older persons with IDD and associated stakeholders on what products and systems work well and produce good, independence enhancing outcomes for this population. There are a wide variety of devices and systems available but little valid info for caregivers on what works best. Finally, there is a concern that these ATs are being developed by many in response to those that see an ageing population as a costly burden on resources, and something that should be and can be at least in part fixed with the application of appropriate technologies. It would be tragic for people with IDD who grow old, if such concerns undermined technology research and development efforts in this field.

Precepts for Effective Application of Technology

This chapter has shown the potential benefits of current and emerging AT for addressing compromises in independence that face the ageing IDD population. There has also been a discussion outlining the difficulties faced by product developers in coming up with effective technical solutions for the functional challenges this population must deal with, and challenges that face an ageing individual with IDD and their family and caregivers with regard to setting up ATs and ensuring they work for them. The technologists and designers have more and more tools at their disposal to address many of the challenges they face, including 3D printers, computer modeling software, and cheaper and cheaper sensor technologies. But family members and caregivers of older persons with IDD need tools of their own to make their

efforts in applying these technologies a success. There are precepts listed and described here that if followed, will increase the likelihood an AT application to enhance the independence and quality of life and to be beneficial for an individual.

1. ***Do not assume that people aging with IDD are less likely to accept technology solutions enabling independent living.***

   Many believe that people aging with IDD are generally less likely to want to use AT in their daily life. While this may have been true 20 years ago, current experience does not support this view. When confronted with the opportunity to try technology that may enhance their lives in some way, most aging individuals with IDD, as per the ageing general population, are quite receptive, and in some cases, eager to engage with technology [10]. Studies have found that the older population of today are just as accepting, or in some cases, more accepting and interested in technology solutions to functional barriers they may face. This may be so for the aging IDD population [8].
2. ***Involve all stakeholders in the discussion leading to a choice and implementation of an AT solution***

   One way to gauge the efficacy of AT interventions for people with IDD is to assess the level or frequency of abandonment of AT products by those who acquire them. While there can be many reasons why someone may choose to stop using a product, a significant contributor to abandonment is because the product's use ceases to benefit the user. One study [11] on AT abandonment found four factors related to this phenomenon; *poor device performance, change in user needs/priorities, easy device procurement* and *lack of consideration of user opinion in selection.*

   These findings suggest that technology-related policies and services need to emphasize consumer involvement and long-term needs of consumers to reduce device abandonment and enhance consumer satisfaction. But the need for consumer involvement applies at least as much, if it is not more so, to those who have a stake in the efficacy of a particular AT application for the benefit of a particular user. It is in this circumstance where not involving all parties who will engage with or be affected by the use of the technology can lead to unforeseen disruptions of its effectiveness and usefulness for the end user.
3. ***Focus the AT intervention on facilitating tasks or activities related to the person's interests and priorities***

   If people are heavily motivated to find a way to perform a task or activity, they will be more likely to exert the effort required to become competent with AT needed to do so [12]. Success can then beget success. Competence and confidence with one technology aid can help mitigate the user's future reluctance to try new, more advanced technical solutions to other issues down the road. Caregivers and other stakeholders should be aware that their take on what is best for the user regarding tasks they can accomplish with technology may not mesh with the priorities of the person with IDD. Efforts should be made to identify the

user's wants and needs, and to meet them in as much as it is safe and practical to do so.

4. ***In addition to the end user, stakeholders should learn how to use the technology as well***

   Stakeholder engagement and support of technology use is often critical to successful adoption and deployment of ATs. The more family, friends, and other caregivers know about the technology, how it is used, and how it can enhance the independence of the person with IDD, the more likely they will be to provide that support. To that end, having someone knowledgeable demonstrate the technology and it's operation to stakeholders, as well as answer questions they may have about its use, maintenance, etc., can be effective in making this happen. This can also be helpful in clarifying and addressing areas related to technology use where stakeholders have differing opinions on what independence needs are most important.

5. ***Have a clear path and process for addressing technical glitches that may (will) come up with the AT***

   Depending on the complexity of the device(s), a stakeholder with technical experience may be sufficient to fix many issues that come up. For more complex problems, contacting the distributor or manufacturer about the problem may be necessary. In any event, this should be clear to the person or persons responsible for ensuring it continues to work for the user.

6. ***Engage a knowledgeable professional who is experienced in assessing the technology needs of people with disabilities for an AT evaluation***

   The previous five precepts make a solid case for technology users to be properly assessed for these technologies [13]. This needs to be done by skilled, knowledgeable professionals who can best determine with the user,

   (a) what needs should be addressed with these technologies,
   (b) what technologies would be best suited to address those needs for this user given their level and nature of cognitive disability, the other stakeholders involved in such use, and funding available for these products.

Conclusion

It is important to note that there is much that needs to be done in research and development concerning the needs of the ageing IDD population discussed here, particularly on how products using the emerging technologies discussed earlier can best be designed to maximize both function and usability for this growing and diverse population. This should be very exciting work, given the tremendous potential these technologies offer to enhance the independence and quality of life for older persons with IDD.

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Chapter 17
Healthy Ageing and Healthspan: Planning Ageing Throughout Life

Flavia H. Santos and Patricia C. Heyn

Introduction

The United Nations estimated in the World Population Prospects 2019 that in the next 30 years there will be 1.5 billion people aged 65 years or over worldwide out of the 9.7 billion estimated to reach, with higher prevalence of women at the older age [1]. This is the outcome of the intricated combination between increase on longevity and decrease in mortality rates at in younger ages. The Sustainable Developmental Goals (SDG) stratify eight geographic regions in the globe, known as SDG regions, including 235 countries demarcated by the *Standard Country or Area Codes for Statistical Use* of the United Nations Statistics Division. This division seems more suitable to account for inequalities and plan customised solutions. The Fig. 17.1 represents the estimated percentage of people aged 65 year or more in these SDG regions contrasting four time points.

In the last two decades ageing become a core topic globally mobilizing researchers all over the world. Right now, there are more older adults worldwide than children under age five. Life expectancy still growing in all eight areas and faster, for instance, Sub-Saharan Africa is estimated to achieve 68.5-year-old in 2050, contrasting with Australia/New Zealand that will achieve 87-year-old in average in the

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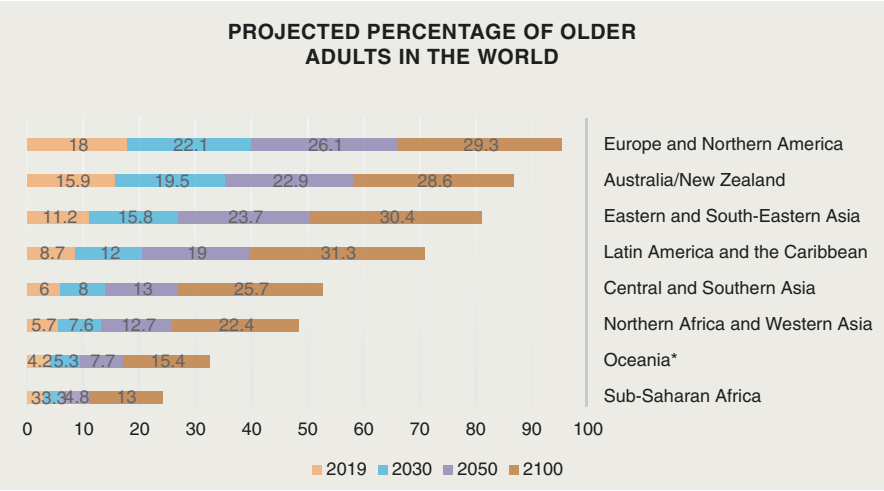

(*) Oceania refers to Oceania excluding Australia and New Zealand territories. Adapted from (1). Estimative based on medium-variant projection.

Fig. 17.1 Growth Projection for the population aged 65 years or over per SDG regions in four moments

same period [1]. The pace of ageing is faster within developing countries than it was in developed countries in the past [2]. Besides, individuals at oldest-older age, i.e., over 80 years of age will also be more frequent [3] and susceptible to geriatric syndromes. But individuals and societies do not seem clearly mindful of its impact. Do you realize that in 30 years, one in five individuals in the world will have 60 years or more? Are our societies developing a supportive environment for older adults? The readiness seems to vary from country to country.

According to Dr. Margaret Chan "*many common perceptions and assumptions about older people are based on outdated stereotypes*" ([4], p. vii). It means that ageing cannot be strictly defined by chronological age it is rather the outcome of the individual trajectory across lifespan. Besides, ageing neither is a synonym of dependence nor it is the absence of diseases. The probability of dying in consequence of cardiovascular disease, cancer, diabetes or chronic respiratory disease in people aged 30–70 years decreased to 18% worldwide in 2016 [5]. It means that the access to health care allows people to achieve quality of life by keeping clinical conditions such as chronical and noncommunicable diseases under control. Besides, certain conditions are socially determined, i.e., endowing people with internal and external resources to allow behaviour change associated with smoking, diet and alcohol which are potentiating further diseases [6].

Ageing is so relevant for the world that taps in 15 out of the 17 the SDGs. Nevertheless, we in this chapter will focus on the Sustainable Goal 3—**Health and Well-being** (SDG3). These terms appear together to highlight that even when there is a disease, i.e., an unhealthy state we should target to the individual's welfare that is feeling comfortable into their socioemotional state. The SDG3 prioritises health for all individuals in all stages of life. Also, the SDG3 aims to ensure that "*all*

people and communities can use the promotive, preventive, curative, rehabilitative and palliative health services they need" ([7], p. 5) having substantial quality without impose financial onus for the users. It is a fact that people are living longer but are they living healthier? If we contrast with the previous decade, the answer is promising, for instance, maternal and neonatal mortality dropped in all regions. However, the major cause of mortality still unsafe drinking water, unsafe sanitation and lack of hygiene. Besides, preventable diseases still uncontrolled causing deaths that could be avoided. The paucity of resources produces these outcomes, for instance, in several places the ratio of physicians, nurses and midwives per inhabitants is far lower than the need [5]. Therefore, these SDG regions have unequal resources, policies, and development consequently we cannot generalise outcomes [8].

The biopsychosocial nature of ageing in undeniable. It is necessarily driven by genetic, environmental, and social factors. Consequently, ageing must be understood as a unique experience, an individual phenomenon. Each transition may vary across domains, for instance, preserved cognition combined with frailty, the domains may have plateaus which might show rapid or lent decline with time. However, race, gender and socioeconomic factors are shaping these transitions. For instance, race and ethnic are predictors of mental health, protecting or potentiating neuropsychiatric disorders. As an example, the incidence of dementia in African Americans and American Indian/Alaska Natives is higher than in Asian Americans among other groups [9]. Of course, these factors overlap the impact of education, culture and socioeconomic factors [10]. Indeed, Siqueira and colleagues [11] pointed the need to enlarge the body of research targeting the screening of mild neurocognitive disorder on illiterate patients and/or patients with lower educational levels to avoid false positives diagnosis.

In fact, education per se is an important predictor of life expectancy. Lutz and Kebede [12] studied life expectancy in 174 countries, among them developed ones, considering the years of schooling of individuals at age 15 or older, merging datasets of educational attainment plus income (GDP per person) and mortality rates for the period 1970–2015. Multivariate analysis indicated that, income becomes insignificant while the education effect remains robust and highly significant. The study also indicated that women's education (aged 20 to 39) is also a stronger predictor of child's mortality than income. Authors concluded that "*that education should be considered a policy priority for improving global health*" ([12], p. 358).

Older individuals are more affected by chronical diseases and disabilities, consequently with higher demands on health care, which require specialised assistance applied for mental and physical health, including the need of long-term care usually at the end of life. Data based on individuals aged 50–64 years from 16 countries predicted that the lower is the education the higher is the exposure to disability risks, with higher impact for women regarding daily living activities, personal care, functional and working. It goes without saying that disability increases with age with worse outcomes for women because older women are more exposed to poverty risks [13].

Low income is associated with worse outcomes on life expectancy and health condition in a late stage of life. Because it is also associated with behavioural risk

factors (smoking and alcohol addiction and poor nutrition) and poor living and working conditions [13]. The lower is the socioeconomic background the higher is the need of support for activities of daily living on ageing [14]. Twenty-three percent of older adults have incomes below the poverty line across the OECD countries; about 10% of women aged 66 year or older had an income below the poverty level, against 7% for men [13]. For instance, globally there are more women than men respectively at ages 65 or over and 80 and over and this trend remains until 2100 projection [1]. Nevertheless, the women longevity does not correspond to ageing healthier [8]. Because in several countries women still having scarce access to sexual and reproductive health care, reduced levels of education and a lack of social security and participation in political process. Moreover, women are a frequent target of domestic violence [1] and usualy assume the role as family caregivers which reduce their own opportunities on career development; becoming digitally illiterate or outdated regarding technology use [15].

Early retirement is no longer a trend. The number of older workers in Europe, i.e., people age 50 years or more has increased since the 1990 decade. In fact, the OECD average rate of employment for the age-group 55–64 in 2016 achieved 59.2% being similar in North American countries, according to data of OECD Employment [16]. Some European countries, such as Sweden, Norway and Switzerland, this rate surpass 70% being equivalent to Japan's rate. However, the hire rate did not increase over the period of 2000–2016 which means that if an older worker is fired is very difficult to obtain a new position (rate variation from 8.9 to 9.2% in the four time points). A common factor among OECD countries is that employers prefer to contract younger employers, even when both candidates share the equivalent qualifications. A systematic review contrasting 17 countries indicated that older workers, regardless if high or low-skilled, need to apply for jobs two or three times more than younger workers [17]. In other words, despite the policies, the ageism prevails in several cultures.

Besides, in countries with low pension coverage workers tend to continue working after age 65 whereas in countries with high pension coverage people retire earlier. In other words, in most countries classified as "very high" human development 89.4% of population receive old age pension contrasting with 9.8 in countries having "low" human development [18]. In this sense, the quality of health on ageing, is strongly modulated by social barriers, such as economic arrangements, distribution of power, gender equity, policy frameworks and the values of society [6]. Consequently, we may change outcomes by changing social constraints during the life course, since perinatal period and even recognising the impact of intergenerational transmission of health inequities [19].

The expression "health inequities" refers to those inequalities that could be dodged. Such "*inequities in health are widespread, persistent, unnecessary and unjust*" ([6], p. xiii). In order to address health inequities is important to target the to the 'causes of the causes': the conditions in which people are born, grow, live, work and age and inequities in power, money and resources that give rise to them. Facing those challenges demand vigorous actions from health and social care systems within and between countries fighting the underlying health inequities.

Therefore, political and societal engagement with this cause is necessary in order to develop the appropriate policies according to the stages of development of each country or SDG region.

It is time to think of ageing as a dynamic process of endogenous and exogenous cumulative circumstances rather than a moment in time [20], consequently, it is possible to plan in individual and societal levels the kind of ageing a person will have. It is difficult to understand the research state of art in this field because most studies are not comparable due to conceptual and methodological reasons. The concept of successful ageing dates from 1990 decade and it was an attempt to reduce "ageism", which is a negative view of ageing process focused on degenerative processes and disease. It reflects the continuity, despite of age, in social and professional engagement sustained by physical and cognitive functionality [21].

Updating terminology, Bryant and colleagues ([20], p. 940) summarise **healthy ageing** as the "*ability to go and do meaningful activity*". The WHO defines healthy ageing as "*the process of developing and maintaining the functional ability that enables well-being in older age*" ([4], p. 5). Key elements of healthy ageing are physical and mental health but taking into account the environment characteristics. The latter includes cognitive, psychological and social aspects. In this sense, achieve a healthy ageing should be a choice and a plan of actions towards an effective transition and living with autonomy and well-being at older age. Moreover, it should be inclusive for individuals who are losing skills at disabling levels.

Thinking that a person may still active and productive during ageing is a watershed. Then, the concept of **Functionality** becomes central. Functionality on ageing is important for individual well-being, but it is also a contribution to society. Since the decade of 1980, the WHO has been concerned to the dialectic between functioning and disability, and the need to emphasise health and functioning rather than disability. It means to focus on what people can do, based on their body function and structure, capacity and performance in activities, despite the presence of disabilities that hinder engagement and opportunities [22]. In the current view, functionality covers individual's physical, mental and psychosocial capacities to interact with the demands of the setting where the person lives [23].

In order to change perspectives on ageing two international policy were released in 2002 (Madrid International Plan of Action on Ageing [24] and the WHO's policy framework on active ageing [25]) aiming to guarantee the right to health and to improve the health of older people. Because these initiatives were not broadly delivered, the global strategy and action plan on ageing and health was traced aiming to make healthy ageing a public health priority.

The **Global Strategy and Action Plan** [26] has five main goals: (1) Commitment to action on Healthy Ageing in every country, (2) Developing age-friendly environments, (3) Aligning health systems to the needs of older populations, (4) Developing sustainable and equitable systems for long-term care and (5) Improving measurement, monitoring and research on Healthy Ageing. These goals are feasible and measurable also they are ruled by principles such as human rights, equality, equity and solidarity. In the follow up of its release evidence-based findings were gathered for 5 years.

These initiatives aim to boost a massive campaign of appropriated actions to be worldwide implemented during the **Decade of Healthy Ageing** or 'the Decade, - from 2020 to 2030 - within individual, societal and governmental levels. It is important to keep in mind that the Decade was the only alternative to accelerate the change in order to achieve the Agenda 2030. Absolutely all sectors must be adapted due to ageing needs: Labour and financial markets, education, housing, health, long-term care, social protection, transportation, information and communication, including family structures and intergenerational ties [14]. Actions impact will be granted if the modus operandi changes across the globe, in other words, actions must transform the ecosystem.

We hallmark that the Decade was proposed exactly because the countries did not develop the readiness and investments necessary despite alerts in previous era. Essentially, there is no Healthy Ageing for all without the accomplishment of all SDGs. Because health is also to live with safety, dignity and equality. In other words, health depends on a legacy of a better planet for all, having the SDGs as pillars. In practical terms, awareness and adaptation of society, geriatric care and economical resources are necessary to health promotion and health care for older adults. Different from other age bands, older adults demand more chronic care—targeting frailty, malnutrition, dementia and geriatric syndromes—rather than acute care. Because older adults will be the majority in the world, we must think of them as a community and as such in adapted environment for their needs. The term environment in this context includes physical, social and economic parameters, which reflect into different sectors such as social protection, housing, transportation, information and communication, apart from the expected health, long-term care [14].

The **first action** is to combat ageism that is *stereotyping, prejudice and discrimination towards people based on age* [27]. The negative attitudes towards ageing influence both public policy (education, labour, health care etc) and public attitudes and behaviours [14]. The ageism has emotional, cognitive and behavioural dimensions, which are observed in the society, organisations and even in the person who is getting old. These views are barriers for employment, because older workers may be seen as inflexible, limited, physically and mentally less capable and these stereotypes are stronger than the positive ones, such as being reliable, friendly, experienced, skilled, etc. [28]. Individuals at older age cannot be considered a burden as they support economy by paying for taxes and social security, as consumers, and even remaining at the work force. In fact, at the workforce is necessary to guarantee to older people opportunities to renovate their professional skills [14].

The **second action** is to boost health care by promoting scientific and medical innovations and developing robust treatments and assistive technologies accessible to all [29]. These conditions will favour people to thrive achieving their maximum potential and keeping autonomy and yield as much as possible. Nevertheless, it must guarantee as well long-term care centred for those who present decline in capacity. Individuals who need supports may also engage in Access to Services and Long-term care. Ageing brings chronical diseases and particularities such as frailty, malnutrition, dementia and geriatric syndromes. Research is targeting to prevent and compensate the cognitive decline, but it is necessary to transform these

scientific findings into services that reach the communities, changing the perspective regarding supports and caregiving practising [14].

The Decade aims to advance in three major areas: (1) create age-friendly communities, (2) provide person-centred integrated care, i.e., that responds to individual preferences, needs and values [7] and (3) develop community-based social care and support. These areas are interrelated and aim to produce programs and policies that will overcome inequities [14]. The key idea is age with good health and in practical terms the actions should: (1) Identify capacities and resources of older adults; (2) meet the needs and preferences of older adults, (3) respect older adult's decisions and lifestyle choices; (4) protect older adults living in vulnerable circumstances; (5) reduce inequities; and (6) include older adults in different areas of community life as active agents [14]. It encompasses different areas where the individuals may explore. The most known areas are: Physical Activity, Diet and Nutrition, Social Inclusion and Participation along with Education and Life-long Learning. But there are also others equally relevant to be considered: Employment and Volunteering, New Technologies and Environment and Accessibility.

The **Platform on Population Ageing** will be the scenario for the communication among all sectors. For example, the European Summit on Innovation for Active & Healthy Ageing 2015 [30] included people from government, civil society, investment and finance, industry and academia to discuss opportunities for economic growth and social development in the embrace of the demographic change in Europe. Delegates agreed that innovation is the way focusing on co-financing projects, allowing rapid transfer among country members and mobilising new public investments crosswise Europe. The Summit defended silver economy[1] strategies in order to facilitate joint initiatives on age-friendly homes, promoting silver tourism and loan and financial schemes to connect and adapt care at home. Another front is to boost the start-ups targeting research, science and innovation which will create new related jobs. Research partnerships is essential for all these developments. Implementation depends on synergy towards technology and essentially research partnerships among scientists and corporations [30]. The platform must also to reach the community, listen to older people, their families and household carers, advocacy and facilitators. This is the only way to make a sustainable change [14].

Finally, the idea of healthy ageing also encompasses the well-being. It means that ageing must assure to all individuals an opportunity to participate in activities which give life meaning. In other words, older people must participate in the economic, social, cultural and political life of their societies [24]. It means that we cannot make decisions for older people but rather we should listen their opinion, in order to build and implement an appropriate Global Strategy and Action Plan. As the saying: Listen to the voice of the experience!

[1] Silver economy is the system of production, distribution and consumption of goods and services aimed at using the purchasing potential of older and ageing people and satisfying their consumption, living and health needs.

Health Promotion, Wellness, and Ageing

Healthcare that is supported by health promotion and wellness services is in great demand; much of this need will be prompted by the ageing of the Baby Boomers, who in 2030 will be 61 million between 66 to 84 years old. In addition to the Baby Boomers, those born prior to 1946—the "oldest old"—will number nine million people in 2030. This demographic change will increase the number of people who will age into disability as well as ageing with disability [31].

As we reconcile the diverse health needs of the growing ageing population, healthcare and residential areas will need to be creative and transformative and it will be easily rammed. Wellness programs will need to advance from current workplace health promotion initiatives to include communities and medical facilities [32]. Wellness can be defined as a positive health behaviour and culture structured around positive health gain and maintenance to support well-being for the prevention of diseases with the overarching goal to achieve a rewarding and satisfying quality of life [33]. It integrates equally important well-being dimensions (mental, physical, social, spiritual, emotional, environmental, intellectual, occupational) based on self-autonomy, self-responsibility and self-motivation [34]. It guides the person to consciously and optimistically adopt good behaviours such as healthy eating, physical fitness, stress management, hygiene, and environmental and safety awareness. The original wellness model consisted of six principal elements (life dimensions), including societal, rational, spiritual, physical, emotional, and occupational wellness [34]. The origin of the "Wellness" concept, especially the term, "High Level Wellness," is credited to Dr. Halbert L. Dunn, who published his "High Level Wellness" book in 1961. He emphasized that the definition of health should be a positive one instead of health meaning the "absence of disease." [35, 36]. Since Dr. Dunn's book release, the wellness movement grew and took greater societal significance, the wellness domains that gave birth to early the wellness movement (Fig. 17.2) were expanded to include additional domains that were recognised to impact one's health and well-being such as money, education, and the environment [37].

One billion people, or 15% of the world's population, experience some form of disability, and disability prevalence is higher in developing countries. One-fifth of the estimated global total, or between 110 million and 190 million people, experience significant disabilities [38]. While these conditions are diverse and some are due to pediatric onset disability (i.e. Down syndrome) or trauma (e.g., spinal cord injury), many are the result of common chronic diseases such as arthritis, heart disease, and Alzheimer's disease [39]. Individuals with disabilities are at higher risk for developing secondary health conditions that exacerbate functional and health decline [40].

Several interventions aimed to alter the trajectory of functional and health decline in individuals with chronic disabling conditions have informed the last 20 years of health promotion and wellness advancements for people with disabilities [41]. Many studies generated innovative interventions to promote the health (rather than

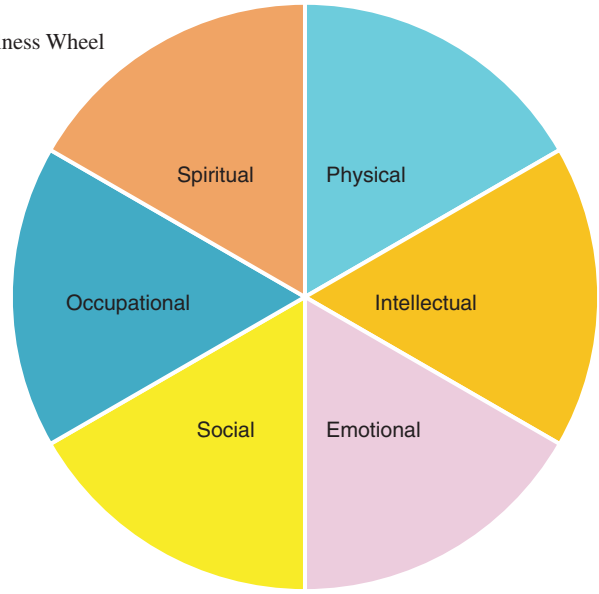

Fig. 17.2 Wellness Wheel

manage the disease or condition) of persons with chronic disabling conditions [42]. Some interventions have focused on a single behaviour such as exercise or stress management, while others have taken a more comprehensive "lifestyle" approach [43]. As a group, these interventions clearly reflect the philosophical perspective of the International Classification of Functioning, Disability and Health that individuals with ageing chronic and disabling conditions (e.g. Intellectual disability, traumatic brain injury) are fully capable of being healthy and experiencing a good quality of life [44].

Unfortunately, advanced age is the main risk factor for many chronic and prevalent diseases such as cancer, cardiovascular disease, diabetes, hypertension as well as neurodegenerative diseases like Parkinson's and Alzheimer's disease [45]. Therefore, it is critical for adults with IDD to adhere to a healthy lifestyle by participating in health promotion activities such as exercise, recreational and outdoor games, and good nutrition [43]. Health promotion approaches should be part of the healthy ageing plan and a standard of care for all individuals with disabilities, especially as they grow older. Starting a healthy lifestyle at any age, including old age, can have powerful effects on well-being and can impact positively a person's independent living, mental health, physical functioning and ambulation as well as societal engagement [46]. Physical activity has many powerful effects in the body, and it can help with many physical changes that occur when a person gets older such as loss of muscle (sarcopenia) and bone (osteopenia), cardiorespiratory weakness, and insulin resistance [47]. In addition, there is strong evidence supporting the beneficial effects of exercise training for cognitive health, including for people with Alzheimer's Disease [48–50].

As people age, they tend to have a harder time falling asleep and more trouble staying asleep than when they were younger, and it is a common misconception that sleep declines with age [51]. In fact, research supports that sleep behaviour needs to remain constant throughout adulthood for proper mental and physical health [51]. Good lifestyle and health behaviours are essential for good physical, mental and sleep health [52].

It is unrealistic to think about healthy ageing, especially for people with IDD, without the learning, practice, and adoption of a healthy lifestyle. Ideally, the focus should be in increasing and sustaining good health throughout life for health-span rather than increasing lifespan with poor health and low quality of life [53]. To properly incorporate wellness into a person's daily lifestyle, a comprehensive health risks and quality of life assessment needs to be incorporated into health programs for people with IDD. The person's current health status and past medical history, current lifestyle behaviours (i.e. diet and physical activity levels), and medications should be adequately appraised to assess what factors and challenges are impacting the person's health [54]. The evaluation can be done by administering standard surveys, or health promotion tools, designed to identify risk factors associated with early disease development, including self-administration of lifestyle behaviour appraisal questions, which also provide clues to health areas needing improvements [55]. The focus should be in understanding oneself and one's capabilities to motivate changes in the health-related areas that need attention and enhancements, such as exercising regularly or eating healthier [56].

Older Adults with Intellectual and Developmental Disabilities and Healthy Ageing

People with IDD are in disadvantage in contrast to the general population regarding access to social and health services and this is particularly relevant once they are older.While deinstitutionalisation is trending worldwide, institutionalisation remains a common practise in many countries, which reduces the visibility of those individuals with severe and profound IDD and their access to health care. Many aspects may be neglected such as housing, rehabilitative services and health care may not be adaptive to account the transition in their needs.

In 1946, the World Health Organisation recognised that everyone has the right to enjoy high levels of physical, mental and social well-being (high level wellness, health and happiness), regardless of their socioeconomic circumstances, age, health status, ethnicity or beliefs [57]. Decades of research have provided a good understanding of the factors that facilitate high level wellness, including healthy food, physical activity, supportive relationships and equitable access to healthcare, education, and resources [46, 58]. However, relatively few people appear to be flourishing [59, 60], suggesting that this growing body of knowledge on wellness determinants

has not translated into 'high level wellness for all'. Unfortunately, people with IDD have even less opportunities to participate in health promotion activities and wellness-based health services than most, as they get older, they get even less prospects to become healthier and happier. Actually, their risk of developing chronic diseases and geriatric diseases increases with age, and it can start as early as the age of 24 years old [40]. Hence, we are a long way from high level of wellness for all, as acknowledged by the World Health Organisation [57, 61].

If "successful ageing" is defined as a long-lived positive and enriched life due to "smart" healthy lifestyle choices that can prevent disease and disability, maintain or enhance physical and mental function, and enrich social engagement [21], individuals with IDD are already in disadvantage to achieve the desirable higher levels of wellness for ultimate healthy ageing outcomes. Individuals with IDD, usually are deprived of opportunities to participate in wellness-based and purposeful activities (physically, cognitively, socially and professionally) due to economic, health, and cultural barriers. Therefore, they are achieving less internal and external resources to assist with their "successful ageing road map" [21]. Individuals with IDD have the same, or even higher, risks of developing geriatric diseases such as dementia and among them, people with Down Syndrome (DS) are at greater genetic risk of acquiring early onset dementia [62]. Therefore, our society is not prepared to embrace people with IDD on health promotion activities that have a healthy ageing paradigm. It is important to understand that older people with IDD have similar needs as older adults without IDD. In addition, there are idiosyncrasies; where some general actions might apply to people with IDD, others must be tailored to their needs, which will vary greatly according to the person's health needs, abilities, social economic status and living arrangements. In addition, physical, mental, and women's health should drive their well-being and be part of their standard of care:

1. ***Physical Health***. People with IDD have secondary health conditions (e.g. seizures, heart diseases, etc), which might be aggravated with ageing. They also might present symptoms related to comorbidities or adverse effects of pharmacological treatment. The severity of these factors in most cases define their living condition, i.e., the degree of autonomy prevalent. Keep in mind that these elements are dynamic and might be potentiated by the time lapsed, hereditary factors, poverty and cultural values. The environment defines the type of experiences they will have for instance, the risk for addiction and contagious diseases, being immersed in a sedentary lifestyle that may lead to obesity—mainly in women—among other clinical complications. Another common observation is that because some of them cannot speak for themselves, detection of pain and other symptoms may be delayed, leading to access to treatment, if any, only in more advanced stages of diseases. Medical, dental and certain laboratorial assessments can be challenging due to behavioural issues. They might have difficulties to understand the treatment or the use of assistive or prosthetic devices. The need of appropriate nutrition, physical activity, and general preventive

health care seems less promoted to people with IDD in comparison to the general population [63].

2. ***Mental Health***. The difficulty to recognise or express emotions and symptoms may lead to inappropriate behaviour. Usually maladaptive behaviours are symptoms of diseases, for instance, a way to express physical pain, they might be a reaction to endogenous and exogenous stressors, for instance, a person with IDD might be afraid of or missing someone, which is a transitory circumstance. However, maladaptive behaviours are also associated to psychiatric and neurological comorbidities. These behavioural disorders are frequent in people with IDD and they might be misinterpreted and improperly treated. In the other hand, they might be the result of a neuropsychiatric dysfunction and represent a major mental illness, for instance schizophrenia or bipolar disorders, which requires specialised treatment. Identify causes of disruptive behaviour requires experience and mainly consider a broader context that includes potential stressors but also comorbidities and residence settings. There is a high risk for polypharmacy in individuals with IDD and the interaction between different medicines may be another source of behavioural disorders, especially for the ones taking neuroleptics [63].
3. ***Women Health.*** Girls and women with IDD should learn about the biological changes in their bodies throughout their life span, and receive regular gynaecological check-up, as other women. However, in some cultures certain topics remain as taboo for women with IDD such as sexual life, marriage and pregnancy. In order to avoid these outcomes most women with IDD receive treatments such as hysterectomy, sterilisation and induced amenorrhea if unable to self-manage hygiene. These irreversible approaches in most cases are not discussed with them, which might be considered another form of violence and violation of rights. In the other extreme they are at risk of sexual abuse, physical violence and prostitution. The screening of breast and cervical cancer is essential but, in most cases, neglected. Control of osteoporosis is crucial to avoid fractures and immobilisation. Most women with IDD are not able to report the transitions such as menopause symptoms, which would lead to proper treatment such as hormone replacement therapy [63].

These events were not thoroughly discussed here, but they were designated to key out the particularities of a lifelong with IDD. Apart from all these health aspects, the ageing in people with IDD can be overwhelmed by a diagnosis of dementia. Its manifestation may be signalised by dissimilar markers in contrast with the general population or detected upon proxy perspectives. Burdened carers, difficulty to access to health system and unprepared professionals to deal with the intersection geriatric and disability areas may be strong barriers to identify and treat progressive cognitive changes and its consequences. As World Health Organisation recommends [64], any national dementia plan should consider the adults with IDD—due to the high risk for dementia, and among them the ones at risk for early onset dementia—aiming to improve quality of life of persons with IDD living with dementia and their families [62].

Health Promotion and Wellness Approaches for Individuals Ageing with Disabilities

The literature supports the adoption of wellness and health promotion approaches for persons with chronic and disabling conditions [65]. Encouraging older adults with or without IDD to become and stay active has developed into an important public health priority [66]. Unfortunately, most of the evidence regarding the feasibility and efficacy of programs that support self-health management and health promotion approaches were conducted with healthy, or cognitively intact adults [67]. There are a few studies devoted to study health promotion approaches for individuals with disabilities [68, 69] and they usually do not address healthy ageing for adults with IDD.

Recently, Santos and colleagues [70] conducted a systematic review evaluating healthy ageing interventions for people with IDD. Only 23 prospective studies worldwide were found, involving 2398 participants both genders, mean age 44 years old, living in eight countries. 73% of the studies were accomplished recently 2010–2019. Four thematic areas were identified: (1) *Physical activity—health nutrition* (n = 10); (2) *Health education and health screening* (n = 6); (3) S*ocial inclusion and community participation* (n = 3); and (4) *Multi-components* (n = 4). Overall, these initiatives were effective, however, there were only five randomised control trials, some studies had small sample size and lack control groups, which limit generalisation of findings. Besides replication and follow up studies were rare.

Heyn and colleagues [71, 72] developed a "Health Passport" tool designed to help individuals with IDD to learn and take control over their health to properly monitor and manage key health parameters involved in chronic disease development. The "Health Passport" is part of the "Cerebral Palsy Adult Transition Longitudinal Study (CPAT)" that has the goal to understand how individuals with paediatric-onset disabilities age after they transitioning to the adult healthcare system [40, 56, 71]. The CPAT study includes a health promotion, self-health management, approach aimed to address the physical activity and health promotion needs of people with disabilities as they grow older [71, 72]. The CPAT self-health promotion component is based on a wellness counselling tool entitled "*Comprehensive Health Assessment and Motivation Program for Restoring Eating and Active Lifestyle (C.H.A.M.P for R.E.A.L.)*". The C.H.A.M.P for R.E.A.L. is designed to evaluate key health and psychossocial domains associate with well-being to inform, guide, and empower a person to make the necessary lifestyle changes to achieve high-level wellness. The C.H.A.M.P for R.E.A.L. evaluates physical function (gait, strength, balance), vital signs (heart rate, blood pressure, body mass index), standard blood laboratory (cholesterol, glucose, triglycerides and insulin); lifestyle habits (diet, physical activity levels, and self-care), risky behaviours (alcohol, smoking, addictions), quality of life (pain, sleep, and participation), and psychological well-being (depression and cognitive function).

The personalized Health Passport is a guide for better health by eliminating health risk factors and enhancing good habits. It includes positive actions for

lifestyle choices and achievable health goals recommendations.The caregivers and healthcare team are also informed and included in the C.H.A.M.P for R.E.A.L program to create a supportive culture and environment for the person to achieve the desirable "high level wellness". The lifestyle recommendations target the health areas that the person needs intervention and health improvements the most [56, 71].

The C.H.A.M.P for R.E.A.L. care team counsels the person and family, or caretakers, about the importance to adhere to a healthy lifestyle and the impact that healthy choices can have on the person's health, function, and quality of life. The person is provided with achievable and sustainable lifestyle adaptations and healthy choice recommendations. The C.H.A.M.P for R.E.A.L was designed to empower the participants to successfully manage their own health accordingly to their ability, lifestyle, and personal life.

Conclusion

Healthy Ageing is core matter for sustainability in the world. Although there is substantial knowledge concerning the need of healthy ageing, governments and societies actions towards its achievement vary from country to country. People with disabilities require tailored healthy ageing initiatives. As for individuals with IDD, the healthy ageing initiatives remain sparse, immature, and infrequent. Therefore, more attention needs to be given to health promotion for all—respecting disability-related idiosyncrasies—as a program practice and public policy.

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Part III
Service System Issues

Chapter 18
Community Mental Health and Support Services

Colin Hemmings and Nick Bouras

Introduction

A major trend witnessed in most high-income countries was the process of deinstutionalisation of services for people with intellectual and developmental disabilities (IDD), occurring mainly in the latter part of the twentieth century. In the United Kingdom (UK), mental health care for people with IDD is now overwhelmingly provided in the community. Previous suggestions of institutionalisation being the major precipitant and perpetuator of mental health problems in people with IDD did not stand the test of time. The overall position of governmental policy in UK has been consistently that people with IDD should have access to generic or 'mainstream' health services, but with additional specialist (specifically for people with IDD) support when needed [1]. Mainstream mental health services refer to existing mental health services available to the general population, though models of care often vary area by area. Specialised services, in contrast, refer to mental health services developed for and restricted to individuals with IDD. The expectation was that mainstream mental health services would provide care for the mental health problems of people with IDD living in the community. The policy of promoting the use of mainstream services for people with IDD wherever possible has not been particularly evidenced based. An as yet unanswered question after decades of community care remains, 'Do

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people with IDD and mental health problems do better in specialist IDD or mainstream services?' [2]. There has been little research to base community services on so there remains no conclusive evidence to favour the use of either [3].

Another enduring question has been, "To what extent should mental illness and behavioural problems be considered separately in people with IDD?" Challenging behaviours, such as aggression, are often the primary reason that people are referred to specialist community IDD services and they can often fluctuate over years and be chronic in duration. Referrals to specialist IDD community services are often made for people with more severe levels of IDD, who tend to have more frequent and more severe challenging behaviour compared to those with milder disability. The overlap between mental health problems and challenging behaviours in people with IDD should neither be over nor under-played. It is not possible to always neatly separate behavioural from mental health problems, nor indeed the mental health needs of people with more severe disability from those with milder disability. In many low-income countries, there is often little provided for the care of those dually diagnosed with both IDD and mental health problems. In high-income countries such as the UK, mental health and IDD services mostly continue to operate separately with quite different cultures. Where there are stand alone, dedicated services for this patient group, the mental health and challenging behaviour components are often split leading to a fragmentation of already scant resources.

It can easily enough be understood how services have varied so much between countries given the enormous differences in income, funding and infrastructure as well as in philosophies and culture. But service provision has also widely varied within countries and even between areas in the UK with similar demographics. This chapter will not include discussion on the community services that have been dedicated solely towards people with challenging behaviours. This chapter will focus on community services in the UK for adults with IDD and mental health problems.

Mainstream Versus Specialised Services

There has been much debate over the adequacy and suitability of mainstream community services for individuals with IDD. While mainstream services have certain advantages, they also come with considerable limitations. The most obvious and arguably significant strength of mainstream services is their capacity to serve and support a large number of individuals with IDD. Mainstream psychiatric services were, however, unprepared to respond to the needs of this population, lacking knowledge and expertise on their assessment and treatment of their mental health needs. Limitations of mainstream services include lack of training and expertise in IDD, inadequate resources to modify or support patients with cognitive limitations, and unsupportive or unhelpful staff attitudes [3]. Mainstream services have also been criticized for their inability to serve patients with IDD who have complex needs (e.g., behavioural challenges, comorbid medical or psychiatric issues, communication impairments, severe disability) and who, as a result, may require more intensive or individualised treatment.

The argument for the provision of mental health care for people with IDD from mainstream services was supported widely [4]. Some argued that specialised services lead to stigmatisation, labelling and negative professional attitudes. Others argued that special expertise is required for the diagnosis and treatment of mental health problems in this population, because although it is theoretically possible to train staff in mainstream settings, the relatively small number of cases gives little opportunity for staff to gain or maintain the necessary skills. Mainstream community services have greater capacity to serve individuals with IDD and may be less stigmatizing, but lack training, resources and expertise in IDD. One of the major advantages of specialised services is their size and staff to patient ratio. Specialised services tend to be smaller and have a higher staff to patient ratio than mainstream services. However, specialised services do not typically have the capacity to service everyone who is referred. Specialised services have greater expertise in IDD but may stigmatise individuals with IDD and lead to longer wait times. Mainstream services have often been considered adequate to provide care for individuals with mild or moderate disability, while specialised services may be better equipped for individuals with IDD who have more severe disabilities and/or even more complex medical or psychiatric needs.

Service Models

A number of different specialist service models were developed post deinstitutionalisation. In the UK the point of access for mental healthcare for people with IDD is usually within generic mental health services with additional specialist mental health services being provided for those with more complex needs requiring a greater degree of support [1]. Policy guidance has though failed to end geographical differences in service provision. It is widely accepted that people with IDD should access generic mental health services wherever possible but there is still a need for specialist mental health services for people with IDD [5], with provision of general mental health services alone being not adequate for those with more complex presentations.

Community services in the UK for people with IDD and coexisting mental health problems remain undeveloped when compared to those for the wider population. The services remain highly variable in constitution and operation. The lack of adequate community services for those presenting with IDD and more complex mental health problems in some areas, coupled with closures of National Health Service (NHS) specialist IDD in-patient units, has led to significant ‘out of area’ populations. There have been a few studies looking at various community services but as they vary so much it has not been easy to compare the findings [6]. It is not clear how best to deliver mental health care within specialist IDD services and it would stretch belief to think that all of the differing service configurations and provision would have equal effectiveness and value for money. There have not been the same sorts of service developments seen in recent years in ‘mainstream’ mental health services.

A major difficulty in overcoming the struggle of balancing between mental health and IDD services is the differing opinions about who should provide mental health care and where mental health of IDD service provision should lie primarily, whether alone or aligned more closely with one of these two larger 'empires'. A problem for dedicated mental health of IDD services is that they are inevitably too small-scale to operate alone effectively. They will always tend to be squeezed between generic mental health and IDD services, even if their clinicians work assiduously at both interfaces. An emerging literature supported the development of integrated models of care which enable mental health and IDD services to work together to provide a range of mainstream and specialised services. More than 30 years ago researchers [7] had reported on a case of specialist staff supporting generic staff in a mental health inpatient bed for an admission of a person with IDD. Hall and colleagues [8] developed and evaluated an integrated model of care where adult mental health services worked collaboratively with IDD services. The foundation of the model was the provision of specific beds for adults with mild disability and mental health issues within a general psychiatric ward and an associated community 'virtual team' comprised of professionals in IDD and other mental health professionals. As with many new service initiatives though, the sustainability of successful new models such as these is not often subsequently reported.

The most common model of services for adults with IDD and mental health problem that emerged in the UK is an IDD community based multi-disciplinary team offering assessment and specialist services to people with IDD. Originally, most of these teams were involved with deinstitutionalisation, carrying out tasks such as identifying appropriately adapted and staffed houses, matching people with IDD to live together and assessing their health and social needs. Most of them have input from clinical psychologists and usually some input from a psychiatrist specialising in people with IDD. Such teams have experienced difficulties in meeting the mental health needs of people particularly those with mild disability and diagnosable mental illness. Previously it had been suggested that Community Intellectual Disability Teams (CIDT) cannot best meet the mental health needs of their patients because of barriers such as lack of teamwork, excessive caseloads, poor eligibility criteria leading to patients falling in between services, lack of staff and inadequate training of staff [9]. Bouras and Holt [10] stated that the provision of mental health services from a specialist CIDT was an historical mistake by transferring into the community an institutional-type model of care. This contradiction remains until nowadays and coupled with ongoing ideological arguments, as to what constitutes challenging behaviour versus a diagnosable psychiatric disorder, has led to a fragmentation of already small specialist services for people with IDD in the UK.

An alternative to the CIDT has been the development of several specialist mental health teams for people with IDD, fully integrated structurally, organisationally and operationally with the generic mental health services and often existing separately from a local CIDT. This service model is compatible with other specialist mental health services in the UK, such as older adults, children and adolescent, substance misuse, homeless and eating disorders services. These are sometimes single-profession teams (e.g. nursing or psychiatry) or more usually

multidisciplinary teams with nursing, psychiatry and psychology components, whose focus is the treatment of mental health problems in people with IDD. They can provide specialist assessment and treatment, along with support in accessing generic services [11]. In contrast to generic services, they usually offer a very specific range of treatments, have a fixed capacity and the staff have very well-defined roles and specific training [12].

A Royal College of Psychiatrists' survey [13] found that the most common model for CIDTs was the generic CIDT (84%). These multidisciplinary teams might include psychiatrists, psychologists, psychiatric/intellectual disability nurses, social workers, speech and language therapists, occupational therapists and physiotherapists. They can be run as a partnership between primary care services, social care services and specialist mental health teams. Next most common were specialist challenging behaviour services (21.5%), stand-alone mental health of CID services (16%) and neurodevelopmental disorders' services (16%). It also suggested that most teams (70%) categorise themselves under one particular model. The majority of services (71%) were not integrated with social care.

Research and Outcomes

Community services for people with IDD in the UK have varied widely in constitution and purpose and research into their effectiveness has been neglected [3, 4]. There have been few multi-site studies and as services vary so much it is not been easy to compare the findings [6]. Many of the studies have been retrospective with their increased potential for bias. Some explored how existing services were being used rather than the actual needs of the service users. Many of these earlier studies were conducted using idiosyncratic diagnostic criteria and outcome measures and so have not been able to make use of the improvements seen in rating instruments and assessments for people with IDD [6].

Variation in the delivery of services, the patient groups studied, and the measures used to evaluate them, has made direct comparison of mainstream and specialised services for individuals with IDD difficult. The numbers of mental health admissions and use of the Mental Health Act for people with IDD in the UK are relatively low, although once in hospital they may require longer admissions than those with more typical IQ [6]. A lesson learned from generic mental health research is that a much wider range of outcomes than days in hospital should be investigated such as quality of life, service user satisfaction, carer burden and cost evaluations of service delivery [14]. While there is general agreement that mainstream services should have the capacity and willingness to provide care for at least some individuals with IDD, there remain circumstances where these services, alone, are not adequate. Many clinicians believe that individuals with more severe disabilities and those with more complex needs may be better served in specialist services, leaving mainstream services to provide care to those with fewer needs or less severe disabilities.

Intensive Community Services

Once community services were established there was increasing interest as to how these might need to evolve. It can be argued that people with IDD with coexisting mental health problems have not had the same access to developments in mainstream community mental health service delivery that have occurred in recent times. Following the introduction of assertive outreach, home treatment and crisis resolution services and other service innovations for the wider population with more severe mental illness, research interest began to grow in more intensive services for those with ID and dual diagnosis [15]. A few Assertive Community Treatment (ACT) services for people with IDD were set up in the UK, but they varied widely in their configurations. For example, some services favoured a "team within a team" model whereby a few professionals tried to adopt a more intensive or assertive approach whilst being part of a wider CIDT. Other services followed a separate team model. Some of these Assertive Community Treatment-type services were aimed at people with IDD and coexisting challenging behaviours rather than severe mental illness.

Two randomised, controlled trials evaluated ACT in people with IDD who present with both a mental health condition and challenging behaviour. One [16] found no significant differences in global function and burden/quality of life outcomes between ACT and standard care. The other [17] found a trend towards better outcomes (unmet needs, carer burden, functioning and quality of life) with standard care over ACT, but these differences were not statistically significant. Standard care and ACT in these studies seemed to differ mostly by intensity of the treatment, with ACT being more intensive. In both studies, the treatment received by the two groups was similar and both could be considered assertive [16]. The ACT models used in these studies differed from each other and the original model [18].

A study of opinions about the ACT model in IDD services among specialist IDD clinicians found much inconsistency regarding how ACT should be applied [18]. A constant theme expressed was that services should be based upon assessed need, and that services should not be provided which duplicate those already available. Maintaining continuity of staff, full-service staffing and a service of sufficient absolute size to consistently provide the necessary coverage and diversity of staff roles were all seen as highly unlikely to be achieved for a hypothetical Assertive Community Treatment in IDD service. The need for services to have clear purpose and care pathways was mentioned frequently. Interviewees often commented that joint working with mainstream mental health services needed to be improved for the best care of the service user. Interviewees frequently talked about the need for those working with people with IDD and mental health problems to have sufficient/adequate training and experience. Related to this they often commented that specialist IDD services need to provide training to mainstream mental health staff, care staff and other services.

What Should Community Services Provide?

Community services for people with IDD and mental health problems vary enormously so it is important to know what they all should provide. One way to do this is by seeking a consensus of expert opinion. Three linked studies investigated this. The first compared views between specialist IDD staff, carers and service users [19]. There were no great differences between the groups as to what they viewed as essential service components. Service users emphasized their need for practical help and for all staff to be trained in IDD and to get to know them. Carers emphasized the need for prompt treatment and for more information to be made available, particularly what to do and where to go in a crisis. The clinicians emphasized the need for all involved to have good understanding of both IDD and mental health problems. They highlighted interfaces between specialist IDD and mainstream mental health services. They wanted to see earlier diagnosis and treatment and access to mainstream services, particularly in a crisis. They were wary of developing new stand-alone services for people with IDD and more concerned that existing IDD services should be able to respond flexibly and more intensively for people in mental health crises.

In the second study a consensus of opinions was generated among specialist IDD clinicians about what service components specialist IDD services should provide using the Delphi survey technique [20]. Service components considered essential for routine services included such items as *regular review of service user and care plans*, *monitoring of mental state*, *monitoring of medication*, *crisis plans* and *out of hours support*. Service components considered as essential for more intensive services, included *can react to a crisis that day* and *provide a comprehensive list of contacts should the service user relapse*. The routine service components considered essential could be broadly considered under a need for a focused approach on the service user and their illness (e.g. *monitoring of mental state)* and the added need to work within the wider context of the service user with psychosis and IDD (e.g. *access to social, leisure or occupational activities)*. Five of the more intensive service components were considered to be essential (e.g. *can react to a crisis that day).* However, the routine service components considered essential already contained many components such as *out-of-hours support* and *crisis plans* also relevant to more intensive services.

Eighteen service components were considered as essential for routine services. These included such items as *regular review of service user and care plans*, *monitoring of mental state*, *monitoring of medication*, *access to social, leisure or occupational activities*, *crisis plans* and *out of hours support* and *support/advice/training around mental health for the person's family, carers and support services*. Five service components were considered as essential for more intensive services, including *can react to a crisis that day* and *provide a comprehensive list of contacts should the service user relapse*. However, several of the routine service components were arguably those that would often be considered a core part of more intensive services also, such as the provision of *out of hours' supports* and *crisis plans*.

In the third study specialist IDD clinicians were interviewed in-depth about how some of these key service components should be implemented [21]. Most were 'expert' in that they had published research or opinions on community services. Their opinions contained several themes such as the need for services to have clarity of purpose and clear care pathways. They often commented that joint working with mainstream mental health services needed to be greatly improved. Participants frequently talked about the need for all staff to have sufficient training and that specialist IDD services needed to provide this. They were concerned that often mainstream mental health services do not meet the needs of people with IDD and they were therefore keen to develop new improved ways of joint working. There was not however support for developing 'super-specialist' services within IDD services such as ACT. Participants were skeptical that there would be the critical mass of staff or service users or the funding levels to make these economically viable, effective and sustainable, even if these might be theoretically the most ideal. There were concerns that these could increase discontinuity in the already patchy mental health care provided. An opinion repeatedly expressed was that specialist IDD services should not seek to provide services that were already available for the wider population, such as a 24-h service.

Conclusion

Service provision for those dually disadvantaged by IDD and coexisting mental health problems is still heavily determined by widespread and enduring dualistic thinking. This leaves it bounced between mental health and IDD services' commissioning [22]. Community services in the UK for people with IDD and coexisting mental health problems remain non-standardised; research into their effectiveness has been lacking. We still have to see a body of research into interventions and services in this specific patient group, including many evaluations of clinical outcomes and cost effectiveness [6]. The mental health of IDD field has tended to have a time lag for the evaluation and the adoption of interventions that have long become standard in generic mental health services. Meanwhile mental health services' research has frequently excluded adults with IDD [23]. Future research into the effectiveness of community services for people with IDD needs to focus on service components rather than whole-service models.

Clinicians did not see the future of their specialist services as always replicating (or as they saw, duplicating) developments in mainstream mental health services. There are frequently voiced concerns that this could cause fragmentation of and lack of 'critical mass' for already small specialist IDD services. In any case many in mainstream mental health services feel that specialisation within those much larger services has contributed to discontinuity of care. There is not widespread support for establishing newer or more intensive mental health services for people with IDD only. It seems instead that the best way forward is in developing new and closer ways of joint working with staff in mainstream mental health services [24]. Mental

health of IDD staff would then be more highly visible to their generic mental health colleagues [25]. Joint or co-working or 'hybrid' service provision needs to be evaluated and evidence-based through research.

The vast majority of people with IDD and mental health problems now live outside of hospitals, including with families in local communities and with increased life expectations. There is an increasingly diverse population of more mild disability, with more complex co-morbidity and increased age expectancy. This along with the issues of ethnicity and gender has meant the need for services to respond to the needs of ever-changing local communities is now greater than it has ever been. Community services for people with IDD and mental health problems are due for a major re-shape as we by now have entered the "meta-community era" [26].

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Chapter 19
Living Arrangement Options

Christine Linehan

Introduction

This chapter aims to outline some of the key issues and research evidence regarding residential living options for older individuals who have intellectual and developmental disabilities (IDD). The United Nation's Convention on the Rights of Persons with Disabilities espouses the rights of all individuals to choose their place of residence, determining where and with whom they live on an equal basis with others [1]. The reality for many individuals with IDD, however, is somewhat different. Data from the US National Core Indicator Set, representing 6778 adults with IDD living in 28 states, revealed that less than half (44.6%) exercised choice regarding where they lived, and fewer again (40.6%) exercised choice regarding who they lived with [2]. Decisions regarding living arrangements continue to be made by others in the 'best interest' of individuals with IDD [3]. Recognition of the limitations that 'best interest' can confer on individuals' autonomy can be observed in Ireland's recent Assisted Decision Making (capacity) Act, where the term 'best interest' was specifically excluded from the legislation.

Globally, most adults with IDD live within the family home, many into old age. In the United States, 75% of older adults with IDD live with family [4] while in the United Kingdom, one third of individuals with IDD are identified as living with a family carer aged 70 years or older [5]. Increasingly, greater life expectancy among individuals with IDD [2], combined with a reluctance by ageing families to plan for the future [6], results in crisis admissions to residential services on the basis of ill-health

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or death of a last remaining caregiver [5]. The capacity of the next generation, often siblings, to provide ongoing residential support is depleting as the 'sandwich' generation juggle ageing parents, young children and careers [6]. Strained relationships with family members are associated with higher numbers of accommodation moves experienced by older persons with IDD in later life [7]. In addition, the higher risk for older individuals with IDD for some health conditions [8, 9] as well as a disproportionate health burden due to lifestyle [10, 11] may also contribute to the need for residential support beyond that which families feel they can provide. For individuals living in service-based or supported accommodation options, similar pressures arise for staff who perceive a 'tipping point' where they deem that better care can be provided elsewhere to accommodate the individual's age-related needs [12, 13].

Residential options have been classified for those with IDD and dementia to three types, albeit these could equally be applied to all those who have IDD with age-related needs; they comprise ageing in place (at 'home'), in-place progression (establishing distinct homes for the care of individuals with similar age-related needs within disability services) and referral out (generally to mainstream nursing facilities for older persons, also termed aged-care) [14].

These options reflect advances in the development of living arrangement options for persons with disabilities from the singular option of institutional care. Over 20 years ago, Hogg noted that increased longevity would be of little interest if people remained in institutional care or were institutionalised with age; remove these options within the context of an aspiration of inclusion and participation and a wide range of issues arise [15]. Theoretical models of disability have moved from a medical basis to espouse notions of choice, autonomy and independence, and contemporaneously, an evidence base has steadily accrued in favour of community living over institutional care [16, 17]. Despite an awareness of the greater life expectancy among people with IDD, a comparable evidence base on quality outcomes in different living arrangements for those who are ageing has been slow to materialize. The need for such evidence is immediate as a disproportionately high number of people with IDD are now living in mainstream nursing homes [18, 19], a practice described by some as 're-institutionalisation' due to ageing [20]. Within the context of the United Nation's Convention on the Rights of Persons with Disabilities, and an ever-growing population of older people with IDD, the need has never been greater for guidance in this area.

The Policy Vacuum

A clear policy directive is needed to guide the development of residential living arrangements that will provide optimal quality outcomes for older individuals with IDD [5]. To date, policies are typically informal arrangements, if present at all [21]. In the absence of formalised policies, there are some broader international declarations and conventions which are likely to influence any future policy consensus.

The most influential instrument to guide policy is the United Nation's Convention on the Rights of Persons with Disabilities which calls on State Parties to protect the

right to live independently and to prevent isolation or segregation from the community. The United Nation's two instruments on ageing may be less influential than the disability convention but set a broad context for State Parties' obligations to those who are ageing. The first instrument, the Vienna International Plan of Action on Aging, was endorsed by the United Nations General Assembly in 1982 (resolution 37/51). Some 20 years later, in 2002, a second assembly on was hosted in Madrid (resolution 57/167). The Madrid Declaration presents over 100 recommendations with an expectation that regional commissions of the United Nations take local responsibility for implementation. One of three over-riding principles of this assembly is the promotion of 'enabling and supportive environments', with a specific recommendation to encourage the development of housing options for older people who have disabilities 'that reduce barriers to, and encourage, independence'. The Declaration promotes 'ageing in place', calling for the *'coordination of multi-sectoral efforts to support the continued integration of older persons with their families and communities'*. A specific call is made to national policy- and programme- coordinating-agencies within the disability field to address issues concerning older individuals with disabilities.

Actions from the Madrid Declaration are reviewed every 5 years, with the most recent occurring in 2018. In addition, an Open-ended Working Group on Aging was established in 2011 to monitor progress against the Madrid recommendations. While there is criticism of the pace of reform [22] the need for action is recognised by the Office of the High Commissioner, United Nations, who in 2014 added to the call for a new convention on the rights of older persons [23]. The development of this convention provides an opportunity for those within the disability field to increase visibility and create greater fusion across the disability-ageing divide.

The United Nations Declarations may be perceived as a broad framework to drive policy development. More specific is a series of international declarations focusing on the intersection of ageing and disability, largely driven by academics within the disability sector. These include the Graz Declaration on Disability and Aging (2006) [24], the Barcelona Declaration on Bridging Knowledge in Long-term Care and Support (2009) [25], and the Toronto Declaration on Bridging Knowledge, Policy and Practice in Aging and Disability (2012) [26]. These declarations note the distinct development of services within the disability and ageing fields, and the inherent challenge this poses when attempting to bridge the two disciplines. The Graz and Barcelona declarations call for significant support from the European Commission, by way of a Green Paper and collaborative efforts, to provide a platform for discussion. The Toronto Declaration seeks an international formal agenda inclusive of self-advocates, family, researchers, practitioners, and policy makers, while emphasising that any 'bridging' across the two fields does not come at a cost of diminution to either.

National policies exist, albeit in varying degrees of specificity. Bigby and colleagues reviewed policies in five countries, Australia, Canada, Ireland, the United Kingdom (UK) and the United States (US) [21]. Ageing policies across the five countries for the general population advocate ageing in place. Australia and Canada reference ageing in place within the context of individuals residing in residential

aged-care facilities, calling for additional supports to facilitate these individuals to remain in situ as their needs change. Ireland's national ageing policy at the time of publication, *Action Plan for Services for Older Persons 1999–2008* [27], is cited as being unique in recommending that the disability sector take responsibility for specialist services for those with Down syndrome and dementia. Since Bigby's publication in 2010, Ireland's policy has been updated with the current national ageing policy, the *National Positive Aging Strategy* [28], no longer containing specific reference to IDD. Ireland's *National Housing Strategy for People with Disabilities 2011–2016* [29], is cited; a document which contains no specific reference to ageing, but advocates *'the promotion of access by people with disabilities to independent living across the full range of housing options'* (p. 2) to enable individuals remain in their homes and communities for as long as possible.

The proposal to identify disability services as the responsible body for the provision of supports to older individuals with IDD remains a favoured option by some commentators who note that the disability field has advantage over the ageing field in its development of a model of support, over many decades, promoting independence, participation and choice [30, 31]. Indeed, the Madrid Declaration cited above notes that older individuals today are comparable to persons with disabilities in the era prior to the UN Convention on the Rights of Persons with Disabilities. Should one field take responsibility, the weight of preference currently may side with the disability sector.

Ireland's departure from specifying one particular sector to take responsibility for older persons with IDD aligns more closely with other jurisdictions who broadly argue that people with IDD should be supported to age in place via a combination of efforts from both ageing and disability sectors. Bigby cautions that the call for joint planning of services provides little incentive for either to take the lead, notably where there is little, if any, reference to additional funding being made available to develop services. Within a history of poor interagency collaboration in policy, there is a concern that shared care may become no care as stakeholders within the disability and ageing fields seek to protect their own distinct funding streams [32]. Tensions already exist where typically disability funding is more generous than aged care due to higher staffing ratios [33]. Some commentators argue that additional 'top up' costs related to ageing should be covered by the aged care sector as individuals with IDD have the same rights to supports in older age as others, albeit the costing of 'additional' ageing needs is complex [34] and would need to be carefully ringfenced to supplement and not replace disability funding [35]. Irrespective of the source of funding, additional funding will be required to support the growing cohort of older individuals with IDD, most likely for increases in staff numbers and for the training of existing staff to provide optimal support for this population as they age [13]. Individuals with IDD, more so than the general population, are reliant on disability-related supports to participate in community living [36] and to avoid sedentary lifestyles which mitigate against successful ageing [37]. Whether from the disability or ageing purse, these additional resources will require funding, and as the deliberations on an appropriate way forward continue, increasing numbers of older individuals with IDD are at risk of receiving sub-optimal support.

Developments in the United States present a collaboration for consideration. In 2012, the Administration for Community Living was founded via the amalgamation of the Administration on Aging, the Office on Disability, and the Administration on Developmental Disabilities. Among many activities, the Administration for Community Living provides guidance to states on how to streamline access to services. Coordination between state and local agencies is promoted to create a simplified process for individuals of all ages, and all abilities, to access services. Research and policy papers guide practice. A common aim that unites activity is the avoidance of long-term care facilities in favour of independence and community living [26, 38].

Notwithstanding advances in the United States, the more typical international picture is that, in the absence of clear policy developments, practice is guided by informal, local policies developed by disability service providers [21]. Differences in the conceptualisation and implementation of ageing in place, a core concept in the field, illustrate the diversity of these local policies. For some, ageing in place refers to initiatives supporting an individual to remain resident in the family home. Where individuals are resident in accommodation owned by a service provider, ageing in place may be defined as remaining within a group home managed by a service provider, or a move to another dwelling managed by same the service provider, or at the more extreme end, a move to any dwelling managed by organisations funded under the disability sector [21]. Progress towards a clear policy will require consensus on a common nomenclature, a development yet to materialise within the ageing-disability arena.

Finally, the voice of older individuals is currently almost completely absent from policy development discourse [20]. A Consensus Statement by the International Summit on Intellectual Disability and Dementia notes this omission and sends a salutary reminder that many of the ‘oldest-old’ who have IDD are likely to have experienced institutionalisation and have expressed resistance to any further attempts at what they perceive to be re-institutionalisation on the basis of age-related need. The harnessing of this lived experience is essential if the long-awaited policy developments in this field are to be truly inclusive.

Succession Planning

Consideration of appropriate living options for older persons with IDD must take cognisance of the fact that the family home is the most common living arrangement [30]. The increased life expectancy of persons with IDD means that some parents and guardians perform a caring role for six or seven decades [39]. In addition to supporting an ageing child with IDD, many of whom may experience accelerated ageing [40], these caregivers engage in ‘compound’ caregiving, providing additional support to ageing spouses and to their own ageing needs [6]. Despite their advancing years and caregiver burden, there is evidence that older caregivers are reluctant to relinquish their caring role [41]. While some caregivers feel that others

would be unable to provide a similar level of care to their family member [41], some caregivers report anxiety about their capacity to continue in a caring role [42]. Older caregivers also report a lack of suitable living options to which their family member could move [42] and evidence supports this concern with some states in the United States needing to expand their residential services 200% to meet demand [7]. Whether related to a desire to continue caring, there is emerging evidence of 'mutual caring' where those with IDD assume a caring role for an ageing parent or guardian, albeit there is little research data on these interactions [30].

There is a general reluctance by families to consider the future support needs of their family member with IDD [43–45] despite expressing an 'unremitting apprehension' for the future [41]. Ideally, family members and others within a circle of support would be included in these discussions before the ageing process begins [40]. Advanced preparation is deemed both possible and practical in situations where an individual is diagnosed with progressive conditions, notably dementia, and yet is not evident in practice [46]. The avoidance of a discussion on succession planning can result in decisions regarding future living arrangements being made in haste, typically during a period of crisis when the main caregiver is unable to continue in the caring role due to ill-health or death [6, 43]. In the UK, one quarter of people with IDD become known to statutory services in later life when such a crisis arises [5].

Traditionally the caring responsibility was often transferred to the next generation, typically a single sibling without children [6]. The capacity of the next generation, however, to take responsibility is diminishing [47], creating a demand for formal services to fill the void [6, 33]. One potential consequence of the lack of succession planning, and the inability of younger generations to step into the caring role, is the possibility that inappropriate residential options may be offered and accepted [44]. Families clearly need support in addressing succession planning. Older adults with IDD similarly need support, not only to recognise the need for this dialogue but also to be appropriately supported to participate [30, 43] as evidence indicates that individuals with IDD experience difficulty articulating the type of support they need with age [35].

A number of studies report that the preferred family option for future living arrangements is for family members with IDD to remain in the family home with paid support, if and when the last remaining caregiver is unable to provide support [6, 41, 48]. This preference is also expressed in numerous studies by older individuals with IDD [45, 49, 50]. Where the option to remain within the family home is not deemed possible, evidence suggests family preferences are for small community-based options within the disability sector rather than nursing home care [42], albeit there is evidence that families feel that the ageing sector may provide better health care outcomes [51].

A level of caution is needed in equating an individual who remains 'at home' with the concept of ageing in place. Watchman notes that ageing in place requires that appropriate supports are forthcoming, such as staffing and adaptations, without which the quality of life of individuals is compromised [46]. This additional support requires funding and moreover, flexible funding systems, which adapt to meet

individual need [52]. Succession planning should provide the necessary time to consider the resources and allied funding that are necessary to ensure that the will and preference of the individual and family are met as the person ages. The failure to support succession planning for all parties, however, has been evidenced for over a quarter of a century [53] highlighting once again the longstanding need for significant policy and practice development.

Ageing in Place

The term ageing in place originates in the field of urban geography, referring to the ageing of neighbourhoods but is now generally understood as individuals living in their own home regardless of increasing care needs [21] close to family, friends and familiar possessions [34]. While the term may be commonly assumed to mean that individuals are supported to live in an 'ordinary' home for as long as possible [54], much of the research on ageing in place has focused on individuals with IDD who have dementia [34] where ageing in place was originally defined as remaining in a group home or moving to another dwelling owned by the same service provider [55]. Supports to facilitate individuals to age in place typically include health and social care directly related to need, training for staff, and environmental adaptations including assistive technologies [46, 56].

The term 'home' may have different application within the disability field than a typical understanding of the concept. Some individuals with IDD will experience a different trajectory in life to the general population, bypassing home ownership, relationships and raising a family [57]. For this cohort, 'home' may equate to their family home, to community-based dispersed housing owned by a service provider which is simultaneously a support staff's workplace, or to an inappropriate setting such as an institution [40]. In addition, for older adults with IDD, home may be a place where the impact of age-related needs must be considered within the context of other individuals with IDD sharing the property [40], whether tenancy, group home or congregated setting. These issues illustrate the inherent complexities in considering an ageing in place policy within the disability field.

Ageing in place needs to take cognisance of the overlay of age-related impairments onto pre-existing disabilities [58]. In combination, ageing and disability may bring increased risk of accidents, mobility difficulties, loss of independence and behavioural issues [59]. The Developmental Disability Aging at Home Project in the United States provides a useful example of how some of these needs may be met within the home environment, reducing a need to move elsewhere to access supports [58]. The project explored the efficacy of a cost-effective intervention to support individuals at the younger end of ageing, average age mid-50s, who were experiencing age-related issues. A task performance assessment completed with each individual identified a number of home modifications including assistive technologies, entrance and egress adaptations, and most commonly, adaptations to bathrooms. For a relatively modest cost of $600 per person, improved or stable performance in 65

daily activities was reported following the installation of these adaptations. Where modifications were not approved, reasons included cost, landlord objections, and health and safety issues. Mindful of the relatively young age of the participating individuals, the demonstration project indicates that future planning efforts may both improve quality of life and be highly cost-effective if resulting in the avoidance of a transfer to more highly staffed, and consequently, expensive living arrangement.

In addition to the introduction of house modifications, a significant contributor to ageing in place is the support of appropriately trained staff. Ageing in place is intrinsically linked to staff knowledge, staff confidence and staff skill. Evidence suggests staff may be unclear about signs of ageing, in particular whether to attribute changes in health or behaviour to an individual's age or disability [54]. Staff also report a lack of confidence, and a lack of training, regarding dementia [60]. Staff training is associated with staff confidence, and higher staff confidence is negatively associated with a likelihood of a staff recommendation that a person should be transferred from ageing in place [13, 61]. The broader staffing context is also influential in these situations, given evidence that staff training is short-lived without the involvement of more senior members of staff [62]. This context is important as evidence suggests staff may feel that even minor health conditions may be sufficient grounds to move someone out of ageing in place [3], in particular those conditions which require 'hands on' procedures such as managing a colostomy bag, dressing 'everyday' wounds, injecting insulin and using a feeding tube [13].

Supporting individuals to age in place takes an emotional toll on staff who have been documented to take on extra shifts, work as waking night staff when employed as sleepovers, and sometimes volunteering to work without pay [63]. These staff members report feeling they have 'failed' where an individual is moved to another residential location, despite also reporting that the issue of ageing is rarely discussed in advance of a move [64]. Staff have also reported that they are reluctant to contact health care professionals in case this would trigger a referral out [9]. In combination, these findings illustrate that support for staff is crucial to facilitate older individuals with disabilities age in place [63].

Decisions to transfer individuals ageing in place from group homes to mainstream aged care are more likely to be made by supervisory staff than by direct support staff [3]. In a series of interviews with ten supervisors, Webber and colleagues identified that while all supervisors agreed with the broad concept of ageing in place, this was qualified by 'as long as possible' and yielded two distinct 'philosophies', 'ageing in place' and 'active engagement' [3]. An 'ageing in place' philosophy was exercised where individuals with IDD were given significant additional supports to remain in situ, even if at a perceived inconvenience to other individuals living in the house. In contrast, 'active engagement' was exercised by supervisors who deemed that a move to aged care was warranted where individuals were no longer physically active, were challenged to participate in day to day activities, or were deemed to be impacting negatively on others in the group home, albeit there is also recognition that the move itself may impact negatively on those left behind [60]. In particular, the need for nursing care was identified as a key driver of a decision to move someone to aged care, albeit that some supervisors automatically

equated a diagnosis of specific conditions with a need for aged care, irrespective of nursing care need.

Ageing in place may not be an appropriate option for all, whether due to the individual's preference or perceived level of need. In particular, the needs of individuals with dementia have received specific attention in this context as the condition places significant challenges on individuals, families and staff, the latter group reporting feelings of being 'overwhelmed and floundering' in their attempts to provide optimal support [63]. Addressing some of the concerns, National Task Group on Intellectual Disabilities and Dementia Practices (NTG) provides specific guidelines on supports for individuals with IDD and dementia [65]. At the outset, NTG recommend that services should aim to effectively support individuals with dementia to remain in their chosen home and community. NTG call for funding and social policies that promote home and community care with investment in training specialised "dementia-capable" staff who implement stage-based supports to reflect the individual's progression in the stages of dementia. 'Home Assist Teams' are identified as staff with diverse specialisms to support ageing in place, and if required, in out-of-home arrangements. NTG state *'In most instances, continued community living is viable and warranted both from a human rights and best practices perspective. With appropriate supports and supervision, most if not all adults with intellectual disability can continue to reside in some type of community living setting and enjoy enhanced quality of life'* (p. 9). NTG recommend that barriers to community living should be considered before exploring alternative residences.

Notwithstanding efforts to support ageing in place for those with dementia, mid-stage dementia is often a trigger for considering a move to alternative accommodation, often due to increasingly problematic behaviours. NTG recommend that where behaviour becomes problematic, compensatory strategies are explored on the basis that the movement of individuals with IDD with mid-stage dementia to unfamiliar residential living options may in fact increase levels of anxiety, disorientation and problematic behaviours. NTG state that the movement of individuals with mid-stage dementia to another setting should only be considered as 'the last resort'. For those with end-stage dementia, or others deemed to have significant care needs, there is recognition that the abilities of staff and family may be exceeded [65]. NTG advises that even in these circumstances, ever effort must be made to honour, where possible, the previously identified preference of the individual.

Where ageing is place is deemed to be an unsuitable arrangement for an individual and a decision is made that a person would be better supported in another residence, planning for the transition is essential [20]. Where 'in-place progression' to a disability specialist home or 'referral out' to aged care are considered, the involvement of the individual in this planning is critical as an understanding of their perspective should ease the transition [20]. Strategies recommended during transitions include a phased transition, introducing new staff, ensuring personal belongings are evident and, if possible and relevant, asking a relative to stay in the new dwelling in the early days. Before such a step is taken however, the current home should be carefully assessed as evidence indicates that staff may be unaware how disabling some existing environments can be for those who are ageing in place [63].

In Place Progression

In-place progression is typically understood to refer to specialised environments where trained staff support older individuals experiencing later stages of dementia [14]. These options are usually observed within organisations that have responsibility for a number of dwellings and therefore have flexibility to transform some of their existing housing stock into small specialised group homes. Early research in this area in the late 1990s advocated that individuals with dementia should be supported in small homes of less than four individuals, where just one individual had dementia [66]. The eventual death of individuals with dementia over time, however, was considered too distressing for others living in these houses resulting in the development of the current model of small group homes where a number of residents, if not all, have dementia [66].

The Preparing Community Agencies for Adults Affected by Dementia Project (PCAD), established by Janicki and colleagues, was designed to examine how disability services might provide support to older adults with IDD and dementia in community settings [14]. Contributors across the US, UK, Canada and Ireland reported that the development of small community-based group homes was becoming the preferred option within the sector and that features can be added to this model to assist individuals with dementia remain the community. The homelike environment prominent in this model of support is deemed to provide a higher quality of life than aged care [67].

While the development of specialised group homes is described as a more reactive than proactive response to the ageing needs of the individuals supported, they are now described as becoming 'viable alternatives' for long term care of individuals with neuropathologies such as dementia [12]. These living arrangements are mirrored by group homes within the ageing sector, termed 'assisted living facilities' which enable older persons to live, with support, in community settings. Costs for both types of group home were reported in 2011 to be almost identical at US$53,000 per annum.

In place progression is defined as a sequential option, in comparison to the linear option of ageing in place. Linear refers to the addition of supports as needed, while sequential refers to the movement of individuals to particular group homes as their needs become greater and require more specialised staff input [12]. Each group home is developed to support individuals who present at particular stages in the development of dementia, and individuals are moved through group homes in a sequential order reflecting their stage of dementia. Staff are required to have an understanding of the phased nature of supports that are likely to be needed over time, and should have opportunities to rotate across the different environments to avoid emotional exhaustion which may result where staff provide ongoing care for individuals with high support needs.

Quality outcomes for individuals living in specialised group homes need to be evaluated on a longitudinal basis [68]. The limited research to date, largely from Janicki and colleagues, indicates that *'it is timely to plan and develop a range of*

community-centred dementia capable services' (p. 382) on the basis that they have been found to be a viable option for community care for older individuals with high levels of need. The ongoing development and evaluation of this model of living arrangement does not come without cost, notably in the greater demands on staff time and effort. Funding and resource allocation for these homes must ensure that they are sufficiently flexible to recognise the changing needs of the individuals supported and the implications these changes have on housing and staff structures [68].

Referral Out to Aged Care

Aged care has been defined as synonymous with senior services, services for the elderly, and older persons services [21], also termed nursing homes. In the United States, individuals with IDD have been reported to comprise 2.4% of a broad nationally representative sample of almost 10,000 nursing homes [69]. Estimates from the United Kingdom were similar across 53 local authorities, averaging 1.7% [70]. In Canada, an audit of individuals with IDD resident in aged care facilities reported this cohort comprised 4.5% of the regional adult population of persons with IDD [19]. This estimate was significantly higher than the comparable rate of aged care admissions for the general adult population in Canada, recorded at 0.9%. This trend highlights the disproportionate admission of individuals with IDD to nursing homes, albeit a figure that was found to be in decline in Canada over a 4-year period.

The Canadian decline in admissions is described by the researchers, Ouellette-Kuntz and colleagues, as 'promising' and reflective of efforts to prevent re-institutionalisation following a programme of deinstitutionalisation. As with other jurisdictions, a practice has emerged over several decades where nursing homes in Canada have become a residential option for individuals who were seeking alternative accommodation following the closure of large institutional facilities [69]. One possible contributor to the diversion of individuals to nursing homes during deinstitutionalisation is the disparity in cost between supports for older persons provided by the disability sector when compared with costs incurred in nursing homes [33, 68]. The UK survey of 53 local authorities identified average costs of nursing homes in 2001 at GPB14,300 to GPB18,300. These costs were considerably less than the GPB43,000 average cost of what was considered, at the same time period, as 'good quality' residential support for people with IDD [70]. The researchers of this audit noted '*there are financial incentives for local authorities to use older people's services rather than developing good quality learning disability provision. Unless this is addressed, the common practice of misplacing people with learning disabilities in older people's homes and then forgetting them will continue'* (p. 5).

There is growing evidence of the profile of individuals with IDD who are resident in nursing homes. This cohort is more likely to be moved to aged care at a younger age, and remain for a longer period than older adults without IDD [3, 32, 71, 72]. Notably, individuals with Down syndrome are reported to be more likely than individuals with other IDD to be placed in aged care [72]. Bigby's survey of

207 individuals with IDD living in nursing homes throughout Australia's aged care system identified an average admission age of 59.4 (compared with 84.3 for those without IDD) and an average stay of 5.6 years (compared to 2.7 years for those without IDD) [32]. The overwhelming majority (90%) of these individuals were under 65 years of age at admission and were reported to have lower rates of ill-health and dementia than the general ageing population. More than one in three moved to the nursing home from a family home due to the death or ill-health of a carer, despite previous research in Australia indicating that decisions initiated by disability service providers to move individuals from disability to aged care services were opposed by family members [18]. These trends have been replicated in other studies conducted throughout the US and Canada using large sample sizes [19, 70, 71].

As Bigby has shown, a proportion of individuals with IDD who are supported in nursing homes are younger than the admittance age, typically 65 years. Evidence suggests that many service providers with a stated admittance age are willing to informally accept individuals with IDD below this age [70]. The accelerated ageing of some younger individuals with IDD may be a contributor to the lack of condemnation expressed for this cohort than that expressed for younger individuals with other disabilities, such as acquired brain injury, who are living in aged care settings [73, 74].

The UK survey of 53 local authorities also explored admission routes into aged care following concerns expressed by the Growing Older with Learning Disabilities (GOLD) Programme regarding residential supports for older people with IDD [70]. The most common reason for referral was the re-organisation or closure of a disability service. For those moving from the family home, the most common reason for referral was the ill-health or death of a caregiver. The researchers conclude that *'meeting age-related needs did not explain why most of the people with learning disabilities were in older people's services'* (p. 16). In fact, less than one third of people who moved to aged care from the family home, and less than one half of people who moved to aged care from a disability organisation, were moved because of the individual's level of support need.

Referral routes to aged care have also been explored in Australia, in a landmark study by Bigby [40]. The study examined the interface of family wishes and service provider' responses to the ageing needs of family members living in group homes. At the time of the study, service providers may have interpreted legislation as allowing them to give notice to quit to residents whose level of need was deemed not to be appropriately met within the group home. While family members were initially of the opinion that the group home would be an option for life, in time they came to an understanding that at some point their family members with IDD would 'cross a line' where their needs could no longer be met within the group home. In contrast, staff had a longstanding assumption that a line would be crossed for older individuals instigating a move to aged care. Although staff commented that they *'wouldn't wish that on anyone'* (p. 82), they felt residents crossed a line requiring aged care if they perceived better care could be provided elsewhere, or if an assessment of the individual's changing needs indicated the group home could not meet need. Bigby

notes that staff's assessment of a line being crossed was not based on objective indicators and varied considerably according to local practice. Discrepancies included an understanding in one group home that a move to aged care was warranted if waking staff were required, yet other group homes employed waking staff as a matter of course.

An acute health issue or period of intense stress within the houses participating in this study typically instigated a referral to aged care. Some 'hasty' decisions arose where families were presented with a 'fait accompli' based on the organisation's argument that they could no longer provide an appropriate level of support, typically due to failing health. This argument was queried where some families noted that the health issues prompting a transfer to aged care resolved over time, raising issues as to the appropriateness of the move in the first instance. Bigby concludes that services should avoid 'major and irreversible' referrals to aged care being made on the basis of health-related issues or staff stress, both of which may be transitory. Despite espousing person centred practices, this study indicated that *'resource and organisational issues that stem from an ill prepared disability system shaped many of the decisions about a move to residential aged care. This situation was fueled by the absence of policies that affirm the right, and funding mechanisms that support, ageing in place for group home residents'* (p. 786).

A follow up to this longitudinal study indicated that lack of sufficient staffing levels and access to nursing expertise in group homes contributed to referrals for aged care [51]. With relatively minor modifications by staff in aged care settings, presenting issues were addressed and resolved. Examples of these modifications included ensuring that individuals with faecal incontinence were regularly supported to access toilets, and individuals prone to falls were supported with adaptable beds. It is arguable that these adaptations could easily be implemented by staff within disability services thereby facilitating ageing in place.

Notwithstanding the skill and professional efforts of staff within aged care settings, there is evidence of diminished quality outcomes for many individuals with IDD resident in aged care, a situation described in the UK as resulting in 'impoverished lives' [70]. Studies of quality outcomes reveal that individuals with IDD who are resident in aged care settings experience less individual attention from staff [75], reduced engagement in activity and reduced opportunities for community engagement [32, 33, 70, 71, 75], reduced access to professionals with expertise in IDD [70, 71], a lack of positive relationships with other residents [32], and poor contact with family and friends [70]. These outcomes are concerning particularly in light of safeguarding issues that may arise for highly vulnerable individuals who are demonstrated to have poor contact from family and from staff members familiar with their specific needs [70, 71]. These outcomes are reflected in the attitudes of some disability staff who argue that the needs of older people with IDD are best provided within the disability sector [54, 64].

Perhaps of most concern are mortality trends. Following the concerns of family members, a Fatality Review Board for Persons with Disabilities was established in Connecticut to examine the deaths of individuals with IDD supported in state residential settings. In total, 32% of these deaths occurred in nursing homes. When

specifically reviewing deaths arising in nursing homes, the Board observed significant lapses in quality of care, a lack of alternative living options, lack of care management, 'faulty communication' between services, and lack of advocacy [76]. Further examination of a subset of these deaths indicated that many individuals with significant medical issues lived for up to 10 years within the nursing homes, raising questions as to why these individuals were not discharged to the community. The reviewers concluded that the development of medical supports within community settings would reduce admissions to nursing homes. These developments, however, require services to examine local policies prohibiting staff to administer certain medical procedures, for example, gastric-tube care and administration of oxygen. The reviewers conclude that *'the failure to use existing resources such as group homes to their potential capacity, and other similar policies should be examined'* (p. 177). Once again, the call for significant policy development is echoed.

Conclusion

Increased longevity of older persons with IDD, combined with the challenges many families experience to provide lifelong support, create an urgent need for the development of services to support this population. A lack of clear policies is matched by, and perhaps contributes to, a reluctance by families to address succession planning. Despite a host of international conventions and declarations calling for collaboration between the ageing and disability sector to agree a way forward, issues of ownership and funding obligations delay progress. Developments in the United States, where ageing and disability sectors are now united under a 'community living' umbrella, may provide a mechanism to jostle stakeholders out of their chosen silo to work collaboratively.

Ageing in place is espoused as the ideal option for all older persons. For individuals with IDD, the concept is more complicated given that notions of 'home' may vary from that experienced by the general population. To age in place with dignity and respect requires additional age-related resources, notably investment in accommodations, staff training and staffing ratios. Yet, too often, modifications are deemed unworkable and staff report being overwhelmed and under-trained. The consequence is a move away from family, friends and familiarity. The available evidence would indicate that considerably more could be done to support individuals age in place, in particular with the addition of some medical expertise to address age-related health issues. It may be that the mere mention of medical expertise harks to an era of a medical model which so many, for so long, have fought to replace with values of self-determination and inclusion. Yet, it is the perceived need for medical support, often temporary, which contributes to the disproportionate numbers of older individuals with IDD residing in aged-care settings, where despite best efforts quality outcomes are diminished.

The lower cost of aged-care in comparison with disability services is evidenced in different jurisdictions and arguably contributes to individuals being moved into

aged-care. The finding of lower quality outcomes combined with lower cost was noted during deinstitutionalisation and is echoed in the commentary by Professor Jim Mansell who observed that institutional care is cheap to run if care is replaced by containment [77]. More recently, Professor Jan Tossebro cautions at the revival of large community-based facilities to support those with high support need, where facilities are developed with a 'take it or leave it' attitude toward consumers [78]. Mindful of these trends, it is now imperative that the disability sector stands by the advances achieved in community living by exploring, evaluating and evidencing quality outcomes in appropriately staffed small community-based housing options that promote ageing in place.

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Chapter 20
Families and Ageing: Working Towards a Positive Partnership with Services

Karen Watchman

Introduction

> *"When I think initially of the very difficult days when he was born, and how we were told he wouldn't achieve anything, just take him home and so forth, when I look at the things that he has achieved it's very very uplifting to look at that." (Parent of adult with Down syndrome)*

This quote from the evaluation of a life story project in Scotland [1] captures a changing perspective among families as their relative with intellectual and developmental disabilities (IDD) ages. It differs from expectations of just a few decades ago, but how far is this positivity reflected in partnership working between services and families where it may be harder to identify the strengths and preferences of people with IDD?

How well we all age depends on a number of factors in earlier life as much as our chronological age. This lifespan approach means that everyone has a role in determining how positive an experience it is to get older. Demographic trends suggest that the population of people with IDD is ageing, consistent with a worldwide increase in the numbers of older people [2]. While there is now recognition of the growing number of people with IDD living to older age, little account is taken of the implications of this demographic trend for the development of appropriate care [3, 4]. As an example, in 1983 the average life expectancy for persons with Down syndrome (DS) was 25 years. This has extended over the years and today for the first time we regularly see people with DS living beyond 60 years of age [5]. However, for the current older generation of people with IDD, especially DS, this has been

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somewhat of a surprise for families who were typically told that their child would not outlive them; we now know that for many this is the reality. This differs from other caring roles where the caregiver is typically of a similar age, usually a spouse or partner, or younger such as an adult child caring for a parent. This leads to worry about the future when family members of people with IDD may no longer be able to provide care. Indeed, studies consistently show that carers are reluctant to plan ahead despite a high number of parents of people with IDD being over 65 years [6].

As people continue to outlive their parents, the role of siblings and extended family members will increase. However, siblings may lack the experience of services or support that their parents have. In some cases, this parental experience comes from decades of struggling, for example for access to services or mainstream education with parents often adopting a 'warrior' role when advocating for their child. Parents 'did their bit' when their children were younger then, using the words of many, did not feel as great a need to advocate as their child grew up, letting other younger parents pick up the mantle [7]. Many families are now finding it difficult to step back into this role in the face of increasing health changes for their, now adult, family member at a time which often coincides with their own later life and health issues. Older people with IDD are not a new population—people have always aged.

The longer life expectancy now enjoyed by more individuals means that increased visibility and numbers pose different challenges to both services, families and people with IDD. If we were to ask what people with IDD want from services when they get older, then the answer would probably have to be that we really do not know, because we tend not to ask. Many people with IDD will not see themselves as getting older and the concept of 'retirement' is a difficult one [8]. This is not surprising given that daytime activity, whether voluntary or paid employment, also provides a significant social interaction for many people with IDD. It also highlights how far many people with IDD rely others to facilitate social engagement or activities.

This chapter reflects on partnership working between people with IDD, their families, and professionals. It is predominantly viewed through the eyes of participants of two studies in Scotland: a family life story project involving parents, siblings, people with DS and service providers, and a dementia study in which co-researchers with IDD engaged with photovoice methodology to share their perspectives of dementia and ageing. This demonstrates the importance of communication within families first in order to understand the views and wishes of the person with IDD, which may differ from family members and might involve difficult conversations. It also highlights how people with IDD are often not given the status they deserve as experts in their own experience, often with self-awareness of being excluded and poorly informed.

What Is Partnership?

Whilst not everyone with IDD will have support from parents, siblings or extended family, social care systems around the world would not function without them. This suggests a crucial role for families in partnership with professionals. Partnership

will look different for families and professionals with potential for disagreement or conflict to occur, for example over safeguarding issues or best interest decisions. Whilst an equal balance of power may be seen as the ideal, this is often difficult. Family may be invited into a partnership but not necessarily on their own terms, indeed that this cannot be a level playing field as family are so deeply involved whereas professionals can walk away [9]. This means that equality requires a shift in thinking as well as power that respects parents expertise. Most definitions of partnership include shared goals and placing the needs and wishes of the person first, i.e. person centred [10]. Frost [11] refers to different levels of partnership: level 1 (cooperation) sees families working towards consistent goals and level 2 (collaboration) involves family helping services to plan ahead towards common outcomes. Level 3 is coordination, seeing families and services working together in a planned and systematic manner towards shared and agreed goals – authors see it as a progression between levels. Findings from both studies in Scotland suggest that even to achieve level 1 there needs to be greater involvement of, and where appropriate control by, people with IDD. Without this there is unlikely to be meaningful progression to levels 2 and 3.

Life Story Work

Developing a life story can give a sense of identity and help an individual to share not only their story, but memories, experiences, life events, details of preferences, and information about family, friends, work, hobbies, holidays and favourite places. Identity is what makes an individual unique. Down's Syndrome Scotland's Family Support Service has previously reported incidences where an adult with DS has been placed with a social careprovider, or received support in emergency situations due to ill health or death of a family member, where nothing is known about the life or preferences of the person with DS. Having a life story can not only ease challenging transitions and help the person come to terms with changes but can also provide a routine and have a positive effect on wellbeing, in additional to recognising the importance of enabling staff in support services to 'know the person'. Life story work can be developed for different reasons in different contexts although those supporting individuals living in an acute hospital, community setting, care home and family home may not have access to the same resources, tools or information.

People with IDD have often been denied the opportunity to reflect on their life events and how this has affected them. The compilation of a life story can be an empowering process helping the person to feel valued and listened to, and most importantly to have 'a voice' in what should be a fun activity. The study at Down's Syndrome Scotland recognised at the outset that the most significant person in the life of a person with IDD may not always, or only, be a family member. Consequently, it was acknowledged that the preference of the person with DS may have been to involve a staff member to engage with their life story activities. Of the 14 families taking part in the project 'My Life, My Future', seven participants with DS lived with

one or more parents whilst seven lived independently with support provided from adult social care services,[1] either alone or in accommodation (group home) shared with up to three other people with IDD. Three families included an adult social care support worker in the development of life story work for their relative with DS.

The study was evaluated by the author [1] with the aim of identifying short-term outcomes (1-year duration of the project) among the 14 participating families, and to consider implications for medium- and longer-term planning beyond 1 year. Participants tried a range of different types of life story, as explained below, at monthly workshops initially to help development and subsequently to support ongoing work. The majority of families ultimately combined several types including a life story box for items that could not be readily stored elsewhere. The format selected was based on individual preference of the person with DS and allowed individuals to draw on their different strengths such as artistic and/or digital skills. The participants who were more creative enjoyed the process of decorating their life story boxes, life story scrapbooks, and folders. Overall, a structured approach to the process was perceived as helpful, with templates being used by members to develop and build their stories and structure the different sections within their chosen format. At the end of the project, there was a suggestion that using more than one type was beneficial with associated conversations and different outputs. Data were collected using pre- and post-project family interviews in months 1 and 12, pre- and post-completion of Warwick-Edinburgh Mental Wellbeing Scale (Tennant et al. 2007) with family/those in the role of family in months 1 and 12, Personal Wellbeing Index – Intellectual Disability (PWI-ID) with participants with DS in months 1 and 12 and end of project semi-structured interview with 2 Down's Syndrome Scotland project workers and 2 volunteers.

Findings identified that families considered how the resource could be used if the person moved out to live independently, moved into different accommodation or was required to stay in hospital.

> *"You do start to think about, as we get older, and what there is for, say, your brother and sister to look at with him, when we're long gone, there'll be something there that will ground him and give a conversation point to somebody else" (Sibling)*

It was highlighted how important the life stories could in theory be in future planning, with social care support workers or other professionals such as Community Learning Disability Nurses, allied health professionals, GPs or IDD psychiatrists gaining greater insight into the person, who they are as individuals, their identities, and what is important to them. However, the reality proved different and there were variable experiences reported of paid social care support staff engaging with the life story process. As a result of taking part, one of social care support workers actively taking part had introduced life story work with other residents receiving support within the same service. However, a number of families reported that support

[1] Adult social care in the UK refers to a system of support designed to maintain and promote the independence and wellbeing of older people, people with IDD and informal carers. This can include provision of formal care services such as shared group homes or help in the home.

workers they had discussed the project with had not engaged as hoped in supporting the participants to develop their life stories, and did not recognise this as a task that they could be involved in.

Nine family members spoke of the potential use of the life story if the person either developed dementia or experienced cognitive changes in the future. One person with DS spoke of how the life story might help her if she developed dementia. Five parents reflected on the importance of the life story as a resource for other people to get to know the person if they were no longer there to provide that information. They described this as reassuring to know that such a detailed resource about their family member was now available to be shared with their son or daughters permission.

There were initial concerns from families that they were leading the process by initiating memories or telling their family member of events that happened when they were much younger, with the potential for ownership of narratives, and therefore life story, to be contested. This was dissipated as the project developed and the person with DS became more familiar with the process. It was also helped by the structured templates which gave the person with DS more control over the topics or items to include or leave out. At the beginning of the project a number of participants were concerned about re-visiting distressing memories in their life stories, mainly of relatives or friends who had died, and expressed worries that this may be too upsetting. By the end of the project, they spoke of how it had been helpful to put those memories into their life stories and to have a means with which to talk about both sad and happy memories of that person. One person with DS experienced a family bereavement just before the project started, and his sibling spoke of how the project had given them an opportunity to spend time as a family reflecting on memories together. Another participant experienced a family bereavement during the project and spoke of how important she had found it to store memories of that person in her life story. Overall, the majority of families volunteered information of how the life story work had provided an opportunity to talk about memories of people who were no longer alive.

In creating life stories some participants identified gaps in their current lives and a key hope was that each would use their life story to take steps towards making the changes they desired. This included participants choosing to share their life stories with others to help them achieve these changes and access the range of support that they wanted in their lives. A further area where it was hoped that the life story might facilitate participants to make bigger changes in their lives was in supporting those still living in the family home to both think about and take steps towards increased independence.

> *"Longer term I would like to hope that it has helped achieve things, just made them think, that it's helped to get the right support that they want. Even if they were having a planning meeting and didn't feel confident enough to talk up in the meeting but were happy to show parts of their story, you get a sense of what this person wants." (Project worker)*

Limitations must be acknowledged as although taking place over a period of 1 year and with a high retention rate, the overall participant numbers were small and

all participants had DS, so were not representative of other types of IDD. The project was aimed primarily at supporting future planning within families and it is acknowledged that the outcomes may be different if the purpose was specifically to improve care or formal support services beyond the family. Similarly, ongoing support was available from Down's Syndrome Scotland which should not be assumed as the typical scenario either for families or services.

Conversely, the high level of support available may have increased motivation and willingness to continue. No participants with DS had profound or multiple disabilities and all involved were able to communicate verbally. There were vast differences between ages of participants with DS with the youngest being 22 and the oldest 58; most participants were in their early 30s. Inevitably, this involved differing life experiences, for example one participant grew up in a long-stay hospital and had different childhood and early life experiences requiring sensitivity in approach.

Photovoice

Photovoice is a highly participatory research method in which the individual observes their everyday realities or the reality of others, and then takes photographs to discuss the meanings behind those images [12]. This is under the premise that it is often impossible to understand the meaning of an image in isolation from the context in which it emerged. Images can give a different perspective on the world than the one that is represented through words.

Photovoice has become increasingly recognised as an accessible method to involve people with IDD in research [13], and this study highlighted the work of an inclusive research team co-led by four members with IDD who used photovoice methodology to capture perspectives of dementia, both their own perspective and those of their peers [14], the first time such a study has been conducted by and with people with IDD.

Co-researchers[2]with IDD took part in seven training and ongoing information workshops over a 10-month period. Basic point and shoot digital cameras were introduced with time spent learning how to use them. A mini printer was provided so that images could be printed out at discussion sessions providing immediacy and enabling the images to be seen more clearly than by looking at the small screen on the camera, or when transferred to a laptop. This gave further control by enabling co-researcher choice in which images to pick up and discuss, again more tangible than scrolling through a range of images on the laptop. The ethical process in advance of the study clarified that no images would be taken of participants. Instead photographs were a visual representation of what the co-researchers saw or felt, used to stimulate later conversation. Whilst all co-researchers took photographs

[2] Co-researchers were involved from the inception of the study prior to funding. One co-researcher had Down syndrome, three others had an unspecified intellectual disability. The age of co-researchers ranged from 31 to 56 including one female and three males.

independently, each required variable support with aspects of the process, such as prompts around taking photographs and talking about the meanings behind the images taken. A flexible approach was subsequently adopted that involved a guided approach [15] as the university-based researcher or support worker assisted co-researchers during photography. A combination of individual and group discussions with semi-structured interviews were used over the period of photovoice data collection. A standard photovoice three-staged approach was taken to analysis: selection of photographs, contextualisation (discussion) of photographs and coding to identify themes or theory.

Findings identified themes related to uncertainties and interrelated worries about dementia for the co-researchers based on both their own personal reflections and as a result of observing participants. They described limited prior understanding of how dementia may progress before their involvement in the project; however, frequently knew of a dementia diagnosis amongst their peers and were aware of several changes that people may experience, including behaviour and memory. Concerns were raised around future care needs, the potential of having to change support teams, and the worry around having to go into what the co-researchers termed 'institutional care' with an insistence that they be involved in decisions made about them if health needs changed. Co-researchers indicated that family and support staff were often not aware of their fears or concerns when a friend developed dementia, when at times they just 'disappeared' (presumably moved elsewhere) with no explanation. Nor was there wider recognition or acknowledgement of the fear among people with IDD especially DS, that they themselves may also get dementia.

> *'I think one of the things… people might be a bit frightened that they might have to go into a long stay hospital, or a long stay institution, if things get really bad…I want to stay where I am as long as I can. I don't want a new team. But I know in the end it's sometimes… That's the thing that bothers me, I don't know how long… different circumstances might mean having to move… It's like a question mark.'*

Such lack of communication, discussion and information with service providers or other professionals can only exacerbate a situation where there is already a lack of future planning. However, this extends to communication within families too where discussion of dementia rarely took place. This is consistent with other research findings where older carers were also often found to be unwilling or unable to make plans for the future or just simply had not got around to it especially those who were still coping well [16]. Yet, many remained fearful of what may happen in the future.

At the end of the study in 2020, two of the co-researchers had already taken forward their learning into their local community by sharing knowledge with their peers through delivery of dementia training within their IDD self-advocacy networks. This was continued via an end of project conference for people with IDD which featured presentations from co-researchers and a display of the images with associated explanatory quotes from photovoice. Additionally, two co-researchers selected images and wrote content before delivering both oral and poster

presentations at an international IDD conference. Both engaged with conference delegates to talk about the project and explain their choice of images on the poster. In this way, recognition of work and ownership enabled the co-researchers to talk to professionals attending the conference and share their views, albeit not the 'policy change' required by the original aims of photovoice methodology. Whilst the underpinning goals of photovoice include under-represented communities being empowered to reach out to policy makers and effect social change [17], this study raised questions around whether this can be achieved under the constraints of a funded project. Other photovoice projects have reported similar barriers in facilitating change, and the need for an on-going collaboration to drive forward social action [18] however, this does not mean that social change cannot be achieved. Whilst this may be more apparent at local or even national level it may not necessarily equate to wider policy reach that effects change. Social change at local level can see families progress beyond level 1 [11], cooperation towards collaboration and coordination and as such should not underestimated.

Conclusion

As people with IDD age, so too will their parents—often siblings or other family members will take on more of a caring role however it is still important that as a family there are proactive plans made. This is to avoid a crisis situation typically caused by illness or disability of parents or older siblings. Unmet need is a common theme underpinning user-focused research, as identified by the two novel studies in Scotland, and is one that any potential partnership with services needs to recognise. The reality is that adults with IDD are often given limited choice in, or information about, issues that directly affect them resulting in divergence between requirements of services, preferences of families and the wishes of people with IDD. However, this does not mean that given appropriate means, persons with IDD are not able to communicate their preferences, for example through appropriate life story work, or advocacy for peers as seen in the photovoice study.

Many older family carers want to continue caring for as long as possible although increasingly recognise that this may not be possible. Similarly, the multiple and complex nature of the physical and mental health needs of older people with IDD and their ageing carers is a significant challenge for services. Future planning for the care of older people with IDD and their families is an essential process and the two studies presented highlight factors to facilitate this. Both studies in Scotland raise implications for partnership between families and social care services recognising that the process takes time, can be increasingly stressful during transition – when people may also change their minds, and does not happen in isolation. It has been demonstrated that a progressive approach is required, beginning with difficult conversations within families to seek cooperation before moving to collaboration and coordination with services.

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Chapter 21
Quality of Life

Lieke van Heumen and Alice P. Schippers

Introduction

The number of people over age 60 in the world will double by 2050. This means that by 2050, more than one out of five people will be over age 60 [1]. The United States' older population is steadily increasing as well. The population over age 65 increased from 37.8 million in 2007 to 50.9 million in 2017 (a 34% increase) and is projected to reach 94.7 million in 2060 [2]. In the 2017 American Community Survey, 35% of people over age 65 years reported a disability [3]. This number included people who experience disability due to aging and a growing number of people with early-onset or lifelong disabilities. Individuals with intellectual and developmental disabilities (IDD) form the largest segment of people living with lifelong disabilities [4]. The number of older adults with IDD in the U.S. is expected to double and potentially triple by 2030 [5]. The same tendency is expected everywhere in the Western world. Despite this demographic urgency, there is a lack of research directly addressing aging of people with IDD [6].

Even though much progress has been made in the last decades to improve the lives of people with IDD, they continue to experience barriers to community participation, social inclusion and self-determination [7]. People with IDD in public life face patronizing attitudes, fear from others and an unwillingness by other people to

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accommodate their difference [8]. There are systematically low expectations of people with IDD [9], and the assumption that they cannot lead meaningful lives or enjoy a high quality of life remains pervasive [10].

Many adults with IDD experience poor social determinants of health and experience negative life events that jeopardize their health and quality of life as they age [11]. Adults with disabilities have lower levels of employment and education and are more likely to live in poverty than adults without disabilities [12, 13]. In the U.S., people with IDD are more than seven times as likely to be the victim of rape or sexual assault compared to people without disabilities. Women with IDD are about 12 times more likely compared to people without IDD [14]. Despite an increased need for healthcare [15], Americans with disabilities lack adequate access to health care [13]. For example, 2010 data from the U.S. Behavior Risk Factor Surveillance System showed that 27% of people with disabilities needed to see a doctor in the past year but did not do so because of cost (compared to 12% of people without disabilities). Additionally, fewer American women with disabilities are current with mammograms and Pap tests than women without disabilities [13]. As a consequence of these circumstances, individuals with IDD have on average twice as many health problems than others without IDD, experience earlier age-related declines in health and function than the general population and are more likely to develop secondary conditions as they age [16–18]. For example, mental illness increases with age in adults with IDD even when excluding dementia [19] and older adults with IDD experience major depression at higher rates than other older adults [20].

Compared to other older adults, aging may be more stressful for adults with IDD because they rely on daily support with (instrumental) activities of daily living. Yet, their social support networks are generally small [21, 22]. Due to communication or cognitive impairments, they may have limited ability to understand the changes that come with the aging process. Furthermore, they lack access to information and support for coping with age-associated losses such as decline in health and the death of close family members and friends [23]. Older adults with IDD are often not included in discussions about death and dying or are inhibited from attending funerals of family members and friends. Some family members and direct support staff fear that adults with IDD do not understand or cannot manage discussions and rituals surrounding death and dying [24].

The increase in the number of people aging with IDD and the challenges experienced by this population have demanded new directions for research, practice and policy that promote social justice and improve this population's health and well-being and overall quality of life. This chapter explores developments in the study of aging with IDD that aim to achieve such improvements. It first discusses the conceptual framework used in this chapter. This chapter then presents three strategies that facilitate enhancing the quality of life for adults aging with IDD: promoting self-determination, facilitating meaningful engagement and supporting social relations. This chapter highlights research that includes the perspectives of adults with IDD on their lives, which have been insufficiently included in previous literature.

Conceptual Framework

The conceptual framework used in this chapter is informed by the literature on Quality of Life, the related development of positive psychology, and the life course perspective. The framework emphasizes the importance of fostering a meaningful and positive ageing experience for individuals in which they perceive to have autonomy over their lives [25–27]. This means the life stories, experiences and the needs and preferences of adults with IDD and their families drive the development of supports and services needed to respond to the unique challenges this population faces.

Quality of Life

The IDD literature has made little distinction between the Quality of Life (QOL) of younger and older people with IDD [28]. Though applying the concept of QOL to the experiences of older adults with IDD is a recent development, the concept itself has been studied since the old philosophers. Aristotle questioned the meaning of the 'good life'. Contemporary scholars place current understandings of the concept in the 1960s' social indicators movement. This movement served to describe the 'state of the states', social measurements and analyses were used at the societal level to improve for example health and school attendance [29]. QOL became an important concept in social policy, as well as an objective for human service provision to measure effects in personal outcomes [29]. Since the 1980s the concept has been widely used in the field of IDD [30], both as an aim and as an outcome measure of human service provision. Although it is hard to find a consistent definition of QOL [31], consensus exists about several characteristics of the concept and its use in the IDD field [32–34].

It is commonly accepted that QOL is a multidimensional concept existing of several interrelated domains of well-being (e.g. physical, material, emotional and social well-being). Because these domains are influenced by personal and contextual factors and mutually influence each other, QOL is considered a holistic concept. In practice this means that improving someone's leisure activities may also improve that person's physical and mental health, for example [32, 34, 35]. Moreover, QOL is considered to be a universal concept, indicating that the life domains are the same for all people, including both subjective (person related) and objective (environment related) components [33, 34, 36, 37].

At a personal level, people differ in their perceptions of the various aspects of life that compose their QOL. People make choices and respond to their unique personal needs, desires and preferences. Moreover, people change in various ways across the course of their lives and therefore their perceived QOL changes over time [29, 32]. Life experiences frame people's perception of their QOL. Across an individual's lifetime there is often a shift in relative importance of one life domain over others. Baltes and Baltes' SOC model (selection, optimisation and compensation)

examines the development of adults across the life course. This model proposes that older adults change their aspirations according to the age related changes they experience, and that they adjust their strategies to realize these aspirations [38].

Early life events influence later life experiences, but do not necessarily predict QOL in later life [26]. The life course perspective is important to understand QOL for ageing people with IDD, because some life domains become more important in old age. For example, changes in health can become more pronounced and retirement can lead one to revalue social relations and leisure activities. Additionally, in the domain of living arrangements age-friendly communities become more important [26].

Worldwide, people with IDD live with their families and (nuclear and extended) families are supporting their relatives with IDD [39]. This is also the case for ageing people with disabilities [26, 28]. Living a quality life as a family when one (or more) of the family members has IDD creates both challenges as well as rewards [26, 40]. This acknowledgement gave rise to the development of Family QOL (FQOL) as a branch of QOL in the early 2000s. In her study on older families, Jokinen [26] found FQOL holds similar characteristics as individual QOL. Relevant core concepts and principles include a holistic approach that reflects on the relationships between family members, a lifespan perspective, life domains, family self-image, opportunities and choice. For families, the dynamic nature of family QOL is also important, individual characteristics and differences as well as the context of the family are relevant [26, 41].

The question: 'whose quality of life is it anyway?' gets posed more frequently in the field in the last two decades [29, 42]. People with IDD should be the primary subjects in QOL assessments and research [28, 43]. The quality of peoples' lives as they themselves experience them should be at the forefront [29, 36]. Methodological challenges to including people with IDD as respondents in research are increasingly addressed and resolved, leading to creative methodologies that unravel and highlight adults with IDD's perceptions of their QOL and ageing experience [44–46]. This development has also advanced emancipatory and inclusive research practices [28, 29].

Positive Psychology

Positive psychology is the 'science of positive subjective experiences, positive individual traits and positive institutions that allow individuals, communities and society to flourish' (page 5) [47]. Positive psychology aims to move beyond the study of pathology to promoting positive qualities in individuals, and relies on the assumption that people can self-direct and organize their lives [48]. Positive psychology as applied to disability aligns with disability studies perspectives that emphasize that different ways of being in the world can be 'sources of knowledge, satisfaction, creativity, and happiness' (page 9) [49]. This perspective moves away from medical model interpretations of disability as an individual deficit in need of cure or

correction and away from the universal experience of people with disabilities with marginalisation and discrimination. Strength-based approaches to understanding disability align with the success of people with disabilities in all areas of life, promoted by the achievements of the international disability rights movement and the implementation of disability rights policies around the world. Historical views of disability as pathology have run their course [50]. The last few decades have seen a similar shift in the conceptualisation of intellectual disability specifically, away from a deficit-focus to an emphasis on the strength and resiliency of people with IDD [51]. Similarly, positive psychology perspectives on ageing move away from framing ageing as a predominantly negative process of loss and lack of engagement towards interpretations of ageing as an opportunity for personal growth and enjoyment [47]. Research on older adults with IDD often focuses on losses in functioning that come with age and on ways to prevent such decline. When applying a positive psychology lens to the experience of ageing with IDD the focus shifts to determinants of positive emotions and strength and on ways of increasing meaningfulness in life as one ages [25].

Life Course Perspective

The life course perspective acknowledges that ageing is a lifelong process, and that circumstances, events, behaviour, and relationships earlier in life influence the ways in which people age [27, 52]. Examples of life course factors that have the potential to negatively impact the quality of life of people with IDD as they age are lack of education, institutionalisation, lack of close and supportive social relations, bereavement, lack of valued social roles, low income and poverty, and changes in support services. These circumstances accumulate and pose a risk to long term well-being and quality of life [53, 54].

Research that examines the experiences of people with lifelong disability from a life course perspective is scarce [55, 56]. A life course approach to disability and ageing addresses the experience of living with disability through various stages of life, and can highlight how disability might be experienced differently in old age versus young age and middle age and across age cohorts [57]. The lives people have lived inform their ageing experiences. A comprehensive understanding of the factors that make up QOL in older people with IDD therefore relies on the application of a life course perspective.

Promoting Self-Determination

Self-determination is a prominent theme underpinning QOL for people with IDD as they age [32], and was one of the earliest constructs a strength-based focus or positive psychology approach to disability was applied to [58]. Self-determination

means “acting as the primary causal agent in one’s life and making choices and decisions regarding one’s quality of life free from undue external influence or interference” (page 177) [59]. The development of self-determination results in a number of positive outcomes for people with IDD. These include improved independence and employment outcomes [60], improved quality of life [61, 62] and improved ability to achieve personal goals for community living and participation [63].

The process of becoming self-determined starts in childhood and continues across the life course. A life course perspective to the development of self-determination acknowledges the role that family, friends and communities have in creating and supporting opportunities for people with IDD to live a life of their choosing [64, 65]. Family members and direct support professionals play important roles in facilitating self-determination [65]. There is an increasing recognition of the right of people with IDD to take control over their lives. Yet, both individuals with IDD and family members experience barriers to choice and control [64]. Adults with IDD have reported overprotective parents [66], having to prove themselves over and over [67] and receiving supports that do not meet their needs [68]. Research has demonstrated that some family members hold paternalistic attitudes about disability. For example, they may lack confidence in the person with IDD’s ability and skill to make decisions [69, 70], and belief that they know what is best for their family member with IDD [69, 71]. Adults with IDD who live in large congregate settings are at the highest risk to experience restrictions in choice making and rules that limit their ability to live their lives on their own terms [72].They may have no choice in selecting staff or housemates [65]. Adults with IDD who live independently with supports usually exercise more choice than those in other living arrangements, including living with family [73, 74]. Research has found that daily interactions between people with IDD and direct support staff may inhibit self-determination [65, 75]. One strategy to promote self-determination of adults with IDD in residential settings, work and other environments is to train the staff [76, 77]. People with IDD need to be provided with adequate support to develop self-determination skills in different contexts throughout the life course, so they learn to set and work towards future goals [58]. A recent study completed in Italy by Di Maggio and colleagues [58] showed that the future goals of adults with IDD related to achieving autonomy (such as to find a job, have a home, being economically independent and making decisions about their lives), and having meaningful social relationships.

Self-determination is particularly salient for older adults with IDD [26, 28], as it is an important part of ageing well [15, 65]. Researchers in the Netherlands who conducted a longitudinal study with older adults with IDD found that older adults experienced less autonomy than younger adults with IDD [78]. Older adults with IDD often experienced limited opportunities to exercise self-determination in their lives [79]. Social, political, and economic realities severely constrained the opportunities of people with disabilities who grew up in the era before the disability rights movement and the passage of major disability rights laws. As individuals with IDD age, major changes occur in their lives. Many of these changes occur at a younger

age for people with IDD compared to the general population and may cause a sense of loss. Examples include a slow decline of physical and mental abilities, retirement, the death of family members and friends, and sometimes moving to a new home. Adaptations to these changes should reflect the choices and goals of the individual [4, 80]. Buys and colleagues [81] observed in their work that older adults with IDD want to have more authority in decision-making regarding their own lives. In the Dutch study by Lehman and colleagues [78] the experience of less autonomy among their sample of older adults with IDD appeared to not affect their satisfaction with life, happiness or loneliness.

Supported Decision Making

At least half of adults with IDD in the US are under some form of guardianship, and most of these are full guardianships [82]. In these arrangements another person makes all legal decisions for the adults with IDD. Adults with IDD under guardianship lose their right to be self-directed in decision-making as their legal capacity to engage in decision making has been determined to be insufficient [83]. The 2019 report from the National Council on Disability [82] found many problems with guardianship in the United States, including misunderstandings about the ability of people with IDD to make autonomous decisions and denial of due process within guardianship proceedings and meaningful consideration of less-restrictive alternatives. Many people with IDD and their families are unaware of options to restore rights and less-restrictive alternatives to guardianship. Many adults with IDD may therefore stay in guardianships that are excessive or unnecessary for most of their lives.

Supported decision making is one recent strategy used to move away from restrictive guardianships. Supported-decision making also holds potential for realizing end-of-life care for adults with IDD that is driven by their wishes and desires [84]. Supported decision making is an individualized process that helps a person to make and communicate decisions with respect to their personal and legal matters. It builds on person centered planning, but focuses specifically on 'identifying and implementing supports needed for decision making, and enhancing legal agency and self-determination' (page 145) [83]. Despite growing attention for supported decision making, a clear definition or agreed upon framework for its use and implementation has been missing from the literature. Additionally, assessment tools have to be developed that identify the support individuals with IDD need to engage in supported decision making processes [83]. Shogren and Wehmeyer [85] suggested that to apply such support there is a need to understand contextual factors and environmental demands for decision making (such as sociodemographic variables, decision making experience, nature and level of disability, emotional factors, accessibility of information, complexity of decisions, living arrangements, opportunities for decision making and family attitudes about decision making), as well as individual supports needed for decision making (such as curricula and decision aids).

Person-Centered and Future Planning

Person-centered and future planning are used to foster self-determination. The terms 'person-centered planning' and 'future planning' are often used interchangeably in the IDD field [27]. Planning is important for people with IDD of all ages and instrumental to support their life course transitions such as from adolescence into adulthood and from middle age into old age [27]. The purpose of person-centered planning is to develop collaborative, goal-oriented supports to promote community participation and positive relationships [86]. Future planning has traditionally been used to prepare for the increasing likelihood of adults with IDD outliving their parents. The goal of future planning is to ensure adequate arrangements are in place that are in line with the wishes of the family and the adult with IDD. Adequately planning for the future is important for all older people, but crucial for people with IDD who need lifelong support. Without future plans in place, people with IDD are at risk of emergency placements in inappropriate settings when they lose their primary caregiver [87, 88].

Person-centered and future planning activities serve to create person-centered goals for education and training, current and future vocational choices, residential options, retirement and leisure-time activities and social supports [55]. Adults with IDD need supports to participate meaningfully in person centered and future planning processes [87]. This includes supporting discussions about end of life wishes, establishing advance directives, creating a will and planning for one's funeral [84].

Although ageing parents express concern about the future of their adult children with IDD, and fear being outlived by their children with IDD, many families have not completed future plans [87, 89–91]. Barriers to planning include a lack of information, lack of time, lack of available services, lack of family members to be caregivers, difficulty to afford an attorney, benefits resulting from caregiving, support provided to the caregiver by the adult child with IDD, and emotional issues concerning mortality [87, 92]. A Canadian qualitative study explored the future concerns of adults with IDD and family caregivers. Adults with IDD expressed concerns for their ageing parents, for their future living arrangements, and about loneliness. Family caregivers were concerned about ensuring the future security of their adult child with IDD, addressing legal issues and financial security, and promoting future choice and self-determination. The study findings point to the importance of early and intentional future planning that prioritizes the needs of the adult with IDD and integrates the desires of family caregivers [90].

The peer support intervention "The Future is Now" supports family members and adults with IDD to plan for the future [87]. The program offers training for legal and financial preparations and hosts small-group workshops that use a peer mentoring co-trainer model. An evaluation of the program found that families who participated in the program were significantly more likely to complete a letter of intent, plan for future living arrangements for the adult with IDD and develop a special needs trust compared to the families in a control group. Caregiving burden also

significantly decreased for families who completed the program. Finally, the daily choice-making of adults with IDD increased.

Life Story Work

Storytelling is a fundamental part of what it means to be human. We tell stories about our lives, who we are and the world around us [93]. Life stories are important for older people, because the longer life is the more there is to be told. Life review and retrieving memories, or reminiscence, promote ageing well [94, 95]. The well-being of older people is not only determined by their current experiences, but also by what happened to them earlier in life, and by their retrospective view on those life events [96]. For ageing adults with IDD it is important that their life stories are included in person-centered planning through life story work [97, 98]. Life story work is a coordinated approach to share life stories between adults with IDD and family members, friends and caregivers [99]. This approach includes activities that create a written record of a life story such as compiling a 'life book' [98]. Life stories can also be recorded through various different media such as a photo-album, an audio or video recording, multimedia, or through the use of a 'memory box' with physical objects representing memories [97]. Creative methods of recording life stories can increase the accessibility of life story work, especially for individuals with significant disabilities and limited verbal communication.

Facilitating Meaningful Engagement

Meaningful active engagement is another important component of QOL as people with IDD age [100]. To consider what meaningful active engagement may look like in the latter part of the life course for adults with IDD, an understanding of their living arrangements and their employment status is useful. Most adults with IDD worldwide live at home with their family caregivers [101]. In the U.S. 71% of the adults with IDD live at home with family caregivers, 24% of these adults live with a caregiver over age 60, and another 35% with caregivers between 41 and 59 years old. Only 13% of adults with IDD live in supervised residential settings [102]. In 2014 almost 100,000 individuals with IDD in the U.S. were on waiting lists for residential out-of- home services and over 216,000 were waiting for any type of long-term services and supports [103]. Many adults with IDD continue to live with their ageing parents until their parents pass away or are no longer able to provide care [102]. A study in Ireland by O′Rourke and colleagues [75] found that older adults with IDD who lived with family were more likely to report being happy with their living arrangement than adults in group homes. However, adults living with family were more likely to report feelings of loneliness than adults living in group homes.

Before the deinstitutionalisation movement, many people with IDD who lived at home with family members were not employed. Individuals with IDD who lived in institutions usually held jobs that were limited to in-service or agricultural tasks related to the running of the institution [27]. With the deinstitutionalisation movement, many adults with IDD have experienced improved access to meaningful daily activities in the community [104]. Yet, even after the deinstitutionalisation movement the majority of adults with IDD continued to experience only sheltered work [105]. Despite the fact that people with IDD value work and want competitive integrated employment, only a small number of people with IDD have achieved such employment [106, 107]. The employment rate for adults with IDD between the ages of 21 and 64 in the U.S. has been reported at 26.1%, which is much lower than for people without disabilities (79.1%) [12]. There is little evidence for the impact of residential setting on employment for people with IDD [106]. McGlinchey and colleagues [108] did find that living in the community was associated with increased employment for older adults with IDD. They also found that increased rates of employment were associated with better self-reported health and less depression. McCausland and colleagues [106] reviewed overall occupational activities for older adults with IDD in Ireland, and found that despite low levels of competitive employment, most adults did participate in occupational activity, most likely a day program. These authors also found that employment was associated with the highest self-rated health outcomes, yet other occupational activities were also associated with improved outcomes.

Because of adults with IDD' different residential and employment experiences compared to the general population, their transition into retirement requires unique considerations. For older people in the general population, retirement often means phasing out of employment and receiving other financial means to support a non-employment lifestyle with more time for leisure activities. Retirement of people with IDD is not financially driven in the same way, as most rely on governmental benefits for income. Additionally, their daily support structures and environments often do not change when they reach retirement age [27]. For adults with IDD who live at home, life might stay very much the same at retirement age. For adults with IDD who live in residential settings, opportunities to engage in a retired lifestyle depend on the policies of the agency that supports them [84].

Older adults with IDD often lack choice in the activities they want to pursue, and some are hesitant to retire because of the importance of employment or day program activities in their social lives and friendships [84, 109]. Yet, a different pace and approach to employment or daily activities may be appropriate and necessary as adults with IDD age [80]. For some adults with IDD this may mean modified tasks or schedules, for others it may mean retiring completely [104]. For many older adults with IDD the meaning of retirement becomes about transitioning from employment or a day program to social and leisure activities. For some older adults with IDD this transition may mean participating in senior centers, which can give them the opportunity to learn and grow in a new and integrated environment [5].

Stancliffe and colleagues [110] developed and tested a program to support older people with IDD in the transition to retirement. The unique component of this

program is that older adults with IDD receive support to participate in local community groups (such as choirs, walking groups, community gardens etc.) by trained mentors who are group members. The evaluation of the program found that the adults with IDD who participated in the program worked less hours, participated more in their community, made new social contacts and were more satisfied socially than the adults in the control group.

Volunteering is another avenue for meaningful engagement for adults with IDD as they age, it can help them build social relationships and develop new skills [111]. A study by Wicky and Meier [112] found that volunteers with IDD valued the social recognition received through volunteering, which was particularly important to them as many experienced discrimination and marginalisation. Volunteering may be underutilized among the population of older adults with IDD [104].

Retirement or engagement in the later stages of the life course should bring new meaning for older adults with IDD and their activities should be in line with their desires and needs as they age. Despite the changes that come with ageing, adults with IDD want to continue to lead active and productive lives and they want to participate and contribute to their communities [81].

Supporting Social Relations

Strong social support networks and overall social well-being are important predictors of QOL and happiness as one ages [113, 114]. Developing long lasting and close relationships leads to positive health outcomes [25] and experiencing social inclusion and engagement in the community throughout the life course promotes healthy ageing [115]. Social isolation and a lack of social support have an opposite effect [113].

Social support networks develop across the life course. The social networks of older adults are reflective of the life they have lived, the choices they made in their life and the opportunities they had to develop and maintain social relationships [116, 117]. Throughout their life course, adults with IDD experience barriers to social inclusion and community participation and the development of social networks. These barriers include marginalizing social and cultural attitudes and experiences of segregation in education, employment, housing, transportation and leisure activities [118, 119]. The social networks of adults with IDD are often small. They do not have many relationships with people who do not have IDD beyond family members and other caregivers [21, 22, 120]. Research has reported that many people with IDD spent a significant amount of their free time alone, and there is evidence for the experience of loneliness in adults with IDD of all ages [121, 122]. When they participate in community activities they tend to be routine and orchestrated for them with limited interaction with the public [123].

People with IDD often are unmarried and do not have children and therefore cannot lean on a spouse or children for additional support as they age [4, 124]. Even though people with IDD do not experience an ‘empty nest’ as they age, they do lose

family members, caregivers, and friends. As older adults with IDD lose those closest to them, they experience an increased risk for social isolation and become more likely to face their own ageing and dying alone, particularly if they do not have involved siblings [55]. Another reason for concern is that older people with IDD have fewer close friendships than other older adults [21, 125, 126]. Formal caregivers such as direct support staff become essential to the social support networks of many older adults with IDD [125]. Yet, direct support staff cannot replace the long-term affection, love and advocacy that can only be provided by family members and friends [125, 127]. Direct support staff should support adults with IDD to build and maintain informal supportive relationships [124]. Facilitating continued engagement with family members and friends is one important part of this type of support. Goals related to an individual's social network and strengthening of social relationships can be included in person-centered and future planning [120]. Another strategy that can be included in person-centered and future planning is social network mapping. Mapping social relations can reveal the history and importance of relationships to an adult with IDD as well as their desires for social relationships in the future [4, 124].

Research by McConkey and colleagues [128] in Ireland found that having access to community activities and maintaining contact with family and friends was important to all adults with IDD in their study, regardless of their living arrangement. Adults with IDD have also indicated that they want to engage in more community activities and have more friends [129]. They value the companionship and support stemming from close friendships [130]. Person-centered and future plans need to include personalized goals for community participation and residential settings need to provide opportunities for older adults with IDD to participate in community activities [131]. Being a member of local community groups or volunteering can help adults with IDD make acquaintances and spend time with others. Such social opportunities are building blocks for social inclusion and the development of social relationships [124].

Conclusion and Future Directions

People with IDD face many disadvantages throughout their lives that multiply and accumulate when they get older and cause them to age with more physical, psychological and social challenges than other older adults [6]. There is an implicit common agreement of the wider public that the lives of people with IDD are of lesser quality than those of people in the general population [132]. On the contrary, people with IDD rate the quality of their lives as quite high, implying that QOL is indeed a holistic framework that applies to all people [133, 134]. The 2006 United Nations Convention on the Rights of Persons with Disabilities prescribes equal treatment of people with disabilities [135]. People with IDD have the same rights as all others to live a good life and societies hold responsibility to respect and protect those rights.

To promote and support QOL among adults with IDD as they age a number of future directions in practice, policy and research can be identified. As parents and other family members pass away or become less available to provide support, the social networks of adults with IDD decrease rapidly. When adults with IDD move to residential services or retire from work or other daily activities, their already small networks become even smaller [136]. In maintaining and fostering existing support networks, it is of interest to invest in timely and explicit communication with relatives other than parents, for instance siblings - if any. Appointing a key person, who knows the person well and can be responsive to changing needs of the individual adds to the perceived QOL of older persons with IDD [6].

With adults with IDD living longer lives than ever before in history, new opportunities arise for personal growth and development in later life. Unfortunately, older people with IDD lack support in preparing for the life phase of retirement and remaining active [6]. It is important to give voice to older people with IDD themselves by including them in the process of future planning and putting them 'in the driver's seat' [35]. Timely future planning should explore individualized strategies to build relationships outside existing support networks, opportunities for engagement in community programs and participation in other personalized meaningful activities. Future planning also needs to integrate life story work that centers the life experiences and preferences of older adults with IDD. Overall, future planning needs to be more broadly encouraged and implemented in ageing and disability social service delivery, including through community agencies.

In order to facilitate implementation of future planning, longitudinal and intervention research needs to be conducted to further encourage the development of evidence-based programs [92]. Future research also needs to investigate the meaning of retirement to and for people with IDD, and identify best practices to plan for and support positive transitions into a retirement lifestyle for people with IDD [100, 104]. The impact of different environments in which older adults with IDD receive support for active engagement (such as employment, day programs, senior centers, or individualized support at home) needs to be investigated [106]. As presence in the community does not automatically lead to inclusion in the community, future research also needs to investigate how a sense of belonging can be fostered in older adults with IDD [7]. More insight is needed into the experience of self-determination among older adults with IDD and its impact on their subjective QOL. Finally, future research needs to investigate coping strategies for people with IDD as they are impacted by age-associated losses [78]. This includes bereavement and their own end-of-life concerns.

The perspectives of older adults with IDD on their lives continues to be a rare topic of investigation. Inclusive research includes people with IDD as co-researchers and draws upon their strengths [137]. Inclusive research with older adults with IDD can facilitate a much needed examination of their lived experiences and those aspects of their lives that bring them happiness, joy and meaning.

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Chapter 22
Disposable Lives: Is Ending the Lives of Persons with Intellectual and Developmental Disabilities for Reasons of Poor Quality of Life an Emergence of a New Eugenics Movement?

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Introduction

With rare exceptions 'eugenics' is, generally speaking, not a term that is favourably used. In the context of this chapter the term refers to terminating human life affected by disability, particularly intellectual and developmental disabilities (IDD), in various medical practices. In support of these practices people say they are justified on the ground that human life can be 'defective', and that the human condition will be improved when IDD can be eradicated. Their critics define this view as 'eugenic', which is not a term that the agents and recipients engaged in such practices use themselves. Instead, they assert that what they are supporting aims at ameliorating human suffering, it is not about improving the human condition. They therefore reject the accusation of being driven by a notion of inferiority of the lives of the human beings involved.

This chapter will refer to these positions as informed by two opposing perspectives that will be presented respectively as 'internal' and 'external' to the contested practices. Before we start elaborating on these two perspectives, it is important to put in a caveat on how they should be read. In actual reality, distinctions neatly separating them are often intertwined, and hybrid variations occur. Regarding some of

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the issues that will be discussed readers may find themselves switching positions, depending on experience and expertise. Mapping the landscape as characterized by two opposing perspectives will be useful nonetheless, in view of the task of the present chapter. The aim is to describe moral issues arising from practices that are questioning the lives of persons with disabilities, particularly people with IDD. In taking this aim the chapter deliberately focuses on one particular interest. It calls on scientists and academics who have devoted their professional life to improving the lives of persons with IDD and their families. In this respect, the chapter takes its inspiration from the *Convention for the Rights of People with Disabilities* adopted by the *United Nations* in 2006 and seeks to support disability advocates in their concern about how 'the new eugenics' may affect its mission. The purpose is to support them in working towards social justice and equality for all human beings with disabilities, and make this position publicly known [1].

The term 'eugenic practices' canvassed the termination of human life in three different contexts [2]. It described current developments that involve ending the lives of infants that are born with a severely disabling condition ('mercy killing'), of future infants that will be born with a disabling condition when carried to term ('preventing suffering'), and of other persons with IDD directly or indirectly subject to practices of euthanasia or physician assisted suicide. This chapter will explore the latter issue in greater depth.

In bringing the termination of human life under the heading of the 'new eugenics', the chapter draws attention to earlier episodes in which this was practised. Eugenics is a program of social and moral reform that aims at improving the quality of human life by improving the human genome pool [3]. In the early twentieth century legislators, government agencies, scientific organisations, and social institutions throughout the western world pursued the goal of sanitizing society by isolating and marginalizing its alleged inferior members, mainly to prevent reproduction. People with IDD have always been a key target group of such programs. By naming practices of terminating human life affected by IDD as the 'new eugenics' the chapter investigates difference and similarity with the 'old eugenics'. Eugenics *per se* does not include coercion. In fact, some of the leaders of the early eugenics movement were opposed to state-enforced reform, while reversely coercion is what gave the first generation eugenicists their bad reputation [3–5]. In our days the practices of medicine addressed in this chapter are not embedded in explicitly laid out policies operated by public institutions or government agencies. As will be discussed later, these practices are much more 'consumer-' or 'market-driven' than they are 'state-driven'. This is particularly true in the area of reproduction. The ideal of enhancing the quality of the human gene pool seems to develop in the twenty-first century bottom-up rather than top-down [6].

Nonetheless, as the chapter will show, there is also a similarity. It regards the justification of terminating the lives of human beings affected by IDD in current medicine. Both then and now the practices at issue are justified because these lives are considered 'defective' and of 'poor quality'. The chapter takes a critical stance in view of descriptions such as 'mercy killing', or 'the prevention of suffering', or 'procreative beneficence' and opposes the utilitarian ethics that generates them.

Historically, ‘eugenics’ refers to a movement that was at the peak of its popular hearing at the turn of the twentieth century. It united people in favour of a social-Darwinian view of the human race. Darwin had taught that life is a competition for survival of the fittest, which implies, for any natural species, that only the healthy and strong among its members will survive in the long run. Social-Darwinists did not only apply this principle to the members of the human species, but to human societies as well. A strong and healthy society could not afford to accept or ignore the presence, let alone the procreation, of members affected by disabling conditions, both socially, physically, and intellectually [7]. The presence of an ‘inferior’ underclass implied a political concern about people whose lives were a mixture of poverty, illiteracy, and ‘feeblemindedness’. It became the main target of the eugenics movement that gained influence in the late nineteenth century; and succeeded in shaping public policy in the early twentieth century [8]. Using legal instruments such as forced sterilisation, society intended to protect itself against being burdened—socially, morally, and economically—by perceived unfit and inferior human beings [9]. The eugenics movement reached its nadir with the Nazi Aktion T4 program, a precursor and testing ground for the wider holocaust to come. Aktion T4 led to the state sanctioned physician led murder of some 300,000 people with disabilities, many on the grounds of ending suffering [10, 11].

To bring contemporary practices in medicine that involve terminating human life under the heading of eugenics, then, is to look at these practices from a historical vantage point. In doing so the present chapter takes what the introduction named an ‘external’ perspective. Its approach is to invite the reader to stand back and see the similarity with what happened in many western countries about a century ago. The moral force of the historical equation with the ‘old eugenics’ would not be as strong as it is, were it not for the fact that the eugenics movement is seen to have provided the pretext for the massive killing of people with IDD in The Third Reich. For that very reason the connection with the original eugenics movement is vehemently opposed by those in support of current medical practices of genetic testing, mercy killing, euthanasia, or physician assisted suicide. In adopting an internal perspective, they will categorically deny that terminating human life in their particular context has anything to do with ‘eugenics’.

In this chapter we do not intend to defame healthcare professionals by the emotivist use of inflammatory language. The term ‘eugenics’ was coined by a historical movement with identifiable characteristics, which makes it possible to ask in which respect the practices discussed in this chapter are similar, and in which respect they are different. It is instructive in this connection to make a distinction between ‘negative’ and ‘positive’ eugenics. The distinction goes as far back as Sir Francis Galton, one of the leading figures of the early eugenics movement [12]. Historically, *‘positive eugenics’* (PE) has meant promoting the idea that healthy, ‘better classes’ of people should have children, or have larger families; introducing institutions and policies that encourage ‘good’ marriages as well as pre-natal health care. Negative eugenics has meant restrictions of freedoms, for example, the restriction of immigration based on putatively eugenically undesirable traits such as race and ethnicity, and, for the same reason, prohibition of marriage and procreation, as well as

sterilisation [13–15]. In our own time positive eugenics aim explicitly at maximizing the quality of human life by means of reproductive techniques. Examples are establishing sperm banks where eugenically desirable traits like 'intelligence' are selected, and the practice of preimplantation selection of 'high quality' embryos in in-vitro fertilisation clinics that produce more 'healthy' and 'talented' children [16]. Or the ethical exhortation to practice 'procreative beneficence' in selected the 'best' children [17].

New Eugenics, in contrast, refers to the same goal of enhancing the human condition, but with a wide spectre of radically different means, for example the 'mercy killing' of new-born children affected by disability whose lives are considered to be an unbearable burden. The present chapter mainly addresses the 'negative' case. It aims at practices in contemporary medicine involving the termination of human life affected by IDD for reasons of 'poor quality'.

As indicated, supporters of these practices strongly reject labelling what they endorse as negative eugenics. They insist that these practices are only acceptable when particular moral and legal 'safeguards' are in place. Accepting that human life affected by IDD may result eventually in defective quality of life (QoL), they do not regard this as a sufficient reason for terminating it. In their view 'poor quality of life' is a necessary, but not a sufficient condition for terminating disabled human life. All existing euthanasia and abortion laws do in fact specify conditions that have to be fulfilled for killing in the context of medicine to be justifiable and legitimate.

Accordingly, from an 'insider' perspective, the morality of terminating human life affected by IDD is guided by a host of moral and legal qualifiers. Such qualifiers enable us to identify when acts of this kind should be disqualified as manslaughter or murder. Frequently mentioned in this connection are distinctions between 'reversible' and 'irreversible', or 'bearable' and 'unbearable', or 'voluntary' and 'involuntary'. Discussions on these acts from an internal perspective address the validity of such distinctions as '*guidelines*'.

'Disability'

For the purposes of the present chapter it is important to understand that both perspectives distinguished above also depend on different views on IDD. The difference plays out at various levels. First, there is a difference regarding the causality of disabling conditions. Those who defend the option of ending human lives in a medical context tend to consider the cause of disability to be inherent to the physical and/or mental conditions of the persons involved. Some human beings are born with, or have acquired, irreversible conditions that bar them from 'normal functioning'.

Their antagonists dispute this conception of disability, because they do not think of disability as an individual trait inherent to a person's biological or psychological condition. In their view disability is caused by the interplay between individual and environmental factors, such that an individual trait may, or may not result in a disability, depending on how a person's social environment responds to its

manifestation [18]. While the first position finds disability in the person, and holds it responsible for a poor QoL, the second position finds disability in the interplay between an individual condition and its socio-historical circumstance. It typically refers to physical or mental traits of the person involved as '*impairments*'; and limits the notion of 'disability' to the social effects of living with a particular impairing condition in one's particular environment.

Second, given these different conceptualisations of disability there is a corresponding difference on how to respond to a verdict of *'poor quality'*. If one takes it to refer to the disastrous effects of 'defective' and irremediable physical and/or mental traits inherent to the person, then the option of ending that person's life presents itself as thinkable. On that view further questions will be raised before seriously considering it as a possible option. Is the condition stable? Are medical interventions feasible to improve the person's QoL? Does it involve a great deal of suffering? Depending on how such questions are answered the verdict of 'poor quality' may be taken as both a necessary and sufficient condition to consider ending that human being's life. On the other hand, if one adopts the conception of disability as the result of social interaction then the most likely response to such verdicts is to ask which factors creating the environmental response are amenable to change. Different conceptions of disability, in other words, invoke different strategies of responding to the person's needs. This is a crucial difference in adopting an 'internal' or 'external' perspective on the issue of terminating disabled human life.

Third, the previous points seem to presuppose that QoL is an uncontested concept. It is not. Particularly in connection with IDD, scientists and 'lay people' may find themselves in dispute about what 'truly' constitutes quality of life. Three different angles can be distinguished. In a medical context the verdict of 'poor quality' will depend on the prospective judgments. The questions raised regard estimated problems in health and development as predicted by healthcare professionals. In the context of support services, however, the concept of quality of life is taken in a much wider sense, in which it not only refers to health conditions, but also to social and material conditions. What both approaches share, however, is the drive towards objective standards to make QoL measurable, even when the actual 'instruments' used are different. In this respect both are different from a third perspective that reflects the world of people who are experienced in living with IDD, or in sharing their lives with someone with such a condition [19]. Personal experience may typically change people's conception of QoL, especially when rewards of overcoming bleak medical prognoses are underestimated. For example, take the remarkable example of a young man whose progressive muscular disease left him deaf and blind, but who with the assistance of support professionals and friends developed his own communication device and managed to graduate from high school, and who judged his QoL to be A minus [20]. It is not just that different approaches to QoL open up a range of different responses to a verdict of 'poor quality', it is also that the verdict itself will not go uncontested. Particularly the use of health-related quality of life (HQoL) as an indicator for end-of-life decisions occurs only in the medical world, and even there is not generally accepted.

Fourth, from the internal perspective '*end-of-life'* decisions are typically framed in respect of doctor-patient relationships. Issues raised about such decisions regard the responsibilities attached to these roles, usually couched in the language of moral obligations, duties, and rights, for example '*the right to die'*. From the conception of disability as conditioned by social environments, the spectre of responsibilities is widened. The individual aspect of 'end-of-life' decisions is abandoned and replaced by questions regarding collective responsibilities for the social and cultural conditions of the debate on these decisions. The way society at large responds to '*disability*' is taken to be a major factor in how individual people arrive at their views on the QoL of the human beings involved. We will see the importance of this point on several occasions in later sections of this chapter. Fifth, following from the previous points, the two perspectives distinguished here tend to be inhibited by people from different academic disciplines. The conception of disability as a trait inherent to the person is more likely to be found among medical doctors, scholars in health law, bioethicists, and scientists from various medical fields. In popular parlance, the *'medical model of disability'* is obviously reflected in positions taken by people at home in disciplines constitutive of that model. In contrast, the socio-cultural conception of disability as the interplay between individual traits and environmental responses is more often represented by people from the social sciences, history, and disability studies. We repeat here that, as indicated before, these demarcations are frequently blurred. For each of the disciplines listed here it is true that in actual reality people may find themselves at cross-purposes with colleagues in the same discipline. Researchers from both worlds are at cross purposes with one another on the concept of disability and its implications. Traditionally people from the medical world—paediatrics, psychiatrists—have contributed much to the knowledge of conditions nowadays named as '*intellectual disabilities'*. In more recent times researchers from other academic disciplines—special educators, psychologists, sociologists—have begun to investigate a wider spectrum of factors and conditions that actually may influence their understanding of people's cognitive abilities, including their scientific instruments for tests and assessments.

In the last couple of decades, the disciplines from the social sciences have gradually come under the influence of critical disability studies. The perspective on disability in society at large has shifted, and the scope of thinking about IDD changed in ways that have influenced governments, politicians, lawmakers—public officers in general. Many are familiar with seeing people with IDD from the perspective of citizenship. Globally the most significant sign of this trend, as noted earlier, has been the adoption by the United Nations of the Convention on the Rights of Persons with Disability in 2006 [1]. To date researchers across social scientific disciplines may find themselves at cross purposes on many issues, both theoretical and practical, but not with regard to their core business, which is to look at persons with IDD and their families in terms of people with support needs. They aim to investigate the factors that can help to improve the effectiveness of the supports that have been developed and implemented.

Another landmark is the universal support for the *International Classification of Functioning, Disability and* Health (ICF) adopted by the World Health Organisation in 2001 [21]. It expresses the view that disability is the product of an interplay

between individual and environmental factors, and so informs the expanding agenda of investigating these factors in a multi-disciplinary community of researchers.

Quality of Life

Given the crucial role of the concept of QoL already addressed above, it is necessary to go in more detail regarding the different scientific approaches. In a medical context QoL is usually specified as health related QoL. In the international community of disability researchers, however, the concept is taken in a much broader sense. A number of *'life domai*ns' is taken into consideration to determine a person's QoL, of which '*health*' is only one, sided by a number of other domains such as relationships, social inclusion, material well-being and rights [22, 23]. The result is that poor results in one domain may be compensated, eventually, by high performances in other domains. A life in good health but without friendships does not evidently outrank a life in poor health that is shared with many good friends.

As a consequence of these diverse conceptualisations, the measurement instruments used in both contexts differ. Health-related QoL instruments provide information about the effects of a specific treatment or therapy on the patient's condition. This approach implies the possibility of using the obtained information in clinical practice to assess end-of-life decisions. Whether they *actually* are so used depends on particular circumstances and local jurisdictions.

In contrast, QoL measurement instruments in the social sciences provide information about the effects of policies, programs, and treatments in a support context. The information in this context is not exclusively related to health conditions, nor is it used in connection with end-of-life decisions. Early in the 1990s a Dutch professor in special education by the name of Ad van Gennep coined the term 'kwaliteit van bestaan' ('quality of existence') to avoid any association with the medical use of '*quality of life'* in the context of end-of-life decision-making. In the world of human services in The Netherlands the alternative term has become the standard use. Particularly in the past 20 years research has established that QoL-oriented services and supports can have significant positive effects on the lives of people with intellectual disabilities and their families [24].

Concerning to differences between disciplinary perspectives on QoL the concept of Quality of Life Adjusted Years (QALY's) also must be mentioned here. The term appeared in the 1980's in the discussion on new and costly medical interventions, first with regard to assessing their benefits for alternative medical conditions, second with regard to respective benefits of alternative interventions for individual patients. The decisive question is whether these interventions are cost-effective in the sense that their life-prolonging benefits outweighed the costs attached to these benefits. The concept of a QALY was introduced to facilitate the calculation [25]. Serious concerns have been raised regarding whether QALYs retain an inherent bias with regards to disabled persons and what the impact of this might be in the context of euthanasia and assisted suicide [26, 27]. In view of its origin it is not immediately

evident how the concept may enter into the process of clinical end-of-life decision-making. But gradually the notion of QALYs seems to trickle down into the clinical practice of *Neonatal Intensive Care Units* (NICUs) regarding the question when life-sustaining interventions on very preterm neonates ought to be withheld. Introducing QALYs in the prospective assessment of these interventions includes considering how foreseeable effects of IDD may determine children's HQoL [28].

The Aim of the Present Discussion

Developments in the field of IDD as indicated suggests that practices of negative eugenics do not easily accord with what social scientists consider to be their core business. Their goal is to improve people's QoL by improving the supports they need, which makes considerations concerning termination of lives affected by IDD appear as counterintuitive. Again, in actual reality members of the same discipline may have different opinions in this connection. For example, both in geriatrics and paediatrics there are those who in principle oppose end-of-life decisions that aim, directly or indirectly at their patients' death. They emphatically will assert that improving QoL of their patients is also *their* core business. At the same time there are social scientists who will accept that end-of-life decisions are in general undesirable, but that incidentally may be unavoidable, perhaps even morally commendable. As indicated, conceptual boundaries drawn to demarcate distinct views on the matter are blurred in reality, and opposing positions are represented across disciplines and perspectives.

The aim of the present discussion, however, is not to target incidental cases or examples of eugenics, however described. Instead, it is to draw attention to expanding eugenic practices as a social phenomenon. It addresses social and cultural trends reflected in this expansion. As we will see, contestable assumptions and presuppositions about what it is to live with a disability largely go unquestioned in the context of decisions to end human life affected by IDD. The practices of medical eugenics appear to be incapable of questioning the presumption about 'normal' functioning bodies and minds. In short, ending human life affected by IDD feeds on preconceptions that in many ways are prejudicial, and as such are questionable from the perspective of professionals and scientists working towards high quality human services. As noted above, our main focus here will be on euthanasia and assisted suicide, however a brief review of the related issues of selective termination and neo-natal euthanasia will help provide some broader context as many of the underlying ethical issues and rationales cut across all three practices.

Neonatology and Pre-natal Testing

To warrant the claim to a potential growth of eugenic practices it is necessary to produce reliable factual accounts.

Historically speaking, reliable accounts from the past are at hand. The eugenic program in Nazi Germany for exterminating people with IDD, for example, has been well-documented [29–32]. So has the legal enactment of programs for forced sterilisation in a variety of western countries [33–35]. However, contemporary programs orchestrated by public agencies are not the main vehicle for the practices under consideration in this paper, which means that building the claim for rising eugenics based on factual evidence will be a more complex task. It requires interpreting actual developments in medicine in view of culturally transmitted preconceptions of disability. What some regard as clear-cut examples of rising eugenics is for others a manifestation of a more benign and compassionate way of practising medicine. Any claim to a description of eugenics in contemporary medicine will therefore be heavily contested. Nonetheless, to describe them in these terms is to claim that with the recognition of important differences there is a relevant similarity with the views underlying eugenics in previous times. Whilst not the primary focus of this chapter, it is nevertheless important to reflect briefly on practices in the field of neonatology and pre-natal screening and testing, as there are similar arguments advanced in the area of end-of-life decisions.

The Groningen Protocol

A case in point is the so-called Groningen Protocol (GP) that has been developed since 2004 in the hospital of the University of Groningen in The Netherlands [36, 37]. It poses a system of rules to determine when the lives of new-born infants with life threatening conditions can be legitimately terminated. ‘Legitimately’ here refers to legal standards. It was an attempt to enable paediatricians to come clean about cases of ‘mercy killing’ they had performed at a time when Dutch law did not provide a legal space to do so. By holding themselves publicly accountable paediatricians wanted to push the courts to draw the line between legitimate and illegitimate killings of new-born infants.

The new-born infants whose lives had been terminated faced a life of ‘unbearable suffering’, according to the authors, while their prospect in life would inevitably be limited to a life of ‘poor quality’. Putting these children—and their parents—out of their misery was depicted as a benign act. The question here is how the claim to unbearable suffering regarding these children was constructed.

The authors of the GP reported three kinds of cases that involved decisions to withhold or withdraw life-sustaining treatment of ‘neonates’, two of which they claimed—without much opposition—to be generally uncontested by pediatricians [36]. Opponents of the GP, too, have argued that there are morally acceptable cases wherein the obligation to preserve life has ended. They point to cases where either the prognosis of death is certain or almost certain, which in their view makes causing death by withdrawing life-sustaining treatment an acceptable outcome [38].

‘Unbearable Suffering’

The GP goes a step further, however, when it considers the third case: disabled infants that will survive their illness on their own, even after life-sustaining treatment is withdrawn, but whose fate in the eyes of their doctors and parents is too horrific to contemplate. These are

> *…. infants with a hopeless prognosis who experience what parents and medical experts deem to be unbearable suffering. (….) This group also includes infants who have survived thanks to intensive care but for whom it becomes clear after intensive treatment has been completed that the quality of life will be very poor and for whom there is no hope of improvement* [36].

The authors of the GP reported that 21 infants in their third group (N = 22) were diagnosed with *spina bifida*, a condition that can be managed and which does not preclude a life of dignity and quality. Persons with spina bifida and families involved in no way consider their lives to be burdened by ‘unbearable suffering’ [39].

An analogous problem holds with regard to the notion of ‘poor quality’. To date there is no theory of QoL that does not include a subjective dimension, which is to say that someone’s QoL is in some sense necessarily related to his or her own perception. It also raises the question of whether the concept is at all applicable to new-born infants as the claim of ‘poor quality’ can only be prospective.

To summarize: while the connection between poor quality of life and unbearable suffering provides the moral basis for terminating the life of severely disabled infants whose condition matches the description, upon closer inspection that connection falls apart. It cannot be applied critically to new-born infants. What remains of the moral justification that the authors of the GP provide for terminating their lives here and now is that they will at some point experience “*what parents and medical experts deem to be unbearable suffering*.”

The Similarity of ‘Old’ and ‘New’ Eugenics

The conclusion presents itself that the prospect of a ‘poor’ QoL for disabled infants necessarily depends on representations of what people believe to be true about the lives of the infants involved, like those born with *spina bifida*. Apparently, these are lives that no human being should be ‘forced’ to live.

The position underlying the GP is seriously flawed, however. Particularly children with congenital disabilities have never known themselves other than with these conditions. The assumption that the child’s projected response to this condition will amount to ‘unbearable suffering’ is unwarranted. To them living with a disability is the ‘normal’ state of being, in which they the experience the world around them. When a child with a disability suffers psychologically, this is most likely induced by negative responses from its social and cultural environment. Anxiety, distress, disappointment, any of these experiences can be part of what it is to live with IDD, but this suffering results from how others react to that condition. ‘Nature’ and biology

may produce the impairing conditions of human life, but they do not produce meanings. That is to say, they do not produce socio-cultural representations of what it means to live with such conditions.

It is at this point that the similarity with the 'old' eugenic practices of killing disabled new-born infants becomes apparent. Ultimately the justification of these killings depends on the assumption that some infants with IDD ought to be spared a life that humans ought not to live. Experiential accounts of quality of life by disabled children do not differ from accounts from within their peer group without similar conditions [40]. Further, doctors tend to underestimate disabled persons' quality of life as compared to self-reports [41] which generally differ with how non-disabled people rate living with a disability; and medical predictions about quality of life do not appear to be very reliable [38].

Finally, it is worth mentioning that the GP has been defended by referencing its particular cultural background in the country of origin, *i.e.*, The Netherlands. Lindemann and Verkerk [37] argued that some misconceptions about the GP can only be understood in reference to its social and cultural context. Contrary to their intentions the authors inadvertently affirm this conclusion when they write: "*It is precisely those babies who could continue to live, but whose lives would be wretched in the extreme, who stand most in need of the interventions for which the protocol offers guidance*." The GP could not have arrived at its current state of recognition in The Netherlands without a cultural environment that shares its negative presuppositions regarding the condition of IDD.

'Tragedy' and Beyond

A second area of potential evidence of a new eugenics is the practice of selective termination of fetuses with Down syndrome (DS) after identification through prenatal screening tests (PNT). In view of the picture emerging from the research on PNT, what stands out is the absence of accurate information about living with the condition of DS [42]. After all, such information has been available for quite a while [43, 44]. The view of a child born with DS as a 'tragedy' is most likely part of this. When information about DS *does* enter into the practices of genetic counselling it often does not surmount the 'medical textbook' type of account.

What renders this suggestion plausible is the fact that from the start the advances in the science of PNT have been sold to the general public as creating an opportunity to avoid human suffering. A powerful example of this sentiment is provided by a comment from Francis S. Collins at the time when he was Director of the National Center for Human Genome Research in the US:

> *The mandate to alleviate human suffering is one of the most compelling of all expectations of humanity. (….) When genetics is seen to fall into that larger mandate, it is hard to argue with its potential goodness. In fact, given that potential, it can be argued that the most unethical approach of all would be to insist that genetic research be stopped; because if it were, those individuals, present and future, who suffer from the ravages of genetic diseases would be doomed to hopelessness* [45].

Striking in this comment is the wholesale opposition between accepting genetics for its potential goodness on the one hand, and the 'most unethical approach' of insisting that genetic research be stopped on the other. No one is arguing that genetic research must be stopped, of course. But it certainly would help if the medical research community would be open to more accuracy in depicting the world of disability experience than Collins' comment encourages it to do.

Euthanasia and Assisted Suicide

The two areas of developing eugenic practices that have been discussed so far are tied in with technological developments in two separate areas, life support for prematurely born infants and PNT respectively. The third, regards medical practices that are more difficult to categorize. They concern the termination of human lives affected by cognitive impairments in quite diverse settings. Perhaps 'euthanasia' will do to categorize most of the cases involved, but certainly not all. Above we looked at ending the lives of newborn infants for reasons of poor QoL, which acts—when they occur—can safely be qualified as 'mercy killings'. The justification given for these acts is that living with certain conditions can be so harsh for these children that it is morally preferable not to 'condemn' them to live. In view of developing practices of euthanasia, however, these killings will fail to quality when a patient's 'autonomous' request is taken to be one of its defining characteristics.

Something similar will hold true for most of the cases to be discussed. In a sense, the problem of categorisation is actually the main subject under discussion here. Generally speaking, practices of terminating human life in a medical context that are mostly discussed under the headings of 'end-of-life decisions or 'euthanasia' and 'physician assisted suicide' (EAS), sometimes also as the 'right to die'. There are exceptions to the qualification of the contexts of these practices as 'medical'. Though the main focus here is on doctors and healthcare professionals, below a case will be discussed that concerned the killing of a girl with IDD by her father. The extent to which this case may represent a larger group of cases is unknown. Usually in such cases, as indicated, a person's own views on how to die as opposed to continue living is an important consideration in medical decision-making. Looking at the facts, however, one finds cases involving persons whose abilities of expressing their own views are uncertain, if not absent altogether, like persons with severe disability, or dementia, or Alzheimer's disease. Even though not 'fitting' the standard euthanasia case, one finds justifications for terminating the lives of such persons, a trend that indicates shifting patterns and perspectives.

The underlying issue here is the well-known 'slippery slope' argument. Supporters of this argument reason as follows. 'End-of-life-decisions' involving the termination of patient lives by their doctors are illegal and should remain so. The fact that there are patients explicitly and repeatedly requesting to have their lives ended is not a sufficient reason to legalize such acts and take them out of the penal code. Legalizing euthanasia may in due course result in the termination of lives of

patients outside this category, for example persons with an advanced stage of dementia. At some point, the argument goes, the legal justification for euthanasia will be extended to people that should be exempted from it because they cannot decide for themselves. Therefore, society had better not embark on this journey at all.

A further aspect of the slippery slope concerns how EAS may come to be an alternative to providing appropriate or acceptable disability supports. The 'suffering' while acutely felt, stems not from the inherent condition but rather from a failure to provide supports which would allow a person to live a life they consider worth living. EAS becomes a cheaper and easier option or there is simply a failure to consider social determinants which may be underlying a request for EAS [46]. There are also concerns about the process of consent and assessment of decisional capacity, particularly in relation to persons with IDD and autism spectrum disorders which, when coupled with negative valuations of lives of people with IDD, create a very dangerous and slippery slope indeed for persons with IDD [46–48]. The slippery slope objection, in other words, questions the viability of safe-guarding the legal justification of end-of-life-decisions and keep them limited to the category of requested killings.

Bad Samaritanism

From the perspective of explicitly limited definitions of EAS terminating people's lives without their explicit request is morally and legally doubtful at best, and unquestionably criminal at worst. But the question is whether this means both kind of cases are unrelated. It regards the connection between the occurrence of particular acts and the social context within which they occur. More specifically, is there a relation between illegal acts of terminating a person's life in a medical context and a socio-cultural environment in which the legitimacy of EAS is openly debated and defended?

On July 25, 2016 *The Guardian* reported the murder of nineteen people in a home for people with disabilities in a village outside Tokyo, Japan. The knife-wielding attacker was a 26 years old former employee of the facility. After his heinous crime the man turned himself in to the police, and declared his motive, explaining that he was angry about being fired and losing his job. "*It's better that disabled people disappear*," the police quoted him as saying, according to local reports. Some 150 people aged between 18 and 75 lived at the facility, according to a public broadcaster, all with IDD and some with additional physical disabilities and mental disorders [49].

About the killer's background the *Associated Press* reported that a few months before his act he tried to get a letter to a local legislator, outlining his intention to attack two facilities. In the letter he demanded disabled people be put to death through "*a world that allows for mercy killings.*" He argued that the government should permit euthanasia for people with disabilities. "*I envision a world where a*

person with multiple disabilities can be euthanized, with an agreement from the guardians, when it is difficult for the person to carry out household and social activities," the letter said (The Atlantic). He went then on to state, "*My reasoning is that I may be able to revitalize the world economy…*" [50].

This was clearly a horrendously criminal act. After the letter had been traced to its author, the killer had been investigated and found mentally disturbed. Only weeks before his crime he had been released from the mental hospital to which he was involuntarily committed. The point of bringing up this story here regards the justification advanced for his acts that linked it explicitly to the wider context of discussing the legitimacy of EAS. Is this a relevant fact about this case?

On November 21, 2017 a Dutch national newspaper reported that a 21-year-old employee of a residential facility for elderly people had killed a woman with dementia by injecting insulin. The man was suspected of two other murders. Only recently—February 2018—he was indicted for seven cases of murder, also by injecting patients with dementia with insulin. Nothing was reported about his motive for these acts, but his lawyer declared they were acts of compassion [51]. The case appeared as similar to the murder of nine residents with dementia reported in The Netherlands in 1996. The declared motive of the killer in that case, also an employee of a caring facility, was that the killer could no longer bear seeing their suffering.

Assuming 'compassion' as the main motive in such cases one could categorize them as 'mercy killings' but it seems wrong to put them in the same box as the killings of new-born infants as described in connection with the Groningen Protocol. At any rate a morally and legally relevant difference is that the latter were done in open and intense communication with these children's parents. Rather than to qualify the acts reported here as 'mercy killings' the description of a criminal version of 'bad samaritanism' comes to mind.

With regard to these cases in The Netherlands one might question their relevance as examples of killings in a medical context in a society that openly discusses and accepts the practice of EAS. Given the time interval of more than 20 years between them, it might seem that such crimes do not frequently occur. One wants to keep in mind however, that these are reported cases, which leaves open the possibility of unreported cases, both in The Netherlands and elsewhere. Second, like in the Japanese case, also here the perpetrators were considered mentally disturbed. Both facts might suggest isolated crimes, then, that apparently have very little to do with the debate over time on the legitimacy of EAS, either in Japan or in The Netherlands. They were not reported as instances of anything like euthanasia. Interesting in the Dutch case however, is the comment from an employee of the caring facility where the murder case of November 2017 took place, when she inadvertently linked these murders to the practice of end-of-life-decisions: "*It is very awkward for the other employees working very hard in that ward (….) It is the place where the medication is kept for the last stage when things are really not working anymore*" [52].

Acts of Killing and Their Socio-cultural Environment

Are lethal crimes against people with diminished capacities that are mendaciously justified as 'mercy killing' in any way related to developing practices of EAS? More specifically, are they in any way related to a socio-cultural environment in which the legitimacy of such practices is openly debated and defended? Put differently, is there a connection between these acts and a public sphere in which the legitimacy of EAS is frequently backed up by popular polls showing that a large majority of the people is in favor of legalizing them?

In the Japanese case, *The Japan Times* reported earlier that voluntary euthanasia was a hotly debated issue in the country, and that both the legislative and judiciary powers tended to be in favor of legislation but thus far had failed to give guidance on when acts of euthanasia could be legal [53]. To be sure there are very significant differences between euthanasia and mercy killing, but these are not the issue here. The issue is rather whether a cultural climate in which terminating the lives of patients is subject to open discussion by respectable people facilitates unstable and misguided individuals in thinking that perpetrating such acts is acceptable. Think about it as an analogy of the dispute on whether excessive violence in children's TV-programs produces more violent kids.

As is well-known, any causal connection of such phenomena is very hard to prove. There are too many people who may hold similar views on people with IDD as the Japanese killer did, and yet don't engage in similar acts. But perhaps causality is not the issue here either. The issue is how particular cultural environments produce structures of moral—and legal—justification that at some point are mistakenly taken to coincide with acts of killing particular people, even when they should not. In defense of EAS opponents of the 'slippery-slope' objection deny any such connection and hence insist on the clear demarcation of distinct categories. Terminating the lives of people with diminished decision-capacity is ill-conceived as a legitimate act because it is involuntary. Killing people without their explicit request should be categorized as murder.

Noticeable, in this connection, is that proponents of legalizing EAS as a distinct and separate category do not necessarily insist on the verdict of 'murder' when such acts are performed by benevolent medical doctors, notwithstanding the fact that occasionally they also engage in involuntary EAS. The relevant difference lies, presumably, in the fact that these doctors are intentionally subjecting themselves to the transparency and accountability of public scrutiny, as we have seen with regard to the Groningen Protocol. In view of such cases the question arises whether EAS can be separated and set aside as a distinct category. In actual fact patient conditions overlap. In cases where there is no request there still can be a patient who is unbearably suffering, including psychological or existential suffering indicated by the frequent reference to 'loss of dignity'. The two kinds of cases are not as distinctly separated as objections to a 'slippery slope' suggest. There seems to be an intermediate range of cases between euthanasia and murder that can be, and in actual fact are identified as 'mercy-killing'.

Also noticeable in the same connection is the repeated assertion that disability advocates have nothing to worry about, because legal safeguards send the message their lives are not at stake. We have seen the same in connection with the prevention of Down syndrome where it was argued that the introduction of non-invasive pre-natal test should in no way be taken to send the message that children with Down syndrome are less welcome as members of their society. Receiving this message appears to be less than reassuring in that disability advocates and self-advocates usually *begin* to worry at this point. A cultural environment that benevolently discusses the legality of terminating disabled lives sends a false message, in their view, namely that seeing this as an act of mercy is a relevant consideration. They would rather not be at the end of other people's benevolence. Self-advocacy groups such as *Not Dead Yet* therefore take assertions that they have nothing to worry about as suspect. The very fact of these assertion is felt as a performative contradiction. If they were truly welcome, it would not have to be asserted. What is beyond doubt needs no confirmation. At any rate, as *Not Dead Yet* in the UK insists, to date there is no single organisation by and for people with disabilities or terminal illness that has joined the argument for legal justification of euthanasia or assisted suicide [54].

So, the remaining question is whether the suspicions of self-advocates can be reduced to a form of collective hysteria in view of open discussions of EAS. Looking at the perceptions of disability in western culture these discussions appear as less than innocent. The qualification of 'western' here should not be taken to mean that other, non-western cultures hold persons with disabilities in higher esteem. The point is simply that the available data on this issue in relation to EAS originate mostly from western countries. Two areas of concern suggest that 'death' has a different meaning in connection with disability than it has with regard to non-disabled people. In a study examining over 200 news reports between 2011 and 2015 from North America about the murder of people with disabilities by their caregivers, the motive routinely given for these acts of killing was 'hardship'. The 'hardship narrative'—describing the life of the disabled person and his/her family as unbearably difficult—is frequently excepted by the media without much questioning. More significantly, the same appears to be true for the courts before these killings are tried. The law does not excuse them, but otherwise they do seem as excusable given the fact that imprisonment for the killers is relatively short, or is not ordered at all [55]. Looking at these reports the author concludes that the drive to explain them as 'mercy killings' produces killer-oriented rather than victim-oriented stories in which the actual lives of the disabled persons involved tend to disappear [55]. It is hard not to see a similarity with how the courts in The Netherlands have dealt with illegal cases of EAS: convictions without punishments equal to the sentences in other murder cases.

Of course, the obvious disparity with EAS is that the medical context is absent in these murders, but the verdict of unbearable suffering as the main justifying reason for EAS is not dissimilar to what the study identifies as the 'hardship narrative'. The conclusion suggests itself that illegal killings of persons with disabilities are more readily seen as excusable, even when criminal, which is a reason to be concerned.

In a high profile 1993 case in Canada, Robert Latimer a Saskatchewan farmer, murdered his 12-year-old daughter Tracy who lived with both physical and intellectual disabilities. He characterized his act as 'mercy killing', citing her pain, quality of life and disability as motivating factors. While the act itself was shocking, what is more concerning for our purposes here was the widespread support he received from both the general public and the lower courts, initially being given a minimum sentence well below the legal threshold for murder. This was eventually overturned in the Supreme Court, though they did note the Government had the power to pardon Latimer [56].

Subsequently he has become a high-profile advocate for 'mercy killing'. In 2012 Latimer appeared on a prominent Canadian public affairs program *16x9* in support of Annette Corriveau, a mother of two disabled adult children who is seeking the right to euthanize them. The promotion for the report in the program indicates the bias of what Perry calls a 'killer-oriented' rather than a 'victim-oriented' story. It announced "*a mother's plea for mercy"* and "*a father who has been down this road before"* for which both have paid a price. Subsequently Corriveau appeared on the popular Dr. Phil show to make her case, an audience vote returned 90% support for her plea for 'mercy' [57].

The second area of concern is from another study from the US regarding the other category within EAS, physician assisted suicide. It notices that since the Oregon Death with Dignity Act [58, 59] various proposals for legalizing assisted suicide have assigned physicians with the authority to decide whose request for assisted suicide is to be acknowledged. With this in mind, the study has looked at responses by healthcare professionals to requests that involve disability [60]. The findings indicate that in the case of people with disabilities the barrier against suicide seems to be much lower. Physicians and other health professionals would assist in suicides of persons with incurable conditions while offering suicide prevention to individuals seen as 'healthy'. Disability rights advocates have charged this practice as unwarranted devaluation of the QoL of people with disabilities [60]. Again, the findings of also this study suggest that death of a person with a disability is of lesser weight than death of a 'healthy' person, which adds to the view of a socio-cultural environment in which the justification of EAS is more readily accepted when it regards people with IDD.

There are opposing views, however. The *Washington Post* published an article in which the author reported the case of a man in his 50s with severe disability who was kept on a ventilator in a hospital in New Hampshire while the prospect of recovery was bleak [61]. The patient was sedated. Normally a physician would ask what kind of care the patient wants, which in fact means to ask about the withdrawal of life-sustaining support. In the present case, the man's legal guardian noticed his QoL was deteriorating without the comfort of his favourite things, and asked to focus on end-of-life care. This was forbidden by law, however. Regarding persons with IDD the law of New Hampshire, as in other states in the US, forbids legal guardians to make decisions about the withdrawal of life-sustaining equipment.

This reported case seems to question the claim that 'death' has a different meaning when IDD is involved. But on a closer look this is seen to be mistaken. Permission

to withdraw life-sustaining equipment is possible on grounds of 'poor quality' but it has to be decided by a judge whose task is to approve of all the details of the decision-making process [61]. Rather than protecting the life of the disabled person it seeks to counter the invasion of someone else's will in determining whether continued medical care is appropriate.

Legalized Euthanasia in The Netherlands

There are other examples of blurred distinctions and categorisations. In the late 1990s The Netherlands has legalized EAS, as is well-known, after two decades of public discussions of pioneering cases. Individual doctors had decided to take matters in their own hands and terminated the life of a patient, sometimes because that patient wanted to die, sometimes because of advanced dementia in a patient who had expressed her will not to have to exist in that condition, and sometimes because the patient was suffering unbearably in the final stage of his/her life. These doctors were prosecuted and convicted by the courts, but the courts occasionally withheld from punishing their crimes [62, 63].

This paved the way for the Dutch version of legalized euthanasia, defined as ending the life of a patient on his or her explicit and repeated request. To be legally acceptable, this act can only be carried out by a doctor, has to be reported to the coroner as an act of killing, and still can be prosecuted if the doctor has failed to meet legally established criteria for 'careful' acting. Regional Review Committees (RRC's) are established to rule whether these criteria have been met in individual cases. The public prosecutor will indict a doctor when a RRC reports legally disputable circumstances. Contrary to what many assume, euthanasia remains a criminal offence in The Netherlands. It can be punished with imprisonment of up to 12 years when a doctor is convicted in a court of law.

However, in the decades since this framework was established some developments indicate a widening scope of legally accepted cases of euthanasia. Not only in the sense of a steep curve of rising numbers: from *1923* in 2005 to *6091* in 2016, a rise of *217%* [64]. Also, in the sense of the changing profile of cases accepted as legally justified by the RCCs.

Based on an analysis of RCC dossiers it has been reported that the interpretation of criteria for 'careful acting' has changed on at least three significant points [65]. The original support for legalized euthanasia in the 1980s and 1990s in The Netherlands was based on the widely shared view that in the final stage of their lives people who are unbearably suffering and for whom no other relief can be given, should be able to ask their family doctor to end their lives. The vast majority of cases concerned terminally ill cancer patients facing imminent death. Twenty years later things have changed significantly. First, the presupposition of a longstanding relationship with a family doctor is no longer in place. This change is due to the emergence of experts who perform euthanasia but have no obligation to consider other options than the termination of life. Second, the life-time expectancy of

patients involved can exceed the period of a few months to go beyond 2 years. This is due to an unqualified acceptance of self-reported unbearable suffering by euthanasia requesting patients. Finally, there has been an expanding range of considered diagnoses. Numbers of euthanasia cases reported in in 2002 show cancer *1658*, heart-vascular *28*, neurological *61*, pulmonary *40*, other (including occasional cases of psychiatry and dementia *95*). Numbers reported 15 years later show cancer *4137*, heart-vascular *315*, neurological *411*, pulmonary *214*, dementia *141*, psychiatry *60*, age related *244*, combination *465*, other *104* [65].

Tuffrey-Wijne and colleagues [47, 48] present a troubling picture regarding the euthanasia of persons with intellectual disabilities and autism spectrum disorder in the Netherlands. They identify six case reports of people with intellectual disability and three of people with autism in records covering 2012–2016. In a subsequent review covering 2017 and 2018, they identified a further four cases of people with intellectual disabilities, one of whom also had autism spectrum disorder, and three with autism spectrum disorder. An earlier paper by Kim and colleagues [66] highlights similar concerns for persons with psychiatric disabilities. They note this is one of the fastest growing populations accessing euthanasia and/or assisted Suicide (EAS) in Belgium and the Netherlands, the two jurisdictions currently allowing EAS for psychiatric conditions as the sole underlying cause of suffering.

In view of these changes the earlier point about how particular structures of moral and legal justification may be transferred to other cases of ending people's lives, appears to have become true for EAS in The Netherlands. To mention just one thing: the development of the Groningen Protocol—as discussed before—would hardly be possible in a society that had remained highly critical towards the justification of terminating human lives of incompetent patients. Of course, it has been noticed that the prime justifying condition of being a voluntary, explicit and repeated request by the patient remained unfulfilled in the cases reported under the GP. As already indicated however, the reason to eventually forego this criterion was already in place. Euthanasia as a voluntary request was introduced in The Netherlands largely as leverage to disallow doctors to decide whether a patient's suffering was still endurable or not. In other words, to end unbearable suffering had been the prime concern of any form of euthanasia to begin with. The necessary but apparently only *intermediate* step had been to tie the evaluation of 'suffering' to the voluntary expression of the will to die by a fully conscious individual patient. Boer [65], a Dutch ethicist, argued that RCC's tend to accept the formal criterion of an explicit and repeated request as sufficient indication for the condition of unbearable suffering.

In view of this intermediate role of the request as leverage for advancing the legal argument, it appears that opening up the discussion about the suffering of infants born with severely disabling conditions would be merely a matter of time. Looking backward Boer [65] signals the failure to categorize and separate the 'traditional case' of euthanasia that elicited the country's support for legalized EAS:

> *Apparently, not even the most careful euthanasia monitoring system in the world has halted developments which many find reason to refer to as a slippery slope).*

A final fact about recent developments in The Netherlands in support of this claim is the opening of the so called 'End-of-Life Clinic' [67]. On its website it is stated that:" *The main focus is on those patients whose requests for assisted dying are more complex and often denied by their own physician: psychiatric patients, people with dementia, or patients with non-fatal diseases*.". The End-of-Life Clinic specialises in cases that were intentionally exempted from the legalisation of EAS in 2004.

Throughout the early years of public debate on legalizing EAS its proponents in bioethics and health law in The Netherlands have been quick to dismiss the 'slippery slope' argument. If there are certain categories of people that need to be exempted from its justification, such as people with IDD or dementia, then that is where society draws the line. In other words, slippery slopes are controlled by safeguarding regulations enacted into law. The obvious shortcoming of this rejection is that the 'slippery slope' objection was interpreted as a prospective concern, whereas in fact it is confirmed in The Netherlands as a justified concern in retrospect.

An interesting question in this connection is why the Dutch parliament decided to retain euthanasia in the penal code. The answer is that doctors needed to be aware that unless it is done according to a set of strict regulations, the act of ending a patient's life is immoral and illegal and thus open to prosecution. After a generation of doctors practicing euthanasia, however, the wider public apparently has become used to the idea and rests assured that it can be done 'safely', so that there is no reason, presumably, not to consider widening its scope by changing the strict regulations. The Netherlands are witnessing what was assured would never happen is now subject of an apparently respectable public discussion.

Canada's Medical Assistance in Dying Act

The observed link between the condition of disability and death arises also in connection with the recent passage of legislation allowing *Medical Assistance in Dying* (MAiD) in Canada. Among the criteria for being 'eligible' for this kind of assistance are found the usual safeguarding requirements (a minimum age of 18, a voluntary request free from external pressure, full information about alternative options). But there is also a demarcation of patient conditions. Eligible are those patients who "*have a grievous and irremediable medical condition*" [68]. The question of how this condition is defined in a way that includes '*disability*':

A person has a grievous and irremediable medical condition only if they meet all of the following criteria:

(a) they have a serious and incurable illness, disease or disability;
(b) they are in an advanced state of irreversible decline in capability;
(c) that illness, disease or disability or that state of decline causes them enduring physical or psychological suffering that is intolerable to them and that cannot be relieved under conditions that they consider acceptable; and
(d) their natural death has become reasonably foreseeable, taking into account all of their medical circumstances, without a prognosis necessarily having been made as to the specific length of time that they have remaining.

The explicit inclusion of disability here raises the kind of questions considered above. A disability can be a 'grievous and irremediable medical condition', apparently independent from an existing illness or disease. Thus defined 'disability' counts as such as 'a medical condition'. The next criterion of an "*advanced state of irreversible decline in capability*" explicitly includes persons with cognitive impairments also independent from illness or disease. Being the cause of intolerable suffering from the patient's own perspective a person's disability provides the condition that makes assistance in dying of that person legal, other things being equal. Disability appears in the purview of conditions for which the law opens the legal option of EAS. The potential impact of the law on persons with IDD and other disabilities has already been foreshadowed with the killing of a man who was unable to secure appropriate supports and hence found his life 'unbearable'. There was also a recent case of a physician providing the unsolicited advice to a mother of an adult woman with IDD and physical disabilities that assisted dying was now legal, despite his interpretation of the current law being completely wrong as it would not currently allow for MAiD in this situation [69].

Also, the Canadian law is intended to safeguard the demarcation of a category of eligible patients, but there is little reason to be assured that this demarcation will prove to be stronger than what is found in the Dutch euthanasia law. As a matter of fact, the new law was recently subject to two court challenges that are asking for a wider scope. In the Truchon case, the judge ruled that the reasonably foreseeable natural death (RFND) criteria was unconstitutional, striking down a key safeguard for persons with disabilities [70].

One of the more concerning aspects of the judgement was that Justice Baudouin rejected two explicitly stated goals in the federal medically assisted dying law (C14) which confirmed the inherent and equal value of every person's life, combined with the prevention of negative perceptions of the quality of life of persons who are elderly, ill or disabled and, the prevention of suicide. By rejecting these two stated purposes Justice Baudouin precluded an analysis of whether the removal of the RFND criteria would in fact impact the equality rights of vulnerable Canadians or whether it would negatively impact efforts to prevent suicide [71]. Perhaps more disturbing is, despite the rejection of stated purposes in the law, the federal government declined to appeal the decision and is currently revising the law to conform with the judgement and removing the RFND criteria.

Furthermore, the new law was also the subject of an expert review panel looking at whether it should be expanded to include advance requests, persons with psychiatric conditions and mature minors [72]. These issues will be the subject of a mandatory review of the legislation in late 2020.

The removal of the RFND coupled with the explicit inclusion of disability as 'eligible conditions' and the potential for both advance directives and eligibility based solely on a psychiatric condition creates a very dangerous potentiality and removes key safeguards protecting disabled and other vulnerable groups. Again, this raises the question of why one can be assured that there will be no slippery slope.

Diminished Capacity for Decision-making

The problem of blurred distinctions reappears in another way in assessing 'requests' by persons with IDD or dementia whose capacity for decision-making is doubtful. The Tuffrey Wijne and colleagues [47, 48] studies of cases of IDD/ASD found that a key problem was to determine in what sense and to which extent there was an explicit and repeated request made by the person involved. The study concludes:

Autonomy and decisional capacity are highly complex for patients with intellectual disabilities and difficult to assess; capacity tests in these cases did not appear sufficiently stringent. Assessment of suffering is particularly difficult for patients who have experienced life-long disability. The sometimes brief time frames and limited number of physician-patient meetings may not be sufficient to make a decision as serious as EAS (2018, p. 1).

The study mentioned above found that in the nine cases of EAS involving persons with IDD it was particularly doubtful to what extent they understood the information regarding alternative options. The circumstance of having these assessments made by doctors based on relatively few contacts left room for doubt about the adequacy of this procedure [47]. These findings concur with those from a study looking at EAS involving psychiatric patients from 2011 to 2014 (N = 66) [66]. Assessing the 'true' expression of the patient's will was found to be difficult and contestable. Granting patient requests in these cases involved considerable insight in complex and chronic illnesses, about which psychiatrists do not always agree. The findings of both these studies are significant in view of the fact that considering the option of EAS for most participants was induced by the refusal of any other treatment. Furthermore, it was also found that the confusing picture resulting from the case descriptions led RCC's to follow the judgments of the physician performing the EAS who typically had no longstanding relationship with the patient. Overall it seems fair to say that foregoing the voluntary aspect of EAS may not seem the biggest threat to Dutch practices, but whether this is actually true depends on how the actual process of assessing patient request in these cases is evaluated. From what both studies report there appears to be much room for doubt.

Conclusion

Describing the various practices of terminating human lives affected by IDD in terms of the 'new eugenics' is not an innocent move. Given the emotivist force of the term 'eugenics' in view of its past, using that term in the present connection suggests a link with horrible crimes against humanity that have been perpetrated in its name, particularly in The Third Reich. Insofar as the proponents of the practices described above are concerned there is no such link. Their ranks are filled with scientists, doctors, healthcare professionals, lawyers, politicians, ethicists, none of whom has in mind, or is engaged in acts that the Nazis had in mind or were engaged

in. When the latter disqualified the lives of human beings with IDD as *Lebensunwertes Leben*, they in fact adopted a collectivist perspective in which their ideal of a purified German race led them to eradicate human beings whose lives contradicted that ideal. In their effort to build their sanitized collectivist empire disabled human beings were seen as 'useless' and their presence was an insult to the aspirations of the 'master race'. Nothing of what we have described in this paper is in anyway connected with these, nor with comparable ideas, nor with the practices of genocide they produced. On the contrary, the underlying concern of people engaged in what has been described above primarily regards the well-being of human beings with IDD and their families. Their perspective on the politics of terminating human life affected by IDD is individualist rather than anything else, and their understanding of well-being is primarily subjectivist. The motivation in support of these practices is driven by 'choice'. The implicitly underlying question is whether persons living with IDD and their families would have chosen their kind of life if they would have had a choice. This is readily seen in the first two practices we have described—neonatology and PNT. The moral imagination leading proponents to support them feeds on the assumption that the answer to that question is negative. The people involved would have rejected living their kind of life. Indirectly, the same underlying assumption is also seen in the third, more complex case. In considering EAS, difficult to read expressions of individual will from persons living with IDD are perceived in light of the wish to die, even though an understanding of the relevant information appears questionable.

Given the need for all these qualifications of our historical equation then, why describe these practices in terms of 'eugenics' anyway? The reason is the similarity that links the kinds of acts that have been reviewed in this paper with the old eugenics. It regards the justification underlying each of them. In one way or another it is based on the preconception of a life lived with IDD as a life of 'poor quality' that is the cause of unbearable suffering for the persons and families involved. In the vast majority of cases however, this preconception is disproven. People with IDD rarely describe their own lives in such negative terms. Even when they do it is often because of the environmental responses of rejection they have to deal with.

If this is empirically true, of which we have no doubt, then the received justification of terminating human lives affected by IDD appears in a different light. Looking at these lives 'we'—as individual people, as professionals, as observers, as 'the public'—are inclined to think that 'they' would rather not exist in the way they do. Seen in this light the medical practices of terminating human lives affected by IDD are driven by a collective preconception that will be contradicted by the persons involved or their advocates when their voice is heard.

Perhaps this is the most pertinent question coming out of our review of developing practices in medicine and the scientific research behind it. If in matters of life and death our society trumps individual views and concerns of the people involved above anything else, why is this not the case here? Is it because given the nature and effect of cognitive impairments one cannot really take seriously what people living with IDD tell us either in words or gestures? In other words, is it because the presumed inability of giving a truthful account of oneself is for many of 'us' precisely

what is means to be affected by IDD? These are by no means rhetorical questions. They indicate the sense in which a conception of what our culture takes to be a human life properly so called appears to be at odds with the conditions of IDD. To the extent that, empirically, this happens to be the case, there is truly a link with what supporters of the 'old eugenics' believed. The link regards the justification of terminating human lives affected by IDD in current medical practices as described in this paper. Ultimately, they appear to be driven by the view that these are lives of poor quality that are in defiance of what a human life properly so called is like.

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Future Directions and Challenges

V. P. Prasher, P. W. Davidson, and F. H. Santos

Individuals with intellectual disabilities (IDD) represent a significant proportion of adults with lifelong disabilities in the developed world. They are an under-represented population that has been the focus of significant legislative activities and mandated services. They constitute a disadvantaged group of individuals with a broad range of co-morbid physical and mental health needs. The aim of this book was to continue to highlight and strengthen good clinical practice and to promote better understanding and management of their mental health.

The information provided in the varied chapters consistently point to the need for more clinical and research data: research to improve our ability to characterise, recognise and treat mental and behavioural difficulties, facts to ascertain best practices for community services and greater training to prepare a competent workforce to manage and also prevent behavioural and psychiatric morbidity.

In recent decades governmental agencies have begun to recognise the need for more research in individuals with IDD and particularly in five key areas in the ageing IDD population, including demography and epidemiology, adaptive functioning, social interactions and family support, intervention strategies, and service and care models. In recent years, the urgency for more reliable information about older persons with IDD has intensified, and developments in drug treatments for dementia, consent and self-advocacy, prevention of health morbidity, individualized care

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are emerging. Since the publication of the 1st edition of this book in 2003, a more contemporary look at mental health and the ageing process in individuals with IDD is in order. The need for accurate information about the epidemiology of mental and behavioural disorders occurring in older persons with IDD remains one of the more fundamental areas of care for adults with IDD.

The International Association for the Scientific Study of Intellectual and Developmental Disabilities (IASSIDD) (https://www.iassidd.org/) has over the last 50 years sponsored many international meetings dealing with health or mental health issues in individuals with IDD. More recently, and particularly since the formation of the Aging and Intellectual Disability Special Interest Research Group, the awareness of emotional and behavioural health in ageing adults has occurred. There remain, however, a number of questions to be fully resolved. For example, what are the real prevalence rates and natural histories of mental disorders in older persons with IDD? There is as yet no definitive international consensus regarding the criteria that should be used to diagnose mental illness among persons with IDD. Additionally, the genetic and biological bases of psychiatric or behavioural disorders in older persons with IDD are not well understood, and we do not have good natural history data on the relative differences between behavioural phenotypes caused by lifelong social and environmental effects.

Once we have settled epidemiological issues, life-span matters must be addressed. For instance, what is the influence of lifelong disability itself on behavioural or mental health? We have almost no data on the causes of changes in mental health status across the age span. What is known about risk factors for morbidity and functional decline on older adults with IDD and when they arise during the lifespan? Most of the research on functional decline has been directed at the association between Down syndrome and Alzheimer disease. But only a fraction of older persons with IDD fall into this category; indeed, functional decline occurs in many, if not all, older individuals with or without IDD who do not have dementia due to Alzheimer's disease. Prevention or slowing of the progressive features of loss of function associated with ageing would go a long way to increasing the likelihood that an older adult can age in community placements more suitable to their particular needs.

There is now more attention to the interplay among the spirit, the mind and the body in individuals with IDD. We have seen more researchers beginning to investigate and understand the mechanisms that lie at the core of the frequently observed relationship between psychological and environmental states and health outcomes. Once thought impossible, there have been numerous demonstrations of the mutual influence of the nervous system (often referred to as the mind or brain) and the immune system, which in turn influences health and disease. Interest in these areas has been intense and has been aided in part by increasingly sophisticated tools for demonstrating changes due to psychosocial factors.

There continues to be paucity of good clinical or research data on the relationship of behaviour and physical health status among ageing persons with IDD. Researchers have shown that there is some interplay between physical diseases and mental or behavioural disorders on persons with IDD and that these

associations may vary with age. Further work is still needed to flesh out the mechanisms that underpin such associations so that interventions and preventative activities may be devised. Clinicians must always be suspect to an underlying physical illness presenting as behavioural or emotional condition.

Stress, and its effect on the immune system and health, has been a major challenge, both to define and to delineate specific causal relationships. Even more so for the IDD population. Stress is mediated by many factors including sex, race, poverty, stressor characteristics (e.g. type, frequency and timing), support systems, availability of coping mechanisms, perceived control and vividness of imagery. In the general population research community, there has been a focus on specific diseases—heart disease, cancer, diabetes, epilepsy, etc.—and the psychological contributions to the development, progression and/or remediation of these diseases. There has been, in recent times, some focus on methods of responding to a variety of problems outside of Western medical tradition, some being distinctly psychological science in origin. Examples of these methods include support groups, biofeedback, hypnosis and self-hypnosis—particularly for pain management- progressive muscle relaxation and imagery—guided or otherwise—cognitive behavioural therapy, Others borrowed from diverse cultures and traditions, such as, meditation, massage therapy, spiritual approaches, traditional Chinese medicine, T'ai Chi, chiropractic, and Ayurvedic medicine. Research evidence for their efficacy, however, is still required.

In the coming decades, one hopes such health practices will also be very much standard in the IDD population as it is striking, how little work has been done in these important biobehavioural health areas with ageing adults with lifelong IDD. Many adults with IDD have risk factors for poor health, such as much higher levels of poverty, lower levels of education, more obesity, less exercise, often low social support, as well as psychological barriers to seeking mental health care on the part of both the individual and service providers, and, for adults with physical disabilities, physical barriers. An illustration of how this may work is as follows. We know that poverty is associated with poor health. A closer examination suggests that low levels of social support, low educational levels and/or depression/anxiety often accompany poverty. Depression in turn has been found in numerous investigations to be associated with a number of health problems, cardiovascular disorders being among the most notable. The physiological mechanisms involved appear to include a higher proportion of sticky platelets, which in turn are associated with clotting and cardiovascular disorders (such as heart disease, stroke). Thus, it might be expected that reducing depression, perhaps by increasing social support or with antidepressants, might also reduce cardiovascular risk. Without specifically designing studies for individuals with IDD, who may well have different reactions to both medications and to psychological interventions, the impact is unknown.

Future challenges in IDD also involve asking ourselves what are the important cultural, ethnic or racial influences on healthy ageing and retention of behavioural or psychiatric health among older persons with IDD? Multi-cultural issues should be addressed by encouraging more cross-cultural research as most research to date has a North American and European bias. A worldwide understanding of ageing in

IDD is not widely acknowledged. While such studies are complex and difficult to finance and conduct, the data from them would go a long way toward identifying the influences of culture on prevalence, treatment and prevention of functional decline and the onset of mental or behavioural disorders. Correspondingly, comparative studies are needed to clarify concordance across cultures in rates of mental and behavioural disorders and treatment efficacy.

Answering these and many other questions through research will require more than the often-momentary enthusiasm of individuals. The role of IASSIDD is essential in defining an international research agenda specific to ageing and mental or behavioural health among individuals with IDD. The agenda should be comprehensive, with a focus on both basic and translational studies. It should be reasonable, identifying a consensus among scientists, practitioners, families and the persons affected by these disabilities. It should clearly state priorities. It should be related to theory and data from mainstream mental health, gerontologic and geriatric research and it should demand inter- disciplinary participation from the ageing and the IDD sectors. And finally, it should be included in the portfolios of agencies and foundations supporting generic ageing and mental health research. Philosophically and practically, our understanding of ageing and mental or behavioural health in persons with IDD is governed by general knowledge about ageing, and about mental health; little will be learned if we isolate our research by adopting only an IDD perspective.

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