A Longitudinal Examination of the Physical Health and Ageing of Individuals with Down syndrome in Ireland

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By

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DECLARATION

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ABSTRACT

Background: Over the past three decades the life expectancy of people with Down syndrome (DS) has dramatically increased with improvements in health service delivery, better knowledge, health care and advocacy. With increased longevity, comes age related change and complex health comorbidities. These health conditions include dementia, epilepsy, thyroid disorder and sensory impairment, some of which are associated with early mortality in this population. With comorbidities comes functional limitations and service providers throughout Irelands need to be informed of the common health conditions and the implications. Staff in services need to be alert for changes and decline that is seen earlier than expected in people with DS compared to people with ID of other aetiologies.

Aim: The aim of this study is to identify and explore the physical health changes that occur among those with DS as they age.

Methods: This study is situated within The Intellectual Disability Supplement to The Irish Longitudinal Study on Ageing (IDS-TILDA) study. Information for this study was extrapolated across the threes Waves of the IDS-TILDA study over a 10-year period. Retention of participants from Wave 1 through to Wave 3 was 86% and this can be attributed to the 'Keeping in Touch' strategy that is employed by the IDS-TILDA Team. Using data from the IDS-TILDA study the key health conditions pertaining to people with DS are examined and discussed. All conditions discussed were stratified by age, gender, living circumstances and level of intellectual disability.

Findings: Life expectancy of those with DS remains considerably lower than people with intellectual disability of other aetiologies. Increasing age has revealed a predisposition for the development of complex health conditions with the prevalence of dementia having more than doubled over the ten years of the data collection. The mean age of onset of dementia was 51.6 years. Other chronic health conditions were identified and they too increased substantially over the 10-years study. The impact of decline in heath has had a considerable impact on functional ability, levels of independence and living circumstances, whereby people have had to move from their place of residence as it had outstripped its capacity to offer safe care with increased level of need. Attendance at healthcare specialists low and many individuals are not being assessed sufficiently. The need to understand ageing and related health conditions for people with DS is crucial. Health care providers need to be alert for health deteriorations earlier than anticipated in the general population, and young adults with DS should be given the appropriate information to better recognize, and recommendations to cope with, changes in their own level of ability or health.

Discussion: Mortality rates were higher in people with DS compared to people with ID of other aetiologies. Preventative health screening needs to be operationalised throughout all services in Ireland and a comprehensive health package should be offered to all people with DS from childhood. The lack of knowledge among service providers and staff around the recognition of the common health conditions in this population is notable by the level of under-recognition of the conditions that were presented. The upskilling of staff is critically important as is the collaboration of the multidisciplinary supports available. While some services in Ireland appear to have excellent multidisciplinary supports for people with DS, with clinics that deal exclusively with people with DS, the findings from this study has noted that most services are poorly prepared for an ageing population.

Conclusion: The under recognition of health comorbidities for people with DS in services remains a critical issue in Ireland today. The key areas that must be developed include health management, quality of life and the promotion of successful ageing.

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ABBREVIATIONS

AAIDD	American Association on Intellectual & Developmental Disabilities
AAMR	American Association on Mental Retardation
AD	Alzheimer's Disease
APP	Amyloid Precursor Protein
CAPI	Computer Assisted Personal Interview
CNSp	Clinical Nurse Specialist
DOH	Department of Health
DS	Down syndrome
GDPR	General Data Protection Regulations
GP	General Practitioner
HRB	Health Research Board
HSE	Health Service Executive
IASSID	International Association for the Scientific Study of Intellectual Disabilities
ICD	International Classification for Disease
ID	Intellectual Disability
IDS-TILDA	Intellectual Disability Supplement to the Irish Longitudinal Study on Ageing
NHS	National Health Service (UK)
NIDD	National Intellectual Disability Database
NIHR	National Institute for Health Research
PET	Positron Emission Tomography
PIQ	Pre Interview Questionnaire
RNID	Registered Nurse Intellectual Disabilities
SPSS	Statistical Package for the Social Sciences
TSI	Test for Severe Impairment
WHO	World Health Organisation

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Chapter 1

Introduction

1.0 Introduction

With advances in health and social care, there has been a steady increase in the numbers of adults with Down syndrome (DS) living into their 60's, 70's and beyond, whilst this is to be celebrated it also presents a challenge for services supporting people with Intellectual Disability (ID) and DS as they age. This trend is set to continue in the coming decades, however understanding the impact of ageing in older adults with DS has had little attention. This chapter begins the investigation of the impact of ageing on those with DS firstly by contextualising the study, providing justification and background to the study. The author then explores the key terms used within the study in relation to adults with DS. This study explores if people with DS experience ageing from a different perspective to the general population, and the possible risk for developing complex health conditions that impact on their wellbeing and overall quality of life. The chapter concludes with the introduction of the aims and objectives of the study.

1.1 Contextualising the study

Increased longevity represents one of the great social achievements of the 21st century. The World Health Organisation (WHO) has predicted the general older population, aged sixty and older, is set to rise by 60% by 2050 (WHO 2007). People with ID are experiencing a comparable increase in life expectancy to those in the general population (Hourigan et al, 2017). Considering those with DS, their lifespan too has increased markedly over the past six decades (Zigman *et al*. 2008; McCarron *et al.* 2010; Strydom *et al*, 2010; Strydom *et al*. 2018; Hithersay *et al*. 2018). People with DS were not expected to live past childhood in the early 1940's with Penrose and colleagues reporting a life expectancy of 12 years in 1949 (Penrose 1949) to reports now of people with DS living well into their 60th year and beyond (Carr & Smith 2018; McCarron *et al.* 2013; Bittles & Glasson 2004).

Down syndrome is the most common of all the described intellectual disabilities and as life expectancy increases, this population are the most commonly represented among older people with an ID (Head *et al*. 2012; Asim *et al.* 2015). Increased longevity overall in people with ID is attributed to improved health and well-being, control of infectious

diseases, improved nutrition and improvements in the quality of health care services and standards for care (Dolan et al 2019, Coppus, 2013). Understanding the variances and parallels between increasing age and complex health morbidities has become an important care consideration in the long term care of adults with DS (Haveman *et al.* 2010, McCarron *et al*. 2011). In Ireland, the majority of people with ID are registered with the National Intellectual Disability Database (NIDD). This database is used to predict service need and inform future policy and planning. While the NIDD does not collect syndrome specific data i.e. data on people with DS, they are included in the overall population figures (Hourigan et al 2018).

Inevitably with longevity there is an increased risk in the occurrence of older aged conditions and health related illnesses. Overall, people with ID and particularly those with DS have a higher prevalence of multimorbidity and health problems than that of the general population (Bayen *et al*. 2018; McCarron *et al.* 2013; Van-Schrojenstein-Lantman-De-Valk and Walsh 2008). Particularly those with DS are considered to have an accelerated ageing physiology with earlier onset of older age conditions and multi-complex needs (Covelli et al 2016). International studies suggest that the health status of people with ID as they age is complex as they are 2.5 times more likely to report health problems than that of the general population who are of the same gender and age (Walker & Ward, 2013; Lin *et al.* 2011; Haveman *et al.* 2010). This is particularly evident for people with DS as research would indicate that increased life expectancy will inevitably result in premature age related comorbidities (Head *et al.* 2010; Esbensen 2010; McCarron *et al.* 2013; Strydom *et al.* 2017; Bayen *et al.* 2018). Responding effectively to the age related conditions presenting in people with DS, particularly dementia, epilepsy, hypothyroidism and sensory impairment is a challenge for the person themselves as well as for family care givers and services.

In Ireland evidence of addressing these complex needs is very much unchartered territory (McCarron 2015). Evidence from European studies suggests that people with ID and DS experience healthcare inequalities such as limited access to services (Englund *et al.* 2013; Cooper *et al.* 2004,). Many have literacy and communication challenges (Morgan *et al.* 2013) which contribute to difficulties in accessing healthcare and healthcare services. Many find it difficult to make their needs, especially their health

needs, known with many finding it difficult to speak with medical staff (McCarron *et al.* 2011). These disparities contribute to a poorer health status and poor lifestyle outcomes for this vulnerable population. It is therefore imperative that adults with an ID and DS should maintain the same approach of preventive health services as that of the general population (van den Driessen Mareeuw *et al.* 2020; Carmeli and Imam 2014). However people with ID and DS often present with significant communication challenges , and service providers must ensure that the voices of people with ID are "heard, wishes respected, and opportunities for choice, engagement and inclusion are maximised" (Bigby 2004; p18). Increased life expectancy for people with ID and DS has highlighted both opportunities and challenges not only for the individuals themselves, but also for family members and for the various services that endeavour to support them as they age. While general policies such as *Valuing People* (Department of Health and Social Care , UK) (2001) and, *Health and Positive Ageing for All* (Health Service Executive [HSE] 2017) can be applied to an ID and DS ageing population, it remains very unclear as to where the responsibility lies.

1.2 Background of the study

Over the past 50-years, the ageing demographic in Ireland has dramatically changed. The population over the age of 65-years is growing by over 20,000 every year, and is expected to reach 111,200 people by the year 2022 (Healthy & Positive Ageing 2015). While these figures are not comparable to people with DS, it is well documented that people with DS too are experiencing increased longevity (Bayen *et al*. 2018; Glasson *et al.* 2014; McCarron *et al*. 2013; Esbensen 2010; Coppus *et al*. 2006; Bittles & Glasson. 2004). This increase in life expectancy can be attributed to advances in medical care, improved understanding of public health issues, education and technology, and a shift in public attitude (WHO, 2000). This changing age profile is highlighted by the NIDD (Hourigan *et al*. 2018) and concern has been expressed that the changing ageing profile of this population will have significant implications for services and care givers alike over the coming years (Haveman *et al*. 2010; Head *et al*. 2012). Similarly, Bittles and Glasson (2004) note that with increased longevity, comes longer time spent in care which is also associated with a longer period of more specific and specialised needs. Greater survival

rates into later life have been reported among people with DS with recent studies reporting averages of 55 to 66 years (Torr & Davis 2007; McCarron *et al*. 2015; Krinsky-McHale & Silverman 2013; Hartley et al. 2015; Glasson *et al* 2016; Carr & Collins 2018). While these figures are to be celebrated, increased longevity in this population has brought increased morbidity in terms of physical health, mental health and neurocognitive challenges (McCarron et al. 2017). There is increasing evidence that multimorbidity and mortality rates are higher in people with ID and even more marked in people with DS (Strydom *et al* 2019; McCarron *et al* 2013; Head *et al* 2012; McCarron *et al* 2005; Moss & Patell 1997).

1.3 Definition of terminology

The purpose of defining terminology is to explain the term and use of the word. Defining the terms used in this study will bring clarity and comprehension of the terminology for the purpose of this research. The following terms will be elucidated; intellectual disability, ageing and, Down syndrome.

1.3.1 Intellectual Disability

In different continents/countries throughout the world various terms are used to describe ID such as, learning disability, mental retardation or intellectual and developmental disability. Historically terms such as idiots, imbeciles and retard were used even within scientific literature and are now thankfully considered unacceptable (Atherton and Crickmore 2011). With the progress of time the term *intellectual disability* was deemed to be a more acceptable and a less derogatory description (Paramenter, 2011). Intellectual disability is a neurodevelopmental disorder characterized by significant limitations both in intellectual functioning (reasoning, learning, problem solving) and in adaptive day-to-day functioning, which covers a range of everyday social and practical skills. This disability originates before the age of 18 (AAIDD, 2017). The individual will have an intelligent quotient (IQ) of 70 or under, with deficits in at least 2 areas of adaptive behaviour (communication; self-care; home living; social skills; self-direction; leisure and work; learning). ID can be categorised into four subgroups; mild, moderate severe and profound ID. Again categorised using the IQ however it must be

remembered that these categories are for descriptive purposes only and not meant to imply an individual's impairment or ability. The categories are described as; Mild ID (IQ 50–69), more often not obvious and not usually identified until children commence school. People with moderate ID (IQ 35–49) are easier to recognise and are usually identified within the first years of life. Those with Severe (IQ 20–34) or Profound ID (IQ 19 or below) require high levels of support and supervision their entire lives. They are unable to live independently or care for themselves and need significant levels of assistance from cradle to grave. The use of the IQ as an approach to establishing the individual's ability has become outdated and is inconsistent with the notion that individuals should not be defined by their individual impairment (Schalock et al 2007). For that reason and for the purposes of this study the World Health Organisation (WHO) (2007) definition of ID will be used and defined as;

> '*a significantly reduced ability to understand new or complex information and to learn and apply new skills (impaired intelligence). This results in a reduced ability to cope independently (impaired social functioning), and begins before adulthood, with a lasting effect on development'* (WHO, 2007)

1.3.2 Down syndrome

Down syndrome (DS) is one of the most common chromosomal abnormalities in humans, and is the most common cause of intellectual disability (Coppus *et al*. 2006). The incidence of DS is influenced by maternal age (Asim *et al*. 2018) and occurs in about one per 319 to 1000 babies born each year, depending on the population (Malt *et al.* 2013; O'Nuallain *et al.* 2009; Canfield *et al.* 2006; Carothers *et al*. 1999;). Since the introduction of antenatal screening in the 1990's, statistics from the United Kingdom, have reported a decrease in the number of babies born with DS (Ballard *et al.* 2016), while the increased number of babies with DS born in the United States of America has been attributed to increased maternal age (Zigman, 2013). In 2013, DS presented in 8.5 million individuals worldwide, and resulted in 36,000 deaths which was a reduction from 43,000 deaths in 1990 (Global Burden of Disease, 2013). DS was first described by John Langdon Down, the British doctor who fully defined the syndrome in 1866 (Hickey

et al. 2012). In his study '*Observations on an Ethnic Classification of Idiots'* (1866) he recorded the pronounced features of people with DS and their associated cognitive impairment (Dekker *et al*. 2015). Trisomy 21 was identified and described by Lejeune later in 1959 (Head *et al.* 2012; Hickey *et al.* 2012). The most commonly identified genetic cause of DS, is triplication of chromosome 21, which was first identified by French researchers in 1959. Trisomy 21 occurs when chromosome 21 fails to separate during fertilisation (Asim *et al*. 2018). Increasing maternal age very much influences the incidence of trisomy (Murthy *et al,* 2007) and a foetus with typical trisomy features is at increased risk of miscarriage (Morris *et al*. 1999). While Trisomy 21 is the most common cause of DS, others causes include Robertsonian translocation and Isochromosomal chromosome, whereby between 2-4% of people will be affected (Asim *et al*. 2018), and here the long arm of chromosome 21 becomes attached to another chromosome causing DS.

1.3.3 Ageing

Ageing is a progressive, predictable phenomenon that involves the maturation of living organisms (Williams, 1995), and characterises the accretion of changes in the human body over time, including physical and psychological health. Environmental dynamics and social issues also influence the length of the lifespan and the time of death (Matteson *et al* 1997). Where exactly a person is born, the family into which they were born, their ethnicity/culture, their level of education and their financial wellbeing will greatly influence the ageing process (Vina *et al*. 2007). Ageing is a multifaceted phenomenon and people age at different rates throughout the world. The processes of ageing are very diverse for different cultures and are genetically determined (Janicki & Dalton 1998). Chronological ageing can be distinguished from biological ageing, chronological ageing representing the amount of time that has passed since you were born by comparison biological ageing is the accumulation of damage to cells and tissue in the body (Philips *et al.* 2010). In summary when we think of ageing as a concept it can be described simply as a process and not an event (Bigby, 2004), however, it is well recognised in the literature that the theories of ageing and the ageing process is not yet understood (Vina *et al.* 2007). This process is summed up by Hagestad and Dannefer

(2001, p7) who suggest that "old age is a part of a life long journey, of individual lives embedded in changing social contexts hence a complex interplay between biographic time and historic time".

There is no such thing as the typical older person. While some 80-year olds enjoy good overall physical health and show no evidence of cognitive impairment, others of a similar age will require high levels of support and care (Adams and White, 2004). Life expectancy has increased considerably in the last century however the incidence of premature death, due to chronic disease accounts for up to 80 percent of age related deaths and disability (Matteson *et al* 1997). Ageing is the biggest risk factor for the prevalence of disease (Belikov 2019) and research continues to search for the answers to the biological basis of ageing. Healthy ageing is defined by the WHO "*as the process of developing and maintaining the functional ability that enables wellbeing in older age*". (WHO 2019 p.1-2)

The Key terms for Health Ageing described by the WHO (2019) are described in Table 1.1

Table 0.1.1 Healthy Ageing (WHO 2019)

Table 1.1 *Healthy Ageing (WHO 2019)*

Functional ability is about having the capabilities that enable people to be and do what they have reason to value. There are five key domains of functional ability, each of which can be enhanced (or constrained) by environmental factors. These are the abilities to: meet basic needs; learn, grow and make decisions; be mobile; build and maintain relationships; and contribute to society. Being able to live in environments that support and maintain your intrinsic capacity and functional ability is key to Healthy Ageing. Functional ability is made up of the intrinsic capacity of the individual, relevant environmental characteristics and the interaction between them.

Intrinsic capacity comprises all the mental and physical capacities that a person can draw on and includes their ability to walk, think, see, hear and remember. The level of intrinsic capacity is influenced by several factors such as the presence of diseases, injuries and age-related changes.

Environments include the home, community and broader society, and all the factors within them such as the built environment, people and their relationships, attitudes and values, health and social policies, the systems that support them and the services that they implement.

Maintaining functional ability enables individuals to maintain a level of independence whereby their quality of life is acceptable.

The notion of successful ageing can be traced as far back as the 1950's and the basis to successful ageing has been described as being the absence of physical health frailties and having good mental/cognitive health (Baltes & Baltes 1990). Various definitions of successful ageing exist including good physical, mental, and psychological health; opportunities to learn and achieve; having a good sense of purpose in life and life accomplishment; having financial security; a sense of humour and some would advocate that physical appearance can influence successful ageing (Bowling & Dieppe, 2005). Rowe and Kahn (1987) proposed that successful ageing involves three critical components:

- Freedom from disease and disability
- High cognitive and physical functioning
- Good overall social engagement and active engagement in life.

These components of successful ageing are closely related. Having opportunities to participate in various social activities and relationships and being satisfied with life is closely associated with good health, longevity and survival into older age (Sarkisian *et al*. 2002). Successful ageing is inextricably linked with health policies and indeed economic policies around the world (Bloom *et al.* 2015; Katz *et al.* 2015; Commission of

the European Communities, 2009). This would include the National Positive Ageing Strategy (DOH 2013), which has highlighted the strategies needed to promote the health and wellbeing of the ageing population in Ireland and improving overall quality of life. Successful ageing it could be said reflects the health and prosperity of a country. However, understanding the concept of ageing is critically important to recognizing the challenges, guiding potential interventions, and perceiving improvement toward the accomplishment of successful ageing (Whitley *et al.* 2016).

In summary ageing is both chronologically defined and biologically defined. Ageing is impacted by a number of components and ageing successfully is seen as a combination of these components. However, for people with ID and especially those with DS successful ageing may possibly be out of their reach. As the years progress those with DS present with ever increasing health challenges and this is specifically associated with ageing chronologically. Therefore, for the purposes of this study a chronological definition has been chosen. In this study ageing refers to those aged 40 years and over with no upper limit. This point has been chosen specifically as it is well documented that people with DS present with age related changes earlier than the general population.

1.4 Philosophy of Ageing

Ageing can be defined as the process of growing older and occurs throughout the life course (HSE 2015). For centuries, the human race has been captivated with the notion of ageing, not only from the organic and emotional perspective, but also from the philosophical perspective of ageing and death (Matteson, 1997). Philosophers have attempted to define ageing, to hypothesize the causes of ageing and to develop measures and cures to postpone the ageing process. To date, attempts to unlock the secrets of the causes of ageing have been futile, and the results of investigative efforts have been speculative (American Federation for Ageing [AFAR] 2011). Ageing is a continuous process which begins from the moment we are born (Heath and Scofield, 1999), and older age is often viewed as a particular stage in life.

Flood (2006) proposed that the concept of ageing well was very much dependent on how people adapted to the functional changes they experienced with increasing age,

while McLaughlin *et al* (2012) proposed that many older people who consider themselves to be ageing well do not meet the defined Successful Ageing criteria This theory maybe particularly relevant for people with DS and ID. Jenkins (2010) in his study proposed that ageing is viewed in rather negative terms and as a significant life transition by people with intellectual disability. However, he also notes that for some older people with ID, ageing and the physical changes that accompany the ageing process often go unnoticed because of their past negative life experiences and lack of opportunities. For people with ID their perception of ageing is similar to the general population. In a study by Burke et al 2014 they identified that people with ID and DS had similar views on ageing, considered ageing to be both demanding and an opportunity, and while people with ID and DS have aspirations of ageing successfully their circumstances impact greatly on this aspiration (Burke et al 2014). It must be remembered that our perceptions of how we age impacts greatly on how we age therefore active ageing is the way to go (Buys *et al*. 2012).

1.5 Ageing in Contemporary Society

Successful ageing is multidimensional and aims to promote positive health behaviours to optimise health across the lifespan (La Plante 2014). The Healthy and Positive Ageing for all strategy Health Services Executive (HSE 2015) aims 'to enhance population health and wellbeing over the trajectory of ageing by the creation and use of knowledge to ensure Ireland is a good place to grow old' (pp 5-6). The IDS-TILDA study measured the perceptions of ageing of the participants with ID and DS to capture their insights and observations as to what exactly healthy ageing meant to them (Burke *et al*. 2014). Key areas recognise that identified with positive ageing were independence, wisdom, life experience and fulfilment, while concerns regarding ageing and getting old were identified as decline in physical health, illness, decline in independence, and death (Burke *et al*. 2014).

Rowe & Kahn (1997) identified successful ageing for the general population in three parts, high physical and cognitive functional capacity, low risk of ill health, and active engagement in life, and note that despite increasing age people can continue to engage in daily life as they grow older. People with ID and DS are living longer, it is important to

understand how they feel about ageing. The Positive Ageing Indicators for People with an Intellectual Disability (McGlinchey *et al*. 2018) explores the vision for ageing for people with ID living in Ireland. This national strategy endeavours to look at ageing exploring healthy aging, housing, social participation, technology and attitudes towards ageing, and in doing this, the goals of the policy is to:

- Provide opportunities to all people with ID to participate, integrate and be involved in all aspects of life in their local communities no matter what their age.
- To encourage and support People with ID to manage and maintain good physical and mental health
- To help people with ID age with dignity and confidence

(McGlinchey *et al*. 2018)

It is envisaged that the Positive Ageing Strategy will help to change the perceptions of ageing with an ID, looking at the positive contribution that people with ID can make to a community and indeed to society.

1.5.1 Ageing from the perspective of people ageing with ID

Understanding the process of ageing from the perspective of people with ID should assist in understanding the needs of people with DS as they grow older. People with ID themselves have described the process of ageing as a time of changes in health, participation and self-identity, associated with decline in physical health, and an opportunity to develop knowledge and wisdom (Kåhlin *et al.* 2015; Burke *et al*. 2014; Kahlin *et al.* 2013). Maintaining health, relationships, having a home, having opportunities to engage in meaningful activities and having choice and control were all associated with a positive ageing experience by Wark *et al.* (2015). Likewise, Haigh *et al.* (2013), reported similar findings and identified that supportive staff and family were key in supporting well-being as people with ID grow older. Staff supporting people with ID who are ageing have described the ageing process in mainly negative terms, relating to physical health decline and need for increased medical care (Kahlin *et al.* 2016). Staff's impact and opinion can have a related influence on how those in their care age. These opinions can be viewed as barriers to ageing well (Lifshitz, 2002). These barriers, along with other aspects such as limited service options, lack of meaningful choice based on

geographical location, lack of options for living situation or lack of transport or finance impact substantially on the positive ageing experiences of individuals with ID. Gilson & Netting (1997) identified that people who enter the ageing process with ID may have their primary identification as disabled person already firmly established and such barriers emphasis the individuals disability rather than focusing on the person themselves (Haigh *et al*.2012, Wark *et al.* 2015).

Ageing with a lifelong disability is a relatively new phenomenon which has come about with improved health care and timely medical interventions being carried out at a younger age (Bigby, 2004). Ageing for this population throws up two diverse issues which are seen and encountered in ID services throughout the world, (Watchman 2014; Udell 2014; Towers & Wilkinson 2014; Head *et al*. 2012; Farriols Danes 2011; Matta *et al.* 2011; Udell 1998). Firstly, as people with ID living at home age the question of who will look after them when their parents become infirm or too old to manage the care is a primary concern for all families. And how to prepare the person with ID to transition from the family home setting to a 'formal' care facility. Secondly, addressing the needs of people with ID as they age and how to age positively. Considering the socioeconomic and service access barriers providing the right supports in an inclusive service that listens to and acknowledges their choices is essential (O'Donovan *et al* 2018; Wark *et al* 2015; Bigby, 2004).

With increasing age comes increased prevalence of functional impairment, morbidity often with early age onset of many common health conditions (Kinnear *et al*. 2018; Evenhuis *et al*. 2001). Similarly it is widely recognised that people with DS present with a higher rate of complex health conditions, compared to the general population (Hithersay *et al*. 2018; Kinnear *et al*, 2017; Strydom *et al*. 2016; McCarron et al. 2014; Head *et al*. 2012; Farriols Danes 2011; McCarron *et al*. 2005; Jansen *et al*. 2004). All the more reason to emphasise how this population require the same health care needs as the general population, including health screening and prevention (Maatta *et al*. 2011; Haveman *et al*. 2010; Smith 2001, Evenhuis *et al*. 2001).

1.5.2 Ageing and Down syndrome

The phenomenon of old age in people with DS is relatively recent and necessitates detailed exploration into the natural progression and longitudinal patterns of comorbidities. In the past, individuals with DS had a low life expectancy, with the median age of death only reaching 25 years in 1983 (Weijerman & de Winter, 2010). Fortunately, due to medical and societal advancements, people with DS are now living well into their 50s, 60s, and even 70s (Moran *et al.* 2013; Zigman & Lott, 2007). However, accelerated ageing and a longer lifespan places adults with DS at an increased risk of developing age-related health conditions (Moran *et al.* 2013; Zigman & Lott, 2007). Despite improved survival rates, people with DS continue to experience atypical lifespan development (Zigman *et al.* 2008). DS is still associated with increased mortality rates, and age specific mortality risk remains higher in adults with DS (Minino *et al.* 2006). A great deal of research has been carried out in an attempt to understand the underlying mechanisms of this significantly accelerated ageing. A common explanation involves the triplication of the approximately 300 genes located on chromosome 21 (Ruparelia *et al*. 2013, Zigman, 2013). This triplication causes a gene-dosage imbalance, which further results in the dysfunction of vital molecular and genetic systems (Ruparelia *et al.* 2013, Zigman 2013). Deficits in the immune system and the underpinning genetic makeup predisposes people with DS to increased risk for poorer health outcomes. As well as these factors up to the age of 35-years, mortality rates for individuals with DS are comparable to people with ID of other aetiologies (Head *et al* 2012; McCarron *et al* 2012; Roizen & Patterson, 2003). However from this general age onwards people with DS appear to present with ever increasing health issues. Strauss & Eyman (1996) found that after the age of 35-years mortality rates double every 6.4 years in people with DS compared to 9.6 years for people with ID of other aetiologies. Therefore, despite improved survival rates, people with DS continue to experience atypical lifespan development (Zigman *et al* 2008). Accelerated ageing and a longer lifespan places adult with DS at an increased risk of developing age-related health conditions (Moran *et al.* 2013; McCarron *et al*. 2012; Esbensen, 2010; Zigman & Lott, 2007; Strauss & Eyman, 1996).

The understanding of the association between chronic disease and increased age has become an important issue in the care of adults with DS and other ID (Haveman *et al*. 2010). Haveman and colleagues focused on examining the physical health of this population and explored the impact of ageing on their overall physical health over a 10 year period. The phenomenon of old age in people with DS is relatively recent and necessitates detailed exploration into the natural progression and longitudinal patterns of comorbidities. Exploration of ageing longitudinally is particularly beneficial when examining lifespan issues and can provide a picture of changes over time. McCarron *et al* (2014 and 2017) and colleagues explored how the relationship between dementia in people with DS was closely associated with other co-morbid health conditions. Both these longitudinal studies examined the health characteristics of people with DS and dementia over 14-years and 20-years respectively and found that the presence of dementia was closely associated with other co-morbid health conditions that greatly impacted on overall quality of life. Ultimately gaining an understanding of chronic disease and its association with ageing in this cohort is an important contribution to developing services and future planning for successful ageing for these individuals.

1.6 Justification for the study

People with DS are often disadvantaged in that they approach the ageing process from a vulnerable standpoint, and that health care professionals often fail to recognise the special health difficulties experienced by this population as they age (WHO, 2000; Haveman *et al*. 2010). It is well documented that people with DS experience the same health care challenges as the general population (McCarron *et al*, 2005), but serious inequalities continue to exist (Haveman *et al*. 2010; Heller *et al*. 2004; Kerr *et al*. 2003; Lewis *et al*. 2002). People with DS, throughout their life, present with a higher risk for complex health conditions (Startin *et al*. 2020; Hithersay *et al.* 2018; Kinnear *et a.l* 2018; McCarron *et al*. 2017). These studies demonstrate how ageing impacts on people with DS. While ageing is inevitable for everyone, highlighting and addressing the unique needs of people with DS as they age is essential to promote the maintenance of

functional ability, to extend competence in later life, to promote successful and productive ageing and more importantly to enhance overall quality of life.

1.7 Outline of the thesis

In the next chapter, the dissertation will detail a literature review synthesising the current research on the common health comorbidities identified in the literature among older adults with ID including those with DS nationally and internationally, which suggests that this population are at increased risk of complex health difficulties than the general population (Van Schrojenstein *et al*, 2000) and in particular the study will highlight the common health conditions pertaining to adults with DS (Kinnear *et al.* 2017). Chapter 3 provides a detailed description of the methods used including sampling, data collection, data protection, the ethical considerations applicable to the study and the analysis undertaken to answer the research question. In chapter 4 the findings are presented including the demographics, an examination of the health conditions, the impact of these health condition with regards the individuals living circumstance and activities of daily living. In conclusion this chapter examines the survival of those with DS over a 10-year period. Chapter 5 discusses these findings in light of the literature and the thesis concludes with the identification of relevance implications to practice, policy education and research.

1.8 Conclusion

With increasing longevity, people with DS are living longer than ever before, living into their 50's, 60's and beyond. This is a true reflection of advances in medical care and successful ageing for this population. Despite this, people with DS die at an earlier age and present with more complex health comorbidities than people with ID of other aetiologies. There is a need for better health surveillance and health care screening in this population. Health care practitioners often fail to recognise the lifelong conditions inherent in people with DS and the conditions particularly associated with ageing are sometimes not diagnosed until they are well established. The next chapter will outline the medical conditions described in the study and lay the foundation for the findings of the study.

Chapter 2

Literature Review

2.0 Introduction

A literature review provides justification for a study and serves to inform the reader of the overall recurring themes that emerge on the topic (Cronin *et al*. 2008). Following a scoping review of the literature the predominant issues arising that impact on the ageing process are very much focused on the complexity of health experienced as people with Down syndrome (DS) age. As DS is the most common cause of Intellectual Disability (ID) and considering that health care professionals often fail to recognise the particular health difficulties experienced by this population it is critical that services understand these complex health issues.

This chapter aims to provide a summation of the current literature surrounding the impact of ageing, specifically health changes, among people with DS. The aims of the study are presented, an overview of the literature strategy is outlined, the inclusion and exclusion criteria are presented along with the screening and eligibility of the chosen literature, the quality assessment applied to the chosen studies is elucidated, and the emerging themes explored.

2.1 Aim of the literature review

The aim of this literature review is to identify and explore the physical health changes that occur among those with DS as they age.

2.2 Developing the question

A well-defined and formulated question ensures the review is focused and clear. For the purposes of this literature review the PICo framework was used (see table 2.2). This assists in identifying the specific concepts that are required in the question so that relevant studies are not overlooked. It facilitates the literature search and avoids collecting a potentially biased result set (Schardt *et al.* 2007).

Table 2.2 Establishing the literature review question

P	I	Co
Population or Problem	**Interest**	**Context**
What are the characteristics of the Population or the patient? • Down syndrome • Intellectual disability • Adults • At least 30 years	**I**nterest relates to a defined event, activity, experience or process • Physical health • Chronic conditions • Comorbidities • Ageing	**Co**ntext is the setting or distinct characteristics • Impact

2.3 Research Question

What are the physical health conditions that impact on the ageing of people with Down syndrome?

2.4 Search Strategy and Parameters

An extensive review of the literature was carried out using 4 electronic databases that included, CINAHL, PubMED, PsychINFO, Academic Search Complete to conduct the search. No time limiters were applied to the study to ensure seminal work was not excluded. Using the PICo frame the following concepts were identified;

1. Intellectual disability (ID) and Down syndrome (DS)
2. Older adults
3. Physical health conditions/comorbidities
4. Impact

The synonyms for each concept are described in figure 2.1. Key search terms used were 'intellectual disability'; 'learning disability', 'cognitive impairment' and 'Down syndrome'. Older terms such as 'mental handicap' and terms familiar to the USA such as 'developmental disabilities' and 'mental retardation' were also utilised. Considering physical health terms included comorbidities, physical health or chronic health conditions. Each concept and the Boolean operators were then applied to each search

within each database. The Boolean operator OR broaden the search and the operator AND combined the concepts and narrowed the search. The reference list of the selected literature retrieved were scrutinised to ensure no seminal work was omitted.

Concept 1: Down syndrome and intellectual disability

Down syndrome OR Downs OR intellectual disability* OR learning disability* OR cognitive impairment OR Mental retardation

AND

Concept 2: Older Adults

Adult OR aged OR old OR older OR elderly OR mature OR senior

AND

Concept 3: Physical health conditions/Comorbidities

Comorbidity* OR morbidity OR multi-morbidity OR chronic health conditions OR physical health OR co-occurring condition* OR illness OR ill health OR sick* OR disorder

AND

Concept 4: Impact

Impact OR concern OR partake OR reach OR effect OR impinge OR impose

Figure 2.1 Concepts and search string applied in the literature review

2.5 Inclusion and exclusion criteria

This review will only consider peer-reviewed studies of full text article that are available in English. No time limiters will be applied. Only studies concerning adults over the age of at least 30 years were included. Studies exclusively on children will be excluded along with studies on individuals with intellectual disability of other aetiology however for comparison purposes these studies will be reviewed and summarised, if appropriate will be included.

2.6 Screening and eligibility

The initial search was conducted for each concept using the Boolean operator OR and the results were combined with the Boolean operator AND to ensure a robust and accurate search. Using the PRIMSA flowchart (see *figure 2.2*) the selected articles retrieved were reviewed against the inclusion and exclusion criteria and included or omitted accordingly. In total N=230 were retrieved from CINAHL; N= 689 were retrieved from PubMed, N=915 were retrieved from PsycINFO, and N=543 were retrieved from Academic Search Complete. These were then combined the duplicates (N=1927) were removed resulting in N=450 going forward for title and abstract scrutiny. In total, N=250 were removed as they did not meet the inclusion criteria. Overall N=65 full articles were retrieved and read thoroughly. Those that did not meet the inclusion criteria (N=50) were at this stage omitted. To support literature decisions and ensure the overall strength of evidence from the literature was appropriate consideration was given to quality assessment and evaluation of the methodological quality of each of the articles chosen. The National Institute of Health guidelines for assessing the quality of studies was used to guide this activity. The researcher then exhausted the reference list of each paper retrieved to identify any seminal literature that may have been excluded within the original search. This included papers that had participants with DS only and participants with ID that included people with DS resulting in 2 further papers begin identified and included. In total N=17 articles were examined for this literature review.

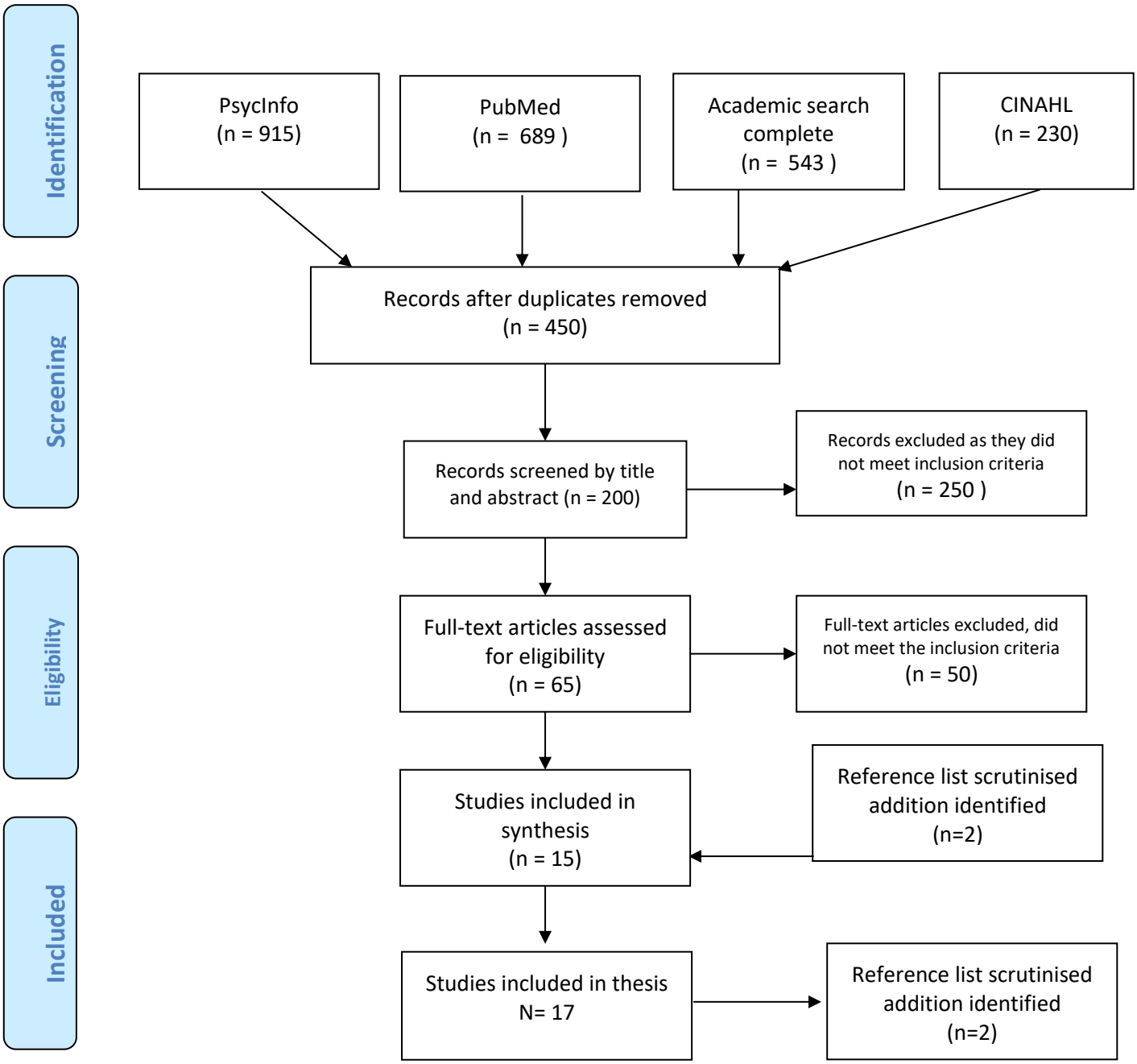


Figure 2.2 PRISMA flow chart for literature search adapted from Moher et al (2009)

2.7 Emerging Health Conditions that Impact on Ageing of those with DS

It was obvious from the literature review that the health of those with DS deteriorates considerably with age. The following recurrent physical health conditions and their subthemes relating to ageing and Down syndrome emerged and will be explored further in the subsequent discussion:

- Chronic health conditions
    - Dementia
    - Epilepsy
    - Thyroid disease
    - Sensory impairment
- Consequences of deteriorating health

The key physical health concerns identified for the purpose of this study are dementia, epilepsy, thyroid disease and sensory impairment. These health conditions have been found to be the most common comorbid conditions in adults with Down syndrome. Other noted conditions prevalent across the life span among those with DS that featured greatly include osteoporosis, diabetes and elevated cholesterol. For the purposes of this review and to answer the overall research question the focus will be on the most prevalent conditions that are emerging from the literature.

2.8 Dementia

The word dementia comes from the Latin words 'De' meaning 'out of' and 'Mens' meaning 'mind' (Livingston, *et al* 2017), and the very nature of this definition can have very negative and stigmatising inferences (Mukadam & Livingston, 2012). Dementia is defined as the development of multiple cognitive deficits, involving memory, aphasia, apraxia, agnosia and disturbance in executive functioning, and "*is the greatest global challenge for health and social care in the 21st century*" (Livingston *et al*. 2017 p.1). It is a neurodegenerative disorder characterised by neuronal death, neuritic plaques and neurofibrillary tangles and is described as clinically progressive with irreversible deterioration in cognition, behaviour and day-to-day functional ability (Jack & Holtzman 2013; McKhann *et al.* 2011). For people with DS the dementia identified as most common is Alzheimer's disease (McCarron *et al* 2015).

Two well described protein combinations are the hall marks of Alzheimer's disease (AD). The first, Amyloid-β deposits (Aβ) are described as neuritic plaques and cerebral amyloid angiopathy (CAA) (Anus *et al.* 2015; Jack & Holtzman,2013). The second protein is tau protein which forms neurofibrillary tangles (NFT). The brain presents with an accretion of pathologies as a consequence of ageing, and with increasing age come the inevitability of complex neurological anomalies. Nelson et al (2011) found most of elderly participants in their study had notable evidence of cerebral changes consistent with the ageing process, and that the plaques and tangles of AD were inevitably associated with increasing age and longevity. In the general population, the earliest clinical presentation of AD is memory loss, this interferes with everyday life functioning, specifically in the ability to retain new information (Alzheimer's Association, 2014, Lopez, 2011). Additional common presentations include problem-solving impairment, decline in ability to complete familiar daily tasks, disorientation of time, language difficulties, social withdrawal, and evident mood disturbances (Alzheimer's Association, 2014). The most common changes in mood and personality are usually apathy, depression, and aggression (Alzheimer's Association, 2014, Lopez, 2011).

Early onset dementia, usually described before the age of 65-years, is uncommon in the general population (Jack & Holzman, 2013) this however is not the case for those with DS where the mean age of onset is 55.39 years (McCarron *et al*. 2014). For the general population a small proportion present with autosomal dominant mutations in one of three genes; the amyloid precursor gene on chromosome 21; the presenilin-1 gene on chromosome 14 and the presenilin-2 gene on chromosome 1 (Goate *et al*., 1991). All autosomal dominant mutations that inevitably result in AD influence the processing of the amyloid precursor protein (APP), and this is true of all people with DS (Holzman *et al.* 2011). An increase in APP will influence the production of Aβ 42 (Annus *et al*. 2015; Holtzman *et al*. 2011) and Aβ appears to drive the development of this progressive disease. Jack & Holtzman (2013) describe the amyloid cascade hypothesis as a disproportionate build-up of amyloid-beta (Aβ) plaques is crucial in the development of Alzheimer's disease. The excessive accumulation of Aβ corresponds with tau dysfunction, whereby the amyloid plaque incites increased tau pathology that spreads through the brain. This accumulation triggers a series of events that results in the

widespread neuronal damage, destruction of brain cells, and eventually Alzheimer's disease. (Jack & Holtzman 2013;Barag & Sonawon 2015), *see figure 2.3* for the amyloid cascade hypothesis.

The overproduction of /accumulation of Aβ42 will result in:

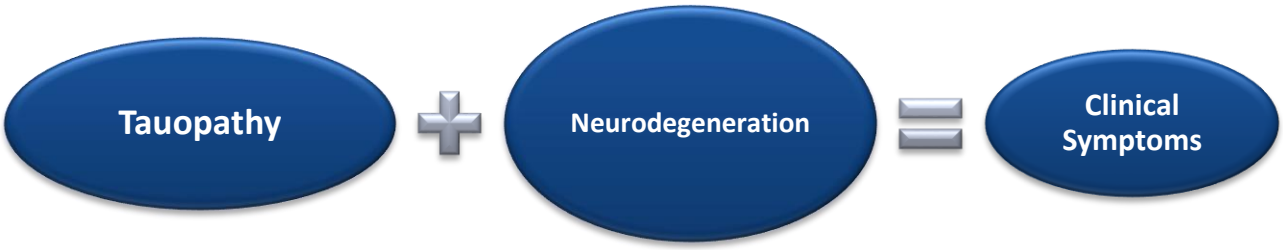


Figure 2.3 The amyloid cascade hypothesis adapted from Jack & Holtzman, 2013

The highest incidence of dementia, particularly early onset Alzheimer's disease is seen in individuals with DS (Hartley et al, 2014), and this population are at high risk for developing early onset dementia (Mann, 1988). The association between DS and Alzheimer's disease has received more attention than any other aspect of ageing among adults with an ID (Zigman *et al.* 2008), and one factor that has been conclusively linked to an increased risk for dementia in DS is advancing age (Schupf, 2002; Bittles & Glasson; 2004; McCarron *et al.* 2014; Ballard *et al.* 2016), with a reported incidence being up to five times greater than the non-ID population (Strydom *et al.* 2013). Various studies have shown that prevalence rates increase from 20% - 50% in the 50-60years age category to 30%-75% in those with DS over the age of 60 years (Schupf & Sergievsky, 2002). Pre senile dementia in adults with DS was first recognised over 135 years ago (Fraser & Mitchell, 1876) and has been described in many studies (McCarron *et al,* 2005, McCarron *et al*, 2017, Strydom *et al* 2018). Down syndrome, which is almost always caused by the presence of three complete copies of chromosome 21 secondary to meiotic disjunction, is the most common chromosomal disorder and is also the most common genetic cause of cognitive impairment (Ness, *et al* 2012, Mann *et al* 1990).

2.8.1 Plaques, Tangles and Down syndrome

A gene known to have a causational association with AD is the amyloid precursor protein (APP) gene and is located on the proximal portion of the long arm chromosome 21 (Zigman 2013; Zigman & Lott, 2007). The APP locus has been described as being within the 'critical region'(Asim *et al,* 2018) of chromosome 21 that causes full DS phenotypic manifestation when triplicated (Zigman, 2013; Zigman & Lott, 2007; Holland, 2000), and it is well established that Alzheimer's disease and DS have a close relationship due to these shared genetic characteristics. It is now well established that those with DS and no diagnosis of dementia have comparable levels of APP, to those without DS and AD (Mann, 1988), with amyloid deposition occurring and developing up to ten years before the clinical manifestations of AD (Sperling *et al*. 2011; Jack *et al*.2013). The result of the triplication of the APP gene is a genetic example of Aβ overproduction (Annus *et al.* 2008). As a result, by the age of 30, people with DS invariably develop amyloid plaques and neurofibrillary tangles and, beginning in their 40's and continuing through their 70's, up to 75% of people with DS will develop dementia (McCarron *et al.* 2017; Zigman *et al.* 1996). Visser *et al.* (1997) described the prevalence of dementia among those aged 40 years and older at 36%, with incidence rates steadily increasing with age. In the general population, it has been established that the delay between amyloid deposition and the presentation of the clinical manifestations of Alzheimer's disease is approximately 20-years. A study by Holland *et al* (1998) would also suggest similar finding for people with DS. Therefore, it can be suggested that a significant number of people with DS, at the age of 50-years, and without a diagnosis of dementia will have the amyloid markers consistent with Alzheimer's disease (Handen *et al*. 2012). Burke *et al* (2014) noted a mean incidence of dementia, in people with Down syndrome of 4.7% per annum, with a significant increased prevalence from 15.8% reported doctors diagnosed in Wave1 to 29.9% in Wave 2 to 35.2% in Wave 3.

Neurofibrillary tangles are another hallmark of Alzheimer's type dementia and are well described in the literature in relation to DS. Tau protein is an intracellular protein that is abundant in the neurons of the central nervous system and works to stabilise the microtubules through isoforms and phosphorylation. Hyperphosphorlation of Tau proteins can cause neurofibrillary tangles, which contributes to the pathology of

Alzheimer's disease. Tau protein has direct effect on the breakdown of a living cell caused by Tangles that form and block nerve synapses. Tangles are clumps of Tau protein that stick together and block essential nutrients that need to be distributed to cells in the brain, causing the cells to die (Alzheimer's Association, 2014). The protein is released on neuronal death, and in conjunction with increased levels of Aβ 40 and Aβ 42 is considered to be neuro-chemically compatible with Alzheimer's disease. The accumulation of neurofibrillary tangles triples, over a 10-year period from the age of 40-years to 50-years in people with DS, echoing the onset of dementia (Margallo-Lana et al, 2007). In 2015, Annus *et al*, reviewed 49 adults with DS between the ages of 25-65 years. Each participant had a positron emission tomography (PET) with Pittsburg compound-B (PIB). This study revealed abnormal PIB bindings in those over the age of 39-years, and this was strongly related dementia and cognitive decline.

2.8.1.1 The end Stages of Dementia

The end stages of AD, brought on by progressive decline of both cognitive and functional abilities, requires complete support for all activities of daily living (ADL) (bathing, eating, mobilising & continence). There is a comprehensive loss of communication abilities, inability to recognize familiar faces, a complete vegetative state, and eventually death (Alzheimer's Association, 2014; Lopez, 2011). A similar picture of end stage dementia in adults with ID has been described by (Startin *et al.* 2020; Strydom *et al.* 2018; Hithersay *et al.* 2018; McCarron *et al*. 2017; McCarron *et al* 2015; Bittles & Glasson 2014; McCarron *et al.* 2010; Esbensen, 2010; Bittles *et al.* 2007; Coppus *et al.* 2006). This stage of dementia requires specialist care and sensitivity. Basic day-to-day activities of living such as eating and drinking are no longer possible unless supported by staff. Weight loss may be apparent, and staff are more often concerned about their abilities and how they can respond to the needs of people at end of life. Mobility is lost and the complications of poor nutrition/hydration coupled with inactivity can increase the risk for infection. Specialist care must be considered to address dehydration, aspirate pneumonia, choking and compromised skin integrity (McCarron *et al*, 2010; Janicki *et al.* 1995).

2.8.2 The Challenge of Diagnosing Dementia

The application of established diagnostic criteria to a population with pre-existing cognitive and functional impairments and additional co-morbidities presents with significant and unique challenges (McCarron *et al* 2013; Krinsky-McHale & Silverman, 2013; Janicki & Dalton, 1998). There is a paucity in information for diagnostic criteria pertinent to people with ID or DS, and the ICD-11 (Salvador-Carulla *et al*, 2011) and the DSM-V (American Psychiatric Association, 2013) only consider the general population. The overall presentation of Alzheimer's disease is complex and more often difficult to diagnose in people with DS not only because of their pre-existing cognitive impairment but also because of poor communication skills (Firth *et al*, 2018; Staunton & Coetzee, 2004). The level of ID of the person with DS as well as pre-existing impairment in memory and executive function increases the complexity of diagnosis (Startin *et al*. 2016), and it is critically important to distinguish the persons established pre-existing impairments from new onset changes and decline (Strydom *et al.* 2018).

The screening and assessment tools used to identify dementia in people with DS and ID are different to the tools used in the general population in that they do not depend of cut-off scores to make a diagnosis. Rather they examine functional and cognitive decline being cognisant of pre-existing cognitive impairment due to ID. Screening provides the opportunity to measure and compare changes to baseline functioning (O'Caoimh *et al.* 2013).

The Test for Severe Impairment (TSI) (Albert & Cohen 1992) was developed for the general population with severe cognitive impairment originally and validated to be successfully used for screening in people with ID (Tyrell *et al.* 2001). The TSI is used in the IDS-TILDA study across the three Waves and was chosen because it is consistent, brief, easy to understand and use, while there is no notable floor or ceiling effects (Tyrell *et al*. 2001; Cosgrave *et al.* 1998). International consensus recommends the all people with DS over the age of 35-years should initially have a baseline cognitive assessment, with which to measure change over time and will improve opportunities to identify changes from previous level of functioning and baseline behaviours (Burt & Alyward, 2000). The importance of baseline information to identify the person's level of

functioning at one point in time cannot be underestimated in identifying change over time (Moran *et al*. 2013), while frequent changes in staff may contribute to cognitive and functional changes going unnoticed (McCarron & Lawlor, 2003; McCarron & McCallion, 2004). Best practice would emphasise baseline screening from 35-years, noting pre-existing impairments with annual neuropsychological assessment thereafter (Moran *et al*. 2013)

Unfortunately, opportunities for people with DS to attend generic memory clinics are uncommon, and the adapted available cognitive tests for people with ID/DS can have a floor effect, whereby people with a severe level of ID would be unable to participate in the neuropsychological test instruments available to them as they would not be able to understand or indeed participate, therefore meeting criteria for dementia despite not having dementia (Strydom *et al,* 2018). Various studies have identified different interpretations of decline in the context of dementia and DS. Some have noted early changes in cognition and decline in memory (Krinsky-McHale *et al*. 2002) while Dekker *et al* (2015) identified personality changes coupled with decline in behaviours and executive functioning. While there is no comparison tool for people with ID to the Mini-Mental- State-Examination (MMSE), the Horizon21 DS consortium is currently carrying out international longitudinal studies examining the early identifiers for AD and DS, and this study is ongoing (Strydom *et al.* 2018).

2.8.3 Diagnostic Overshadowing

Access to an accurate and timely diagnosis is crucial, however people with DS can more often present with communication difficulties, and dementia is more likely to be detected in the mid stages of the disease process. This can on occasion be due to diagnostic overshadowing, when dementia symptoms are attributed to other physical health conditions or the early signs of dementia can go undetected due to the persons underlying ID (Stanton & Coetzee, 2004). Unlike the general population memory loss is not typically seen as the early signs of dementia in people with ID, rather, changes in personality and decline in function are more apparent. Gates and Barr (2009) reported

that diagnostic overshadowing was particularly relevant when new behaviours present or existing behaviours escalate. Diagnostic overshadowing occurs when family members, support staff or medical professionals, ignore the changes in the person with ID or DS, assuming that changes in personality and new behaviours are part of the ID (Javaid, 2019) never considering that other possible factors may be attributed to cognitive decline, physical or organic causes (Javaid, 2019; Emerson & Baines, 2010), leaving the underlying health condition untreated.

Contrastingly, it is often assumed that when a person with DS is in their 50's, and presents with changes in personality and behaviour, that they are experiencing the early manifestations of dementia. Emerson and Baines (2010) assert that the comorbid health conditions common in people with DS, hypothyroidism for example, can be mistaken for dementia. It is vitally important to rule out any reversible causes for dementia, and in particular hypothyroidism (Stanton & Coezee, 2010). It is for this reason that annual baseline screening is invaluable, and informs the medical professionals of previous level of functioning. A thorough physical health work up is vital in order to out rule a differential diagnosis and to ensure an accurate and informed diagnosis is made. Offering the person with DS and their support network a timely diagnosis is vitally important in order to strategically plan ahead for emerging needs, involving multidisciplinary and post diagnostic supports through the continuum of the disease and will ensure better outcomes and enable the person with dementia to live a full life (O'Caoimh *et al.* 2013).

2.8.4 Neuroimaging and Dementia

The presentation of dementia in people with DS is however somewhat different to that of the general population. There is a dearth of information describing the cognitive and personality changes prior to diagnosis of Alzheimer's disease in people with DS and this is more often attributed to poor knowledge base among carers, who are generally unaware of the prevalence of dementia in this population or what they should be vigilant for (Bittles & Glasson 2004). Lautarescu *et al* (2017), in their study of people with DS, describes a preclinical presentation with a prolonged prodromal phase which on examination do not meet ICD-10 criteria for a diagnosis of dementia. In MRI and PET

scanning carried out by Mann *et al* (1990) changes were observed in hippocampal volume and in what is described as amyloid burden in a large range of participants. Similar findings, by Haier *et al* (2008) reported, people with DS (between 34 – 52 years of age), had reduced grey matter identified on MRI and elevated cerebral glucose metabolism on PET, despite never having a diagnosis of dementia. Consequently, the early stages of the disease often go unnoticed or are not diagnosed, and a definite diagnosis is generally established in the early to late 50's. Sabbagh *et al* (2006) highlighted increased amyloid burden and hippocampal atrophy when comparing people with DS with and without a diagnosis of dementia. These particular studies highlight the importance of neuroimaging in identifying cerebral changes, to enable the identification of the early stages of the disease (Haier *et al* 2003). This lack of diagnosis or late identification leads to individuals being well advanced into the disease and so early intervention medication, Donepezil and Memantine for example, are redundant. These medications known as cholinesterase inhibitors were developed to improve cholinergic function and neurotransmission and have shown some degree of success in improving cognition and overall quality of life in the general population (McShane *et al.* 2006). People with DS are rarely included in clinical trials for anti dementia drugs (Eady *et al*. 2018), making it impossible to be decisive regarding their effectiveness in the early stages of the disease process (Livingston *et al*. 2015; Hanney *et al*. 2012). Eady *et al* (2018) in a study of 145 adults wit DS, 47% of the study population, who were taking Memantine had a greater survival time compared to those with DS who were not prescribed the drug.

2.8.5 The Importance of Cognitive Reserve

In the general population it is well reported that despite having the neuropathological manifestations of dementia, many people do not have the disease and are cognitively unimpaired (Cholerton *et al*. 2016), and this has led to the notion of cognitive reserve (Livingston *et al.* 2017). Cognitive reserve enables individuals who have Alzheimer's type neuropathology, to resist the typical presentation of cognitive and functional decline and as a result the development of the disease process is much slower than typically seen (Stern 2012). This theory would therefore indicate that people with poor

cognitive reserve are at greater risk for the development of dementia (Livingston *et al*. 2017), and cognitive resilience with increasing age appears to be closely associated with continually building and enhancing the brains cognitive reserve at an earlier age through ongoing education, be it formal or informal (Borenstein & Mortimer, 2016; Larson, 2010). One has to therefore consider if this is one reason why people with DS develop dementia at an earlier age (Strydom *et al,* 2007).

Various studies have examined how exactly brain reserve can be enhanced and improved through the lifespan, and the general consensus points to three key areas; physical exercise, intellectual motivation, and social and leisure activities, appear to be heavily associated with the reduced incidence of dementia with increasing age, even in those with the genetic risk factors for dementia (Wang *et al,* 2017). Healthy lifestyles and the notion of a healthy body equals a healthy mind cannot be ignored. Annus *et al* (2015) highlighted that amyloid accumulation appeared to manifest in people with DS in their early 40's, and that the short dormant period between accumulation and cognitive decline, could be attributed to poor cognitive reserve. The importance of increasing brain reserve and promoting increased cognitive resilience for people with DS cannot be ignored for future generations.

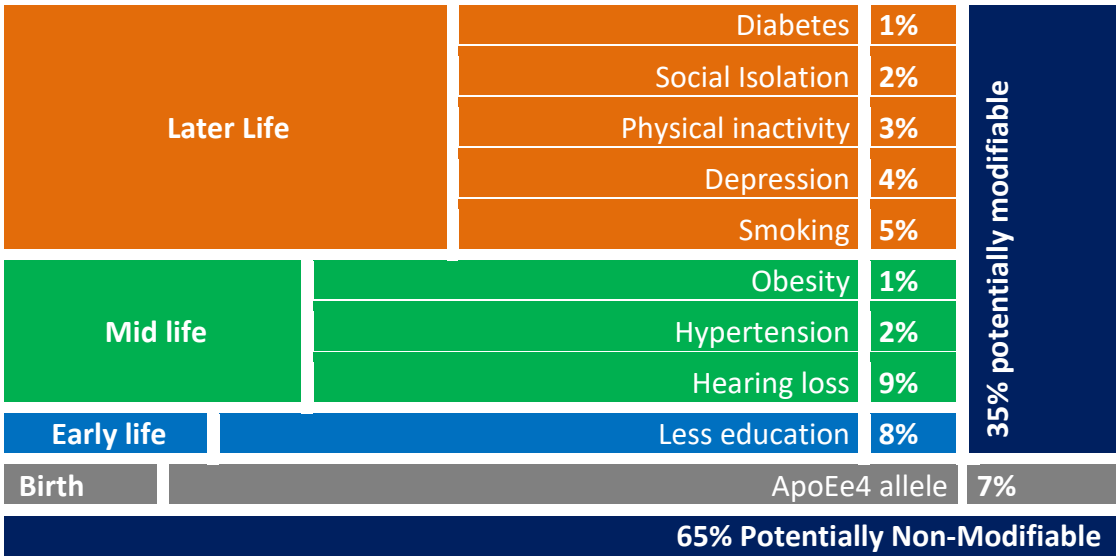
2.8.6 Prevention of Dementia

While the prevalence of dementia continues to increase throughout the world, there continues to be no cure or treatment to offset the clinical manifestations of the disease process (Dodge *et al*. 2012). Wu *et al* (2017) proposed that increased exposure to protective factors and less exposure to risk factors could offer greater resilience to the development of the disease. Livingston *et al* (2017) proposed a theory for prevention of dementia (see figure 2.2). Livingston and colleagues takes note of the widely published research that highlights the relevance of hypertension, obesity, sedentary lifestyle and smoking in the development of dementia, and that by addressing these health risk factors on the development of dementia the impact could positively addressed. Other risk factors included social isolation, age-related hearing loss and associated loneliness. The association between good health and an active lifestyle is

highlighted by Livingston *et al* (2017) who also recognises that poor general health and obesity are risk factors for dementia. Mangialasche *et al* (2012) noted that high education; a good socio economic status, good social networks and social engagement, mentally stimulating activities and an active lifestyle were protective of dementia. McGlinchey *et al* (2018) reported that 85% of people with ID in Ireland were underactive, while 43% of those over the age of 40-years were obese. Similar finding for people with DS were reported by Bertolli *et al* (2011) noting that their overall health needs, opportunities for meaningful employment and social engagement were not being met. Bertolli (2011) and colleagues highlighted the urgency for more wide-ranging inclusion in society of adults with DS and for the establishment of support services to create an enabling environment for inclusion. Educating People with DS about healthy lifestyles and building brain reserve (through brain training) is key to promoting better health outcomes and may be a way to improve cognition and delay dementia in this vulnerable population. The BEADS study (Brain Exercises for Adults with Down syndrome) McCarron & McGlinchey (2015) has proposed that using brain training games with a group of adults with Down syndrome without dementia, should positively influencing levels of executive functioning such as planning, attention and memory, and can have a protective effect for healthy ageing.

Physical health conditions and lifestyle choices are associated with dementia through the life span. **Table 2.2** below is a life course model of contribution of modifiable or non-modifiable risk factors to dementia put forward by Livingston et al (2017).

Table 2.2 Modifiable Risk Factors for Dementia



2.8.7 Dementia Prevention and Modifiable Risk Factors

The idea of prevention and cure for dementia is widely recognised and millions of Euro's are spent by the large Pharma corporations around the world to find the elusive breakthrough (Livingston *et al* 2017). But the cure for dementia is not yet in the pipeline and we must therefore look at modifiable risk factors (Livingston *et al* 2020; WHO, 2019; Livingston *et al* 2017).

Regular exposure to physical exercise, challenging cognitive stimulation and consistent social interactions over the life course is closely allied to increased cognitive reserve and reduced risk of dementia in later life, including those with the genetic risks for the development of dementia (Wang *et al* 2017), and people with ID especially those with DS who generally have poor cognitive reserve develop dementia much earlier in life (Strydom *et al* 2007). A person's overall health status coupled with their lifestyle can have a positive or indeed negative impact on their risk for dementia (Norton *et al.* 2014). With increasing age and a propensity for obesity there is a 19% increased risk for dementia reported by Loef & Walach (2013), while a study by Sofi *et al* (2011) found that regular high-level physical activity appeared to be protective against cognitive decline. A regular exercise regime not only improved cognition, but also reduced the

risk of falls in later life as well as improving mood and overall function (Almeida *et al.* 2014). The association between obesity and diabetes is well documented, and the link between obesity, diabetes and dementia is associated with low concentrations of cerebral insulin, increased glucose concentrations which can weaken amyloid clearance and impair cognition (Luchsinger & Gustafson, 2009).

Hearing loss confers a greater risk for the development of dementia, whereby it can lead to social isolation, depression and increased atrophy, which in turn increases the cognitive load in an already susceptible brain (Gopinath *et al.* 2009). Hearing loss, as will be described later in this review, is highly prevalent among those with DS. Depression and dementia are inextricably linked though whether depression is a symptom of dementia or a precursor to dementia is still strongly challenged (Livingston *et al.* 2017). Depression influences stress hormones, neuronal development and hippocampal capacity and therefore must influence dementia risk factors (Alexopulos 2003). Similarly, the question of social isolation and whether it is a symptom of dementia or indeed a precursor to dementia is challenged (Livingston *et al.* 2017). In a study by Kupier *et al* (2015) they reported that social isolation would inevitably lead to reduced cognitive activity leading to increased risk for accelerated cognitive decline and poor mental health, all of which increase the risk factors for a diagnosis of dementia. While promoting optimal physical health, the importance of engaging in a varied social activities programme cannot be underestimated (Livingston *et al* 2017). However it is known that social networks and physical activity among those with ID including those with DS are limited (McCausland *et al.* 2016; Burke *et al.* 2014;McCarron *et al*. 2011).

Following the Mediterranean diet (fruit, vegetables, fish, olive oil) will lessen the vascular risk factors and reduce glucose and insulin levels (Scarmeas *et al.* 2009). Dairy free options are also available. People who participate in regular exercise, do not smoke, drink alcohol moderately, eat a healthy Mediterranean based diet will inevitably age from a better standpoint and increase their life expectancy (Khaw *et al*. 2008). For people with DS however this type of diet could pose a challenge, many are lactose intolerant and Burke and colleagues (2014) reported people with ID including those

with DS did not consume the recommended daily allowance of fruit and vegetables which would inevitably contribute to overall poor health (Burke *et al*. 2014).

2.9 Epilepsy

The word epilepsy comes from the Greek word *epilamabanien* which means to seize or attack. It is the most common neurological disorder (after migraine) affecting 50 million people throughout the world (WHO, 2005), with a prevalence rate of 0.5 – 1.0% among the general population (Forsgren *et al.* 2005). Irish prevalence figures would suggest there are 36,844 people (over the age of 5) in Ireland affected by the disorder (Linehan *et al.* 2010) . Seizures can be caused by any type of brain injury, and in some circumstances seizure activity can cause actual brain injury (Holmes 2002). Epilepsy is the most common medical condition affecting people with ID (Chang & Lowenstein, 2003), and prevalence rates in people with ID are significantly higher than in the general population 16% to over 30%. (McCarron *et al.* 2017; McCarron *et al*. 2011; Morgan *et al.* 2003; Deb 2000). In people with DS the prevalence of epilepsy is generally lower than that in people with ID from other aetiologies, however, as people with DS age and particularly as they develop dementia a very strong association has been reported between new onset seizure activity and epilepsy (Robertson *et al*, 2015; McCarron *et al,* 2014, De Simone *et al* , 2010)

In recent years neurodegeneration and epilepsy have been inextricably linked in older adults and the association with Alzheimer's disease is well documented, with 10%-22% of patients presenting with new onset epilepsy requiring anticonvulsant therapy (Colom 2006). Epilepsy associated with dementia can have important outcomes on the prognosis of the underlying dementia and can result in decline in cognitive ability, particularly in language and comprehension, as well as decline in day-to-day functional ability, and increased risk of mortality (Hommet *et al.* 2008; Robertson *et al.* 2015). In their study McCarron *et al* (2017) McCarron et al (2014) found that the prevalence of epilepsy increased with age in people with DS and was predominantly associated with Alzheimer's disease.

Age, epilepsy and dementia are inseparably linked (Friedman *et al.* 2012; Hirtz *et al.* 2007; Stephen & Broadie, 2000). For people with dementia the incidence of epilepsy is up to 10 times greater than those without dementia (Hommet *et al*, 2008), and can contribute to an accelerated compressed decline in cognition and memory impairment (Scharfman 2012). A study by Palpo *et al (*2007) who examined epileptic activity in mice noted that epileptic seizure activity had a significant impact on hippocampal reserve and memory impairment was inevitable. It is now accepted that epilepsy is a frequent accompaniment to people with DS as they age and is associated to the development of neuropathological changes of Alzheimer's disease (McCarron *et al.* 2014; Lott *et al.* 2012; Prasher & Janiciki, 2002). While 10% - 22% of people with Alzheimer's disease in the general population will go on to develop epilepsy (Szot, 2012; Colom 2006; Hauser *et al.* 1986), for people with DS and Alzheimer's disease the figures are much higher and reported to be up to 74% (McCarron *et al.* 2014; McCarron *et al.* 2010). Alzheimer's disease and progressive myoclonic epilepsy are both linked to chromosome 21 (McCarron *et al.* 2014; Menendez, 2005). This has received varying emphasis in the literature and Strydom *et al.* (2010) espouses that epilepsy and myoclonus are inextricably linked with dementia and DS. It has been noted that this presentation is particularly evident in those with a severe level of ID (Cosgrove *et al*. 2000, Tyrell *et al*. 2001). Lai & Williams (1989) in their study followed 96 adults with DS longitudinally over 10 years and of the 51% who had a diagnosis of dementia, 84% of that cohort had epilepsy, and 92% of this group had their first seizure following dementia diagnosis. A study by McVicker *et al.* (1994) looked at establishing the prevalence of epilepsy in 191 adults with DS aged 19 years and over. They found an overall prevalence of 9.4%, which increased with age, reaching 46% in those over 50, and up to half of people with DS over the age of 50 years presented with new onset epilepsy (Bittles & Glasson, 2004). More recently, a systematic review by Strydom *et al.* (2010) noted that in those with DS over 60-years there appears to be decreased prevalence of epilepsy, but this is associated with increased mortality rates in those with dementia.

While the prevalence of epilepsy and dementia are widely reported in people with DS, so too is dementia and thyroid disease (Startin *et al*. 2020; Kinnear *et al.* 2017; McCarron *et al* 2017; Farriols Danes, 2012; Esbensen, 2010). The prevalence of thyroid

disease in people with DS is also well recognised with increasing age and hypothyroidism in particular is often mistaken for the symptoms of dementia (Lavigne *et al.* 2017). Diagnostic overshadowing in relation to thyroid dysfunction and the importance of routine thyroid testing cannot be overstated (Javaid & Michael 2019Fergeson *et al.* 2009).

2.10 Thyroid Disorder

Thyroid disorder is common in the general population and is reported in more than 10% of people over the age of 80-years (Boelaert, 2013). The thyroid gland is responsible for producing thyroxine, which helps regulate the body's metabolism. The most common form of thyroid disease for people with Down syndrome is hypothyroidism, an under active thyroid gland. Hypothyroidism is an endocrine disorder in which the thyroid gland is not functioning to produce enough thyroid hormone, and leads to excessive tiredness, poor tolerance to the cold, constipation, depression and increased weight (Chakera *et al*, 2012). Thyroid disease can occur at any age but is more common with increasing age and is particularly common in people with DS (Prasher 1999). Thyroid disease can be difficult to diagnose in any group of people and particularly people with ID. It is well documented that the prevalence of hypothyroidism in people with DS is high, with prevalence close to 50% (Lavigne *et al*. 2017; Farriols Danes, 2012). This can cause weight gain, dry skin, abdominal discomfort with associated constipation, general slowing down, tiredness and what can appear to present like cognitive decline (Perry *et al.* 2011). The prevalence of hypothyroidism is more apparent with increasing age and this is clearly highlighted by Startin *et al* (2020) who report a prevalence of 7% in children up to 5 years of age, 8.6% in children 5-15 years of age; 30.6% in young adults 16-35 years and 42.5% in older adults of 36+years. Lavigne (2017) and colleagues also noted that increasing age in people with DS was associated with increased prevalence of thyroid disease.

The prevalence of hyperthyroidism among those with DS is not as well known (Goday-Arno *et al.* 2009). The symptoms of hyperthyroidism are usually difficult to recognise. Hyperactivity, restlessness, anxiety and fatigue, the most common symptoms, are the

result of an overproduction of thyroxine hormone, which can lead to marked weight loss and an irregular heartbeat. For healthy adults with DS there is a gradual increase in thyroxine and possible gradual decline in thyroid-stimulating hormone with age (Prasher *et al*. 2011). Prasher's longitudinal study over 15 years notes that many studies have overestimated thyroid disorder in people with Down syndrome, and that the incidence of thyroid disorder is in fact low. This argument remains unclear to the writer. However, it is envisaged that additional research may contribute to clarifying the issues. Despite the contradictions, early diagnosis and treatment of any thyroid disorder is critically important, for ageing adults with DS, because of the effect of circulating thyroid hormone levels, which can have a significant effect on the heart, the skeletal system and the neurological system (Boelaert, 2013). Ongoing surveillance of thyroid function is recommended every two years. Henderson's (2007) study found that only 39 percent of people with DS had their thyroid function checked in a two-year period while 44 percent had gone untested in a five-year period, and this is also reported in Lavigne *et al* (2017) study where they found that not all people with DS were up to date with their thyroid function tests. Henderson (2007) concluded that it remains unclear as to who is responsible to provide primary health care checks and standardise care for this population, and the possible reason why routine thyroid function tests were not being routinely carried out was due to non compliance on the part of the person with DS or their carers (Lavigne *et al*. 2017).

2.11 Sensory Impairments

Vision and hearing are critical to how we learn to understand the world. "*Information received through the senses can be considered as the basis for an individual's learning about and acquiring a conceptual understanding of the properties of the physical world*" (Warren, 1984, p.59). The literature reports that people with DS are at greater risk for visual impairment (Jensen & Bulova 2014; van Splunder *et al* 2006; van Splunder *et al* 2003; Evenhuis, 2001). As far back as 1912 Ormond reported ocular abnormalities in people with DS (Ormond, 1912), while Cullen (1963) noted that if a person with DS survived to the age of 30-years, eye surgery would be uncertain, and possibly life threatening. Visual deficits can have negative implications for learning; can impact on cognition and influence behaviours of concern for people with DS (Krinsky-McHale *et al,*

2014). Furthermore, poor vision can impact on independent living skills, ability to communicate, day to day social skills and on mobility (Silverman *et al* 1998; Evenhuis *et al* 2009). In his study, van Splunder *et al* (2006) observed and examined 1598 people with ID in an epidemiological study of visual and hearing impairment. Here the study noted alarming under recognition of visual impairments. Visual impairments were not recognised in 42.6% of participants and 35.9% of people with ID in the study were blind despite having no diagnosis. The study focused in particular on how the prevalence of visual impairment was related to increasing age, level of ID and DS. Young people with mild ID of other aetiologies represented 2.2% (least affected subgroup) of those with visual impairment, while 66.7% of people with DS and over the age of 50-years had the disorder. This study found that the relationship between increasing age and visual impairment in people with DS was much stronger that the relationship with the level of ID (van Splunder *et al.* 2006). Whereas, Evenhuis *et al* (2001) did find an association between level of ID and visual deficits. Here, she reports that the risk for visual impairment was lower in people with mild to moderate ID with a prevalence rate of 4.5% in people with DS, while 74% of those who had severe/profound ID and DS had a visual impairment. She did identify an association of visual impairment in those with DS over the age of 50-years, whereby an increase from 4% to 31% in visual impairment is recognised and only found in those with mild/moderate ID and DS. This, she concluded, was most likely associated with age-related cataracts (Evenhuis *et al.* 2001). Findings described by Georgalas *et al* (2014) reported that visual impairment in people with DS was as high as 77%, with deficiencies in pupil size, corneal thickness and volume. This study described a prevalence of 25% of people with DS presenting with refractive errors and 21% with strabismus, while cataract disease had the highest prevalence rate at 42%. In his 2003 study, Van Splunder *et al* noted a higher prevalence of myopia and low distant acuity in people with DS compared to people with ID of other aetiologies. Considering these findings overall it can be noted that the prevalence is higher for those with DS and what is of more concern are the findings that it can go unrecognised. Identifying risks for visual impairment among those with DS is imperative and the associated outcomes of poor eye health.

In ascertaining the risk factors for visual impairment in people with ID over the age of 50-years, DS itself was the strongest risk factor for visual impairment (Van Splunder *et al.* 2006). Evenhius *et al*. (2001) also had similar findings investigating 672 people with ID and noted that poor recognition of visual impairment was a concern for people with ID. This study noted that people with DS (n=97) over the age of 50-years particularly were at high risk of visual impairment. All studies noted the importance of regular ophthalmologic reviews as part of routine health checks for people with DS, as poor surveillance would inevitably lead to permanent visual impairment which would certainly impact on overall quality of life (Georgalas *et al* 2014; van Splunder *et al* 2003; Evenhuis *et al* 2001). Evenhuis *et al* (2009) reported that visual impairment contributed to additional disability for people with ID and DS. In her study Evenhuis and colleagues asserted that visual impairment was linked with decline in independence; increased difficulty in communication; decline in mobility; social isolation and disruptive behaviours which were very influenced by progressive visual impairment. Anthony *et al* (1989) reviewed 187 medical records of people with DS over a 10-year period for strabismus, myopia, hyperopia, astigmatism, nystagmus, cataract, glaucoma, and other significant eye findings. This study showed that a higher proportion of these individuals than reported in previous studies had strabismus (57%). Again, the high prevalence of visual impairment and the possible under recognition is concerning. This study noted that the primary care physician needs to be aware of the specific eye problems of individuals with DS so that he or she may initiate or refer the patient for appropriate ophthalmologic care, because most of the eye findings in DS are treatable. Van Splunder *et al* (2013) noted that significant visual loss, a usually avoidable event in Down syndrome, should occur rarely. However, for those with DS and visual impairment especially if there is a gradual onset the risk of diagnostic overshadowing is possible without the presence of a robust comprehensive health assessment eye conditions may be missed and individual's reluctance to walk for example, be attributed to onset of dementia.

A population based survey by Maatta *et al* (2011) noted that 70 percent of adults with DS had significant eye concerns, including eye disease and refractive anomalies. Poor eye health was associated with increasing age, with 67 percent of the older population presenting with eye concerns compared to 50 percent of the younger cohort in the

study. This Finnish study reported that corrective glasses were given where prescribed and the uptake of regular eye examinations was relatively good. Contrastingly, Evenhuis (1999) found that visual impairment in an ageing population of people with ID including those with DS was invariably poorly managed. In her study she found that most people had never been seen by an ophthalmologist. Some of the participants refused to cooperate with eye examinations, or indeed were too fearful to attend. As a result, prescription glasses were not recommended, and this was especially true of people with DS and dementia. Woodhouse *et al* (2000) in a study involving 154 adults with ID, found that 52% of participants had distance refractive errors and 56% had poor near vision acuity. This cohort of people did need corrective lenses, and would benefit from wearing spectacles. However, only 20.8% of this study group were wearing corrective lenses. These findings are further supported by McCarron *et al.* (2013), who notes that while 87.5% of people with ID without dementia presented with visual impairments and 92.8% with a diagnosis of dementia reporting significant visual difficulties very few had had corrective surgery or wore glasses. Evenhuis (1999) also noted in her study that many ageing people who need to wear glasses would more often refuse to comply. Other visual abnormalities seen in people with DS and documented in various studies include keratoconus (whereby the cornea protrudes) cataracts (causing clouding of the eye lens) and blepharitis (inflammation of the eyelids) (Krinsky-McHale *et al.* 2012; Matta *et al.* 2011; Evenhuis *et al*. 2001). Overall the prevalence of eye disease and impairment is notable among those with DS and ensuring robust assessment and annual ophthalmic health checks are conducted is imperative to overall health and wellbeing of people with DS.

Hearing impairments are also common in adults with DS, with up to 70 percent affected with varying degrees of hearing loss (Gallagher *et al.* 2012; Smith 2001). This population often have narrow ear canals, which increases the risk of recurrent infection and wax impaction. Because of associated intellectual disability and poor communication skills, whereby self-reporting is often difficult, individuals find it difficult to communicate that they are having hearing difficulties, and as a result will more likely go untreated. Presbycusis (age related hearing loss), is common in most adults with DS, which significantly impairs receptive skills, complicate verbal skills and consequently the ability to communicate well (Evenhuis, 1999). In her study in 2001, Evenhuis reported that the

prevalence of hearing impairment in people with DS was significantly higher (64%) than in those with ID of other aetiologies (21%), and increasing age was a significant factor here for this population. McCarron *et al.* (2013) reported significant differences in hearing loss when comparing those with or without dementia. Presbycusis was significantly more common in those with dementia at 62.3 percent, while 37.5 per cent of those with no evidence of dementia had no obvious hearing difficulties. Studies by Deal et al (2016) found that hearing loss was a significant factor in dementia in the general population, while other studies also highlight the significance of hearing deficits and incident dementia (Gallagher *et al.* 2012; Lin *et al.* 2011).

Hearing loss is often unrecognised in people with ID and in those with DS. Issues and concerns arise whereby the individual is deemed uncooperative, if they do not respond to requests, when in fact they have poor hearing function and significant sensory deprivation as well as their underlying intellectual impairment. Suspicion should be raised when the individual is not responding in the usual way, or if they have obvious difficulties with new learning, or they become withdrawn (Perry *et al*. 2011). Kiley *et al* (2013) and Kuiper *et al* (2015) identified hearing loss and associated social isolation as a key risk factor for dementia but noted that hearing loss is modifiable. While auditory testing every two years is recommended for people with DS (Smith, 2001), various studies throughout Europe show that people with DS fail to receive regular health checks and consequently treatment is more often delayed or not undertaken until the problem becomes clinically apparent (Glasson et al 2014). This is supported by Ouellette-Kuntz et al (2005) who reported that while it is recognised that people with ID do have a higher than average prevalence of hearing loss, they are more likely to experience 'diagnostic overshadowing', with presenting symptoms being attributed to their underlying intellectual disability.

The National Institute for Health Research (2020) reported that there are long standing difficulties for people with ID in accessing mainstream health services, for physical health supports and sensory impairments, and consequently, with many easily remedied conditions, such as visual and hearing impairments, becoming barriers that reduce social/community participation, and overall quality of life of the individual (Emerson & Baines 2010). Similarly, Henderson (2007) reported high rates of

preventable and treatable medical conditions in adults with DS, with negligible medical involvement. The diagnosis of both hearing and visual complications in people with DS, is complex and frequently overlooked, due to poor communication and diagnostic difficulties all the more reason for vigilance and care (Evenhuis *et al*. 2001). The need for regular auditory and visual testing needs to be highlighted across all services, particularly for those with DS over the age of 50-years as recommended in the IASSID International Consensus Statement (Evenhuis & Nagtzaam, 1998).

2.12 Consequences of Deteriorated Physical Health Conditions

The patterns of physical health conditions in people with DS continue to rise with increasing age (Kinnear *et al.* 2018; Burke *et al*. 2014; Malt, 2013; Farriols-Danes, 2012;Esbensen, 2010; McCarron *et al*. 2005; Bittles & Glasson 2004; McCarron & Lawlor 2003), and with increasing health comorbidities comes the need for increased levels of support for declining skills, particularly associated with dementia, and more specialised support services (Hogg *et al.* 2001; Bittles & Glasson, 2004). With the progress of dementia comes cognitive decline and marked decline in day-to-day functional ability. People with DS and dementia who were previously independent require increased levels of support with dressing, continence and personal care, nutrition and hydration, mobility and a high level of healthcare support (McCarron & Lawlor, 2003) which ideally would be multidisciplinary to include nursing support, social care, psychiatrist, physiotherapist, physician, speech and language therapist, psychologist who are specialists in ID and have a high level of understanding of the health complexities of DS (Farriols-Danes, 2011).

Increased challenges exist for families and services supporting people with DS, dementia and functional decline. Ideally the 'Ageing in Place' model of care (Wilkinson and Janicki, 2002) is the preferred care model, but with progressive physical deterioration associated with dementia, this can be difficult to sustain (McCarron *et al*. 2010). With advanced stage dementia it is often the case that people with DS and dementia have to move home setting to high nursing support environments (McCarron *et al*. 2014; McCallion *et al*. 2012). Specially designed environments or modified homes

are required with large spaces to facilitate ease of movement with wheelchairs and hoists (McCarron *et al.* 2010), and this model of care works well.

The life expectancy of People with DS is similar to that of age matched peers with ID of other aetiologies until the age of 40-years (Esbensen, 2010). This pattern in mortality changes radically from the age of 50-years in people with DS when comparing them to the general population or people with ID of other aetiologies. Some studies have reported that decline in function is a predictor of mortality in people with DS (Esbensen *et al* .2007; Chaney and Eyman, 2000) and (Eyman and Call, 1991) reported that declining mobility or feeding skills were strong predictors of an early death in people with DS. Having a clear understanding of the complex health comorbidities common in DS is particularly important for providing accurate information for future care needs and in the provision of appropriate post diagnostic supports (Hithersay *et al*. 2019)

2.13 Overview of the problem

It is clear from this literature review that the main health conditions to emerge from the literature that impact on the overall ageing process of individuals with DS are dementia, epilepsy, thyroid disease and sensory impairment. Whilst there are other conditions such as osteoporosis, respiratory disease and diabetes, this literature review specifically sought out the more common health conditions that can impact on ageing in those with DS. Individuals with DS emerge with higher risk and prevalence than those with ID of other aetiologies and of the general population (Startin *et al*. 2020; Jensen & Bulova 2014; Esbensen 2010; Henderson *et al*. 2007; McCarron *et al*. 2005; Cooper *et al*. 2004). It was also evident that healthcare utilisation and assessment is sporadic and infrequent this could possibly be as a result of the complexities of the presentation among those with DS or indeed the difficulties of arriving at a diagnosis due to communication challenges with the individual or simply the lack of recognition of these conditions among this cohort.

2.14 Gap in the research

There exists a vast array of literature on those with DS especially in the area of dementia however investigations on specific health conditions longitudinally that impact their ageing as a whole is particularly sparse especially in Ireland. Subsequently, the researcher has developed a conceptual frame from the literature examined to guide the exploration and investigation of the impact of health conditions on the ageing of a cohort of individuals with DS. The Intellectual Disability Supplement to the Irish Longitudinal Study on Ageing (IDS-TILDA) will facilitate this exploration and by mapping this frame to the IDS-TILDA conceptual frame appropriate data will be identified. This will be explored in the next chapter.

2.14.1 Conceptual frame for the investigation of the impact of physical health conditions on the ageing of those with DS overtime.

From the literature the main concepts were extracted and the following conceptual frame was developed considering our subject of interest ie DS ageing over time, the chronic health conditions that are experienced and the outcome of those conditions on the individual.

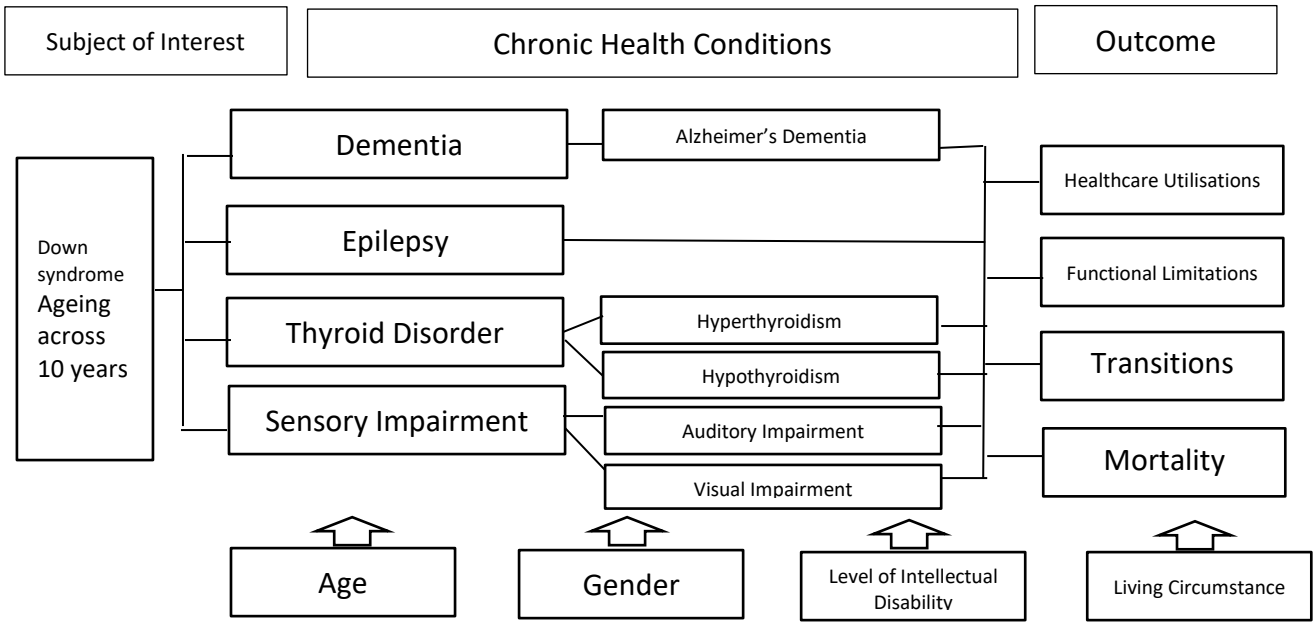


Figure 2.4 Conceptual frame guiding the investigation

2.15 Conclusion

Health encompasses a lifestyle that upholds the physical health and well-being of the person through every stage of life, and is more than the absence of disease. This is the vision of '*Healthy Ireland*', (Health Service Executive, [HSE], 2015) a strategic policy framework that promotes the health of all citizens of Ireland to ensure health and wellbeing is valued at every level of society and by all citizens. For people with an ID including people with DS this can pose challenges because of pre-existing cognitive impairment and increased risk for specific health conditions (Harris, 2010). Down syndrome is among the most commonly identified causes of ID (Sherman *et al*. 2007), and in developed countries accounts for 12 to 15% of the population of those with ID (Bittles *et al*. 2002). The life expectancy of people with Down syndrome has increased exponentially over the past 50-years. The life expectancy of this population has increased to a point whereby, it is suggested that within the next generation people with DS will be living as long as the general population (Bittles & Glasson 2004). However, as a result this longevity it is also obvious from the literature that individuals are experiencing similar complications of ageing as the general population however there are specific health conditions that are more prevalent (Kinnear *et al.* 2018; Burke *et al*. 2014; Malt, 2013; Farriols-Danes, 2012;Brown *et al.* 2010; Esbensen, 2010; McCarron *et al.* 2005). Gibbs *et al* (2008) recognise that people with an ID including those with DS have more unmet and unrecognised health needs than the general population, and the Department of Health (DOH 2007) would suggest that people with an ID have a reduced life expectancy as a result (Read 2006). It is considering these anomalies that the researcher poses the following research question, aims and objectives for this study:

2.16 Research Questions

Considering the high prevalence emerging in the literature of specific conditions among those with DS the following questions is posed:

> *What are the predominant health conditions emerging among those with DS as they age? What is the impact of these conditions on their ageing with respect to functional ability, healthcare utilisation and mortality?*

2.17 Research Aim

The aim of this two-fold; i) to examine the health profile of those with DS across a 10-year time period with specific consideration to the most prevalent health conditions emerging. ii) To establish the impact of ageing with respect to functional ability, healthcare utilisation and mortality.

2.18 Research Objectives

To achieve these aims the study will have the following objectives:

- Identify the physical health conditions prevalent among those with DS across 3 waves of data from the IDS-TILDA study [Wave 1, 2011; Wave 2, 2014; Wave 3, 2017].
- Examine dementia, epilepsy, sensory impairment and thyroid disease across 3 waves of data collection in the IDS-TILDA study in respect to age, gender, level of intellectual disability and living circumstance.
- Identify the impact of these conditions on healthcare utilisation, living circumstances and functional ability.
- Identify the associations between health condition and mortality

The next chapter will elucidate the methods used to explore how these aims and objectives will be met.

CHAPTER 3

Methods

3.0 Introduction

Methodology implies the comprehensive research plan for the overall study and methods refer to the techniques to format and design the study data collection and analyse the appropriate data collected (Polit & Beck 2018, LoBiondo-Wood & Haber 2017). This chapter presents the research design and the methods used to explore the physical health of individuals with DS as they age. The research process will be presented with consideration to the aims and objectives for the study, and the research design used by the IDS-TILDA study. The IDS-TILDA sampling frame, sampling strategy and recruitment will be described to include sample selection and inclusion criteria for this study. The measurement tools used to retrieve the data pertaining to this thesis will be outlined. As this study is part of a large longitudinal study the researchers' development and contribution to the study is elucidated. The ethical principles and their application within the study are explored along with the data protection requirements for participants involved in this study. The analysis applied within the study is presented. In conclusion an overview of the chapter will be summarised.

3.1 Research Methods

3.1.1 Research Design

Research design refers to the actual planning and designing of strategies for answering the research question and specifies the fundamental form the research will take (Polit & Beck 2018). Parahoo (2006) notes that research design describes a plan that outlines how, when and where data will be collected and analysed. It is important to consider what strategies will best suit the research question to ensure the correct information is extrapolated and that the information gathered is accurate and understandable. The design of the study should include:

- The approach (qualitative or quantitative research)
- The method of data collection and ethical considerations for the study
- The time, place and source of data
- The data analysis

(Parahoo, 2006)

3.1.2 Exploration of the research paradigms

Qualitative and quantitative methods are very different approaches to research. Both qualitative and quantitative research examines relationships or the connection between phenomena and identifies different types of information. It is important to select the most appropriate design to suit the research question so that the aims and objectives of the study are met, and to ensure reliability of the data. Both methods of research create scientific information in different ways providing different types of information and different answers (Polit & Beck, 2018).

Qualitative research is a form of social inquiry the basis of which lies in the interpretive approach and in the description of the lived experience (Holloway & Wheeler, 2002). By comparison quantitative research largely involves measuring outcomes that can be converted into numerical data in either an experimental or non-experimental fashion and is embedded in descriptive statistics, which will describe the data (Polit and Beck 2018). The quantitative approach can be used to objectively test theories and analysis associations between variables. It is a scientific method of collecting and examining empirical data. It is considered to be the most appropriate approach to document the health status of individuals which it therefore the most suitable design to apply to this research. Using the steps proposed by Polit and Beck (2018) the researcher has adopted the quantitative approach to explore the physical health changes among those with DS as they age. These steps, which are now complete include:

- Reviewing the relevant literature
- Formulating and determining the question
- Defining the theoretical framework

By employing a quantitative approach, the research will be able to portray the traits and frequency of the data under scrutiny, the associations and changes over time which is in keeping with the longitudinal studies (Polit and Beck 2018).

As this study is situated within The Intellectual Disability Supplement to The Irish Longitudinal Study on Ageing (IDS-TILDA) study. The researcher will present the design

for the IDS-TILDA study with emphasis on those elements pertaining to this study specifically.

3.1.3 The Intellectual Disability Supplement to the Irish Longitudinal Study on Ageing (IDS-TILDA)

The Irish Longitudinal Study on Ageing (TILDA) examines the health and wellbeing in Ireland's of over 50's from 2006, and was designed to provide an evidence-base for addressing current and emerging issues associated with population ageing in Ireland across health, economic and social systems (Kenny *et al,* 2019). It focuses on the prevalence and incidence of age-related disease, disability, health service utilisation or economic and social data. TILDA has changed this landscape, enabling both cross-sectional and longitudinal evidence-based research (Kenny *et al,* 2019). The inception of The Intellectual Disability Supplement to the Irish Longitudinal Study on Ageing (IDS-TILDA) followed TILDA as they were not specifically seeking individuals with ID to be part of their research. The IDS-TILDA study identified that there was a unique gap in research on ageing and ID and this would provide an opportunity to address that gap.

The IDS-TILDA study following people age 40 years and over with an ID in Ireland is the first longitudinal study on older adults with ID being run in tandem with the general population study TILDA. IDS-TILDA was designed with the aim of understanding the specific characteristics and experiences of the ID population as they grow older which allows direct comparison between the two studies (McCarron *et al.* 2011). This makes the study unique both nationally and internationally. The overriding premise of IDS-TILDA is that people with ID approach ageing and experience ageing that is comparable to the general population, and that their participation and contribution to the study will be invaluable to the promotion of better health outcomes and living circumstances in years to come (McCarron *et al.* 2011).

3.1.4 IDS-TILDA Conception, recruitment and development

IDS-TILDA commenced in 2007 and collects data every three years, this is referred to as a data Wave. The IDS-TILDA principal investigator (PI), co-PI and the IDS-TILDA team worked in close collaboration with an international scientific advisory committee and a

number of advocacy groups (people with ID) on all aspects of the study development. The National Intellectual Disability Database (NIDD) provided the sampling frame from which 1800 individuals registered with an ID were randomly selected. The NIDD, established in 1995, is a large administrative service planning database that gathers information on people with ID accessing specialised health services in Ireland to provide for future planning of these services (Kelly *et al.* 2013). It provides the database of all individuals with all levels of ID in the Republic of Ireland that are eligible or receive services (HRB 2013) and is unique to Ireland. In 2011 (Wave 1) there were 27,324 registered on the NIDD database, in 2017 (Wave 3), these numbers had increased to 28,388 individuals with ID registered on the database. Based on the 2016 Census of Population figures, this represents 5.96% per 1,000 populations. Of these, 59.1% were males and 40.9% were females (Hourigan *et al*, 2017). Census data indicates that the prevalence of ID nationally has increased by 15% since 2011, to 66,611 people or 1.4% of the population in 2016 (Bruton *et al*. 2020). Looking at the population with moderate or severe ID, the percentage aged 35 years and over increased from 28.5% in 1974 to 37.9% in 1996 to 49.1% in 2017 (see Appendix 1). Gatekeepers in the form of the regional database controllers from the NIDD were used to distribute the invitation packs to the potential participants in IDS-TILDA. This included an easy to read information booklet, easy to read consent, a family information leaflet and family agreement form. When the participant agreed to engage in the study they signed and returned the consent form, and family members (where applicable) returned the agreement form. For those who were unable to provide signed consent their carer explained the study and determined if they wished to take part. Carers then signed on behalf of the participant. The consent was returned to the IDS-TIDLA office in the stamped address envelope provided with the invitation pack. On receipt of the consent, the participant was assigned an exclusive identity number.

The IDS-TILDA study chose a 10% representative sample of all adults over the age of 40 years, across all levels of ID and living in all types of accommodation from independent living to residential type accommodation. Whereas the TILDA study had a higher age limit of 50-years (Kenny *et al.* 2019) , IDS-TILDA set the lower age limit at 40-years as people with ID develop complex heath conditions at an earlier age compared to the

general population (Haveman et al, 2010). The Wave 1 (2011) sample included 753 persons, which represented 46% of the sample drawn from the NIDD, comprising 8.9% of those registered on the NIDD who were 40 years and older (McCarron *et al.* 2011). For Wave 2 IDS-TILDA data collection, researchers contacted those who had completed Wave 1 (N=719, individuals living at time of wave two participant recruitment) and invited these individuals to take part in Wave 2. In total, 701 people agreed to continue with their participation in the IDS-TILDA study for Wave 2 data collection (Burke *et al.* 2014). For Wave 3 IDS-TILDA data collection, researchers contacted participants who had completed Waves 1 and Wave 2 (N=699 living at the time of Wave two recruitment) and invited them to take part in Wave 3. Those who agreed to engage in Wave 3 of the IDS-TILDA data collection protocol included 609 in total (McCarron *et al.* 2017). Retention of participants from Wave 1 through to Wave 3 was 86% and this can be attributed to the 'Keeping in Touch' strategy that is employed by the IDS-TILDA Team. Participants are sent Christmas cards and Easter cards and receive a biannual newsletter to keep them up to date with the study; they are encouraged to attend various roadshows and information sharing forums throughout the year (See Appendix 2 for an example).

A comprehensive conceptual framework which examines the areas of health, behaviour and well-being underpins all data collected in the IDS-TILDA study (see Appendix 3). The IDS-TILDA conceptual framework gathers a vast array of data across a wide variety of domains including physical health, cognitive health, psychological health, behavioural and mental health, healthcare utilisation, social participation and connectedness, and other areas related to health, well-being and quality of life of adults ageing with ID in Ireland. The strong quantitative design, representativeness of the sample and focus on ageing with ID meant that IDS-TILDA held the best available data for examining the common health conditions associated with ageing and DS. The study is also underpinned by a values framework that summarises the philosophy of person-centeredness in the study and reinforces the rationale behind IDS-TILDA (see Appendix 4). People with ID were involved throughout the whole research process, in planning, preparation, piloting, data gathering and dissemination of IDS-TILDA. Consultation workshops took place with various advocacy groups representing people with ID when

planning for next wave of data collection. These workshops took place in various centres around the country in disability services. During these workshops, people with ID were able to identify the key issues they believed were important and should be included in the next wave of data collection. They were invited to give their thoughts about proposed new topics to be included in the new questionnaire. All ideas were documented and shared with the team.

3.1.5 Population sampling and participant distribution

Before any decision is made regarding a study design it is important that the researcher knows exactly what data is required and from whom it should be collected. The target population is the group of people whom the researcher aims to draw the sample from (Parahoo, 2006). The target population for this study included people with DS. The IDS-TILDA sample was drawn from the NIDD which includes those with DS, the IDS-TILDA study included a question asking the participants the aetiology of their ID, this identified those with DS and ensured those with DS could be selected. The sample for this study comprises of all the participants with Down syndrome who participated in Waves 1, Wave 2 and Wave 3 of the IDS-TILDA study. In total N=147 participants with DS engaged in Wave 1, N= 138 participated in Wave 2 and N=109 engaged in Wave 3 (see *figure 3.1*) for sample of those with DS included in this longitudinal study over the three waves of data collections.

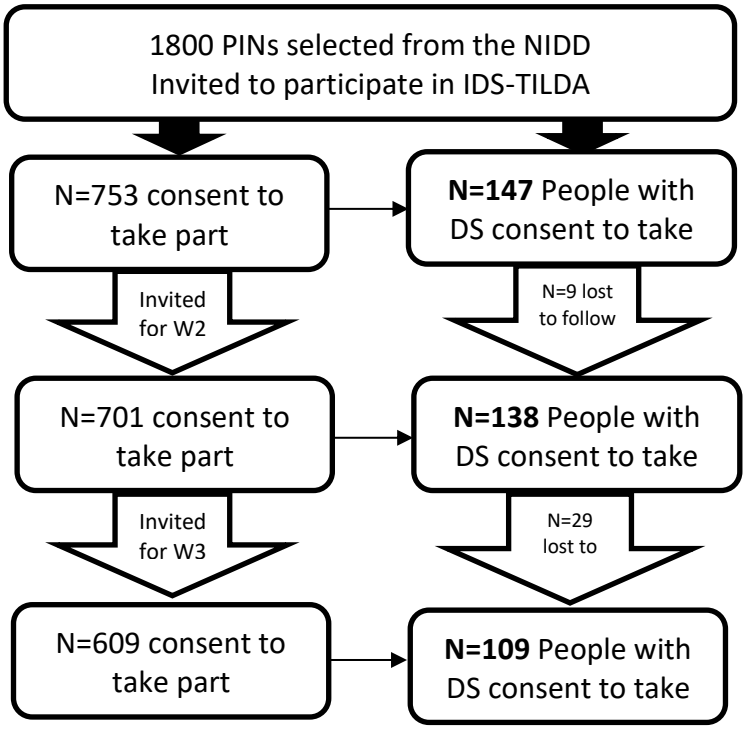


Figure 3.1 Flow chart of those with DS participating in the study

The sample was selected from over 130 individual service throughout the country see *figure 3.2* for the distribution of the sample.

Figure 3.2 National participant distribution

3.2 Data collection process

As one of the field researchers this researcher undertook a comprehensive field research training over three days (see Appendix 5). On completion the researcher was assigned a case load of participants. It was the field researcher's responsibility to manage that case load which consisted of contacting the participant, arranging the face to face interview and sending out the pre-interview questionnaire (PIQ) a minimum of one week before the face to face interview. Sending the PIQ in advance was to ensure the participants had enough time to gain support to assist with completion and time to source the information. The interview was conducted at a time and place to suit the participant. When the appointment was arranged an appointment card with the interviewer's picture and their contact details were sent to the participant, in that way they knew who was going to come to see them and do the interview (Appendix 6).

For the purposes of the interview there were 3 styles offered to the participants depending on their ability.

1. Self report whereby the participant answered all the questions independently.
2. A supported interview whereby the participant would be supported by their carer or keyworker.
3. Proxy interview whereby a proxy would answer the questions on behalf of the participant. The proxy had to be someone who knew the participant well, either a family member or a key worker who had supported the individual for a minimum of 6 months. For the purposes of a proxy interview self-reported modules and questions were skipped for example questions that referred to how the participant felt.

3.2.1 Researcher contribution

The researcher had a case load of 40 participants to complete. Each interview lasted between 90 – 120 minutes. Prior to undertaking this the research completed the comprehensive training which included interview techniques, building familiarity with the protocol, data protection, all ethical considerations involved in the study and the informed consent process. The researcher also delivered the comprehensive training on the administration of the neuropsychological test instruments included in the IDS-TILDA. Practice guidelines were delivered to provide a framework to ensure a common understanding amongst all researchers and to ensure consistency. The overall philosophy of IDS-TILDA was instilled, and the importance of confidentiality discussed, particularly in the context of carrying out research with people with ID. The researcher was also responsible for creating her own dataset, adhering to the data management protocol of the IDS-TILDA study (see Appendix 7), data cleaning and variable construction with the supervision of the IDS-TILDA data manager and her supervisor. The researcher conducted all analysis, development of graphical and tabular representations and write up. The analysis is explored further below.

3.2.2 Data collection tools

The IDS-TILDA data collection instruments, pre-Interview questionnaire (PIQ) and the computer assisted personal interview (CAPI) were developed in consultation with a national Steering group, an international scientific advisory board and self-advocacy groups of people with ID and experts in the field of intellectual disability. These groups were involved throughout the piloting, design and evaluation of IDS-TILDA questionnaires and study processes. This helped to ensure greater accessibility, validity and quality of the instruments and the data collection processes for participants and researchers (McCarron *et al.* 2011).

The Pre-Interview Questionnaire

The Pre-Interview Questionnaire (PIQ) was sent to participants approximately seven days prior to the interview. This enabled participant time not only to gain assistance to

complete the document but also time to gather the information from their medical files or doctors to ensure the information was as accurate as possible. At the time of the interview the researcher collected the PIQ and completed a face check to ensure responses were recorded for all questions. This PIQ gathered demographic information, health conditions, health care utilisation and medication use. The PIQ included questions pertaining to specific chronic health conditions, including "Do you have a doctor's diagnosis" of any of the following including:

- Dementia
- Epilepsy
- Thyroid Disorder
- Sensory Impairment

Computer assisted programme for interviewing (CAPI)

The face to face interview was administered using a Computer Assisted Personal Interviewing Programme (CAPI) using an encrypted laptop. The main questionnaire was built into the system. All answers were directly inputted into the computer negating the need for transcription. The CAPI system uses built in programming to apply the questions filtering in accordance to the answers provided by the participant. All interviews in the IDS-TILDA study where completed in this manner. The main face to face interview consisted of a number of modules which included health, social, psychological, mental health, ageing perceptions, loneliness, community participation and social networks, friends and family connections as well as activities of living and functional limitations. In Wave 3 the data on age, gender and address were re-verified as part of the consent process before the Wave 3 interview commenced.

Neuropsychological Cognitive Assessments

The following measures were used by IDS-TILDA to detect cognitive decline. The Test for Severe Impairment was used across Waves 1, 2 & 3. The Brief Praxis was introduced in Wave 3 however as this has only one wave of data collection it has not been considered for this study. These instruments were chosen to assess cognition, behaviour and clinical global functioning.

Test for Severe Impairment (TSI)

The TSI, a neuropsychological test instrument was developed in 1992 by Albert and Cohen, and its scores correlate with the Mini-Mental State Exam (MMSE). It is a validated and reliable test instrument that measures cognitive function over eight domains (including language, memory, executive function and motor performance), and was specifically developed to assess people with severe cognitive impairment and deemed appropriate to use with people with Intellectual Disability (Cosgrave *et al.* 1998). The TSI has been included in the IDS-TILDA protocol for all three waves of data collection this will enable comparison across waves and track changes over time. (See Appendix 8 for TSI instrument).

3.2.3 Feedforward

In Wave's 1 and 2 the main physical health condition questions, health care utilisation and medication usage were contained in the PIQ. At Wave 3, previously identified health conditions were pre-programmed into encrypted files only accessible through the participant individual identifier (PIN) known as feedforward. Feedforward is the means through which data obtained from individuals from Wave 1 and Wave 2 was carried forward into their interview at Wave 3 to enable more efficient and higher quality data collection. By enabling the feedforward system of health conditions, the interview was also streamlined as it identified within the CAPI programme what questions could be skipped thus creating a more individualised experience for participants and reducing any possible burden. It also enabled any previous misinterpretations to be identified and clarified at the time of the Wave 3 interview. This ensures a more valid and accurate dataset.

3.3 Ethical considerations

Codes of ethical conduct in research guides good practice and promotes highest standards for researchers to adhere to in order to engage in safe and ethical research that will benefit and protect those participating. Ethical conduct in health research is guided by the Nuremberg Code of Ethic (1947), the Helsinki Declaration 1964 (World

Medical Association 2013) and by the Code of Professional Conduct and Ethics for Registered Nurses and Registered Midwives (NMBI 2014). Every research process presents with a plethora of ethical repercussions, particularly medical/healthcare research. NMBI (2014) observes the right of an individual to participate or not in a research study. Full disclosure about the study is imperative in order to avoid any sense of ambiguity. The benefits and burdens of the study must be disclosed in order that the person can make an informed choice as to whether they wish to participate or not, and indeed are also made aware that they can withdraw from the study at any time. There is great potential for nursing research to inform all areas of practice and policy. In undertaking this area of research, the ethical principles proposed by ABA are used to guide the researcher through the research process. When conducting research with individuals with intellectual disability ethical principles must be considered carefully and rigorously to ensure this clarity, accessibility and maintain privacy. As Flaskerud and Winslow note;

> '*Research with vulnerable populations challenges us to consider once again ethical principles basic to research. Issues of providing informed consent, maintaining confidentiality and privacy, weighing the risks and benefits of a study and paying attention to issues of fairness are all especially important with working with groups who are vulnerable*'.
>
> (Flaskerud and Winslow 1998, p. 69)

Adhering to ethical standards implies good ethical conduct and the six ethical principles put forward by International Council of Nurses (2005) and NMBI (2014). This research is embedded in these principles to ensure the highest possible standards in the research process. Ethical approval for all 3 Waves of the study has been granted by the Ethics Committee, Faculty of Health Sciences, Trinity College Dublin. Ethical approval has also been granted by all the service providers participating in the study (see Appendix 9 for ethical approval letters from the FHS, ethical approval letters from services are available on request, too numerous to include N=138).

3.3.1 Respect for the Dignity and Autonomy of the Person

Autonomy can be defined as the ability of the participant to make their own decisions. It refers to the capacity to be one's own person and to live a life of their own choosing.

The right of the participant to make sound decisions about their future care needs without being unduly influenced by outside forces or indeed the right to participate in research without fear of coercion. Participants were well informed and educated about the research through the use of easy read information and accessible documentation (see Appendix 10). All questions were supported through the use of flash cards which were a pictorial representation of the concept of the question to assist with the understanding. Through the use of an accessible consent form (see Appendix 11) all the participants rights were made clear and they were informed that they could withdraw from the study at any time without any consequences.

3.3.2 Beneficence

Beneficence is the duty to do and to maximise good. It is a concept in research ethics which implies that the research should be of benefit to all participants and to the wider general public and should above all do no harm to any individual or group. Beneficence implies 'to do good' and above all else to never impose any risk or harm on a participant. The IDS-TILDA study research team made it clear to all participants that the outcome of the study would not directly impact on them but would have positive outcomes for the future of people with ID living in Ireland. The study would provide detailed information for an ageing population with ID and how services are responding to the increasing needs of this cohort. Participants were informed of how the study was progressing and how their participation and the information they provided was used to inform policy and future planning. Newsletters and cards were sent throughout the year to ensure participants were made feel appreciated, respected and a valued member of the IDS-TILDA team which incorporated the 'keeping in touch strategy'.

3.3.3 Non-Maleficence

Freedom from harm is paramount in any research study and while the physical care aspects of research are more often obvious, the psychological consequences of participating in a study must also be recognised (Polit *et al,* 2001). Harm includes: "*not only hazardous medical experiments but also any social research that might involve such*

things as discomfort, anxiety, harassment, invasion of privacy, or demanding or dehumanising procedures" (Bailey, 1978, 113). This is well illustrated by Parahoo (2014) who notes that research participants will often experience more attention and 1:1 human contact during the research interviewing process, something which can be regarded as very positive for the individual, but once the study is complete, the participant may feel secluded, which could have negative consequences. It is through the 'Keeping in Touch strategy' employed by the IDS-TILDA study that these potential difficulties are overcome.

3.3.4 Confidentiality

Building on a relationship of trust between the participant and the researcher is vital. Polit *et al* (2001) notes that "*a promise of confidentiality is a pledge*" that all participant information will be kept secure and will not be shared publicly. Personal information gathered from all participants for the study must be kept confidential, and a person's identity is never published. If the researcher is put in a position whereby, they feel the participant is at risk in some way they must put the participants' best interest to the fore, even if this means compromising the objective of the study. The outright safety of the participant is paramount. In all material provided to the participants in the study the principle of confidentiality is emphasised. All data collected is on a double encrypted laptop and participants are reassured that at no time will their name or the name of the service they attend be published directly related to the information they provide. All data is pseudonymised within the dataset as each participant is assigned a PIN for the purposes of the interview. The key to this PIN linking it with the participant name is held on a secure server within Trinity College and no one outside the data manager, project manager and PI has access to this key.

3.3.5 Informed Consent

Informed consent ensures that participants have the correct information, are well informed and have a clear understanding about the research. It is a process for getting permission before conducting a healthcare intervention on a person or disclosing personal information. Considering the Assisted Decision-Making Capacity Act (2015), and its implications for people with ID, this legislation reinforces the case to make certain that people with ID are actively supported to make their own decisions where

they can. Initially participants received an invitation letter, an information booklet about the study, a consent form and a letter of support from their service provider. All this material was furnished in easy to read accessible format. The invitation pack also contained a family information booklet and agreement form. Ethical approval and consent have been reiterated with all participants at each Wave of data collection. The study also uses a system of process consent throughout the interview duration, which means reiteration of consent with the participant as each section of the questionnaire commences. Thereby reassuring the participant they have a right to withdraw at any time without consequences.

Informed consent is said to have been given according to guidelines from the fields of medical and research ethics. Informed consent can be assumed based upon a clear understanding of the facts and implications of the study and the basic elements of the consent process include:

- Full disclosure regarding the nature of the research and how this will impact on the participant
- Adequate comprehension and understanding of the research
- The participants ability to consent to participate in the study
- The participant's voluntary choice/consent to participate on their own volition.

(Burns & Grove, 2003)

Consent to participate in Waves 1, Wave 2 and Wave 3 of the IDS-TILDA was recorded on the day of the interview. The field researcher used a battery of easy read materials to explain the study, the interview process and then to obtain consent. Many people with ID may find it difficult to indicate their willingness to participate in the research process and provide consent and in these instances, a key support person or family member was asked to act as proxy and delegate the preference of the individual as to their willingness to agree to participate in the IDS-TILDA study. Throughout the 3 waves of IDS-TILDA, process consent was carried out to ensure reaffirmation throughout the interview process, and ensuring that the participant is kept well informed at all stages through the interview.

3.3.6 Data Protection

The researcher at all times abided by the strict data protection protocol set out by the IDS-TILDA study. The study uses a PIN system to maintain confidentiality of the participant data and all data is uploaded following completion of the interview onto an encrypted server held within Trinity College. The researcher completed this step at the end of each interview and all hard copy material was securely couriered back to the IDS-TILDA office for secure inputting and storage. All hard copies of information, the PIQ and the consent form, are stored separately in locked cabinets within a secure storage room in the IDS-TILDA offices. These were available to the researcher for data cleaning purposes however with permission and under the supervision of the data manager thus ensuring that confidentiality is maintained. The researcher was provided with a double encrypted laptop with the encrypted software, CAPI to conduct all the interviews. No information about the participant's identity is ever published in any reports of the study, in accordance with the Data Protection Act (2018).

At no time did the researcher have full access to the complete IDS-TILDA dataset. The researcher completed a data request form (see Appendix 12) which was reviewed by the data manager and the PI. The data request outlined the data required and for what analytical purpose. The researcher in collaboration with the data manager identified the variables on a separate excel spread sheet. These variables were then provided to the research in a secure folder on the IDS-TILDA secure drive. The researcher had only access to her own folder and no other. The data folder was only accessible within the IDS-TILDA office and duplication or creating copies on an external USB drive was not permitted. The researcher signed an agreement form to maintain these principles (see Appendix 13). Data protection guidance was provided by the IDS-TILDA team and the research completed this training which ensured all aspects of data protection were maintained at all times. For the purposes of this study, the researcher abided by the data protection protocol of the IDS-TILDA study which takes into consideration the new GDPR regulations (see Appendix 14).

3.4 Statistical Analysis

An outline of the statistical analysis plan is described here to explain how the data was analysed using Wave 1, Wave 2 and Wave 3 of the IDS-TILDA dataset. Quantitative research data is measured by statistical analysis which include the use of a statistical computer programmes (Polit & Beck 2016). In this study the software computer program used is Statistical Package for the Social Science (SPSS) Version 22 (Pallant 2013, Kinnear& Gray 2009, Field 2009). This package allows the study to perform a wide range of statistical operations suitable to analysis its data. There are three basic steps used in analysing data from the SPSS program (Pallant, 2013)

1. Data Entry, as this was automatically conducted by the CAPI system the researcher did not have to engage in this aspect. However, the research contributed to the entry of the PIQ data at Wave 2. This step also entails the data preparation, data cleaning detailed below.
2. Analysis, described in detail below.
3. Inspection of Results.

3.4.1 Data preparation and cleaning

The aim of the statistical analysis in this study was to ascertain key independent variables and model the associations among those variables. The statistical analysis in this study describes:

1. The baseline health profile of people with DS at Wave 1 (2011) and follow up through Wave 2 (2014) and Wave 3 (2017), focusing on the more prevalent health conditions identified in people with DS.
2. How the changes from baseline health impact on functional ability and living circumstance.
3. And the examination of the expected survival rate of those with DS over the age of 40-years during the 10-year period.

All data was analysed using the statistical computer package SPSS version 22 (SPSS Inc., 2014). Continuous variables were summarized using descriptive statistic including means, standard deviations, range and median. Categorical data was summarized using frequency counts and percentages. For all tests a p-value below 0.05 was considered significant. Missing data was taken into consideration by presenting only valid percentages throughout the analysis. Overall relationships between variables was explored through the application of chi-squared test of independence, stratified by age, gender, level of ID.

3.4.2 Identifying the variables

Utilising the conceptual frame developed from the literature the concepts required were mapped to the IDS-TILDA data available over the 3 waves of data collection. The following provides an overall description of the variables analysed within this study mapped from the IDS-TILDA study;

Table 3.1 Identifying study variables among IDS-TILDA and identifying data transformation

Exploring Physical Health Conditions among those with DS	**IDS-TILDA Equivalent Data** [Unless otherwise stated these questions were asked at Waves 1,2 3]	**Study data questions and transformation applied** Identify prevalence/incidence/scoring
Down Syndrome	What caused your intellectual disability – Down syndrome, cause unknown, don't know, other.	Down syndrome; Yes or No
Level of intellectual disability	What is your level of intellectual disability – Not verified, mild, moderate, severe, profound, don't know.	Categorised into 3 groups Mild; Moderate; Severe/Profound
Older age	What is your date of birth	Categorised into 3 groups 40-49 years; 50-64 years and 65 years+
Gender	Male; Female [tick box option]	Male or Female
Overall health	Self rated health: Is your health - Excellent, Very good, Good; Fair and Poor	Excellent/very good/good=Positive rating Fair/Poor = Negative rating
Dementia	Doctors diagnosis of Dementia/Alzheimer's	Has the doctor ever told you… you had dementia? Yes or No
Sensory Impairment	Doctors diagnosis of Cataracts	Has the doctor ever told you… you had cataracts? Yes or No
	Self-reported eyesight; Is your eyesight - Excellent, Very good, Good; Fair and Poor	Excellent/very good/good=Positive rating Fair/Poor = Negative rating
	Self-reported hearing; Is your hearing - Excellent, Very good, Good; Fair and Poor	Excellent/very good/good=Positive rating Fair/Poor = Negative rating
	Have you ever had cataracts Y/N Have you ever had cataract surgery Y/N	Have you ever had cataracts Y/N Have you ever had cataract surgery Y/N
Epilepsy	Doctors diagnosis of epilepsy	Has the doctor ever told you… you had epilepsy? Yes or No
Thyroid condition	Doctors diagnosis of thyroid disease [Wave1]	Has the doctor ever told you… you had thyroid disease? Yes or No
	Doctors diagnosis of hypothyroidism [Wave 2, 3]	Has the doctor ever told you… you had hypothyroidism? Yes or No
	Doctors diagnosis of hyperthyroidism [Wave 2, 3]	Has the doctor ever told you… you had hyperthyroidism? Yes or No
Communication challenges	How well are you able to make yourself understood? No difficulty/ Some difficulty/Much difficulty/Cannot do at all	How well are you able to make yourself understood? No difficulty/ Some difficulty/Much difficulty/Cannot do at all
	Are you understood by healthcare professionals? Completely/ Partially/Not at all	Are you understood by healthcare professionals? Completely/ Partially/Not at all

Assessment/accessing health services usage	Healthcare utilisation In the last year have you attended any of the following services GP; optician services, hearing services, Endocrinology services or neurological services	Have you attended any of the following services for your health condition: General practitioner Endocrinologist Neurologist Ophthalmologist Auditory specialist
Dementia assessment	Have you ever been screened or assessed for memory impairment/dementia?	Participants were asked if they had an assessment for dementia with the following response options: 1. Yes, within the last 2-years 2. Yes, over 2-years ago 3. N0
	Test for Severe Impairment (TSI	TSI Score – mean score
Activities of daily living difficulty	Please indicate the level of difficulty you have with Bathing, dressing, getting in and out of bed, No difficulty, some difficulty, a lot of difficulty, cannot do at all	Recategorized to a dichotomous variable No difficulty – cat.1 No difficulty Some/a lot of difficulty/cannot do at all – cat. 2 Difficulty
Transitions	Tell us if you have moved home since your last interview. From where did you move?	Moves categorised into Lateral move: participant moved from one type of setting to a similar type of setting for example from one community house to another Less supported move: participants moved from a more supported setting to a less supported setting for example moved from an institution or residential care setting to a community home independently or as a group. More supported setting: participant moved from a more independent setting to a setting requiring more support for example from a community group home to fully supported dementia setting.
	What was the reason for the move? People were offered a selection – physical health changes, staff shortages. Etc	All options used to describe reasons for moving between Waves 1 and 2, and between Waves 2 and 3.

3.4.3 Testing for longitudinal change

Following on from this the three waves of data was used to identify change over time of the impact of ageing on the health trajectories of the sample. McNemar's test was used to compare paired nominal data or paired proportions. It tests for consistency in response across two points in time. McNemar's test compares the proportions for two correlated dichotomous variables. These two variables may be two responses on a single individual at two different time points. McNemar's test was used for within-subject designs where the **change of a dichotomous categorical baseline measure is assessed across two time points or two within-**subjects' **observations**. With McNemar's test, the proportion of individuals that switch from one level to the other across time dictates statistical significance. See figure 3.x which provides a graphical representation of the McNemar's test.

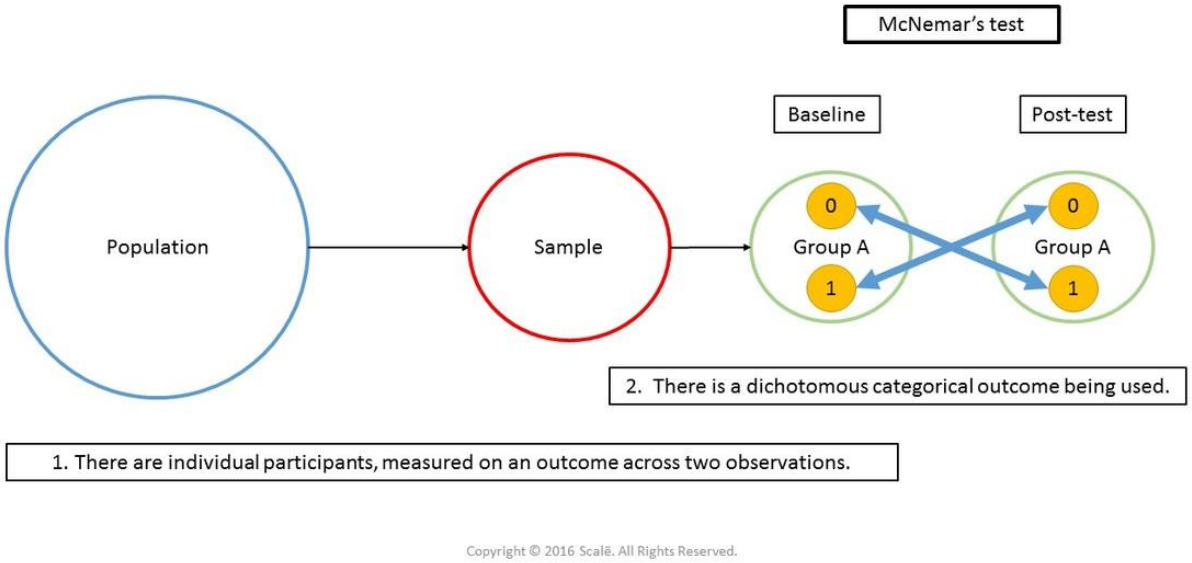


Figure 3.3 McNemar's Test graphical representation
Source: https://www.scalelive.com/mcnemars.html

McNemar's assumptions

The McNemar's test has three assumptions that must be met.

- Assumption **1:** There is **one categorical dependent variable with two categories** (i.e., a **dichotomous** variable) and one **categorical independent variable with two related groups**
- Assumption **2:** The two groups of the dependent variable must be **mutually exclusive**.
- Assumption **3:** The cases (e.g., participants) are a random sample from the population of interest.

*Therefore the following hypothesis are applied***:**

Null hypothesis H_0: *The **null hypothesis** is that the percentages or proportions at Wave 1 and at Wave 3 are equal in the population.*

Alternative Hypothesis H_A: *The **alternative hypothesis** is that the percentages or proportions at Wave 1 and at Wave 3 are not equal in the population.*

3.4.4 Test for differences in paired proportions between Wave 1 and Wave 3

The Paired sample *t* Test

The Paired Samples *t* Test compares two means that are from the same individual, object, or related units such as a measurement taken at two different times (e.g., pre-test and post-test with an intervention administered between the two time points). The TSI is administered in each wave, and therefore a paired t test is used to test for significant differences in scores between two waves. A significant result is determined by a p-value < 0.05. The null hypothesis is one of no difference and the alternative is that there is a difference in scores across two waves.

3.4.5 Survival Analysis

Survival analysis is concerned with data where the time to an event is measured and the outcome of interest is time to that event. It relates the time taken for an individual to reach a certain event. The participants with DS in IDS-TILDA were followed over a 10-year period and the survival time is considered in terms of the duration of the study, i.e. 10-years. The recorded observations are 'censored' when the information regarding survival time is incomplete, referred to as 'right censoring' whereby the survival time for

the person is as long as the study. Censoring means that an individual is still alive at the end of the study or that they withdrew at some point during the study. If the person drops out of the study before same is complete, they are considered 'right censored' as the event (death) did not happen for them during their course of observation. When examining survival analysis for this study, censoring is important as it represents missing data (Kleinbaum, 1996).

3.4.5.1 Kaplan-Meier

The Kaplan-Meier method (Kaplan & Meier, 1958), also known as the "product-limit method", is a nonparametric method used to estimate the probability of survival past given time points (i.e., it calculates a survival distribution). Furthermore, the survival distributions of two or more groups of a between-subjects factor can be compared for equality.

The time starting from a defined point to the occurrence of a given event, for example death, is called as survival time and the analysis of group data as survival analysis. This can be affected by subjects under study that are uncooperative and refused to be remained in the study or when some of the subjects may not experience the event or death before the end of the study, although they would have experienced or died if observation continued, or we lose touch with them midway in the study. We label these situations as censored observations.

The Kaplan-Meier estimate is the simplest way of calculating the survival over time in spite of all the difficulties associated with participants or situations. The survival curve can be produced presuming numerous situations. It involves calculating of probabilities of incidence of event at a certain point of time and multiplying these successive probabilities by any earlier computed probabilities to get the final evaluation. This can be calculated for two groups of subjects and also their statistical difference in the survivals (Goel *et al.* 2010).

The survival probability at any particular time is calculated by the formula given below:

$$S_t = \frac{\text{Number of subjects living at the start} \_\_ \text{Number of subjects died}}{\text{Number of subjects living at the start}}$$

Goel *et al* (2010)

A Kaplan-Meier plot displays survival probabilities (cumulative probability of an individual remaining alive at any time). The cumulative survival probability is the product of the survival probabilities up to that point in time.

3.4.5.2 Assumptions for the Kaplan-Meier method

1. Censoring is unrelated to the outcome. The Kaplan Meier method assumes that the probability of censoring is not related to the outcome of interest.

2. The survival probabilities are the same for participants recruited early and late in the study. Circumstances that can affect survival (better or different treatments) are not assumed to change the baseline risk of survival in the patients as a group.

3. The events occurred at the times specified. However, sometimes we do not know the exact date of an event, but only its status at each visit.

3.5 Conclusion

IDS-TILDA is a unique, longitudinal study of ageing among people with an intellectual disability. Over 750 people with ID, representing 8.9% of the ID population, over the age of 40-years consented to participate in the study. It provides data on the health, social, economic and environmental circumstances of the study participants and the study design facilitates direct comparison of health characteristics and status of persons with intellectual disability with those of the general population study (TILDA). The study

commenced in 2007. An integral component of IDS-TILDA is the health and well-being of people with DS. This research is positioned within the IDS-TILDA study taking into consideration data collected at 3 time points from 2007 – 2017, a ten-year period.

This chapter has provided an overview of the IDS-TILDA study and a detailed description of the research methodology adopted for this study by the researcher. The sampling frame and the sample for this study has been presented as well as the data collection methods and the data analysis to be applied. Clear description of the ethical process and principles applied in this study have been described along with the data protection processes applied. The chapter concludes with the statistical analysis applied in this study, the choosing of the variables working with the IDS-TILDA conceptual frame in conjunction with the researcher's conceptual frame developed from the literature, the data preparation, cleaning and the tests applied to answer the research questions. In the next chapter, chapter 4 the findings for this analysis will be presented.

Chapter 4

Results

4.0 Introduction

Chapter 4 reports the findings of the study with regards to the specific aims and objectives outlined in Chapter 2. The Statistical Package for Social Science (SPSS) version 22 was used for all analysis. The applicable data from Wave's 1, 2 and 3 of the study were analysed and prior to conducting the analysis, the data was examined for outliers and irregularities, by use of descriptive statistics. Only valid percentages are presented to account for missing data. The results are presented in the following sections:

Section 4.1 An overview of the demographics of the participants across the three waves of data collection from Wave 1 [2011], to Wave 2 [2014] ending with Wave 3 [2017]. For the purposes of description, the years identified are the years the data collection ceased.

Section 4.2 Presents an overview of the self-reported health of those with DS across the three waves of data collection.

Section 4.3 Presents the prominent chronic health conditions as identified within the literature. The areas of dementia are explored in relation to prevalence and incidence. The occurrence of dementia is then stratified by gender, age and level of ID. Considering the cognitive assessment conducted in this study the TSI results are then presented. The incidence and prevalence of epilepsy is presented as well as epilepsy stratified by the three co-variates, gender, age, level of ID. The incidence and prevalence of thyroid disease *across* the three waves with hypothyroidism and hyperthyroidism denoted at waves 2 and 3. Thyroid disorder is examined and presented stratified by gender, age, level of ID. *S*ensory impairment is explored and stratified by the three co-variates, gender, age, level of ID.

Section 4.4 This section presents an overview of the functional limitations' individual's experience.

Section 4.5 The section explores the movement in living circumstances reported by individuals with DS in this study and why they moved.

Section 4.6 Examines healthcare utilisation by individuals with DS in this study.

Section 4.7 This section examines the communication difficulties individuals have and how clearly they make themselves understood to healthcare professionals.

Section 4.8 Using the McNemar's test this section examines change over time

Section 4.9 Examines attrition and survival over the 10-year period of the study.

4.1 Demographic profile of participants

The overall profile of the participants was examined by gender, age range, level of ID and living circumstances. As outlined in Table 4.1 at Wave 1 the mean age of this cohort was 49 years (±6.4 yrs.). This had risen to 53.8 years (±5.3 yrs.) at Wave 3. Considering the age categories, the majority at Wave 1 were in the younger age bracket with 56.5% (n=83/147) aged between 40-49 years and by Wave 3 this had changed to having the majority in the middle category (50-64 years) (73.4%, n=80/109). This would be expected considering it was 10 years later however what is quite noticeable and interesting is that the older age group only shows a slight change across the three Waves from 1.2% (n=2/147) to 3.4% (n=4/109) at Wave 3. Across all waves there were more females than males in the study with 56.5% (n=83/147) at Wave 1, 55.8% (n=77/138) at Wave 2 and 51.4% (n=56/109) at Wave 3. Whilst there is a difference of 19 between males and females (n=64 males versus n=83 females) at Wave 1 by Wave 3 this difference is substantially reduced to 3 individuals (n=53 males versus n=56 females respectively). The greatest proportion of people with DS participating in the study have a moderate level of intellectual disability followed by severe/profound level of ID and finally participants with a mild level of ID are lowest which is reflective of the overall sample of the IDS-TILDA study. In Wave 1 the largest number of participants lived in community group homes (CGH) at 42.5% (n=62/147), while Residential placement supported 37.7% (n=55/147). The numbers living independently or being supported by family members (parents or siblings) were 19.9% (n=29/147). At Wave 2, and similar to Wave 1 the largest number of participants lived in CGH representing 48.6% (n=67/138), while residential placement supported 31.9% (n=44/138). The numbers living independently or being supported by family members were 19.6% (n=27/138). There was little evidence of movement from one type of residence to another to support increasing age or increased level of need. At Wave 3, similar to Wave 1 & 2, the largest number of participants lived in CGH representing 46.8% (n=51/109), while residential placement supported 33.9% (n=37/109). The number of participants living

independently or being supported by family members (parents or siblings) were 19.3% (n=21/109).

Table 4.1 Demographics of sample over three waves of data collection

			Wave 1				***Wave 2***				***Wave 3***
			N = 147	N=109* at wave 1			N = 138	N=109* at wave 2			N = 109
Age (Years)											
Mean (yrs.)			49	47.9			51.5	50.5			53.8
St Dev (yrs.)			6.4	5.3			5.7	5.3			5.3
Median (yrs.)			48	46			50	49			52
		%	***(n)***	***%***	***(n)***	***%***	***(n)***	***%***	***(n)***	***%***	***(n)***
Age											
	<50	56.5	(83)	66.1	(72)	47.8	(66)	56	(61)	22.9	(25)
	50-64	42.2	(62)	33	(36)	49.3	(68)	42.2	(46)	73.4	(80)
	65+	1.4	(2)	0.9	(1)	2.9	(4)	1.8	(2)	3.7	(4)
Gender											
	Male	43.5	(64)	48.6	(53)	44.2	(61)	48.6	(53)	48.6	(53)
	Female	56.5	(83)	51.4	(56)	55.8	(77)	51.4	(56)	51.4	(56)
Level of ID***											
	Mild	14.7	(20)	17.8	(18)	15	(19)	17.8	(18)	17.8	(18)
	Moderate	55.1	(75)	57.4	(58)	55.9	(71)	57.4	(58)	57.4	(58)
	Severe/Profound	30.1	(41)	24.8	(25)	29.1	(37)	24.8	(25)	24.8	(25)
Living Circumstances[>]											
	Indep. / Family	19.9	(29)	22.9	(25)	19.6	(27)	21.1	(23)	19.3	(21)
	CGH	42.5	(62)	48.6	(53)	48.6	(67)	55	(60)	46.8	(51)
	Residence	37.7	(55)	28.4	(31)	31.9	(44)	23.9	(26)	33.9	(37)

**Survivors across all waves N=109*

***Not all participants provided their level of ID (observations missing 11/147 and 8/109)*

> Not all participants provided their living circumstances (observations missing 1/147)

4.2 Self-reported health of those with DS from Wave 1 to Wave 3

Self rated health was described as being excellent/ very good/ good or as being fair/poor. People were asked at each wave to self report their health. As can be seen in table 4.2 the majority of people across all waves rated their health as either excellent, very good or good. Wave 1 through Wave 3 (Wave 1: 81.5% (n=119/147); Wave 2: 73.7% (n= 101/138); Wave 3: 77.6% (n=83/109)). However this positive rating decreased with time and conversely the percentage who reported their health as fair or poor increased over the three waves from 18.5% (n=27/147) to 22.4% (n=24/109) .

Table 4.2 Self reported health among those with Down syndrome across waves 1,2 and 3

Self rated health	Wave 1 N = 147		Wave 1 (survivors to Wave 3) N=109		Wave 2 N = 138		Wave 2 (survivors to Wave 3) N=109		Wave 3 N = 109	
	%	n	%	n	%	n	%	n	%	n
Positive	81.5	119	87	94	73.7	101	83.3	90	77.6	83
Negative	18.5	27	13	14	26.3	36	16.7	18	22.4	24

4.3 Prominent chronic health conditions

4.3.1 Dementia of the Alzheimer's type

As outlined in *figure 4.1* the number of participants in Wave 1 with a doctor's diagnosis of dementia (of the Alzheimer's type) represented 15.8% (n=23/147). In Wave 2, the numbers of people with DS and dementia had almost doubled to 29.2% (n=101/138), and in Wave 3, from a total sample of 109 participants with DS, 35.5% (n=38/109) reported a Doctor's diagnosis of dementia.

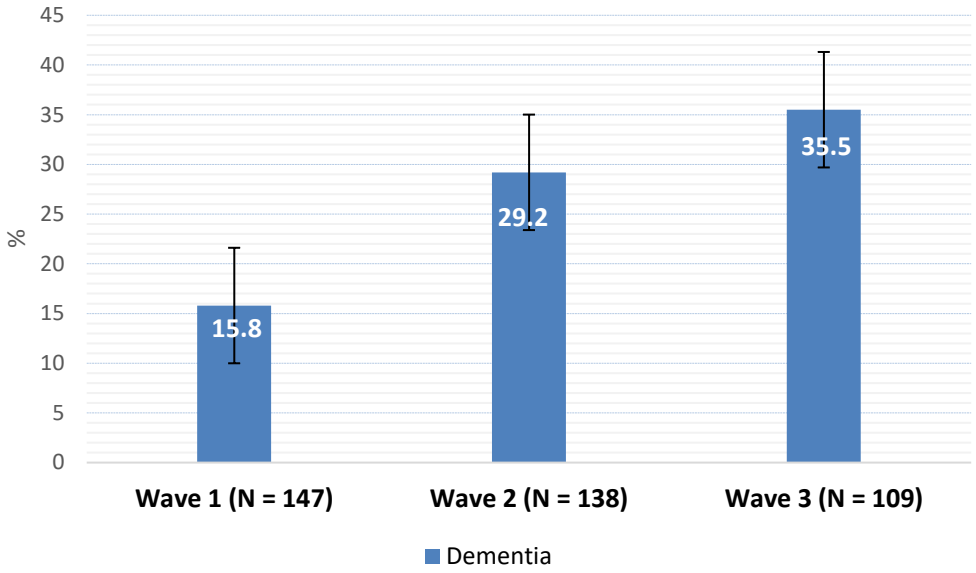


Figure 4.1 Prevalence of dementia among those with DS across three waves

Examining the incidence rate, the number of new cases between waves 1 and 2 was 15.3 per 100 (n=21) and for the 109 who survived across the three waves the incidence was identified as 7.4%. Between waves 2 and 3, the observed new cases were 18.7 per 100 (n=20). This is graphically presented in *figure 4.2* below.

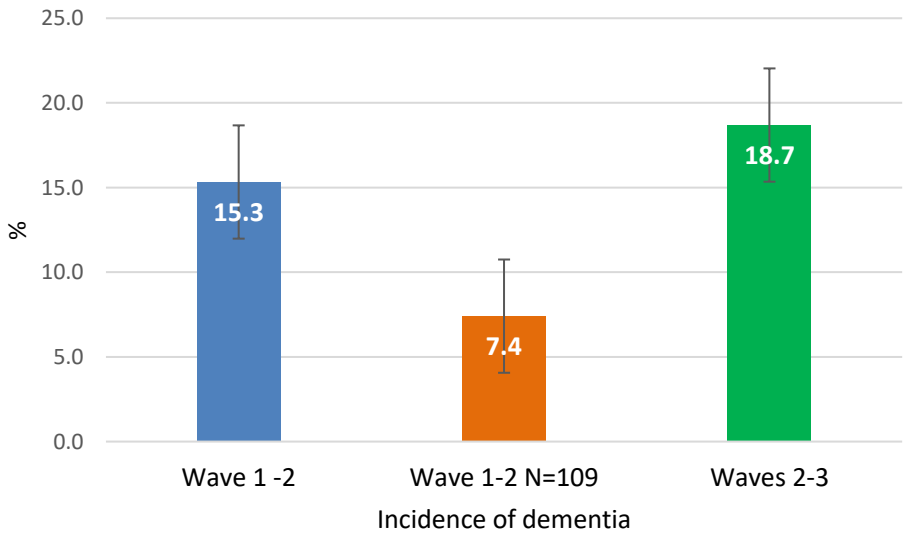


Figure 4.2 Incidence rate of dementia across the three waves of data

4.3.2 Presence of doctor's diagnosis of dementia stratified by gender, age and level of ID

Those who reported a doctor's diagnosis were examined with consideration to their gender, age and level of ID. It can be seen in **Table 4.3** that overall females reported a higher prevalence than males across the waves. However, it is evident as individual's age, no matter what their sex, the prevalence increases. It is notable that from Wave 1 to Wave 3, the prevalence of dementia for females doubled, while the prevalence in men tripled from 10.9% to 30.8% from Wave 1 to Wave 3. With consideration to age, the highest prevalence is observed among those in the middle age group (50-64 years) across the waves (Wave 1: 26.2% (n=16/147); Wave 2: 48.2% (n=27/138) and Wave 3: 55.6% (n=20/109). This ageing trajectory is interesting considering that there were no individuals who reported a doctor's diagnosis over the age of 65 years. With respect to level of ID in Wave 1, those who had a doctor's diagnosis were predominantly within the severe/profound category (24.4%, n=10/147) and this figure almost doubled by Wave 3. In the other categories, with respect to level of ID, substantial increase in the prevalence of dementia also occurred. This is particularly evident within the mild level of ID category which showed an increase from 5% (n=1/147) in Wave 1 to 33.3% (n=6/109) by Wave 3. Please see full stratification in **table 4.3**.

Table 4.3: Presence of doctor's diagnosis of dementia stratified by gender, age and level of ID.

Dementia		Wave1 N = 147		Wave 1 (survivors to Wave 3) N=109		Wave 2 N = 138		Wave 2 (survivors to Wave 3) N=109		Wave 3 N = 109	
		%	*n*	*%*	*n*	*%*	*n*	*%*	n	*%*	*n*
Gender	Male	10.9	7	5.7	3	21.3	13	15.1	8	30.8	16
	Female	19.5	16	8.9	5	35.5	27	18.2	10	40	22
Age	40-49	8.4	7	5.6	4	16.3	13	9.9	7	25.7	18
	50-64	26.2	16	11.1	4	48.2	27	30.6	11	55.6	20
	65+	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0
Level of ID	Mild	5	1	5.6	1	15.8	3	11.1	2	33.3	6
	Moderate	12.2	9	6.9	4	25.7	18	17.5	10	33.9	19
	Severe/Profound	24.4	10	12	3	40.5	15	20	5.0	40.0	10

4.3.2.1 Assessment for memory impairment/dementia among those with DS and no doctor's diagnosis of dementia.

The data was examined for those who did not have a diagnosis of dementia to establish if they had received a cognitive assessment in the last two year, as recommended, to screen for dementia (Burt and Aylward, 2000). As can be seen in **Table 4.4,** of those who did not have a diagnosis of dementia at Wave 1, 75.7% (n=56/123) did not have a cognitive assessment in the last 2-years (consisting of 8.1% who had an assessment over two year ago and the remaining 67.6% (n=50/123)never having had a cognitive assessment). Of those who did not have a diagnosis of dementia at Wave 2, 63.4% (n=57/97) did not have a cognitive assessment in the last 2-years (7.8% having had an assessment but over two years ago and 55.6% (n=50/97) never had a cognitive assessment.) Finally, of those in Wave 3 who did not have a diagnosis of dementia 58.9% (n=30/69) did not have a cognitive assessment in the last 2 years (11.8% had an assessment over two year ago and 47.1% (n=24/109) never had a cognitive assessment).

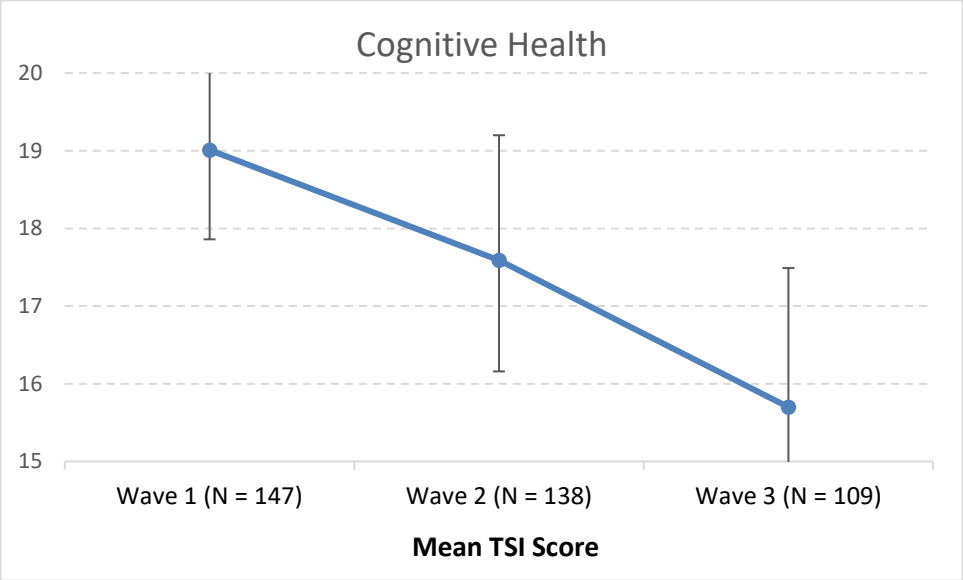
Table 4.4 Cognitive assessment of those with DS who did not have a doctor's diagnosis of dementia

Did you have a dementia assessment	Wave 1 N=147		Wave 1 (survivors to Wave 3) N=109		Wave 2 N=138		Wave 2 (survivors to Wave 3) N=109		Wave 3 N=109	
	%	n	%	n	%	n	%	n	%	n
Yes, within last 2-years	24.3	18	22.6	14	36.7	33	34.9	29	41.2	21
Yes, over 2-years ago	8.1	6	4.8	3	7.8	7	8.4	7	11.8	6
No	67.6	50	72.6	45	55.6	50	47	56.6	47.1	24

4.3.3 Test for severe impairment (TSI) scores Wave 1 through Wave 3 with or without a doctor's diagnosis of dementia.

The scores described in *figure 4.3* below highlight the TSI score which noted imperceptible decline from Wave 1 through to Wave 3 in the cohort of all the participants with DS over the 10-year period, stratified by those with or without a

doctor's diagnosis of dementia. At Wave 1, the mean score was 19.01 (SD±5.69). At Wave 2 the mean score dropped to 17.59 (SD± 5.93). At Wave 3 the mean score dropped further to 15.70 (SD± 7.50). The decline in score on the TSI over the 10-year follow-up indicates a 4-point drop from 19 to 15. These figures however do not differentiate between the population with DS with a doctor's diagnosis dementia and without a doctor's diagnosis of dementia.



Not all individuals did the TSI assessment Missing obs: n=51 at W1; n=68 W2; n= 39

Figure 4.3 TSI Scores for all participants with DS

Examining and comparing the scores of those with dementia and those without dementia the decline in score appears more noteworthy. See *figure 4.4* for those with and without doctor's diagnosis of dementia compared. The mean score for people without a doctor's diagnosis of dementia (n=35/123) at Wave 1 was 21.1(SD± 2.93). The TSI score declined to 20.3 (SD ±3.2) at Wave 2 and declined further to 19.3 (SD± 5.3) at Wave 3. Examining the scores of those people with DS and a doctor's diagnosis of dementia who completed the TSI across all three waves (n=11), the decline in the TSI score is particularly interesting compared to the cohort with no diagnosis. People with DS and dementia presented with a score of 21.6 (SD± 1.91) at Wave 1 and this score declined to 18.8 (SD± 4.04) at Wave 2. Over the following 3-years the score for this

cohort declined to 14.1 (SD± 8.4). Also the variability of scores increased as shown by the SD.

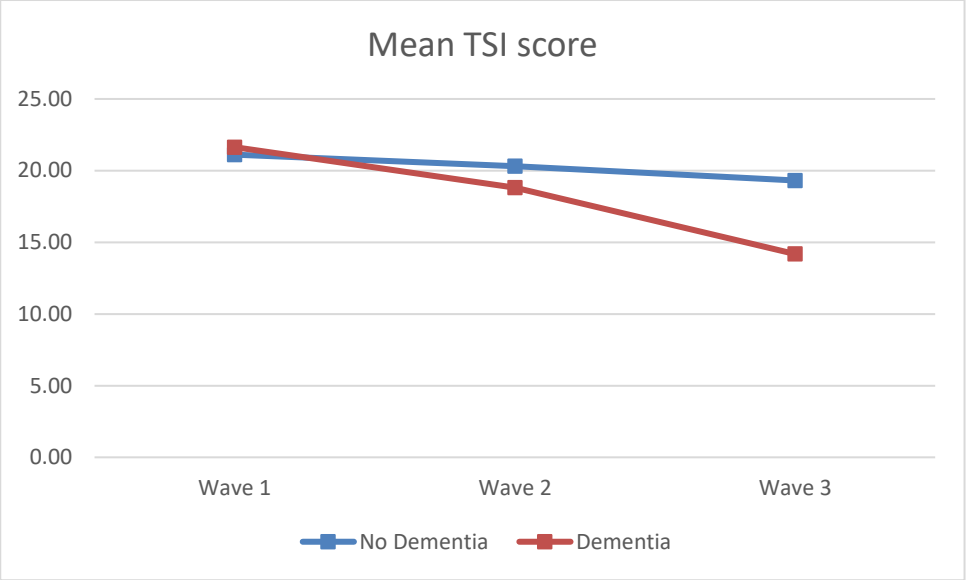


Figure 4.4 TSI Scores comparing those with and without Dementia

Considering the age of onset of dementia people were asked at what age did they received a doctor's diagnosis. The mean age of onset was 51.6 years (SD± 5.94) with a median age of 51 years and a range of 34-63 years for all those diagnosed across the three waves who reported an age of onset. See *figures 4.5 and 4.6* for overview of age of onset.

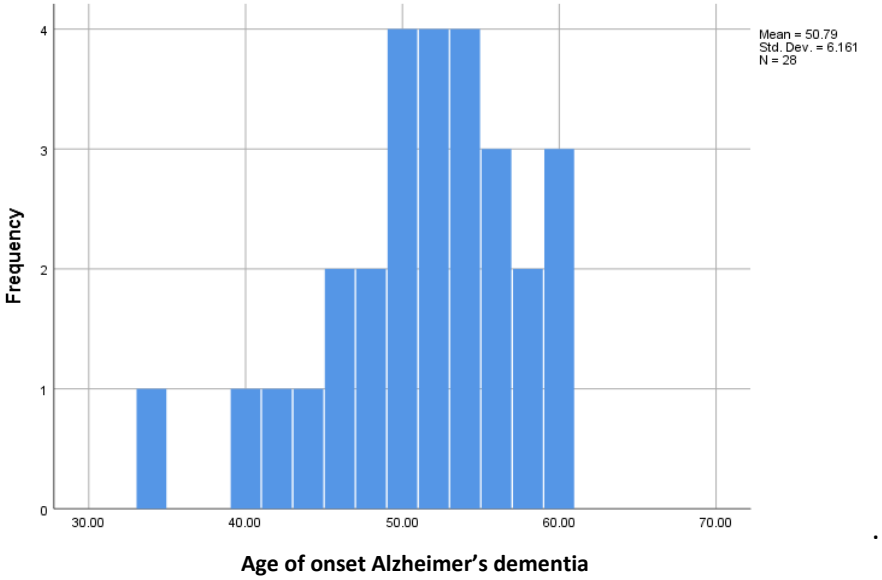


Figure 4.5 Frequency of age of onset among those who reported at Wave 1

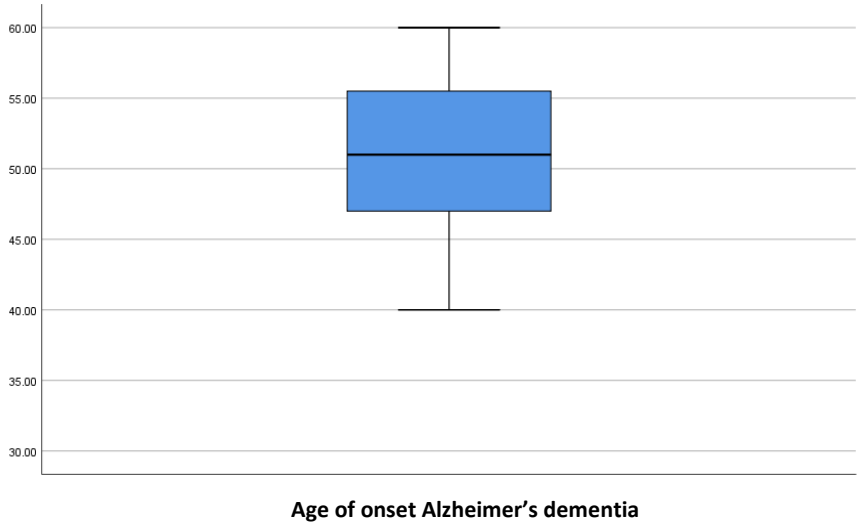


Figure 4.6 Mean age of onset of dementia

4.3.4 Prevalence and incidence of epilepsy

People were asked to identify if they had epilepsy. In total, the number of participants at Wave 1 with DS and epilepsy was 19.2% (n=28/147). By Wave 2 this had increased to 27.7% (n=38/138), and at Wave 3, from a total sample of 109 participants with DS, 28%

(n=30/109) had a diagnosis of epilepsy. See figure 4.6 for a graphical representation of the prevalence of epilepsy among those with DS across the three waves of data collection [2007-2017].

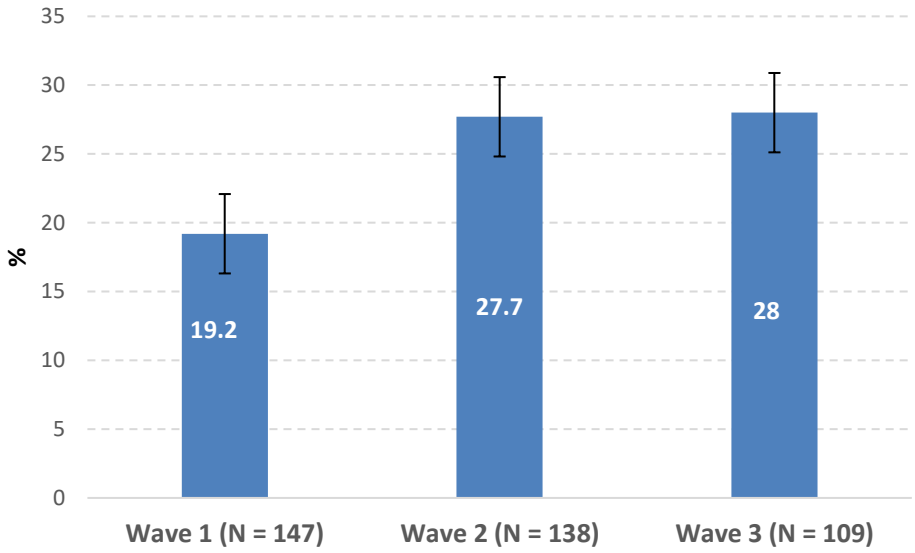


Figure 4.7 Prevalence of epilepsy across Wave1, 2 and 3 among those with DS

Considering the incidence of epilepsy across the three waves of data collection there was a 10.3% increase between waves 1 and 2 which represented 10 new cases and between waves 2 and 3 a further 11.5% increase was observed representing new cases. Considering those who survived across all waves there was a 4.7% increase representing 5 new cases (from Wave 1 – Wave 2). See *figure 4.8* below for an overview of the incidence of epilepsy across the three waves of data collection.

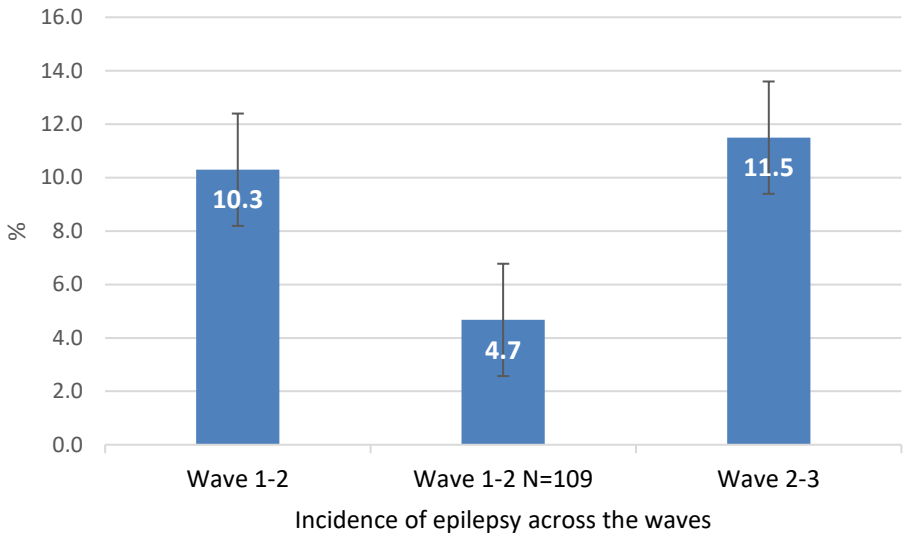


Figure 4.8 Incidence of epilepsy among those with DS across Waves 1,2 and 3

4.3.5 Epilepsy among those with DS stratified by gender, age and level of ID

Those who reported a doctor's diagnosis of epilepsy were examined with across gender, age and level of ID. It can be seen in **Table 4.5,** that similar to dementia, overall females reported a higher prevalence of epilepsy than males across the 3 waves. However, it is evident as individual's age no matter what their sex the prevalence of epilepsy increases. With consideration to age the highest prevalence of epilepsy is observed among those in the middle age group (50-64 years) across all waves [Wave 1: 29% n= (18/147); Wave 2: 42.9% n= (24/138) and Wave 3: 44.4% n= (16/109)]. Participants surviving to the age of 65 years and over had no diagnosis of epilepsy. With respect to level of ID, the highest prevalence of epilepsy is observed in those with a severe/profound level of ID across all 3 Waves [Wave 1: 31.7% n= (13/147); Wave 2: 48.6% (n=18/138) and Wave 3: 52% n=(13/109)]. Those with moderate level of ID demonstrated a substantial increase between Wave 1 and Wave 2 with a prevalence of 14.9% (n=11/147) at Wave 1 and this figure increased to 47.1% (n=33/138) at Wave 2. The association between increasing age, level of ID and gender, and a diagnosis of epilepsy in people with DS across all three waves of data collection is described in **Table 4.5** below.

Table 4.5 Epilepsy among those with DS stratified by gender, age and level of ID

Epilepsy		Wave1 N = 147*		Wave 1 (survivors to Wave 3) N=109		Wave 2 N = 138		Wave 2 (survivors to Wave 3) N=109		Wave 3 N = 109	
		%	*n*	*%*	*n*	*%*	*n*	*%*	n	*%*	*n*
Gender	Male	17.2	11	18.9	10	21.3	13	20.8	11	26.9	14
	Female	20.7	17	8.9	5	32.9	25	16.4	9	29.1	6
Age	40-49	12	10	11.1	8	17.5	14	14.1	10	20	14
	50-64	29.5	18	19.4	7	42.9	24	27.8	10	44.4	16
	65+	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0
Level of ID	Mild	10	2	11.1	2	36.8	7	16.7	3	16.7	3
	Moderate	14.9	11	10.3	6	47.1	33	12.3	7	19.6	11
	Severe/Profound	31.7	13	28	7	48.6	18	36	9	52	13

**Missing data: not all participants answered all questions*

4.3.6 Prevalence and incidence of thyroid disease

The prevalence of thyroid disorder at Wave 1 for those with DS was identified as 37.4% (n=55/147). This increased to 46% (n=63/138) by Wave 2 with a further increase noted by Wave 3 to 50.5% (n=54/109), see *figure 4.9* below. Thyroid disorder at Wave 1 was not categorised into hypothyroidism or hyperthyroidism in the questionnaire. However, this differentiation was introduced at Wave 2. In Wave 2, the number of participants reporting hypothyroidism was 27.9% (n=38/138); with a further 5.9%% (n=8/138) reporting hyperthyroidism. In Wave 3, similar findings were reported with 35.5% (n=38/109), reporting hypothyroidism and a further 7.5% (n=8/109) reporting hyperthyroidism.

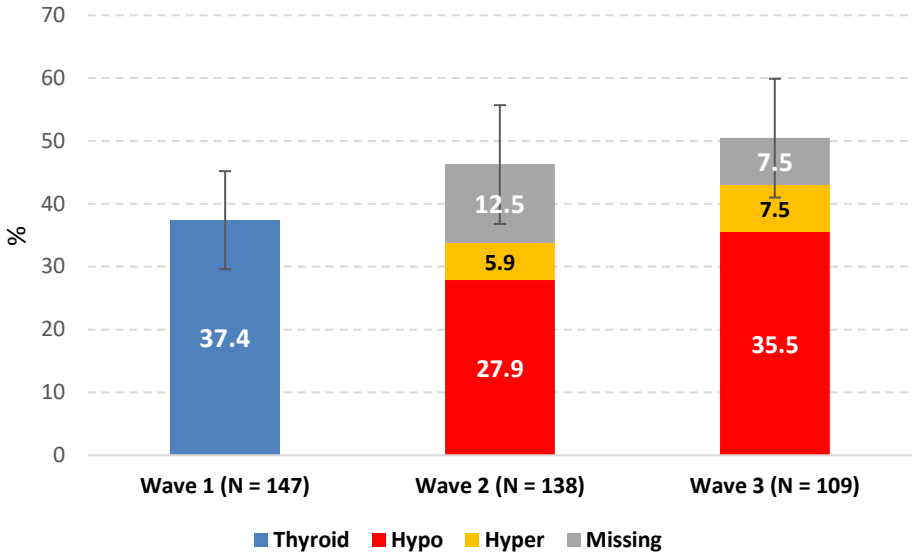


Figure 4.9 Prevalence of thyroid disorder across the three waves with hypothyroidism and hyperthyroidism denoted at waves 2 and 3

Considering the incidence of thyroid disorder across the three waves, between waves 1 and 2, there were 8.8 new cases per 100. For the subgroup who survived across all three waves, N=109 there were 7.4 new cases per 100 from wave 1 to wave 2. Between waves 2 and 3, there were 8.6 new cases per 100. See figure 4.10 for a graphical representation of incidence of thyroid disorder.

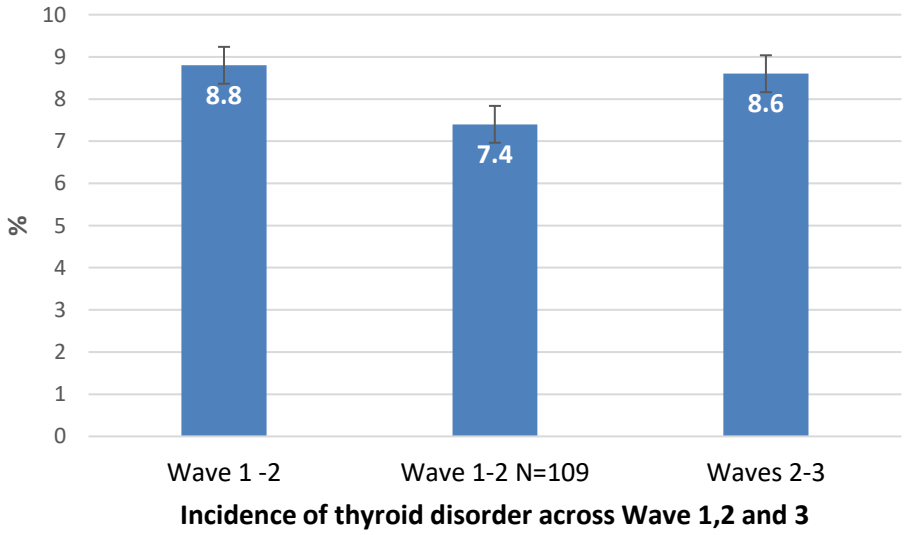


Figure 4.10 Incidence of thyroid disorder across all three waves of data collection

4.3.7 Thyroid disorder stratified by gender age and level of ID

Those who reported a doctor's diagnosis of thyroid disorder were examined with consideration to their gender, age and level of ID. It can be seen in **Table 4.6** that, similar to the other conditions, that females a higher prevalence of thyroid disorder than males across the waves. However, it is evident as individual's age, no matter what their sex, the prevalence increases. With consideration to age, the highest prevalence is observed among those in the middle age group (50-64 years) across the 3 Waves (Wave 1, 48.4% n=30/147; wave 2, 58.9% n=33 and Wave 3, 58.3% n=21). This aging trajectory is interesting considering that there was only 1 person surviving to Wave 2 (100% n=1/138) and Wave 3 (100% (n=1/109) who reported a doctor's diagnosis over the age of 65 years. Nobody in this age category reported thyroid disorder at Wave 1. With respect to level of ID in Wave 1, those who had a doctor's diagnosis of thyroid disease were predominantly within the severe/profound category (41.5%, n=17/147). This figure increased to 52% (n=13/109) by Wave 3. The other categories of level of ID also demonstrated a substantial increase in prevalence of thyroid disease, including those within the mild level of ID category who presented an increase from 35% (n=7/147) in Wave 1 to 44.4% (n=8/109) in Wave 3. Similarly, of all those with moderate ID, the prevalence of thyroid disease increased from 37.3% (n=28/147) at Wave 1 to 53.6% (n=30/109) at Wave 3. See full stratification in **Table 4.6**.

Table 4.6 Thyroid disorder stratified by gender age and level of ID

Thyroid		Wave1 N = 147*		Wave 1 (survivors to Wave 3) N=109		Wave 2 N = 138		Wave 2 (survivors to Wave 3) N=109		Wave 3 N = 109	
		%	*n*	*%*	*n*	*%*	*n*	*%*	*n*	*%*	*n*
Gender	Male	26.6	17	28.3	15	37.7	23	36.9	21	46.2	24
	Female	45.8	38	46.4	26	52.6	40	50.9	28	54.5	30
Age	40-49	30.1	25	30.6	22	36.3	29	38.0	27	45.7	32
	50-64	48.4	30	52.8	19	58.9	33	58.3	21	58.3	21
	65+	0.0	0	0.0	0	100	1	100	1	100	1
Level of ID	Mild	35.0	7	33.3	6	36.8	7	38.9	7	44.4	8
	Moderate	37.3	28	37.9	22	47.1	33	47.4	27	53.6	30
	Severe/Profound	41.5	17	40.0	10	48.6	18	48.0	12	52.0	13

*Missing obs: not all participants answered all the questions

4.3.8 Sensory Disorder

4.3.8.1 Visual impairment

Participants were asked to self-rate their vision as being positive or negative, full results can be seen in table 4.7. In Wave 1, 76.3% (n=106/147) of participants rated their vision as being positive (Excellent, very good or good) while the remaining 23.7% (n=33/147) rated their vision as being negative (fair or poor). In Wave 2, a total of 66.4% (n=85/138) rated their vision as being positive and similarly at Wave 3, 66.3% (n=67/109) rated their vision as being positive. The positive rated responses can also be reviewed in *figure 4.11* below. Considering those who survived across the three waves, their dominant choice was to rate their vision as positive. However, in saying that, there is still almost a third of those with DS who rate their vision negatively, see **table 4.7** for full self rated vision.

Table 4.7 Self rated vision of those with DS across Waves 1- 3

Self rated vision	Wave1 N = 147*		Wave 1 (survivors to Wave 3) N=109		Wave 2 N = 138		Wave 2 (survivors to Wave 3) N=109		Wave 3 N = 109	
	%	**n**	**%**	n	**%**	**n**	**%**	n	**%**	**n**
Positive	76.3	106	78.6	81	66.4	85	65.6	70	66.3	67
Negative	23.7	33	21.4	22	33.6	43	31.4	32	33.7	34

Not all participants answered all questions; Missing obs. W1 n=8, W2 n=10, W3 n=8

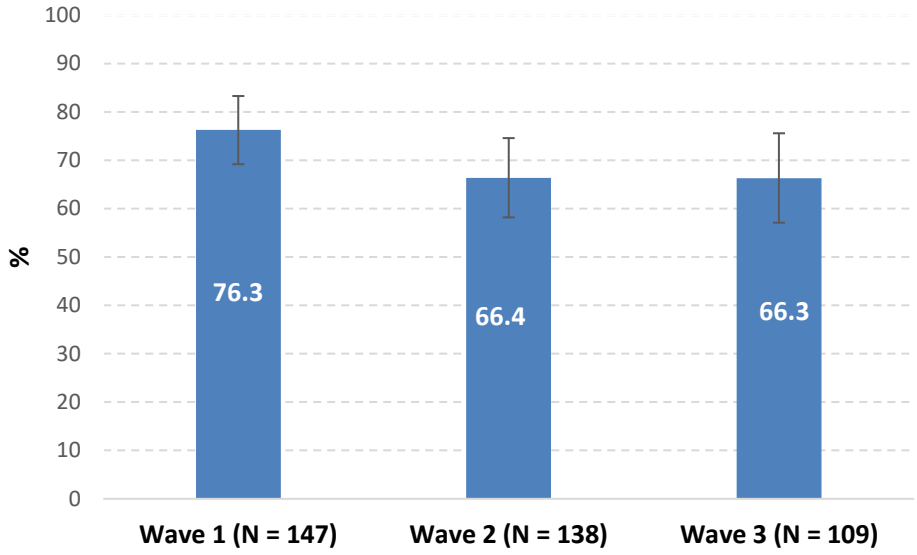


Figure 4.11 Positively self-rated vision across Waves 1,2 and 3

4.3.8.2 Fair/Poor vision stratified by gender, age and level of ID

Those who self-reported their vision as poor were examined with consideration to their gender, age and level of ID. It can be seen in **Table 4.8** that overall females reported a higher prevalence of self reported poorer eyesight than males across the 3 Waves. However, it is evident as individual's age regardless of their sex, the prevalence of poorer self-reported eyesight increases. With consideration to age, the highest prevalence of poor self-reported is observed among those in the middle age group (50-64 years) across the waves (Wave 1: 30.5% (n=18/147); wave 2, 38.8% n=19 and Wave 3, 45.5% n=15). Here, again the ageing trajectory is interesting considering that there were no individuals who reported poorer self rated vision over the age of 65 years. With respect to level of ID, participants with a severe/profound level of ID, were reported as having fair/poor eyesight (Wave 1, 40.5% n=15/147; Wave 2, 37.9% n=11/138; Wave 3, 42.9% n=9/109). The figures for all those participants in the moderate level of ID who self-reported their eyesight as fair/poor increased considerably from 13.9% (n=10/149) in Wave 1 to 36.2% (n=25/138) at Wave 2 and reached 33.3% (n=9/109) at Wave 3. Please see full stratification in **table 4.8.**

Table 4.8 Poor vision among those with DS stratified by gender, age and level of ID

Vision Poor		Wave1 N = 147*		Wave 1 (survivors to Wave 3) N=109		Wave 2 N = 138		Wave 2 (survivors to Wave 3) N=109		Wave 3 N = 109	
		%	n	%	n	%			%	n	%
Gender	Male	20.0	12	20.4	10	23.2	13	22.9	11	26.5	13
	Female	26.6	21	22.2	12	41.7	30	38.9	21	40.4	21
Age	40-49	19.0	15	20.6	14	30.8	24	27.5	19	28.4	19
	50-64	30.5	18	23.5	8	38.8	19	40.6	13	45.5	15
	65+	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0
Level of ID	Mild	25.0	5	27.8	5	21.1	4	16.7	3	11.1	2
	Moderate	13.9	10	16.4	9	36.2	25	37.5	21	33.3	18
	Severe/Profound	40.5	15	30.4	7	37.9	11	35	7	42.9	9

Missing obs: not all participants answered all the questions

4.3.8.3 Doctor's diagnosis of cataracts among those with DS across Waves 1,2 and 3.

People were asked if they ever had cataracts. As outlined in **table 4.9,** at Wave 1 the number of participants reporting cataracts was 30.3% (n=44), at Wave 2 44.5% (n=61) and at Wave 3 46.7% (n=50) reported cataracts. People were also asked at Waves 2 and 3 if they ever had cataracts and as can be seen there is a slight difference between cataracts reporting at one point in time and verifying if they ever had cataracts. In Wave 3 particularly there is an obvious difference noted, 32.7% versus 46.7% respectively.

The uptake of cataract surgery over the 10-year period is also outlined in **table 4.9**. Participants who answered 'yes' to having a doctor's diagnosis of cataracts were then asked if they had cataract surgery. In Wave 1, 41.9% (n=18/44) had surgery. Of great concern is the fact that 58.1% (n=25/44) reported they did not have surgery for their cataracts. In Wave 2, 46.0% (n=29/63) participants with cataracts had surgery, and again the figure for those not receiving surgical treatment was concerning at 54% (n=34/63). In Wave 3, 63.3% (n=31/49) of participants with cataracts had cataract surgery, leaving a total 36.7% (n=18/49) of this cohort not having surgery.

Table 4.9 Prevalence Cataracts and Cataract surgery

	Wave 1		Wave 1 (survivors to Wave 3) N=109		Wave 2		Wave 2 (survivors to Wave 3) N=109		Wave 3	
	%	**n**	**%**	**n**	**%**	**n**	%	**n**	**%**	**N**
Cataracts (at point in time)										
Yes	30.3	44	26.6	29	43.8	60	40.7	44	32.7	35
No	69.7	101	73.4	80	56.2	77	59.3	64	67.3	72
Have you ever had cataracts										
Yes	30.3	44	26.6	29	44.5	61	41.7%	45	46.7	50
No	69.7	101	73.4	80	55.5	76	58.3	63	53.3	57
Surgery* (includes only those who said yes to cataracts)										
Yes	41.9	18	46.4	13	46.0	29	48.9	22	63.3	31
No	58.1	25	53.6	15	54.0	34	51.1	23	36.7	18

Missing obs: Wave 1 n=2, Wave 2 n=1, Wave 3 n=2
**Missing obs cataract surgery Wave 1 n=1, Wave 2 n=0, Wave 3 n=1*

To graphically review the overall results, for those who self rated their vision as positive, those who ever had cataracts and those who had surgery across the three waves of data collection see *figures 4.12* below. Overall people generally reported their vision positively. However, there was a notable decrease in the number from Wave 1 through Wave 2 to Wave 3 (76% to 66.4% to 66.3% respectively) reflecting the fact that as people got older, they generally report their eyesight as poorer. It is interesting to also note that the presence of cataracts appears to be rising year on year. However, so to is the number of people having cataract surgery.

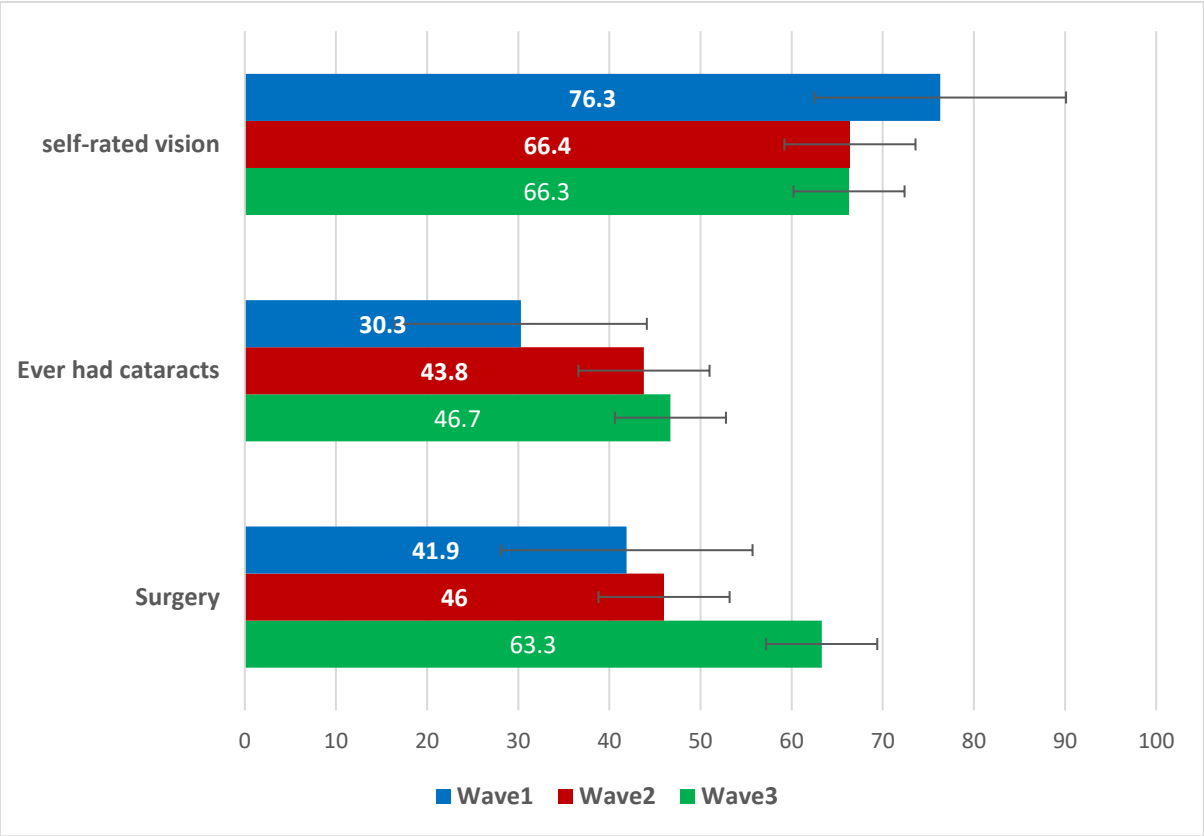


Figure 4.12 Overview of those who self rated their vision, ever had cataracts and had surgery for cataracts

4.3.8.4 Auditory Impairment

Participants were asked to self-rate their hearing as being positive or negative. Overall more participants rated their hearing as excellent/Very good/good, in Wave 1 76.9% (n=113/147) rising to 79.9% (n=110/147) by Wave 2 with a slight drop which could be expected with advancing age by Wave 3 (74.3%, n=81/109). Participants who survived across all three waves, again, the majority rated their hearing as positive. However, across the three waves almost a quarter of individuals with DS rated their hearing as being poor (Wave1, 23.1% n=34/147; Wave 2, 18.0% n=25/138; Wave 3, 22.0% n=24/109). See **table 4.10** for full details of self reported hearing.

Table 4.10 Self reported hearing of those with DS across Waves 1, 2 and 3.

Hearing	Wave1 N = 147*		Wave 1 (survivors to Wave 3) N=109		Wave 2 N = 138		Wave 2 (survivors to Wave 3) N=109		Wave 3 N = 109	
	%	n	%	n	%	N	%	n	%	n
Poor/Fair	23.1	34	21.1	23	18.0	25	16.5	18	22.0	24
Excellent/Good	76.9	113	78.9	86	79.9	110	80.7	88	74.3	81

**Missing obs: not all participants answered all the questions*

4.3.8.5 Hearing self-rated poor/fair stratified by gender, age and level of ID across Waves 1, 2 and 3

With consideration to their gender, age and level of ID participants who self-reported hearing as poor/fair was examined. It can be seen in **Table 4.11** that overall females reported a higher prevalence of self-reported poorer hearing than males across the 3 Waves. With consideration to age, among those in the middle age group (50-64 years) across the waves reported their hearing as poor or fair (Wave 1 38.7% n=24/147; Wave 2 27.3% n=15/138 and Wave 3 35.3% n=12/109). Again, it can be seen that only 1 person at Wave 2 (100% n=1/138) in the 65+ years cohort reported poorer hearing. With respect to level of ID in Waves 1 and 2 those with a severe/profound level of ID had the highest reported poorer hearing and this changed by Wave 3. At Wave 3 those within the moderate category of ID reported a higher prevalence of self reported poorer hearing (26.8%, n=15). Please see full stratification in **table 4.11**.

Table 4.11 Self-rated hearing stratified by gender, age and level of ID across Waves 1,2 and 3.

Hearing rated poor /fair		Wave1 N = 147*		Wave 1 (survivors to Wave 3) N=109		Wave 2 N = 138		Wave 2 (survivors to Wave 3) N=109		Wave 3 N = 109	
		%	*n*	*%*	*N*	*%*	*n*	*%*	n	*%*	*n*
Gender	Male	15.6	10	13.2	7	13.3	8	13.5	7	23.1	12
	Female	28.9	24	28.6	16	22.7	17	20.4	11	22.6	12
Age	40-49	12	10	12.5	9	11.4	9	12.9	9	17.1	12
	50-64	38.7	24	38.9	14	27.3	15	22.9	8	35.3	12
	65+	0.0	0	0.0	0	100	1	100	1	0.0	0
Level of ID	Mild	10	2	11.1	2	21.1	4	22.2	4	11.1	2
	Moderate	22.7	17	22.4	13	14.3	10	12.3	7	26.8	15
	Severe/Profound	26.8	11	24	6	28.6	10	30.4	7	26.1	6

4.4 Functional changes/ decline in activities of daily living

While examining activities of daily living (ADL's) it was evident that participants with DS presented with marked functional decline with increasing age. Levels of independence deteriorated remarkably over the 10-year period. With consideration to **Table 4.12** the ADLs; bathing, dressing, eating, getting in and out of bed, using the toilet and walking across the room were examined in respect of those who reported some, a lot of difficulty or could not do the activity at all. In Wave 1, a number of individuals (36.6%, n=52/147) had difficulty with bathing. This figure dropped slightly in Wave 2. However, difficulty with bathing increased to 34.0% (n=36/109) by Wave 3. Similarly, increases in difficulty across all activities of living are evident. For example, difficulty with dressing almost doubled from 17.6% (n=25/147 in Wave 1 to 28.3% (n=30/109) by Wave 3. A slight increase with the difficulty in eating is observed between Wave 1 and 2. However, difficulty with getting out of bed, using the toilet and walking across a room increased substantially from Wave 1 to Wave 3, see ***table 4.12***.

Table 4.12 Difficulty with ADL's from Wave 1 – Wave 3

Activity of Daily Living	Wave 1 N= 147		Wave 1 (survivors to Wave 3) N=109		Wave 2 N=138		Wave 2 (survivors to wave 3) N=109		Wave 3 N=109	
	%	n	%	n	%	n	%	n	%	n
Bathing	36.6	52	28.3	30	31.6	43	24.3	26	34	36
Dressing	17.6	25	14.2	15	21.9	30	14.8	16	28.3	30
Walking across Room	8.2	12	1.8	2	13.1	18	5.6	6	19.5	17
Eating	17.2	25	12	13	21.2	29	13.9	15	14.9	13
Using the Toilet	9.9	14	3.8	4	14.6	20	6.5	7	18.4	16
Getting out of Bed	9.0	13	2.8	3	11.7	16	3.7	4	16.3	14

4.5 Transitions, moving residence prevalence and type of move

People were asked at Wave 2 and again at Wave 3 if they had moved home in the previous three years. As previously described, these moves were categorised as; lateral moves, less supported move and increased supported move.

As can be seen in **Table 4.13,** overall at Wave 2 18.25 (n=25/138) moved home and by Wave 3 this figure had increased to 28.4% (n=31/109). However, the majority remained in their own home setting across all waves of data collection. What is interesting is the type of moves that people engaged with. A total of 56 people with Down syndrome moved from their home over the 10 -year period from Wave 1 through to Wave 3. In Wave 2, the number of people with DS who made a lateral move, that is moving to a similar type of setting, represented 58.3% (n=14/138). This type of move may have been within a campus setting whereby the participant moved from one area of the campus to another or was living in a community group home and moved from one house in the community to another. At Wave 3, the number of people with DS who made a lateral move to a similar type of setting represented 58.1% (n=18/109). With consideration to moving to a less supported environment, in total 12.5% (n=3/138) at Wave 2 moved to a home setting that enabled a more independent way of living. At Wave 3, 9.7% (n=3/109) of this cohort moved to a less supported home setting. With consideration to a more supported home setting at Wave 2, 29.2% (n=7/138) of those with DS had moved to a home that provided increased levels of assistance. Three years later at Wave 3 there was a notable change in the proportion of people who had moved to a more supported setting with 32.3% (n=10/109) of this population moving.

Table 4.13 ***Types of Moves between Wave 1, Wave 2 and Wave 3***

Moved residence	Wave 1* N=147		Wave 2* N=138		Wave 2 (survivors to wave 3) N=109		Wave 3 N=109	
			%	n	%	n	%	n
Yes	**NA	NA	18.2	25	14.8	16	28.4	31
No	NA	NA	81.8	112	85.2	92	71.6	78
Type of move								
Lateral	NA	NA	58.3	14	62.5	10	58.1	18
More support	NA	NA	29.2	7	25	4	32.3	10
Less Support	NA	NA	12.5	3	12.5	2	9.7	3

*Missing observations: W1 = 1 ; W2=1 **NA= not applicable to wave 1

4.5.1 Reasons why people with DS moved home setting

As can be seen from **table 4.14,** people choose a variety of reasons why they moved residence between waves 1 and 2, and also for those who moved between waves 2 and 3. Overall the main reason was because of physical health changes at 52% (n=13/25) from Wave 2, and 45.2% (n=14/31) to Wave 3. By Wave 3, individuals changing needs emerge as an important issue with 16.1% (n=5/31) moving because of this. What is interesting across the waves, is that some people moved to accommodate services. All reasons for moving can be reviewed in table 4.14 below.

Table 4.14 Reasons for moving residence between waves 1-2 and waves 2-3

Reasons for moving	Wave 2 N = 25		Wave 2 (survivors to Wave 3) N=16		Wave 3 N = 31	
	%	n	%	n	%	n
Physical health changes	52.0	13	31.3	5	45.2	14
Loss of primary carer	0.0	0	0	0	0.9	1
Changes in service policy	1.4	2	6.3	1	0	0
Moved to accommodate services	2.9	2	25.0	4	12.9	4
Not happy with living situation	4.0	1	6.3	1	3.2	1
Staff/funding shortages	0.0	0	0.0	0	0.0	0
Skill mix did not meet need	0.0	0	0.0	0	0.0	0
Lack of accessibility	8.0	2	0.0	0	3.2	1
Changing needs (e.g. no downstairs facilities)	0.0	0	0.0	0	16.1	5
Lack of nursing support	4.0	1	0.0	0	3.2	1
Lack of 24-hour care	8.0	2	0.0	0	3.2	1
As part of transition process	16.0	4	18.8	3	3.2	1
Personal choice	12.0	3	18.8	3	3.2	1
I don't know the reason	4.0	1	6.3	1	0.0	0
Other	36.0	9	50.0	8	64.5	20

Missing obs; Wave 1 n=0; Wave 2 n=1; Wave 3 n=0

4.6 Healthcare utilisation of those with DS in relation to the most prominent chronic health conditions identified in the literature.

People were asked if they attended their GP and other specialist health services in the last year for assessment and for review. Overall, the majority of people with DS have at least one visit to their GP on an annual basis (Wave 1, 88.4% n= 130/147; Wave 2, 97.1 % n=132/138; Wave 3 97.1% n=100/109). Attendance at other specialist services appears sporadic with fewer people reporting attendance in the last year particularly to endocrinology services where by overall less than 4% received services in the year prior to the time of their interview across the waves of data collection. See figure 4.14 for a graphical representation of healthcare utilisation of those with DS across the three waves of data collection.

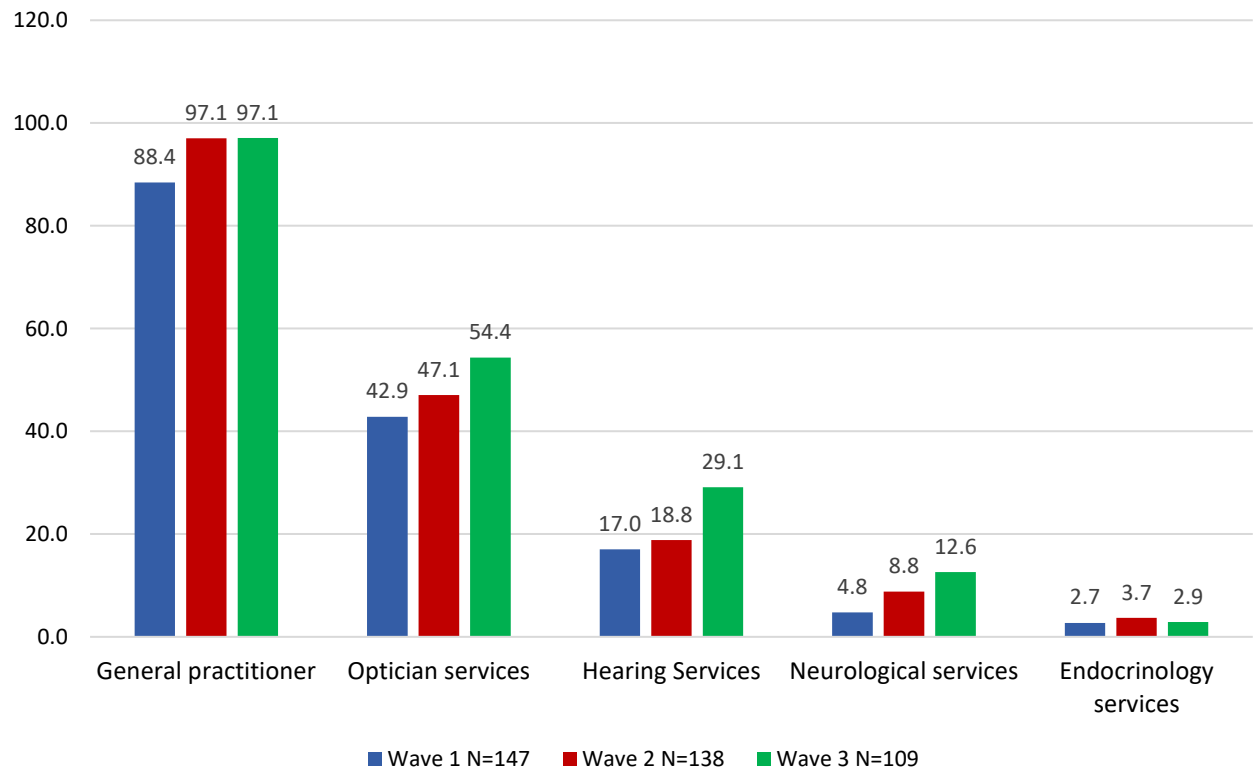


Figure 4.14 Specialist healthcare utilisation

4.7 Challenges in communication

People were asked what level of difficulty they had when communicating i.e. speaking or making themselves understood generally. As can be seen in *figure 4.15,* across the three waves of data collection, a lot of people had some level of difficulty communicating. At Wave 1, 23.8% (n=35/147) could not communicate at all, with 45.6% (n=67/147) individuals with DS reporting they has some or a lot of difficulty communicating. In Wave 2, 21.9% (n=30/138) could not communicate with a further 54.7% (n=75/138) having some or a lot of difficulty. And finally, in Wave 3, 18.5% (n=20/109) could not make themselves understood with a further 54.6% (n=59/109) having some or much difficulty making themselves understood.

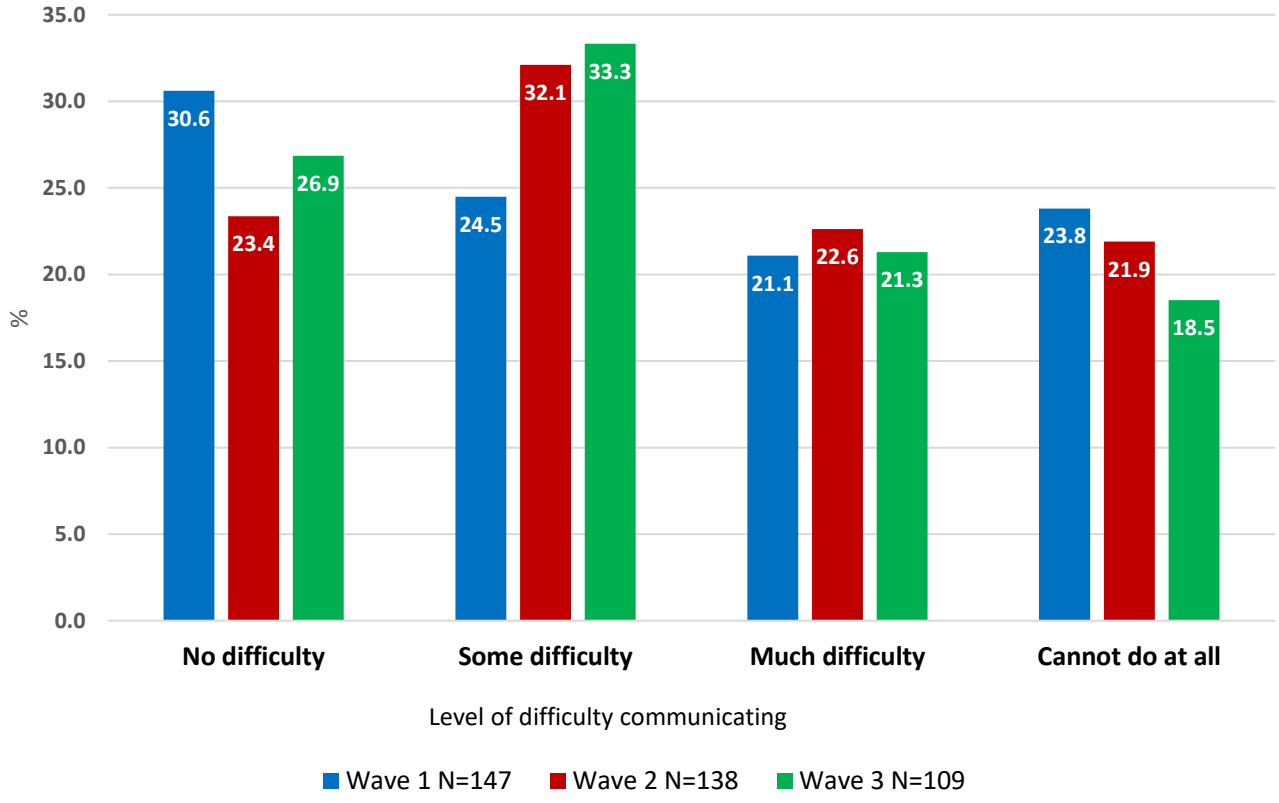


Figure 4.15 Difficulty in communicating

4.7.1 Difficulty making themselves understood by health care professionals

People were then asked what level of difficulty they had when communicating i.e. speaking or making themselves understood, with their healthcare professional (HCP), examples given included, with the doctor, nurse, physiotherapist or social worker. As can be seen in figure 4.16, a small number of people could not converse with their HCP at all. However, a large number could only partially make themselves understood by HCP (Wave 1 41.4%, n=61/147; Wave 2 66.7%, n=92/138; Wave 3, 64.4% n=70/109).

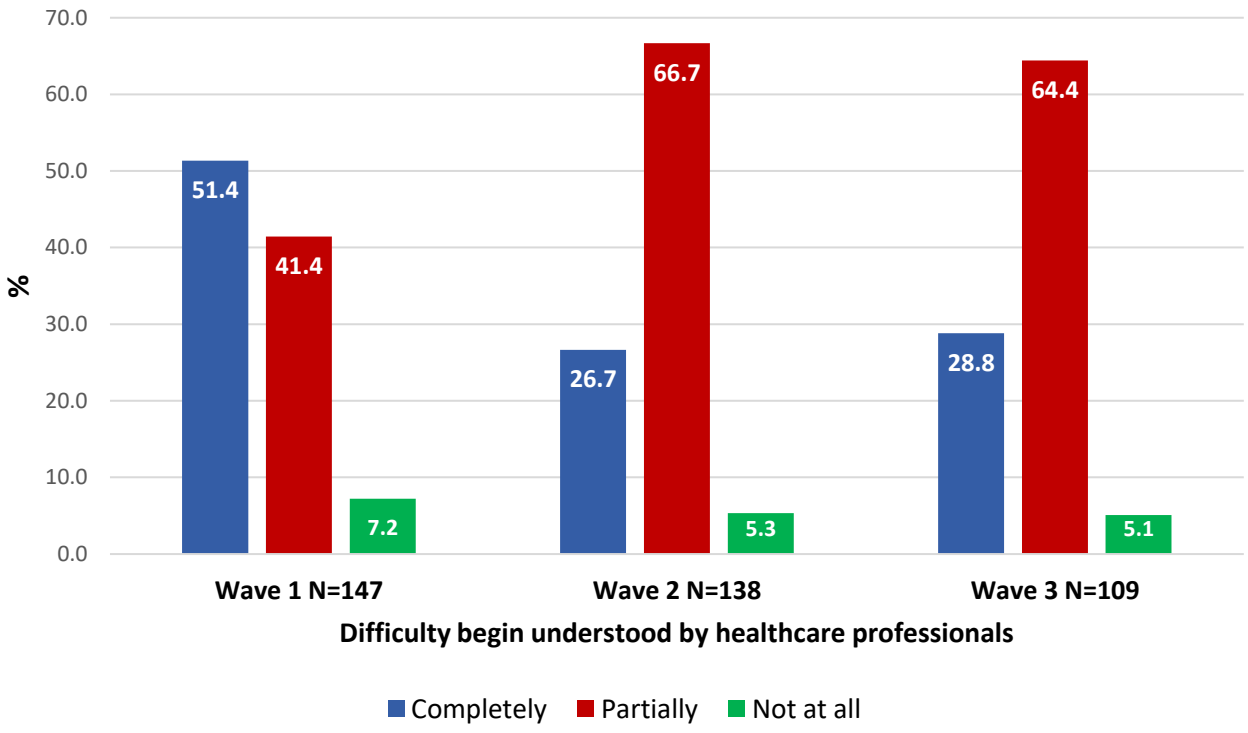


Figure 4.16 Difficulty being understood by healthcare professionals

4.8 Examining decline in conditions across the 10 years

4.8.1 McNemar's test for significant change in proportions longitudinally

As previously noted, the McNemar test as a repeated measured version of a chi-square test of independence. It is used to test for consistency in responses across two variables. If the statistical significance level (i.e., *p*-value) is less than .05 (i.e., $p < .05$), a statistically significant result is observed and the proportion of participants showing a particular characteristic at Wave 1 compared to Wave 3 is statistically significantly different. Alternatively, if $p > .05$, a statistically significant result is not observed and the proportion of participants showing a particular characteristic at Wave 1 compared to Wave 3 is not statistically significantly different (i.e., the proportion does not change over the course of the study). Note, this test follows only those individuals with an observation at both points in time.

Table 4.15 shows that proportions presenting each characteristic of interest in Wave 1 changed significantly by Wave 3. For example, the proportion of those reporting a doctor's diagnosis of dementia at Wave 1 changed significantly by Wave 3. As shown above in section 4.3.1, this significant change reflects an increase in the prevalence of dementia. It can be seen in table 4.15 that all characteristics changed significantly from Wave 1 to Wave 3, all physical health conditions have increased, TSI scores have declined (indicating cognitive decline), and functional limitations have also increased significantly as the population has aged over the three waves of the study, indicating that increasing age in people with DS impacts significantly on overall physical health, and functional ability.

Table 4.15 Mc Nemar's Test for differences in paired proportions between Wave 1 and

Variable	*p-value*	*NN*
Dementia	<0.005	107
TSI*	<0.005	59
Epilepsy	<0.005	107
Have you ever had cataracts	<0.005	107
Cataracts Surgery	<0.005	107
Self rated vision	0.037	96
Self rated hearing	0.034	83
Thyroid	<0.005	107
Functional limitations	<0.001	103

Wave 3

4.9 Attrition and survival analysis

Of the 147 participants with DS recruited at Wave 1, a total of 109 completed all 3 Waves. The sample attrition was 26% (n=38/147) for this cohort which was largely due to deaths in the sample (30 deaths and 8 withdrawals) (see **Table 4.16**). The mean age at last interview for the deceased was 57.2 years and 73.3% (n=22/30) of these were female. A total of 59.3% (n=16/27) % of the deceased were from the severe/profound group and 76.7% (n=23/30) lived in residential care at the time of their last interview. Health conditions were highly prevalent in the deceased group with 80% (24/30) having dementia, 70% (n=21/30) having a diagnosis of epilepsy, 60% (n=18/30) have had cataracts and 53.3% (n=16/30) had thyroid disease).

Table 4.16 Demographics and health conditions across the 3 Waves for non-active participants with Down syndrome

		Deceased N = 30		*Withdrawals* N = 8	
Mean age at last interview		57.2		50.25	
Median age at last interview		57.5		50	
Age at last interview		***%***	***N***	***%***	n
<50		10	3	50	4
50-64		80	24	50	4
65+		10	3	0	0
Gender		***%***	***N***	***%***	n
Male		26.7	8	37.5	3
Female		73.3	22	62.5	5
ID		***%***	***N***	***%***	n
Mild		3.7	1	12.5	1
Moderate		37	10	87.5	7
Severe/Profound		59.3	16	0.0	0
		Missing = 3			
Residence at last interview		***%***	***N***	***%***	n
Indep / Family		6.7	2	50	4
CGH		16.7	5	25	2
Residential		76.7	23	25	2
Chronic Health Conditions		***%***	***N***	***%***	n
Dementia	**Yes**	80.0	24	75.0	6
	No	20.0	6	25.0	2
Cataracts	**Yes**	60.0	18	25.0	2
	No	40.0	12	75.0	6
Thyroid	**Yes**	53.3	16	25.0	2
	no	46.7	14	75.0	6
Epilepsy	**Yes**	70.0	21	12.5	1
	No	30.0	9	87.5	7

4.9.1 Examining those who had deceased and the emerging health conditions (N = 30)

The participants with DS for this study were followed over a 10-year period and the survival time is considered in terms of the duration of the study, i.e. 10-years (see **table 4.17** for mean age N=147). Of the 145 in the sample, with outliers removed, 65 (44.8%) had a diagnosis of dementia over their lifetime and 80 (55.2%) had no diagnosis (see **table 4.18** for mean age with outliers removed. Of the 65 people with dementia, 24 (36.9%) experienced the event of interest (i.e. they had died) and in the non-dementia sample only 5/80 (6.3%) experienced the event of interest. This implies that 75 (93.8%) in the non-dementia sample are censored (i.e. alive at the 10 year follow up) compared to 41 (63.1%) surviving in the Dementia sample. This shows that the mortality rate is higher in the dementia group compared to the non-dementia group (see table 4.18). Examining the most common health conditions in participants with DS who are deceased (n=30) since the inception of the IDS-TILDA study in 2009 as shown in *Figure 4.17* dementia is the most prevalent health condition with 80% of the deceased previously reporting the condition. Prior to death, 70% of participants had developed epilepsy. The number of participants with cataracts who were deceased accounted for 60% while 53.3% had a diagnosis of thyroid disorder.

Table 4.17 Descriptive statistics for Age (N= 147)

Variable	N	Missing	Mean	StDev	Minimum	Maximum	Median
Age	147	0	54.64	6.06	44	84	53

Table 4.18 Descriptive statistics for age after outlier removal (N = 145)

Variable		N	Missing	Mean	StDev	Minimum	Maximum	Median
Age		145	0	54.29	5.284	44	68	53
Age	Censored	116	0	53.41	4.94	44	68	52
	Deceased	29	0	57.83	5.2	46	67	57
Age	No Dementia	81	0	52.54	4.53	44	68	52
	Dementia	64	0	56.50	5.37	46	68	56

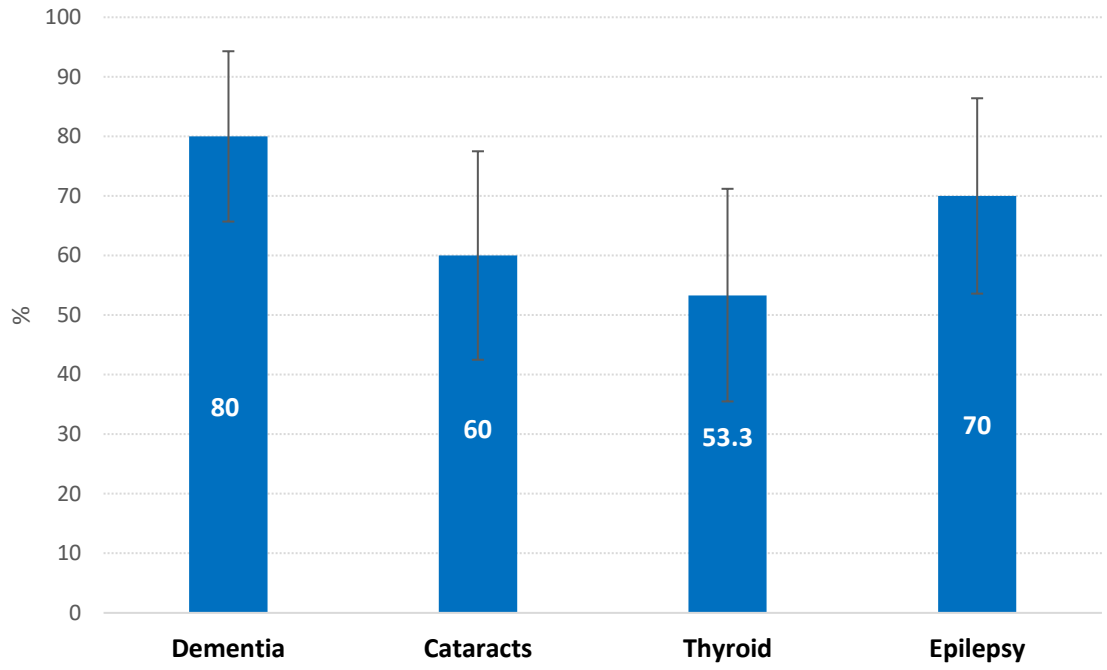


Figure 4.17 Prevalent health conditions in deceased participants

4.9.2 Survival Analysis for sample with DS (N = 147)

The variable age used is the age at death for participants who died with the exception of 6 missing dates of death. For these six people, the age at last active Wave was used to estimate age at death. For the active participants, their age at Wave 3 is used. The dot plot below, *figure 4.18*, *4.19* shows the maximum age recorded for each participant. The maximum age reached by any participant was 84 years with only 5 participants in total surviving to 66 years or above.

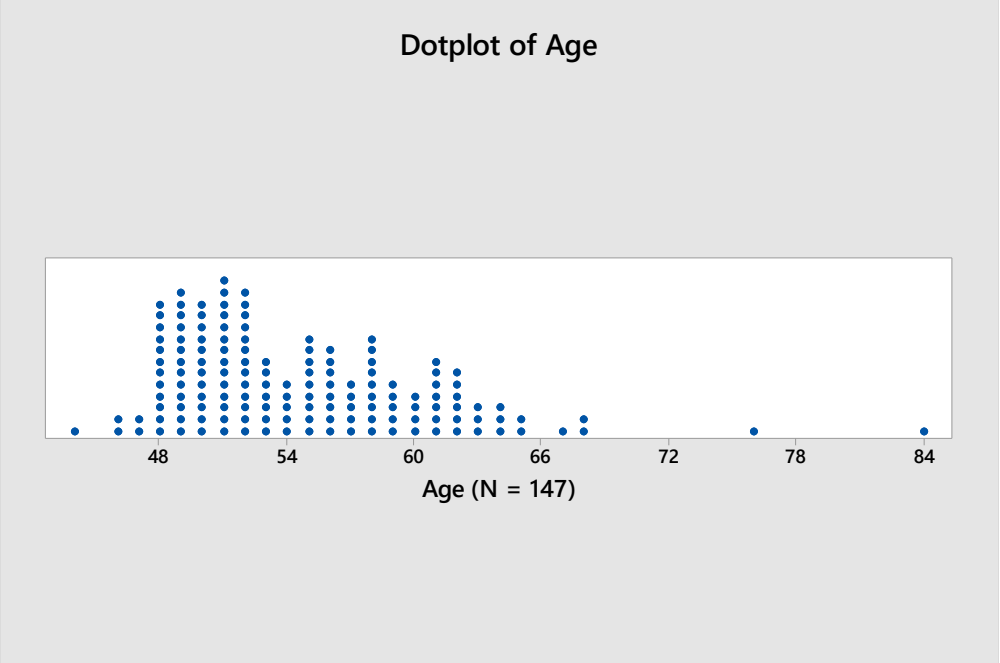


Figure 4.18 Dotplot of Age (n= 147)

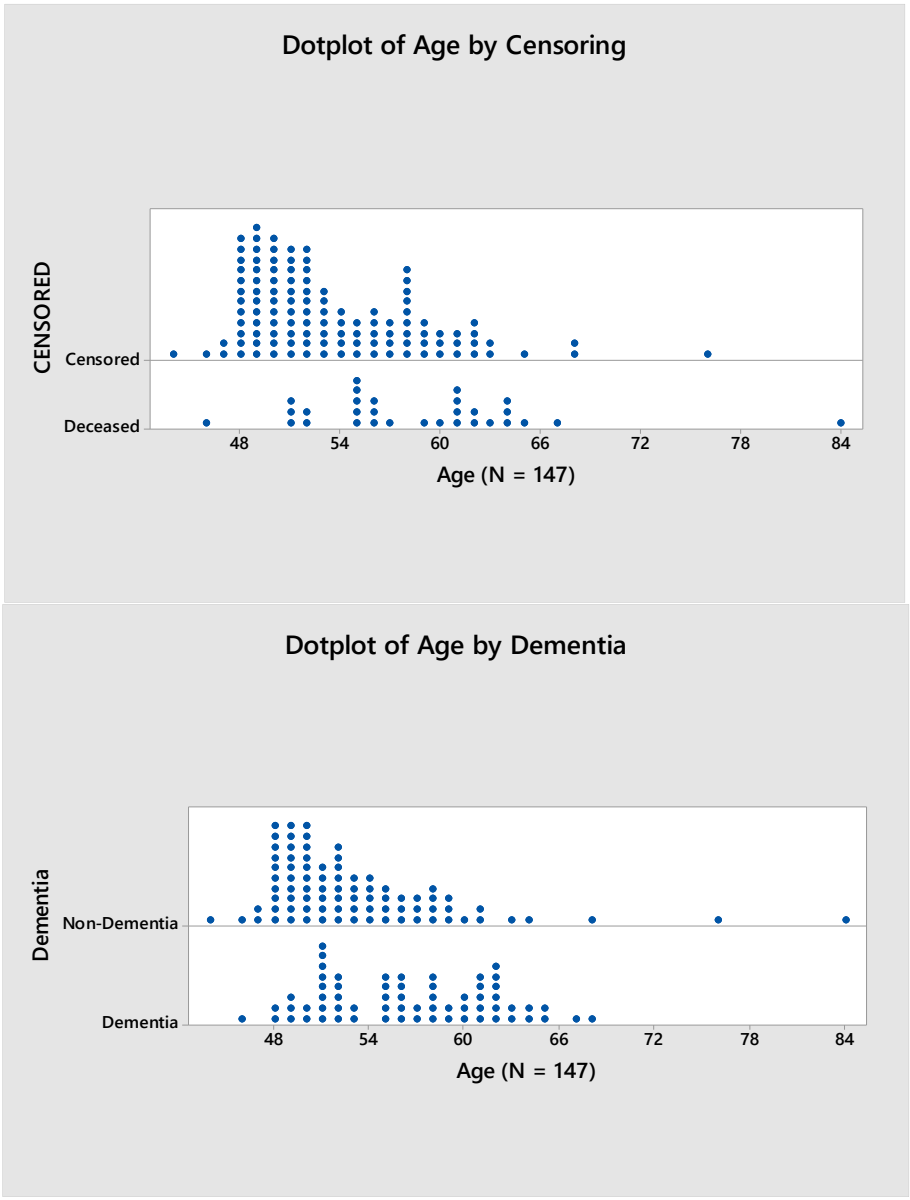


Figure 4.19 Dotplot of age by censoring and dementia

A boxplot of age identifies the two outliers, both in the non-dementia group, aged 84 and 76. As these were subsequently found to be inflating the mean and median survival time in the non-dementia group a decision was made to remove them after the running of the analyses with and without the outlier. The maximum age after outlier removal is 68 years with a mean of 54 years. The mean age in the censored group (surviving or withdrawn at the 10 year follow up) is 53.4 years compared to 57.8 years in the non-censored group (deceased within the 10 year time frame). See figure 4.20 and 4.21.

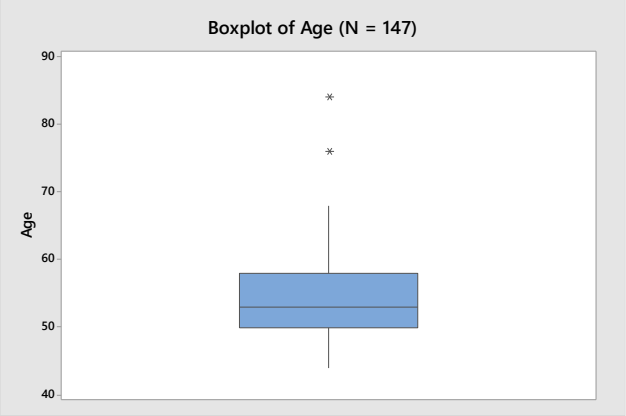
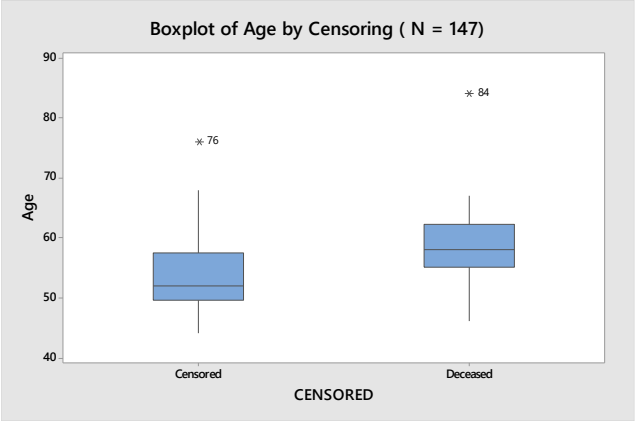


Figure 4.20 Boxplot of age



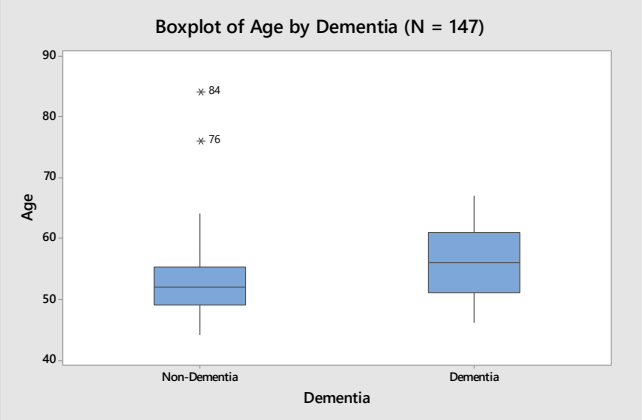


Figure 4.21 Boxplot of Age by Censoring and Dementia (N = 147)

4.9.3 Kaplan Meier survival analysis for N = 145

Of the remaining 145 after outlier removal, a total of 29 people (20%) experienced the event of interest i.e. death. The remaining 80% (n=116) were censored i.e. alive or withdrawn at the end of the study (wave 3). A total of 74.5% (n = 108) were still active and 5.5% (n=8) had withdrawn. **Table 4.19** shows the estimated mean survival time for the total sample with DS is 62.25 years (95%CI = 60.1 – 63.6 years) and the median survival time is 64 years (95% CI = 62.3, 65.7 years). Although, it should be noted that due to the high number of censored observations (n=116, 80%) that the vast majority of the sample have not yet experienced the event of interest i.e. death. This means that the survival analysis is an ongoing piece of research and more time is needed for the sample to mature to give a fuller picture of the mortality rate and survival analysis. *Figure 4.22* displays the survival curve for the 145 participants.

Table 4.19 Kaplan Meier means and medians for survival time (N= 145)

Mean[a]				Median			
Estimate	Std. Error	95% Confidence Interval		Estimate	Std. Error	95% Confidence Interval	
		Lower Bound	Upper Bound			Lower Bound	Upper Bound
62.25	0.68	60.93	63.58	64	0.86	62.32	65.68

a. Estimation is limited to the largest survival time if it is censored.

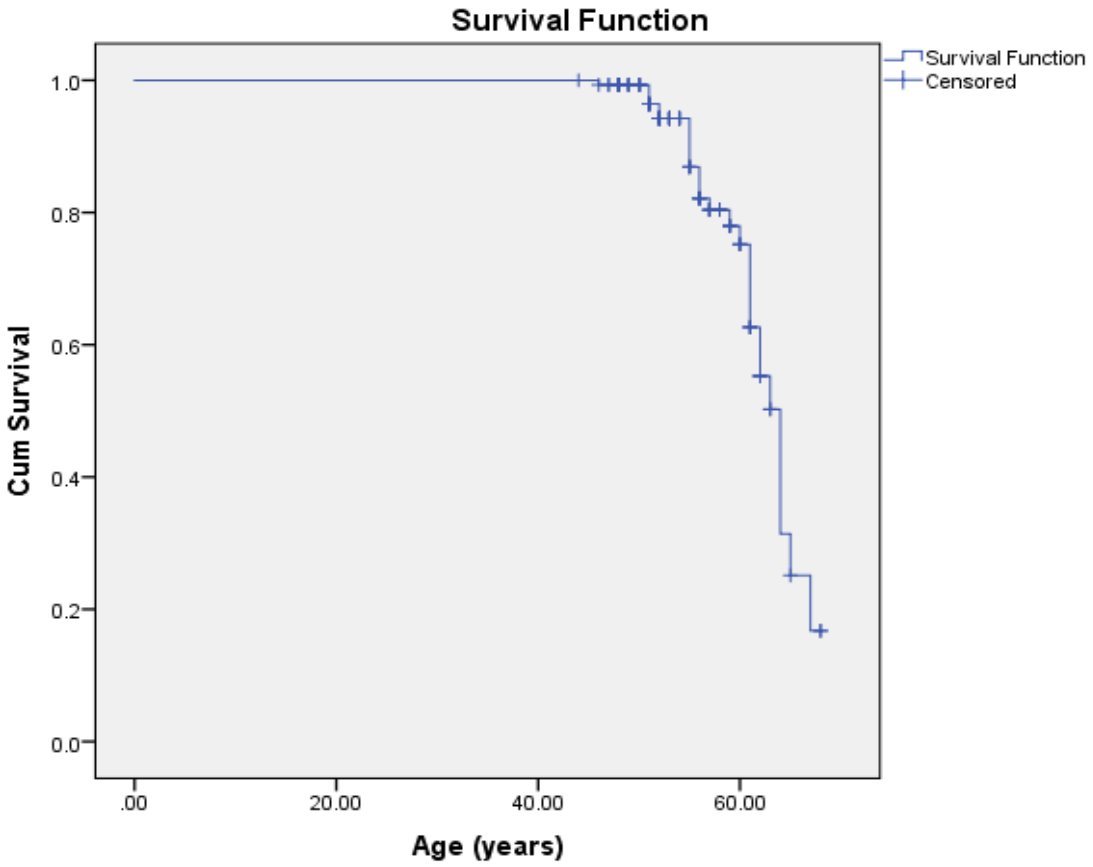


Figure 4.22 Kaplan-Meier Survival Curve (N = 145)

4.9.4 Kaplan-Meier for sample with DS stratified by dementia (N = 145)

Of the 145 participants in the sample with outliers removed, 64 (44.1%) had a diagnosis of dementia over their lifetime and 81 (55.9%) had no diagnosis see **table 4.20**. Of the 64 people with dementia, 24 (37.5%) died and in the non-dementia sample only 5 (6.2%) had died. This means that 76 (93.8%) in the non-dementia sample are censored (surviving/withdrawn) compared to 40 (62.5%) censored (surviving/withdrawn) in the Dementia sample. As almost 94% of the non-dementia group are still alive at Wave 3, it is evident that survival cannot be fully explored at this time and remains a point for future research. However, this analysis will focus on the data available to date and act as a preliminary look at an evolving situation.

Table 4.20 ***Dementia by Censoring Descriptive Statistics (N = 145)***

Dementia	N	N of Deaths	Censored N	Percentage Censored
No	81	5	76	93.80%
Yes	64	24	40	62.50%
Total	145	29	116	80.00%

The estimated mean survival time for those with dementia is 61 years and higher at 64 years for those with no doctor's diagnosis of dementia. The log rank test tests the hypothesis that there is no difference in survival times between the group with a doctor's diagnosis of dementia and those without a doctor's diagnosis of dementia. A p-value of 0.074 is not less than 0.05, so it therefore can be concluded that there is not significant evidence of a difference in survival times for those with and without dementia (see **Table 4.21**). Those without a diagnosis of dementia have an increased duration of survival of 3 years but as most of this group is censored the true survival time is not yet known. The median survival time is calculated as the smallest survival time for which the survivor function is less than or equal to 0.5. The median survival time for those with dementia is 62 years compared to 64 in the non-dementia group. (*Figure 4.23*)

Table 4.21 Kaplan Meier Means and Medians for Survival Time by Dementia Status (N = 145)

	Mean[a]				Median			
	Estimate	Std. Error	95% Confidence Interval		Estimate	Std. Error	95% Confidence Interval	
			Lower Bound	Upper Bound			Lower Bound	Upper Bound
Dementia								
No dementia	64.03	1.44	61.21	66.85	64	2.23	59.63	68.37
Dementia	61.51	0.80	59.95	63.08	62	1.25	59.54	64.46
Overall	62.25	0.68	60.93	63.58	64	0.86	62.32	65.68
Log Rank (Mantel-Cox)								
Chi-Square	3.21							
p-value	0.073							

a. Estimation is limited to the largest survival time if it is censored.

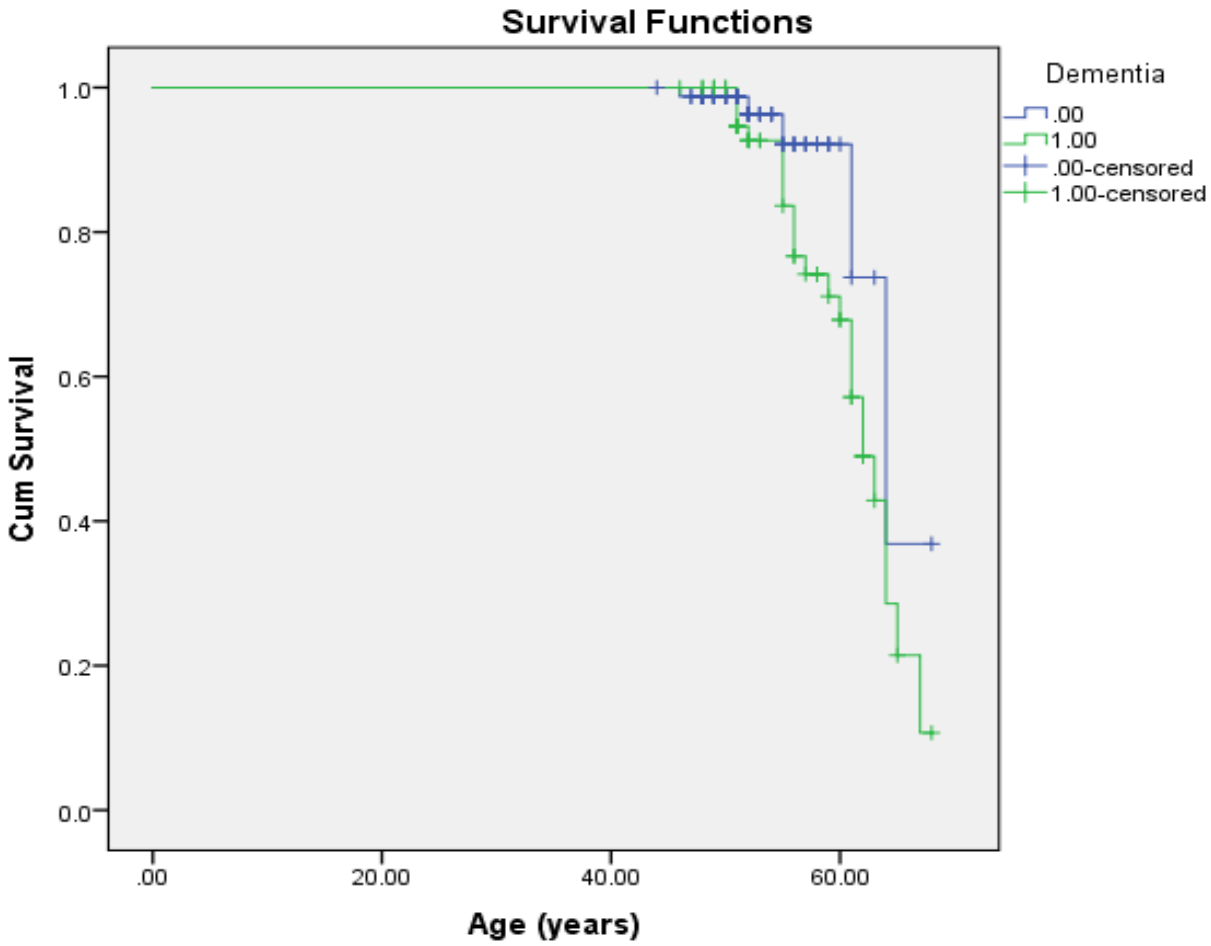


Figure 4.23 Kaplan Meier Survival Curve by Dementia (N = 145)

4.10 Conclusion

In this chapter the demographic characteristics of people with Down syndrome, their physical health, stratified by age, gender, level of intellectual disability and living circumstances were examined in relation to how ageing has impacted on their life. Descriptive and inferential statistics were used to analyse the data. It is evident from the findings of this study, that the impact of ageing is notable for people with Down syndrome. Adults with Down syndrome are at higher risk of complex comorbid health conditions which appear to accelerate both their cognitive and functional decline, and these described health conditions show an ominously elevated prevalence with increasing age.

The prevalence of dementia has increased significantly over the three Waves of IDS-TILDA, in the ten year period and this study has shown that the age related health conditions described are manifesting at an earlier age than the general population. It will be important to continue to follow this cohort to assess ongoing physical and cognitive changes over time and to compare the data to other studies. This information will inform and guide future planning and supports for people with DS as they age. Supporting people with DS as they age to maintain and manage their current level of functioning, to manage and possibly improve their physical and cognitive health will be key to improving overall wellbeing and better outcomes in the future. These findings will be discussed in chapter 5.

Chapter 5

Discussion

5.0 Introduction

In recent decades there has been an unparalleled increase in ageing, in the general population throughout the world. The WHO (2012) has estimated that this trend is set to continue and by 2050 the numbers of people over the age of 60 years will have increased to over 2 billion (Beard *et al*. 2011) This picture is also evident in people with ID and DS. Greater survival rates into later life have been documented among people with DS with recent studies reporting people with DS living into their fifties, sixties and beyond (McCarron *et al*. 2013; Head *et al*. 2012). In this chapter the researcher will discuss the findings considering current literature. The ageing experience of those with DS will be explored and considering the findings regards dementia the findings will be elucidated in relation to the literature. Specific health conditions emerged as highly prevalent among those with DS and consideration of these chronic health conditions will be explored, especially in light of the findings regards females. The impact of chronic health conditions is evident in the literature and similarly in this study chronic health conditions impact on individual's independence and their ability to age in place. Finally, with consideration on how to address the overall issues arising especially in relation to communication, assessment and mortality the researcher will consider the impact and propose possible suggestions on how to address these issues.

5.1 Down syndrome, ageing and dementia.

Ageing and life expectancy for individuals with DS has improved over the past decades (Bittles and Glasson, 2004). Positive changes in ageing may be attributed to better health surveillance and medical care, improved socio-economic entitlements and better education (O'Donovan *et al* 2018; Burke *et al*. 2014). Great improvements have been seen, with individuals with DS living well into their 60's a trend that will continues (Hourigan *et al*. 2018; McCarron *et al*. 2014). However, in this study it noted that those surviving into older age i.e. over 65 years, is a considerably small number and perhaps this could be attributed to the fact that the overall number participating in the study (N=109 surviving across all waves) is low.

It is noted that people with DS do approach ageing from a position of vulnerability, compared to adults with intellectual disability of other aetiologies, they present with

more complex health comorbidities and consequently present with a higher mortality rate at a younger age (Head *et al*. 2012). People with DS over the age of 40-years display the neuropathological manifestations typical of Alzheimer's disease and are predisposed to developing dementia due to an over expression of the amyloid precursor protein (APP) gene (De Simone *et al.* 2010). It can be seen from this study that the prevalence of dementia in those with DS over the 10-year period increased substantially from 15.8% to 35.5%, over double the prevalence in this short period. Considering that the prevalence of dementia in the general population in Europe is estimated at 6.8% in those over the age of 60 years (Ali *et al*. 2015), while the prevalence in people with an ID is 18.3% in those over the age of 65 years (Strydom *et al*. 2009) this finding in concerning however not unexpected bearing in mind that the pathological manifestations typically seen in Alzheimer's type dementia is seen in most people with DS (Godfrey & Lee, 2018). The data in this study highlights an increase in the prevalence of dementia over the 10-year period however the overall numbers are small. That said this two-fold increase overtime is reflective of diagnostic trends among those with DS. However, the prevalence rate would appear to be much lower than other research carried out throughout Europe, USA and Australia, for example Sheehan *et al* (2015) followed 85 people with DS and found that 64 of the participants (73.5%) had been diagnosed with dementia.

It is recognised that the diagnosis of dementia in people with DS is quite complex due to pre-existing cognitive impairment, while the standardised cognitive tests used in the general population are entirely unsuitable for people with underlying cognitive impairment (McCarron & Lawlor, 2003; Hithersay *et al*. 2019). In this study at Wave 1 those who did not have a diagnosis of dementia, 75.7% (n=56/147) did not have a cognitive assessment within the last 2 years and of those, over 67% had never had any type of cognitive assessment. This trend continued across the remaining waves of data collection, with over 63% in Wave 2 and 59% in Wave 3 who did not have a diagnosis of dementia had not had a cognitive assessment in the last 2 years from the time of their interview, with 55% in Wave 2 and 47% in Wave 3 never having any cognitive assessment. It is likely therefore that there may be much greater numbers of people with DS and dementia, but they simply have not been diagnosed. This study also

considered the objective assessment, Test for Severe Impairment (TSI) and identified a decline across the 10-year follow-up indicating a 4-point drop from 21 to 19 however these figures did not differentiate between the population with DS with a doctor's diagnosis dementia and without a doctor's diagnosis of dementia. When considering those with a doctor's diagnosis the scores indicated a drop from 18-14 across the 10-year follow up. This would indicate a notable change for those with a diagnosis. When you consider that those with a more moderate range of ID this 4-point drop would be somewhat alarming, and they ought to be assessed and have a full work up and comprehensive health and cognitive assessment. The TSI is a relatively easy test instrument and changes are indicative of some cognitive decline, so it is safe to say looking at the 4 point drop along with other manifestations such as decline in function, changes in personality, emotional lability amongst others would be a cause for concern. The 4 point drop alone with no other symptoms may just be that they are having a bad day and have no interest. It is important to look at the full clinical picture. With consideration to the results of the TSI in this study the numbers of individuals who completed the TSI across the waves was however low, which is a limitation of this particular observation. This test was made available to all participants in the study however with the low uptake the outcome has to be considered with caution. A possible explanation for this lack of participation could be that those who completed the test were individuals deemed capable of doing so by staff and those who had a pre-existing dementia may have been deemed too unwell to participate in the overall study along with this it is also possible that those with a more severe profound level of ID may have been inadvertently omitted as again their carer may have deemed them unable.

When considering diagnosis and assessment of dementia among those with DS another concern is the lack of specialist service consultations identified in this study for those with or without dementia. Just 4.8% in the first wave attended neurological services and whilst there is a year on year increase observed the consultation rate at 12.6% by Wave 3 is still low considering the levels of dementia and other complications of this condition, namely epilepsy. There are many reasons why dementia is so under recognised in this population and McCarron & Lawlor (2003) reported that caregivers are frequently not aware of the implications and consequences of cognitive or

functional decline in people with ID and DS. This is concerning as it is known that the early recognition and diagnosis of dementia is crucial in order to facilitate appropriate care packages, and to ensure individuals have access to the necessary supports they may require at that moment in time (Cahill *et al.* 2009). Considering the predisposition of those with DS to dementia and the fact that early recognition is critical to delaying onset and build reserve it is concerning that standards for assessment and care development are not policy throughout services. These do exist, however are not widespread (McCarron and Reilly, 2010). Lack of cognitive assessment and overall comprehensive assessment leads to poor outcomes, the onset of the dementia going unrecognised and overall poorer quality of life for the individuals with DS a fact supported by Lautarescu *et al* (2017) who asserted that the early symptoms of dementia often go unnoticed. Frequently, changes as a result of the onset of dementia are usually attributed to increasing age and are not challenged. Bittles and Glasson (2004) noted that services and primary carers lacked the basic knowledge for risk of dementia in people with DS. It is estimated that 1 in 3 people in the general population with dementia are not diagnosed (Pink *et al*. 2018) and approximately 50% of those with a formal diagnosis have no access to post diagnostic supports (Alzheimer's Society, 2015). These findings would appear to be also true of people with DS and would highlight the need for Memory Clinics to be established in the various services through the country of Ireland. Considering the overall trends in the literature and the findings of this study the prevalence of dementia is high among those with DS however there appears to be a lack of confirmed approach to assessment and diagnosis. Therefore, there is a need for public awareness of the prevalence of this disease in people with DS and the adoption of standards; overall this area warrants further exploration and discussion.

Successful ageing and increased longevity for people with DS has been well described in the literature. The mean age of survival in this study is 62 years which could be considered as a growing success considering that 50-years ago the life expectancy of a person with DS was 10-years of age (Hithersay *et al*.2019). There is a difference in mean age of survival and life expectancy and in this study the numbers are too small to draw conclusions, further research is required. That said life expectancy of people with DS has

extended and with advances in medical intervention and future research on health, especially in the area of dementia, the possibility of people with DS living a long successful life is conceivable.

Successful healthy ageing has a number of dependent factors one of which is having a positive attitude to one's own ageing (WHO 2019; Burke *et al.* 2014; Buys *et al.* 2008). Having a positive attitude bodes well for overall health and wellbeing. In this study overwhelmingly people self-rated their health as positive (from 81% in Wave 1 to 77% in Wave 3). Positive ratings have been recognised as being very important for the individual themselves and for professional health care providers, these attitudes can influence the person's quality of life and overall health status (Vina *et al*. 2007). However in this study there is a decline over the 10-year period reflecting a change as people got older, something that service providers need to be mindful of no matter how slight. Harnessing those positive attitudes, encouraging and promoting positive ageing overall does lead to better health and a more positive ageing experience (Burke *et al*. 2014; Buys *et al*. 2008). Walker (2015) noted that ID services had an advantage in that they have for decades promoted a model of increasing independence with greater community involvement and choice. In her study Walker (2015) proposed that ageing for people with ID should be addressed across the life span, planning for potential health anomalies, with particular attention to improve skills, independence and health outcomes, this is particularly true for those with DS. Having a positive attitude to how their overall health experience is a positive start on improving the quality of life for those with DS.

5.2 Down syndrome and chronic health conditions

Ageing successfully incorporates a number of factors from maintaining a positive attitude to staying healthy and mentally well. In a study by Bowling & Dieppe (2005) in the United Kingdom following 854 people over the age of 50-years, they observed that 75% (n=631) of their participants noted that they felt they were "ageing very well", and this cohort perceived successful ageing as being free from chronic illness and having a high level of independence and functioning. However, chronic illness or the presence of chronic health conditions is commonly a feature for those with DS. Startin *et al* (2020)

observed in their study, an association with increasing age, DS and complex health conditions; highlighting dementia, epilepsy, hypothyroidism, cataracts and hearing loss as being the most noteworthy conditions in the study, and these conditions were also central in studies by Glasson *et al* (2014), McCarron *et al* (2013), Krinsky-McHale (2012) and Alexander *et al* (2004). These conditions were identified as the conditions of interest in this study with an increasing prevalence across the 10-year period. Understanding the link between increasing age and chronic disease is of significant concern for people with DS, their family, caregivers and services who provide supports to this population (McCarron *et al*. 2013; Haveman *et al*. 2010; Esbensen, 2010).

In this study epilepsy rates increased from 19.2% in Wave 1 to 28% by Wave 3. Considering that the prevalence of epilepsy among the general population is less than 1% these increases are concerning as epilepsy has been significantly associated with the onset of dementia (Gholipor *et al.* 2017; De Simone *et al*. 2010). Epilepsy is a well-documented phenomenon from childhood to adulthood in people with DS and is an added cause of morbidity in this cohort (De Simone *et al,* 2010). With longer life expectancy comes the increased risk of Alzheimer's type dementia and seizure activity (Bittles *et al*. 2007). The triple copy of chromosome 21 and the overexpression of amyloid precursor protein are also documented as having noteworthy influences on neuronal susceptibility and late onset epilepsy (Bittles *et al*. 2007). With consideration to late on set epilepsy the findings from this study would demonstrate that people with DS do have a considerably greater risk of late onset epilepsy, where the incidence of this neurological disorder increased from 10.3% at Wave 1 to 11.5% at Wave 3. Perhaps the most interesting finding in this study in relation to epilepsy and DS is the possible link with dementia. Those who survived into that older cohort (65 years +) with DS are possibly healthier as the finding here indicate that those over the age of 65-years had no diagnosis of epilepsy or dementia. Studies by Sharma *et al* (2015); Bittles *et al* (2006); Menendez (2005) and McCarron *et al* (2005) would all agree that the incidence of epilepsy in people with DS & dementia was notably higher than those with DS without dementia. Considering the data from this study, findings would possibly indicate that increasing age is a prodromal to the onset of epilepsy. What is also interesting is that there is a definitive age trajectory of new onset epilepsy among those

with severe profound level of ID where rates increased from 31.7% to 52% across the 10-years of the data collection. Also, for women with DS the prevalence of epilepsy was notably higher than men in the first two waves however by Wave 3 the gap in the difference between both genders had decreased considerable from a 7% gap in Wave 1 to 2% by Wave 3. In fact, women with DS in this study appear to present with higher prevalence of conditions across all waves of data collection. A possible explanation for this could be the fact that it is reported that females with DS frequently present with amenorrhea or early menopause. In a seminal study by Schuf *et al* (1997) they reported 87% of women with DS experienced cessation of menstruation by the age of 46 years. Where there is a loss of sex hormones it has been suggested that this would encourage the development of accelerated ageing phenotype which could lead to the development of brain hypometabolism which is a feature frequently identified in menopausal women and prodromal Alzheimer's disease (Zarate *et al.* 2017). Overall this possible connection between lack of sex hormones could possibly present a link with poorer health for women with DS an area requiring further investigation. People with DS and epilepsy have a markedly greater risk of mortality (Robertson *et al*. 2015). It is imperative therefore, that service providers and health care professionals are not only made aware of the prevalence of epilepsy in this cohort but also provisions must be made to upskill staff to understanding and managing epilepsy for this vulnerable population (De Simone *et al.* 2010). Marriott *et al* (2014) demonstrated that this can be done with rational and realistic changes that should improve informed epilepsy care and improve the overall quality of life towards end-of-life for people with DS.

Thyroid disorder is common in the general population and is reported in more than 10% of people over the age of 80-years (Boelaert, 2013). Hypothyroidism is the most commonly reported endocrine problem in people with DS (Chakera *et al*. 2012; Perry *et al*. 2011). In this study 37.4% of participants with DS had a doctor's diagnosis of thyroid disorder at Wave 1. With increasing age, the prevalence of thyroid disorder increased to 46% at Wave 2 and to 50.5% at Wave 3 ten years later and these figures are comparable to other studies (Startin *et al*. 2020; Farriols-Danes, 2012; Chakera *et al*. 2012). However, when incidence of thyroid disorder is considered in this study there is very little change observed, in fact the incidence was slightly lower by 0.2% between

waves 1 and 3. This is an interesting finding considering a 15-year study by Prasher *et al* (2011) examining the prevalence of thyroid disorder. They concluded that there was little evidence to suggest that thyroid hormones changed in people with DS as they aged, and that other studies relation to thyroid disorder and DS were grossly exaggerated. They claimed that the association between DS and hypothyroidism was very much overstated and noted that annual screening was unnecessary and recommended 5-year screening. However, the evidence that thyroid disease among those with DS is more common with increasing age is overwhelming (Kinnear *et al.* 2018; McCarron *et al.* 2017; Head *et al*. 2012; Chen *et al,* 2007; Prasher & Haque 2005). Hypothyroidism can occur at any time from childhood through to adulthood. Thyroid disease can be difficult to diagnose in any group of people and particularly people with intellectual disability and DS. Many people with DS may not be compliant for blood testing and it has been reported that thyroid screening in DS is low (Fergeson *et al*. 2009). While most participants in this study reported they visited their GP on an annual basis very few, less than 4% across the three waves, were in receipt of endocrinology services. Having specialist consultation could possibly contribute to improved care and better diagnoses. It has been reported that a high proportion of people with DS had undiagnosed hypothyroidism which can present like cognitive impairment (Lavigne *et al*. 2017; Prasher & Gomez, 2007). This is very concerning considering the already predisposition of this cohort to dementia. Another aspect to also consider is diagnostic overshadowing whereby the symptoms of thyroid disease may be mistaken for dementia. Considering that hypothyroidism result in symptoms of fatigue, mental sluggishness which looks like cognitive decline, poor tolerance to the cold, constipation, depression and increased weight (Chakera *et al*, 2012; Perry *et al*, 2011) it is a condition should be regularly monitored among those with DS.

There are extensive studies reporting the incidence of visual impairment in adults with DS (Georglas *et al* 2014; van Splunder *et al.* 2006; van Splunder *et al,* 2004; Evenhuis *et al*, 2001; Van Buggenhout *et al,* 1999; van Schrojenstein Lantman-de Valk *et al*, 1994), and van Splunder *et al* (2006) identified that in people aged over 50-years , DS was the strongest risk factor for visual impairment. It was evident in this study that over the 10-year period visual acuity decreased with individuals reporting a 10% drop of positive

self-rated vision over the three waves. Visual deficits can have significant implications for people with DS, affecting levels of independence, cognitive functioning and day-to-day behaviours (Krinsky-McHale *et al*. 2015). The ability to communicate and decline in social skills is widely reported in people with DS and failing vision (van Splunder *et al.*2004; Evenhuis *et al*. 2001; van Schrojenstein Lantman-de Valk *et al.* 1994). The co-occurrence of visual impairment and DS is reported by van Splunder *et al* (2006) and the relationship between increasing age and poor vision was established in the study, while Evenhuis *et al* (2009) identified that visual impairment in people with ID led to additional disability. Visual impairment can impact on level of independence and overall quality of life, and the importance of annual routine eye examination cannot be overlooked. Considering this it is concerning that attendance at optician services across the three waves of data collection is not higher. At Wave 1 43% of individuals with DS had attended optician services in the previous year and this increased to 54% by Wave 3, whilst this increase is welcome it still is not sufficient as depletion in vision is linked so strongly to loss of independence and poor quality of life (van Splunder *et al.*2004). It has also been linked to increased behaviours of concern (Prasher, 1994).

Cataracts are the most widely reported eye condition in this population and are more prevalent in later years (Krinsky-McHale *et al*, 2012; Georgalas *et al,* 2014). The association between the onset of cataracts and ageing is evident in this study with the prevalence of cataracts increasing from 30.3% at Wave 1 to 46.7% at Wave 3. These findings are supported by Georgalas *et al* 2014 and Kinskey-McHale *et al* (2012) whereby they found that cataract disease highly prevalent among those with DS especially as they aged. What is concerning in this study is the uptake of surgical correction for cataract whereby 58% at Wave one who reported having cataracts did not have any surgical correction, this had positively decreased by Wave 3 to 36% however this still appears high. With surgery in mind there could be several possible explanations for the low intervention. Treatment options for cataracts vary and while surgery is an option, post-operative compliance must be considered. In her (2012) research, Krinsky-McHale and colleagues found that almost half the people with DS (43.5%) with cataracts did not have any medical treatment, and cataract surgery was carried out in only 15.6% of the study participants which is considerably lower that the

rates in this study at 63% at wave 3. A study by Li *et al* (2013) examined the outcomes of cataract surgery in 33 eyes of 20 patients with DS. While the surgery was successful and the outcomes were positive, the overall success was deemed suboptimal in patients with DS due to other ocular comorbidities. It is evident in the literature that cataract surgery does have a high rate of success and is recognised as being a safe procedure. However, it does not come without complication and compliance for post-operative management may be challenging for some people with DS. Overall when considering visual impairment, it must be remembered that it can contribute to accelerated cognitive decline associated with dementia in adults with DS (Rizzo *et al* 2000) so is therefore an aspect of concern in relation to the care of those with DS as they age. The impact of visual impairment on individuals with DS can be detrimental to their overall quality of life, and ophthalmology consultations should be carried out as a matter of routine health care screening (Georgalas *et al*. 2014; Evenhuis *et al*. 2009).

People with DS have a predisposition to hearing impairment due to the accumulation of adenoid tissue in the hypopharynx which can result in marked hearing loss (Trotter-Ross & Oleson, 2014). Evenhuis *et al* (2001) found that the prevalence of hearing loss in people with ID was 21% while contrastingly, those with DS had a much higher prevalence at 64%. The prevalence of hearing loss in the general population doubles with each passing decade and it is estimated that up to two thirds of people over the age of 70-years will present with significant hearing loss that will impede daily communication skills (Lin *et al*. 2011), while Tay *et al (*2006) reported that moderate hearing loss in those over 50-years did result in lower scores on the Mini Mental State examination (MMSE). Age related hearing loss is well recognised in people with DS and McCarron *et al* (2005) noted that 44.4% of adults with ID presented with hearing impairment. In this study the majority of participants rated their hearing as positive however for almost a quarter (from 22% in Wave 1 to 21% at Wave 3) their hearing rate was negative. Considering that this was self-reported and not objectively measured it could potentially be underrating the problems that exist especially with consideration to the predisposition for those with DS and the high prevalence rates of other studies. With consideration to this the uptake of hearing services overall is poor across the waves of data collection with just 17% at Wave 1 reporting attending hearing services

and this had positively increased to almost 30% however that means 70% of those in the study with DS were not attending hearing services which again indicates that there is possibly a higher prevalence of hearing difficulties, if we consider those who cannot self-report, and possible undiagnosed hearing impairment. Highlighting those who did rate their hearing negatively the majority were female, in the middle age group (50-64 years) and had a severe profound level of ID which reflect the results of self-rated hearing across the study, however by Wave 3 the differences between gender and level of ID were minimal. Age is interesting as again we can see that there was no one over the age of 65 years who reported poor hearing, this is because there were very few still living over this age and those that were, were probably healthier individuals. Of importance to consider is that hearing impairment and decline in cognition are well recognised in the ageing population (Wong et al, 2014). Lin *et al* (2013) noted that older adults with hearing impairment presented with 24% increased risk of decline in cognition and would typically have up to 40% faster decline than those with no hearing deficits which has been described as a "second hit" on the brain (Lin & Albert, 2014). Mulrow *et al* (1990) measuring cognitive function following the introduction of a hearing aid did highlight some improvement in cognitive scores while similar findings were noted by Acar *et al* (2011) who reported marked improvement in a group of adults over 70-years of age. This is an interesting aspect to consider for those with DS and it is suggested for further studies especially considering the levels of dementia identified in this study and perhaps unrecognised hearing deficit as hearing in this study was not objectively measured.

5.2.1. Health Disparities for People with ID and Down syndrome

The combination of age-related comorbidities and positive improvements in survival statistics over the last 50 years necessitates a more detailed investigation into the natural progression and longitudinal patterns of chronic health conditions in individuals with DS (Glasson *et al.* 2014). When people with DS are examined, a particular set of co-morbid health anomalies become apparent as does a different pattern of ageing (Mc Carron et al 2005, McCarron *et al.* 2013). Studies around the globe show that people with ID have a greater variety of health care needs compared to the general population (Haveman *et al.* 2010), and these needs are often unrecognised and unmet (O'Donovan

et al 2018; Cooper *et al.* 2004). The Disability Rights Commission (2006) reported on its findings that people with an ID had fewer health screenings and investigations which resulted in accessing specialist healthcare services at a later stage than the general population, which is evident in this study. Furthermore, it found that people with an intellectual disability are four times more likely to die from preventable diseases. Mencap's (2007) Death by Indifference report poignantly showed how six people with an ID died from treatable conditions. The LeDeR report (2019) provides the largest body of evidence in the world about the deaths of people with an ID. The report highlights the high incidence of deaths, issues around access to appropriate health care for people with ID and more importantly has contributed to the enhancement of quality of care and health outcomes for people with ID. This report has set out to address premature mortality and to highlight the health inequalities experienced by people with ID. A consequence of the LeDeR report is that the National Health Service (NHS) UK is fully committed to strengthening their commitment to the future care needs of people with ID.

Mason & Saor (2004) found reports of diagnostic overshadowing occurred for people with ID. It was felt that healthcare professionals may interpret changes occurring for people with an ID as being part of their disability rather than an indication that the person was ill. It is evident in this study that the chronic health conditions investigated are highly prevalent with an evidential ageing trajectory. Specialist healthcare screening is poor however everyone appears to go to their GP annually, therefore these visits to the GP ought to be approached with great care and planning. For people with DS to age successfully and healthily health surveillance cannot be underestimated. Robust health screening as a matter of routine for individual with DS across their lifespan is imperative and should be constantly highlighted as a matter of urgency with all those involved in their care otherwise deleterious consequences could unfold (Maatta *et al.* 2011).

5.3 Functional and residence change overtime.

Considering the chronic health conditions that people with DS experience with increasing age, the impact on their day-to-day life must be considered. Planning for the future and making decision about future care needs must be addressed. Considering the prevalence of dementia, epilepsy, thyroid disease and sensory impairment and critically

their association with increasing age in people with DS, service providers must think and plan long term (Towers & Wilkinson, 2014). In the first wave of data collection for this study 36.6% of participants with DS reported difficulty with bathing a figure that remained fairly constant up to Wave 3 with 34% reporting difficulty. The increase in difficulty across all ADL's is evident and problems with dressing had almost doubled from 17.6% at Wave 1 to 28.3% at Wave 3. Perhaps the most noteworthy increase in difficulty was seen in using the toilet and mobility. The number of participant with DS who reported that they had difficulty in using the toilet at Wave 1 was 9.9%, coupled with increasing age and prevalence of comorbid health conditions this figure increased to 18.4% at Wave 3. The National Positive Ageing Strategy (2018) highlights the vision for ageing in Ireland and supports the notion that all people with ID should be supported to age (physically and mentally) to their full potential, removing the barriers to participation in a normal life and providing opportunities for continued involvement in everyday life, in line with their preferences and needs. Considering that decline in mobility was the most widely reported difficulty for this population this could be considered the impediment that would impact the most on successfully ageing. In this study participants were asked if they had difficulty walking across the room and at Wave 1, 8.2% reported that this activity was problematic by Wave 3, 19.5% of the participants reported decline in mobility reflecting a two-fold increase. This is concerning as the impact on independence, getting around the community and networking with friends is greatly impacted by loss of mobility and it is known that having that social engagement, connecting with family and friends has a positive impact on ageing well (McCausland *et al* 2018). The trajectory of decline in activities of daily living and the complex health conditions identified in this study would suggest that with increasing age people with DS experience marked decline which will require increased levels of staff/carer support. This decline will have considerable implications for future care and in particular their place of residence.

This study found that over the 10-year period of investigation 32% of participants had moved their home between Wave 1 and Wave 3. During the period of data collection Ireland has been going through policy change in the form of decongregation (HSE, 2011). This strategy advocates the closure of residential or institutional type settings

and integration into the community. There is movement evident in this study however the vision of the strategy does not transpire. The majority of the moves were lateral, that is, moving from one type to a similar type of living circumstance, which does not specifically meet the vision of '*Time to Move on from Congregated Settings*' (HSE, 2011). The concept of ageing in place continues to be an aspiration for many services and many service providers continue to be challenged and have not yet provided a workable and acceptable response to an ageing population with ID (Wilkinson & Janicki, 2002). More supported care was also an option that was evident in this study, at Wave 2, 29.2% of this population moved to a home that provided increased levels of assistance to support the increased level of need, associated with increased age and/or the possible development of complex health co-morbidities. Three years on and at Wave 3 there was a significant change in the levels of support required by this population. In total, 32.3% of this population moved to a home that provided increased levels of assistance to support increased level of need, associated with increased age and/or the possible development of complex health co-morbidities. With increasing age, the health status of people with DS in the study changed over the 10-year period which is a possible explanation for these moves. Dementia and epilepsy; decline in mobility and perceptual difficulties; increased complex physical health conditions and the need for increased nursing support would obviously become a priority with the answer being more supported care. However, what is interesting despite these potential outcomes is the fact that those that lived at home with family/independently remained constant at 19% across the study. This could imply that those with DS living at home are either healthier and living well into older age or the burden of care is being carried by their families. This is an aspect of care highlighted by Brennan *et al* (2018) who noted that sibling care was becoming the norm with families as individuals with ID outliving their parents.

5.4 Challenges in Communication

It is well recognised that individuals with ID have both expression and perception difficulties with communication (Smith *et al.* 2020). Communication is a fundamental concept to live and function well in society, basically communicating well enables people to interact with each other. However, communication challenges do exist and do

impact on the health and wellbeing of individuals with ID including those with DS (Smith *et al*. 2020). This study asked participants about their communication challenges and how they spoke and made themselves understood. Between 23-30% reported no difficulty in communicating however what is very notable is that the majority of individuals had some level of difficulty communicating with almost a fifth of individuals unable to communicate at all. This is particularly important in terms of health care screening and assessment. Being able to discuss or communicate, by whatever means that the individual is able, is important when it comes to healthcare. In the study the participants were also asked if they had difficulty in communicating with healthcare professionals and again a large proportion had some level of difficulty with the majority noting they could only make themselves partially understood. Expressing their health concerns and anxieties about their health with the health care professional (HCP) is important for good health care outcomes and quality of life. While it is important for the person with DS to describe health concerns to the HCP is equally important that they are understood.

5.5 Conclusion.

Throughout their life people with DS are at higher risk for specific health conditions particularly the most widely documented conditions of dementia, epilepsy, thyroid disorder, visual and hearing impairments (Startin *et al*. 2020; Evenhuis *et al.* 2001). The high prevalence of these conditions requires a multi-disciplinary approach to care but unfortunately it was evident that health screening and health care surveillance was not sufficiently accessed in this study. Increased life expectancy was also noted however the life expectancy remains behind their non-disabled peers. Whilst life expectancy has changed for people with DS the focus of research and service development must shift in recognition of these age-related health problems of later life. There is an inherent need to understand their ageing processes and the complex health comorbidities that they present with at an earlier age than the general population (Esbensen, 2010). It is critical for service providers and for family/carers to be aware of the health disorders common in this cohort and to be aware of the importance of not only health care surveillance but also of heath care management of the common conditions associated with ageing and

Down syndrome. More work is needed to understand how healthcare must respond given that age related problems begin earlier than in the general population, and typically the common comorbidities affecting adults with DS are dementia, epilepsy, thyroid disorder, and sensory disorders, at specific age intervals (Glasson *et al.* 2014), and these inevitably result in additional disabilities (McCarron *et al.* 2014). Being proactive and being alert for decline at an earlier age and putting processes in place to manage these conditions will have positive implications for the individual's overall quality of life. The long-term chronic health conditions that adults with DS typically present with coupled with age related changes in physical health status, all of which impact on the person's ability to manage daily activities independently, have very important implications for not only service providers and family members but also for public policy (Walker, 2015; Esbensen, 2010). Services must respond to these issues for adults with DS in a timely fashion, they must look at their service user profile, plan ahead and ensure that ageing and DS is not turned into a crisis (McCarron *et al.* 2014). It is apparent that failing health has had a notable impact on the person with DS overall quality of life, affecting the levels of support required the types of support required and ultimately where they reside. It was notable that individual's needs change and there is need for improvements to enable ageing in place. Enabling this population to age with dignity and confidence in their own homes for as long as possible ought to be the vision of goal for all individuals as they age.

The next chapter, the final one of this thesis, will conclude the study and present recommendations and implications to clinical practice, service policy, education and research.

Chapter 6

Conclusion

6.0 Introduction

The association between DS and complex health comorbidities is well established (Coppus, 2017; McCarron *et al*. 2017; Jensen & Davis 2013), and the Convention on the Rights of Persons with Disabilities asserts that people with ID have the absolute right to receive the highest possible level of health care (UN, 2006).

As people with DS are now experiencing increased longevity compared to previous years, there is a critical need to understand their patterns of ageing and the comorbid health conditions that they experience. These conditions as highlighted in this study and common age-related changes in people with DS have vital implications for their future health care. Service providers throughout Ireland need to be informed of the common health conditions observed in people with DS as they age. Staff in services need to be alert for changes and the decline that is seen earlier than expected in people with DS compared to people with ID of other aetiologies especially with regards to onset of dementia. The Health Inspection Quality Authority (HIQA) (2019) has recommended that people with ID should have a comprehensive health work up annually including hearing test and ophthalmology. Based on the finding of this study and recommendations made by Bittles & Glasson (2013), Maatta *et al* (2011), Bertoli *et al* (2011) , Torr *et al* (2010), Henderson *et al* (2007), there is a need for these findings and the recommendations of these other studies be operationalised with in services. The improvement in healthcare surveillance and clear direction for services needs to be defined to ensure those who are responsibility for ensuring these physical investigations knows their role and conducts this role in an efficient and appropriate manner (Maatta *et al.* 2011).

Following the findings from this study, services need to think about precautionary health screening, that ideally should be routinely carried out on an annual basis to capture health conditions in a timely fashion and critically, people with DS should undergo well planned health investigation throughout their lives. This study has highlighted the high prevalence of multiple comorbidities including dementia, epilepsy, thyroid disorder and sensory impairment, and people with DS require a multidisciplinary

approach to their care. Unfortunately, many of these conditions are undiagnosed and Services need to strategically plan for the emerging and complex needs of an ageing population. The LeDeR report (2019) has highlighted that services need to be vigilant and be aware of the need for timely health care screening. It is crucial that the treatment, outcomes and experiences of people with ID can be understood and used to motivate advances in care. The provision of a structured health care programme will improve outcomes for this population.

6.1 Recommendations for Clinical Practice.

The key finding in this study highlighted the lack of knowledge among service providers and staff around the recognition of the common health conditions in this population, and this was seen in the under recognition of the conditions that were presented. While some services in Ireland appear to have excellent multidisciplinary supports for people with DS, with clinics that deal exclusively with this population, the findings from this study has noted that most services are poorly prepared and are not meeting the changing needs of people with DS as they age. There is a need for relevant training for staff in regard to the ageing and DS. It is important that the management structures in these services support staff not only to be facilitated to receive the necessary training to support an ageing population, but they must also plan for better integration of the necessary health supports to support people with DS as they age.

Dementia is the most widely reported condition among people with DS. Despite this, this study has highlighted that 47% of people with DS with no diagnosis of dementia have never had a dementia assessment. The American Association on Mental Retardation (AAMR) (1995) proposed a three-step process for the assessment of people with ID including those with DS. The first step is to recognise what changes are normal and what changes are deemed uncharacteristic. The second step involves conducting appropriate assessments and evaluating the findings of same. The third step is to devise a plan of care that best suits the person's needs, examining their preferences and future care needs.

Using the ICD-11 criteria for dementia, the assessment compares current level of functioning to previous level of functioning. Capturing and documenting change over time is vital. Taking a detailed history of the individual is possibly one of the most important aspects of the assessment process (Pink *et al*. 2018). Building familiarity with these criteria is crucial for all staff working with individuals with DS. Along with this the importance of reliable informants cannot go unrecognised during this critical process (Pink *et al.*2018). Best practice would recommend that people with ID should have annual screening for dementia from the age of 55-years (Lautarescu *et al.* 2017; Silverman *et al.* 2004; McCarron & Lawlor, 2003; Devenny *et al,* 2000; Alyward *et al*, 1997), while people with DS should be screened from the age of 35-years (Burt & Alyward,2000) This process of annual screening will inevitably supply a plethora of premorbid information providing detailed baseline knowledge of the individual whereby changes in cognition and function can be detected and measured over time (Lautarescu *et al.* 2017). Services have the responsibility to facilitate the development of staff to fulfil these roles and consequently contribute positively to the ageing of those with DS.

The assessment of cognitive decline and the subsequent diagnosis of dementia in people with DS with pre-existing intellectual impairment is challenging. Intellectual disability services are familiar with addressing the cognitive impairments associated with intellectual disability and many have developed programmes and multi-disciplinary responses to promote and support increasing independence and community participation, with many other services facilitating the training of staff to be familiar with support needs. Dementia symptoms challenge this service model because the symptoms are often masked until decline is quite substantial, there may be co-morbid health conditions, and the trajectory is for steady decline in memory and functioning. As a result, changes that the person is experiencing may not be diagnosed or understood. Programming models may no longer be appropriate, and staff may not understand how and why the care and support provided needs to change. Models of care need to be reviewed and revisited frequently. Additionally, the home where the person with DS lives may now out strip its capacity to give care with comfort and with safety. Some small pockets in the wider ID community throughout the country have well recognised

specialist memory clinics for people with DS. These memory clinics have been hugely successful in providing annual screening for this population, providing critical baseline data with which to measure change over time and supporting staff to support individuals with DS and dementia to age in place. However, this is not nation-wide. A systematic services wide approach to dementia identification, diagnosis and treatment is called for and a memory clinic offering a structure for the delivery of such an approach is required especially for those with DS.

No matter what the level of ID, these memory clinics ought to have clinical nurse specialists in dementia (CNS) who are trained to observe and document the typical changes in not only memory and function, but also in overall health status, personality and day-to-day behaviours of those with ID and DS. The need to have specialist support who are expert in comprehensive evidence-based screening, diagnostic work and assessment for people with all levels of intellectual disability is essential. Equally as important to assessment and diagnosis these CNS are would then be in the position to offer post diagnostic support and guidance to the person with dementia, their peers, staff and family caregivers.

Memory Clinics have been identified as a best practice model for screening and assessment, offering accurate diagnosis and post diagnostic support to people who receive a diagnosis (Jolley *et al*, 2006; Nagamatsu *et al,* 2013). The key functions of a memory clinic are identified by McCarron & Reilly (2010) and Cahill et al, (2009) as a model for:

- Screening and follow-up for individuals with Down syndrome over the age of 35 years.
- Improve the early detection of cognitive disorders and support comprehensive diagnostic work up for dementia
- Recognising and promoting remaining strengths
- Pharmacological and non-pharmacological treatment
- Training – to include brain training and improving cognitive health
- Training for families/staff – promoting innovations in care and best care practices

- Understanding the early, middle and late stage symptoms of the disease process
- Understanding the Behavioural and Psychological Symptoms of Dementia (BPSD)
- Preparing the individual, their family/carers for further decline
- Addressing Palliative Care

Dementia assessment and the development of a baseline report needs to be recognised by service providers as being essential. Pre-existing intellectual impairment means that the changes associated with symptoms of dementia are likely to be subtle. Therefore, detecting changes over time in day-to-day global functioning is imperative. Diagnosis is only possible if there is a baseline of optimal functioning for each individual against which to compare subsequent changes. There is a growing consensus that there should be annual screening for dementia offered to all persons with Down syndrome over the age of 35 years. The completion of the cognitive screen will assist the memory clinic team in clinical decision making, including a timely response to refer for comprehensive diagnostic work-up. Service providers need to realise the importance of and develop an evidence-based screening protocol consistent with international guidelines proposed by Burt and Alyward (2000); and choice of instruments are influenced by a number of factors including:

- Level of intellectual disability-comprehension and understanding, and attention.
- Language and communication competence.
- Availability of reliable informant who knows the individual well.

This study used the TSI and once a baseline score was established at Wave 1, a decline in scores was apparent over the following 2 Waves. Providing a timely diagnosis provides opportunities to talk to the person about their preferred health options and their future plans going forward. It enables the multidisciplinary team to meet and agree on a consensus plan of care for the individual and consider post diagnostic supports. It also helps family and carers to strategically plan ahead for future care needs including providing opportunities to age in place. Using tools like this provides services with the mechanism of creating the opportunity for timely diagnosis.

As part of the overall care package, services also need to consider post diagnostic supports. A comprehensive approach to diagnosis is likely to lead to the identification of both dementia symptoms and other health concerns including thyroid disorder and sensory impairment. Thyroid disorder can exacerbate the symptoms of dementia if left untreated. Appropriate referrals need to be made for co-morbid health concerns with recommendations for follow-up and additional testing. Services need to empower their highly trained CNS staff to make these referrals which may include more specialised testing e.g. neuroimaging, and/or for additional assistance from other multi-disciplinary team members e.g., social worker, occupational therapy, physiotherapy, speech and language therapist, dietitian, geriatrician, counselling, and palliative care.

A key activity of the specialist memory clinic includes providing training for all staff who work in the facility. It is recommended that all staff working with adults with DS are 'Dementia Aware' (McCarron *et al* 2016) This training would prove invaluable for services who provide care for those with DS and dementia as it enables staff to recognises change and the early signs of dementia, to make appropriate referrals, to advocate for the person when the HCP adopts a 'wait and see' approach to care. This training will inevitably lead to better health outcomes and a better quality of life overall for the person with DS. From a service point of view it is not enough to just understand the needs of adults with DS as the age. It is critically important that management structures and frontline staff work together to meet these needs, improve the goals of care and ultimately improve the overall quality of life of the adult with DS.

The association between dementia and new onset seizure activity for adults with DS is well described (Gholipour *et al* 2017; Sharma *et al* 2016; De Simone *et al* 2010; McVicker *et al* 1994) and the prevalence of epilepsy escalates with increased longevity (Sharma *et al* 2016). This study highlighted that in the 40-49 years cohort the prevalence of epilepsy grew from 12% - 20%; while in the 50-64 years cohort the prevalence of epilepsy grew from 29.5% - 44-4%. New onset epilepsy has significant implications for adults with DS and continues to be under recognised (Gholipour *et al.* 2017). The high prevalence of epilepsy for adults with DS coupled with progressive cognitive and functional decline, associated with dementia, puts this population at

increased risk of mortality (Kinnear *et al.* 2017; Robertson *et al.* 2015). The true incidence of epilepsy in adults with DS is not widely reported (Sharma *et al* 2016) and diagnosing epilepsy in people with DS and dementia is particularly difficult. Service providers need to acknowledge that adults with DS will more often necessitate changes of 'normal' healthcare practices (Gholipour *et al*. 2017; Evinhuis *et a.l* 2001). Diagnosing epilepsy requires detailed EEG investigations. Friedman *et al* (2012) suggested the use of mobile EEG systems, whereby the EEG equipment can be used in the familiar home setting, making the experience less daunting and upsetting for the person and their carers. It is important that all services are prepared with the knowledge and abilities required to recognise and manage epilepsy for this vulnerable group of people.

The prevalence of thyroid disorder increases with age and hypothyroidism is the most common category of thyroid disorder (Lavigne *et al.* 2017; Esbensen 2010). Various studies have highlighted the high incidence of undiagnosed hypothyroidism which can impact on cognitive health in adults with DS (Lavigne *et al.* 2017), and the high rates of hypothyroidism reported in this study highlights the need for regular blood testing and monitoring. Health care providers need to ensure that adults with DS have access to a robust annual health assessment that ensures routine blood testing. Collaborating with health care professionals with a specialist interest in DS and endocrinologists can be invaluable and can lead to an increase in diagnosis of this very treatable condition.

The prevalence of visual deficits and associated cataracts for people with DS is greater than the prevalence (8-50%) observed in older adults with ID without DS (Izquierdo 2015; Li *et al* 2014; Esbensen 2010), and the association between declining vision and ageing is well established (Santoro *et al* 2017; Georgalas *et al.* 2014; Jensen *et al.* 2014; Krinsky-McHale *et al.* 2012; van Splunder *et al.* 2006). Cataracts are the most widely reported ophthalmic disorder and are associated with increasing age (Krinsky-McHale *et al.* 2012). If visual impairment is not addressed the results for the person with DS can be devastating, and may impact not only on there visual acuity but also on their ability to manage their day-to-day life skills and inevitability will impede mobility. Services need to be alert to the high prevalence of ophthalmic disorders for adults with DS and routine annual screening should be a part of their annual health check.

Age associated hearing loss is also very common and up to 70% of adults with DS will present with degenerative hearing loss (Farriols Danes 2011). Hearing loss can have negative implications for the persons overall quality of life, and is noted to be more prevalent in adults with DS and dementia (McCarron *et al.* 2005). Social connections become increasingly more difficult to maintain with difficulty in understanding verbal cues and information. Social isolation may lead to the person becoming more withdrawn and the lack of stimulation may inevitably lead to decline in cognition (Farriols Danes 2011). The incidence of dementia increases considerably with hearing impairment (Liu *et al* 2019; Fritz *et al* 2016). Kinnear *et al* (2017) noted that carers and health care providers are more often unaware of the high prevalence of sensory impairments in people with DS, attributing decline in function and cognition to the persons underlying ID. It is imperative that services are aware of the prevalence and subsequent management of sensory impairment. The importance of regular health surveillance cannot be underestimated and can only contribute to improving the lives of adults with DS.

6.2 Recommendations for Changes in Policy

The findings in this study noted that a large proportion of the participants with DS had dementia and other associated health conditions when they died. These findings support the need to include people with DS in clinical trials going forward examining treatment options and exploring possibilities for the prevention of dementia in this population.

Promoting healthier lifestyles and in turn healthy ageing in people with DS will help to inform national strategies, promoting a better quality of life for older adults over the age of 40-years with DS. Service Providers need to be aware of the complex comorbidities that adults with DS present with and to be alert for earlier onset of these complex conditions (Esbensen, 2010), while McCarron *et al* (2014) went further and noted that understanding survival in adults with DS was critically important for their future care needs from a resource perspective in terms of services availability, environmental planning and staff competencies, in terms of changing needs of the client group. Startin *et al* (2020) highlights the importance of identifying people with DS who are at risk of various health comorbidities early on in the disease process will help

with future care planning and personalised care and will provide opportunities for earlier interventions. Promoting a better quality of life for adults with DS can only be achieved if carers and service providers are alert to the common health conditions of this vulnerable population (Head *et al.* 2012; Haveman *et al*. 2010). One of the more challenging presentations for this cohort is the increased risk for dementia, yet services are not adequately prepared for this challenging condition (Head *et al.* 2012). Most services throughout have no memory clinic to provide accurate diagnostic support and indeed most generic memory clinics are not yet ready for people with ID. Policy needs to be strengthened to meet these requirements.

More work is needed on understanding the manifestation of dementia and co-morbid health conditions in people with DS, potential markers that will predict later health compromises, preventative strategies, pharmacological and non-pharmacological treatment approaches and appropriate end of life care will provide services with the evidence they require to ensure evidence based care and improved service planning. This will not happen without policy change and improvement. Services need to strategically plan for the future with a multidisciplinary approach to this end of life of an individual. Future policy planning ought to guide service delivery and ultimately highlight the emerging and complex needs of adults with DS. For the individual with DS, policy planning will ensure accurate and timely diagnosis will facilitate appropriate investigations to rule out differential diagnosis and will enable an effective plan of care going forward.

6.3 Recommendations for education

Overall considering the lack of assessment, the high prevalence of chronic health conditions and the changing needs of individuals as they age many services are not prepared therefore it would be prudent for services to seek out appropriate education for their staff on the specific areas associated with ageing and intellectual disability particularly those with DS. Staff need to be confident practitioners, and this only comes with knowledge, therefore supporting staff to engage in further education is best practice for service providers. The findings from this study could be embedded in undergraduate and postgraduate curriculum for registered nurses in intellectual

disability to support a better understanding of the clinical manifestations of ageing in adults with Down syndrome.

6.4 Recommendations for research

This study examined specific area of the physical health of those with DS as they age, further research is required to examine all domains of the individuals lives. Exploration is required to specifically examine how social influences and aspects change for individuals, how their community connections and networks change and what kind of impact has this on both the individuals themselves and those who care for them. Their relationships with peers is frequently an overlooked aspect of individuals with DS as they age, with early onset poor health and the possibility of moving residence what impact is this having on their peers and those friends in their lives who may not understand. The need for further investigation in this area is also essential to build a complete picture of the lives of those with DS as they age.

6.5 Study Limitations

No study is without limitations and while the research engaged in every effort to overcome limitations they do exist. Across all the Waves of this study participants were asked to self rate and report their doctor's diagnosis of dementia, epilepsy, thyroid disorder, visual impairment and hearing impairment. Self-reported health questions allow the person to report on their own assessment of health according to their own definition of same. While the answer to the self-reported question is based on what the person thinks, and therefore is subjective, it is considered to be a statistically powerful to predict morbidity (Latham & Peek, 2013). In the study every effort was made to ensure accuracy by sending the PIQ in advance, encouraging individuals to seek the information from their GP and seek support in completing the questionnaire. However, as it is a subjective measure the accuracy of this method of reporting must be considered. Going forward, using objective measures to measure change over time would perhaps provide more accurate findings. Some participants had communication difficulties or perhaps changes in overall health status throughout the 3 Waves of the study made self reporting too difficult. Proxy reports were provided by a person who

knew the person with DS well and was deemed to be accurate however this ultimately would out rule the voice of some of the individual in the study. The sample for the study was selected from the IDS-TILDA study who randomly selected their participants from the NIDD. This database holds only those who are registered therefore excludes those who are not, it could be said that the numbers of unregistered individuals with ID were consequently missed.

6.6 Conclusion

The under recognition of health comorbidities for people with DS in services remains a critical issue in Ireland today. The key areas that must be developed include health management, quality of life and the promotion of successful ageing. The health conditions described in this study can have a profound impact on the overall quality of life on the person with DS. Some of the health conditions described are modifiable with good surveillance, reporting and follow-up. The study noted the lack of recognition of specific conditions and that differential diagnosis was a concern. Changes in current practice across the services to ensure early detection and accurate diagnosis of these conditions will improve outcomes for people with DS.

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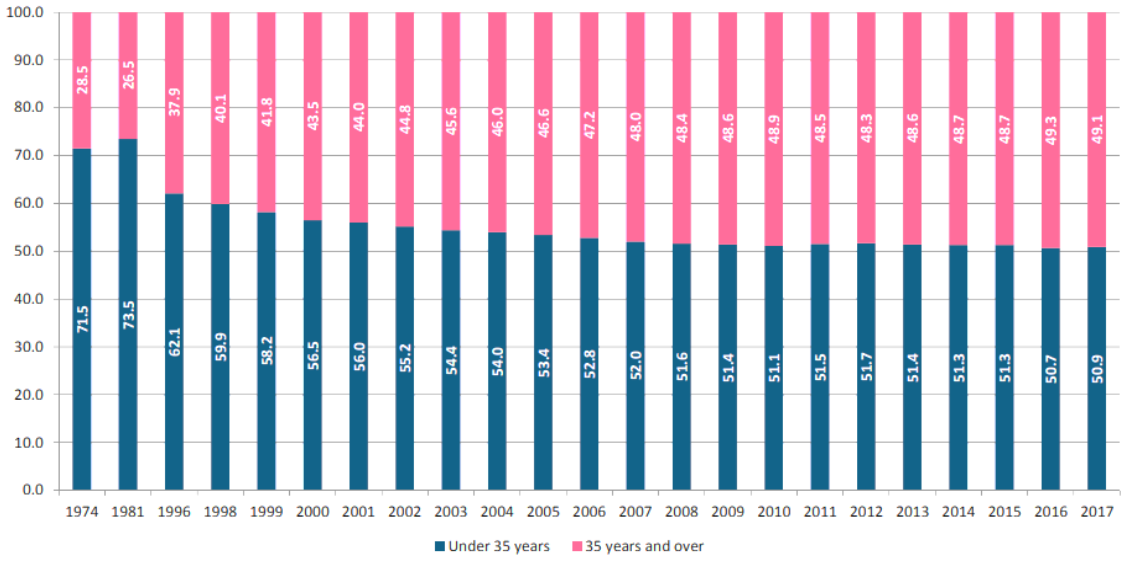
Appendices

Appendix 1: NIDD



Sources :

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National Intellectual Disability Database, Health Research Board, 2017

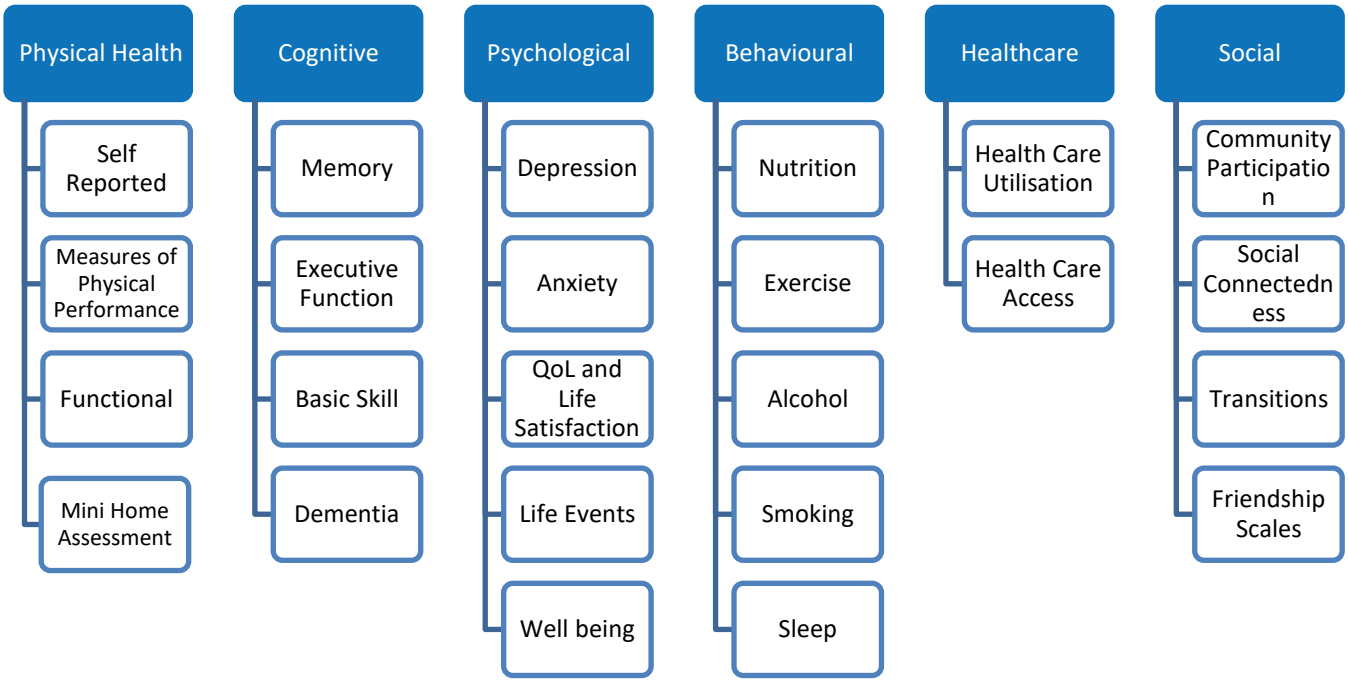
Proportion of individuals registered on the NIDD 1974 – 2017
Over age 35, level of ID – Moderate Severe or Profound

Source:

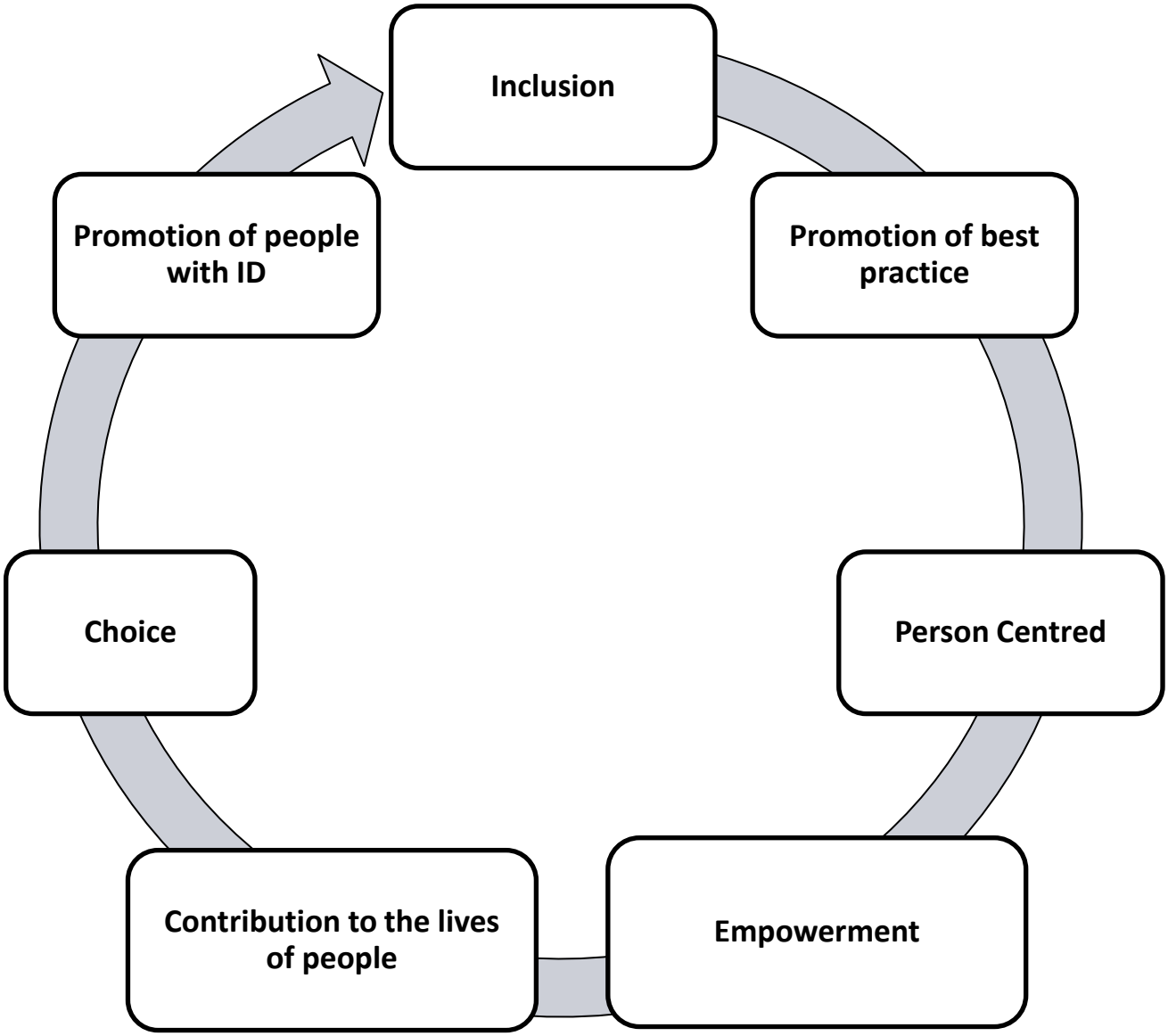
https://www.hrb.ie/fileadmin/2._Plugin_related_files/Publications/2018_pubs/Disability/NIDD/NIDD_2017_Annual_Report_Tables_Figures.pdf

Appendix 2: Keeping in touch

Appendix 3: IDS-TILDA conceptual frame



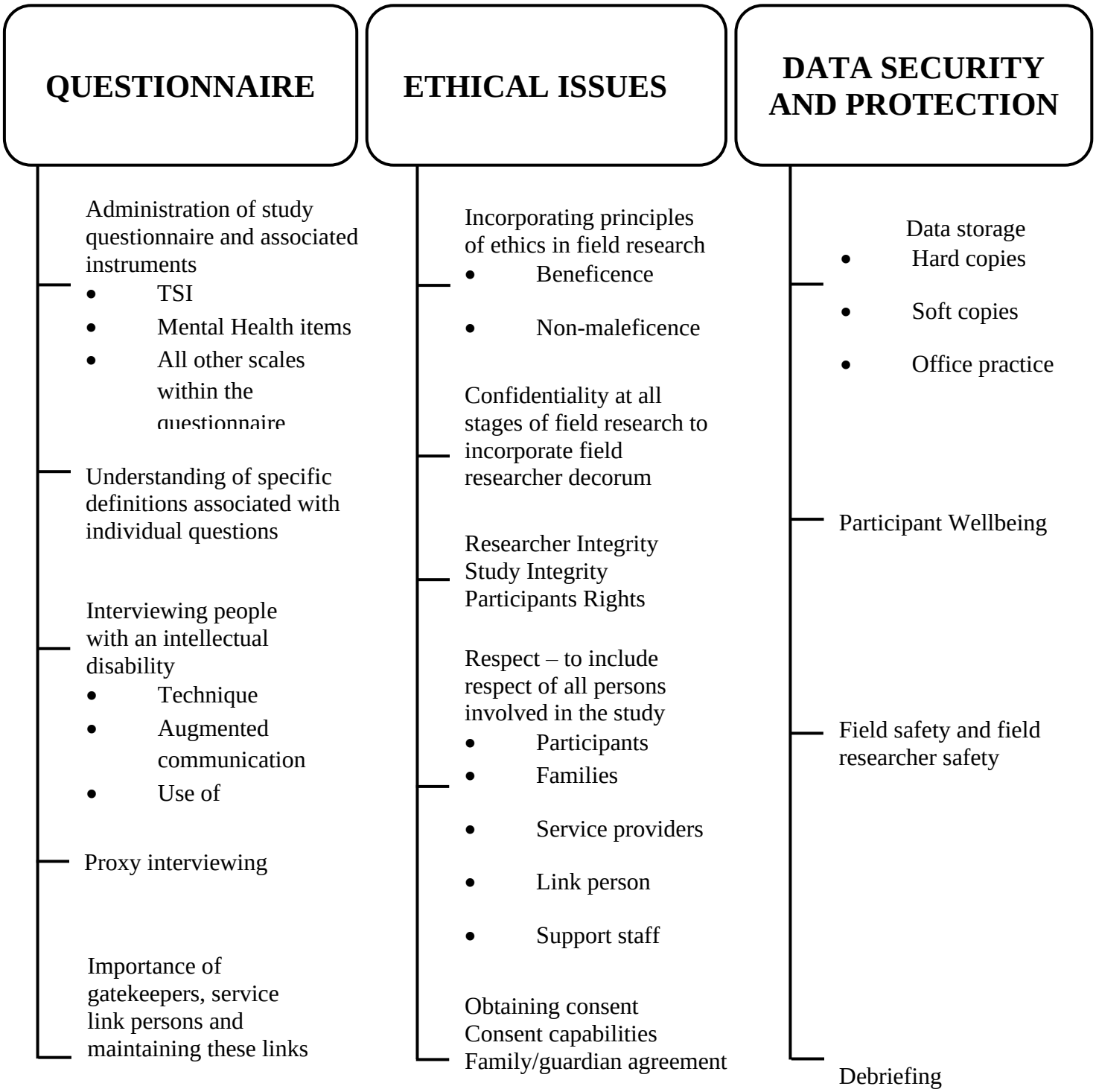
Appendix 4: IDS-TILD Values framework


Inclusion
Promotion of best practice
Person Centred
Empowerment
Contribution to the lives of people
Choice
Promotion of people with ID

Appendix 5: Field researcher training

Field Research Training Package – The Intellectual Disability Supplement to the Irish Longitudinal Study on Ageing

As part of the three day comprehensive training package the field researcher receive input on the background and justification of the study, the findings to date and the justification for each module included in the study. The training package incorporates the following broad elements.



Appendix 6: Appointment card

Intellectual Disability Supplement to TILDA
The University of Dublin, Trinity College,
School of Nursing & Midwifery, 2 Clare Street, Dublin 2

Telephone: 01-8963186/3187 Fax: 01-8963001
Email: danng@tcd.ie / haighm@tcd.ie

Thank you very much for your help with this important study.

If you cannot keep this appointment, please phone on 0831234567 to make another appointment

Appointment card

Evelyn Reilly will visit on

	Mon		**Tues**		**Wed**		**Thurs**	✓	**Fri**		**Sat**

Date: _____ **Time:** _____

At the beginning of the interview, you will be able to find out more about the study and to ask any questions you may have.

The interview will take about 90 minutes.

You can stop to have a break at any time.

We are going to do some health measures. Please bring a support person with you if you like.

Appendix 7: IDS-TILDA data management

Data Management Guidelines

The Intellectual Disability Supplement to The Irish Longitudinal Study on Ageing (IDS-TILDA)
August 2018

Intellectual Disability Supplement to TILDA
Trinity College Dublin, The University of Dublin
School of Nursing & Midwifery
2 Clare St
Dublin 2
Ireland

Telephone: (01) 896 3186 / (01) 896 3187
Fax: (01) 896 3001
Email: idstilda@tcd.ie

Contents

1.0 Purpose of these guidelines

The purpose of this document is to assist members of the IDS-TILDA team, other researchers, students and collaborators to fulfil their responsibilities with respect to the collection, storage and retention of data and records associated with, and arising from, their research activities.
This document establishes uniform data management standards and identifies the shared responsibilities for assuring that IDS-TILDA has integrity and that it efficiently and effectively serves the needs of the study.
These guidelines incorporate the day-to-day access requirements of IDS-TILDA paper data, electronic data and the equipment that gives access to the data.

2.0 Objectives

- Raise awareness that data constitute an important resource and that the value of data as a department resource is increased through its widespread and appropriate use while its value is diminished through misuse, misinterpretation or unnecessary restriction to its access
- Help ensure that data comply with all relevant legislation such as the General Data Protection Regulations (GDPR) and the Freedom of Information Act
- Raise awareness of data access vs. privacy issues and formalise procedures for access management
- Improve ease of access. Assure that data is easily located, easily accessed once located, and that people have enough information about the data to understand what they have found
- Facilitate database integration
- Reduce the redundancy of the data by defining an official record of reference

3.0 Data Classification

All data to be collected, processed and stored in IDS-TILDA should be identified and classified using the college data classification table below or similar.

Data Classification	Information	Description	Examples	Handling
Non Confidential	Public	Such data is available to anyone to see, and is often made available to public via the college website,	Publications, Articles, Presentations	Access to this data is not usually restricted.
	University Internal	Such data is generally available to all staff and students in College	Publications, Articles, Presentations	Access is usually restricted to members of College Staff
Confidential	Restricted	Personal data This data is only made available to authorised members of IDS-TILDA	Participants Name, address, telephone numbers,	Access is restricted to authorised members of IDS-TILDA only
	Critical	Sensitive personal data	Information relating to the mental & physical health of individuals	Access to such data is tightly controlled

3.1 Datasets related to individual study participants

Where data sets are collected relating to individuals you may also want to label data sets with regard to whether the data is.

- **Personal data** means any information relating to a living identified or identifiable natural person; an identifiable person is one who can be identified, directly or indirectly, in particular by reference to an identification number or to one or more factors specific to his physical, physiological, mental, economic, cultural or social identity, which is in, or likely to come into, the data controller's possession.
- **Anonymised data** is data prepared from personal data but from which the person cannot be readily identified by the recipient of the information.
- **Coded data** is identifiable personal data in which the details that could identify someone are concealed in a code, but which can readily be decoded by those using the personal data. Such coded data is not anonymised data.
- **Linked data** is typically used when it may be necessary to refer back to the original recodes for further information, or for verification. Unlinked data usually ensures confidentiality but prevents follow-up, verification or feedback.

With both linked and unlinked anonymised data it is sometimes possible to deduce an individual's identity through combinations of information. It is therefore essential that no data is removed, shared or copied without prior knowledge of the PI, Project Manager and Data Manager.
The most important identifiers are:

- Unusual disease or treatment
- Height (very tall/ short),
- Any text data,
- Partial address, geocode or similar
- Details of health professionals responsible for care
- Specific/unusual occupation or place of work
- Combinations of details or any of the above e.g. birth date, place of birth

4.0 Data Handling Procedures

Once data has been classified, appropriate handling procedures should be agreed on and documented. The researcher must ensure an agreed procedure is in place when linking data such as in the examples above, for examples aggregating the variables which can de-identify the specific variables concerned.

#	Data Type	Handling Procedures
1	**Critical Data**	• Is authorised for use by the PI, Project Manager, Data Manager, Project director & research staff only. • May not be removed from premises • Must be encrypted at all times • Must never be transported on insecure USB media
2	**Restricted Data**	• Is authorised for use by the PI, Project Manager, Data Manager & research staff • Must be encrypted at all times • May not be removed from premises
3	**Coded Data**	• Is authorised for use by the PI and Project Manager, Data Manager & research staff • Must be encrypted at all times • May not be removed from premises
4	**Participant Contact Records**	• Is authorised for use by the Participant's signed consent form. • Must be encrypted when stored & transferred • Can be transmitted by Email but only if fully encrypted

5.0 Daily Data Management

It is the responsibility of all members of the IDS-TILDA team, other researchers, students and collaborators to adhere to the following office policy.

- All members of IDS-TILDA are issued with their individual secure login and password.
- Each member is responsible for their login and password and should never give this out for other people to use. Passwords should be changed every three months.

- Members should lock their screen when their PC or laptop is unattended.
- Members should never leave personal data on their desk unattended.
- All members have access to an encrypted laptop or desktop. These must not be removed from the IDS-TILDA office.
- The laptops are in a secured press in office 1. It is the responsibility of each individual to ensure they leave the laptop back in the press when they are finished. The press is to be locked and the key put back in the designated area.
- The Mews has five hot desks which have five encrypted desktops for members of IDS-TILDA and associated researchers to use. IDS-TILDA has a secure and dedicated link for accessing the required data.
- At no time can the researcher copy, download, transfer data or datasets from the original dataset variables secured on the IDS-TILDA secure drive.

6.0 Responsibilities

Principal Investigator & Project Manager

- Responsible for ensuring project compliance with all relevant legislation
- Responsible for Data Management Guideline content
- Responsible for ensuring compliance with data management guidelines

Data Manager

- Responsible for backup and recovery of all project data
- Responsible for promoting ongoing awareness of data management guidelines among project staff
- Responsible for ensuring the integrity of dataset

Researchers

- Responsible for compliance with Data Management Policy

Administrative Staff

- Responsible for compliance with Data Management Policy

Appendix 8 TSI

Test for Severe Impairment

NAME: _____ DATE:_____

RATER:_____

Write down all responses verbatim that are different to those on the sheet.
You may repeat a question 3 times to gain subjects attention.

Max Actual

1. MOTOR PERFORMANCE Score

Score

A. Comb

"SHOW ME HOW YOU WOULD USE THIS COMB". Hand person the comb.

Correctly demonstrates combing (1)

____

B. Pen and top

"CAN YOU PUT THE TOP ON THE PEN?" Remove the top from the pen in full view of S.
And hand the pen and top to S.

Correctly puts top on pen (1)

____

C. Pen and paper

"WRITE YOUR NAME" Hand S. pen without top and place paper on table/desk
in front of S.

Writes name correctly (first or last name legible) (1) ____

TOTAL (3)

____

2. LANGUAGE-COMPRENHENSION

A. "POINT TO YOUR EAR"
"CLOSE YOUR EYES"

Correctly points to ear (1)

____

Correctly closes eyes (1)

____

B. Pens – red, blue and green

"SHOW ME THE RED PEN---THE GREEN PEN".

Place the 3 pens on the table spread out so that they have some space between them.

Correctly points to red pen (1)

____

Correctly points to green pen (1)

____

TOTAL (4)

____

3. LANGUAGE PRODUCTION

A. "WHAT IS THIS CALLED?" Point to your nose.

Correctly names nose (1) ____

B. Pens red and green

"WHAT COLOUR IS THIS PEN?" One at a time hold up red/green pen in front of S.

Correctly names red (1) ____

Correctly names green (1) ____

C. Key

"WHAT IS THIS CALLED?" Show S. the key.

Correctly names the key (1) ____

TOTAL (4) ____

4. MEMORY IMMEDIATE

One large paperclip - "WATCH CAREFULLY". Place clip in your hand so person can see.
Hold hands open

A. With hands open - "WHICH HAND IS THE CLIP IN?"

Correctly points to clip (1) ____

B. With hands closed – "WHICH HAND IS THE CLIP IN?"

Correctly points to hand with clip (1) ____

D. Move hands behind back - "WHICH HAND/SIDE IS THE CLIP IN/ON?"

Correctly points to the hand with the clip (1) ____

TOTAL (3) ____

5. GENERAL KNOWLEDGE

A. "HOW MANY EARS DO I HAVE?"

Correctly states two (1) ____

B. "COUNT MY FINGERS AND THUMBS: Place hands in front of S. Credit given even if no 1 to 1 correspondence between fingers and numbers. If S. only gives final answer ask "CAN YOU COUNT TO 10 STARTING AT 1?"

Correctly counts to 10 (1) ____

C. "HOW MANY WEEKS ARE IN A YEAR?"

Correctly states 52 (1)

____

D. "I AM GOING TO SING A SONG. IF YOU KNOW THE WORDS I WANT YOU TO SING ALONG WITH ME"

Softly sing 'Happy Birthday'

Correctly sings most of words (1)

____

TOTAL (4)

____

6. CONCEPTUALIZATION

A. Two large paperclips and one pen (Spread objects out on table)

"WHICH OF THESE IS DIFFERENT?"

Correctly points to or states pen (1)

____

B. 2 red pens 1 green pen

Place one red pen and one green pen down and hand the person the other red pen

"Put this next to the pen that is the same colour"

Correctly places the red pen (1)

____

C. One large paperclip.

Place hands out in front of person. Alternate the clip between your hands 4 times.

"WATCH ME MOVE THE PAPERCLIP, WHICH HAND WILL I MOVE IT TO NEXT?"

Correctly points to correct hand (1)

____

After person responds place clip in correct hand. If person was incorrect say "I WOULD PUT IT IN THIS HAND. NOW WHICH HAND WILL I PUT IT IN NEXT?"

Correctly points to correct hand (1)

____

TOTAL (4) ____

7. MEMORY DELAYED

Thread, key and paperclip - Place objects on the table.

"WHICH OF THESE HAVE WE NOT WORKED WITH ALREADY?"

Correctly points to the thread. (1)

____

8. MOTOR PERFORMANCE

"THANK YOU FOR SPENDING TIME WITH ME".

Extend hand to shake hands.

Correctly shakes hands. (1) ____

OVERALL TOTAL (24) ____

Appendix 9: Ethics approval

THE UNIVERSITY OF DUBLIN
TRINITY COLLEGE

SCHOOL OF MEDICINE
FACULTY OF HEALTH SCIENCES

Trinity College, Dublin 2, Ireland
Tel: +353 1 896 1476
Fax: +353 1 671 3956
Email: medicine@tcd.ie

Professor Dermot Kelleher, MD, FRCPI, FRCP, F Med Sci
Head of School of Medicine
Vice Provost for Medical Affairs

Ms. Fedelma McNamara
School Administrator
Email: fmcnamar@tcd.ie

Prof. Mary McCarron
School of Nursing and Midwifery,
Trinity College Dublin,
24 D'Olier Street, Dublin 2

10th July, 2008

Study Title: An Intellectual Disability Supplement to the Irish Longituddinal Study on Ageing (TILDA)

Dear Prof. McCarron,

Further to the meeting of the Faculty of Health Sciences Research Ethics Committee on 27th May 2008, I am pleased to inform you that the above project has been approved without further audit.

Yours sincerely,

Dr. Orla Sheils
Chairperson
Faculty of Health Sciences Ethics Committee

TRINITY COLLEGE DUBLIN
The University of Dublin
Research Ethics Committee
Faculty of Health Sciences,
Chemistry Building,
Trinity College,
Dublin 2, Ireland.
T:- +353 (0)1 8964255

Professor Mary McCarron
Dean of the Faculty of Health Sciences and Principal Investigator of IDS-TILDA,
Chemistry Building,
Trinity College Dublin,
Dublin 2.

7 February 2013

Study: Intellectual Disability to the Irish Longitudinal Study on Ageing (IDS-TILDA) – Wave 2

Dear Professor McCarron,

Further to the review of the modifications submitted for the IDS-TILDA Study Wave 2, I am pleased to inform you that the continuance of your ethical approval has been re-affirmed by the Faculty of Health Sciences Research Ethics Committee.

Wishing you the best of luck with your study.

Yours sincerely,

pp C Baff

Dr. Ruth Pilkington
Chairperson
Faculty Research Ethics Committee

Coláiste na Trionóide, Baile Átha Cliath
Trinity College Dublin
Ollscoil Átha Cliath | The University of Dublin

Prof. Mary McCarron,
Intellectual Disability Supplement to TILDA
School of Nursing and Midwifery
2 Clare Street
Trinity College Dublin
Dublin 2.

Ref: 151208

Title of Study: WAVE 3 - AN INTELLECTUAL DISABILITY SUPPLEMENT TO THE IRISH LONGITUDINAL STUDY ON AGEING (herein referred to as IDS-TILDA)

Dear Prof. McCarron,

Further to a meeting of the Faculty of Health Sciences Ethics Committee held in January 2016, we are pleased to inform you that the above project has been approved without further audit.

Yours sincerely,

pp Brian Brown
Prof. Brian O'Connell
Chairperson
Faculty Research Ethics Committee

Dámh na nEolaíochtaí Sláinte
Foireann na Coimise,
Coláiste na Trionóide,
Ollscoil Átha Cliath,
Baile Átha Cliath 2, Éire.

Faculty of Health Sciences
Chemistry Building,
Trinity College Dublin,
The University of Dublin,
Dublin 2, Ireland.

www.healthsciences.tcd.ie

Appendix 10: Easy read information booklet

TRINITY COLLEGE DUBLIN
COLÁISTE NA TRÍONÓIDE
THE UNIVERSITY OF DUBLIN

An Intellectual Disability Supplement to The Irish Longitudinal Study on Ageing

Intellectual Disability Supplement to The Irish Longitudinal Study on Ageing (IDS-TILDA)

WAVE 2
INFORMATION BOOKLET

This study is being carried out by Trinity College Dublin

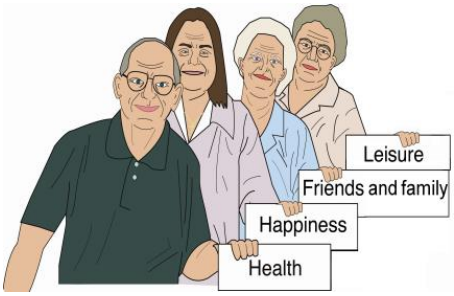
Dear Participant,

We are delighted to tell you we will be starting the interviewing again from April. This booklet reminds you about IDS-TILDA and tells you about some of the new things we would like to ask when we visit.

1. What is this study about?

This study is about growing older with an intellectual disability in Ireland.

Trinity College Dublin is doing this study.

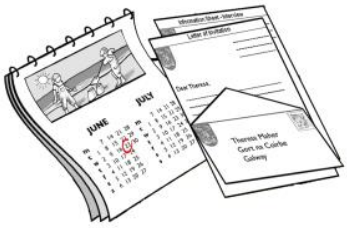


This study will help us to understand what is important to people

It will help us to learn about the

- health
- well-being
- and lifestyles

of people as they get older.

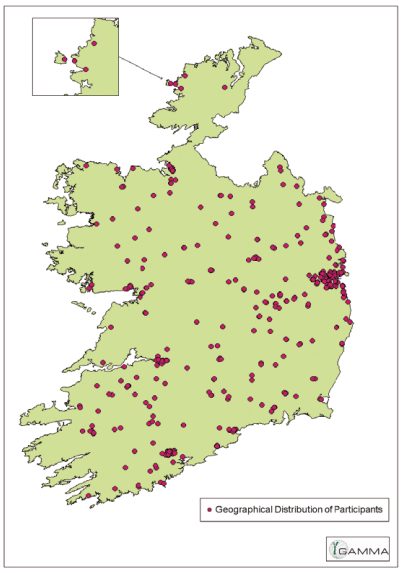


This is your second visit. We will see what has changed for you since your first interview.

2. Who is taking part in the study?

People with intellectual disability aged 40 years and over who were chosen for the first interview.

3. Who is taking part?



People from all over Ireland are taking part in the study.

You were one of the people selected to take part.

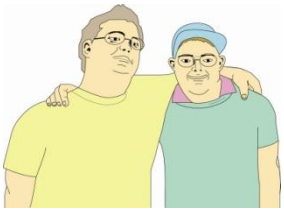
Thank you for answering the questions when we last visited.

4. What will I be asked this time?

You will be asked some of the same questions we asked before and some new questions about

- Activities you do everyday

- Your health and well-being



- Your friends

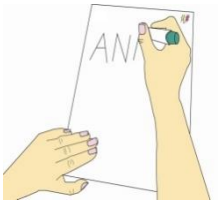
- Your family

- Interests and hobbies

- Work and retirement

- Your home

You can choose someone to help you answer questions



You will be asked to do some things like write your name

The researcher will put your answers into a laptop computer at the interview.

We will also be doing a health assessment.

This involves taking measurements, for example your height, your weight and your blood pressure.

We will also measure your bone strength and your grip strength.

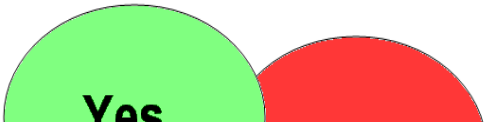
A researcher will be in touch with you to tell you more about the health assessment.

5. Who will visit you?

Trinity College Dublin

Researchers working on the study from Trinity College Dublin will visit you.

6. Do you have to take?



No. It is your choice to take part in the study. You can also change your

mind and stop taking part at any time.

Your decision will not affect the support you receive now or in the future.

7. How long will it take?

The interview will take about 90 minutes.

The Health Assessment will take about 30 minutes.

If the interview is too long you can stop and take a break

If you want we can call back another day

8. Are there any risks involved?

We do not know of any risks to you being involved in this study.

9. Will anyone else know what you tell us?

No. We will keep all your personal information private. Your name will not be in any report.

10. What should you do next?

A researcher will be in contact with you soon to see when it suits for us to come and visit you.

11. Is there anything else you need to think about?

Yes. You need to think about:
Where and when we can meet for the interview.
If you want to bring someone with you to the interview.

12. Who do you contact if you have any questions?

If you need more information or if you have any questions, please contact:

Professor Mary McCarron
Principal Investigator

Phone: 01-8963187 or 01-8963186

Email: mccarrm@tcd.ie

Eilish Burke
Project Manager/PhD Student

Phone: 01-8963187 or 01-8963186

Email: eburke7@tcd.ie

CONSENT FORM

IDS-TILDA PARTICIPANT ID W 2

Please read the information below and sign this consent form if you wish to take part in this second wave of the study.

Or write to us at:

Intellectual Disability Supplement to TILDA
The University of Dublin, Trinity College,
School of Nursing and Midwifery
24 D'Olier Street, Dublin 2.

Email: idstilda@tcd.ie

Intellectual Disability Supplement to TILDA
The University of Dublin, Trinity College,
School of Nursing and Midwifery
24 D'Olier Street, Dublin 2.

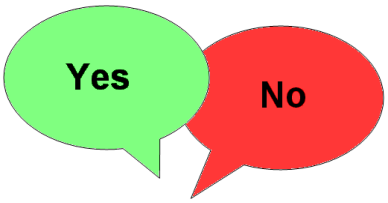
I agree with the following statement

Please tick √

I have gone through the information about this study ☐

I know who to contact if I have any other questions. ☐

Any questions that I might have had were answered. ☐



I know that it is my choice to take part in this study. ☐

I understand this study is for ten years and I will be visited again by a researcher from Trinity College Dublin. ☐

Trinity College Dublin

I understand that I will be asked questions about my:

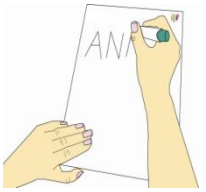
- life
- health
- work
- friends

☐

and things I like to do.

I have gone through the information about this study ☐

I understand that I do not have to answer questions I don't feel happy with. ☐



As part of the study, I know that I will be asked to try and do some things such as write my name. ☐

I do understand that I can stop taking part in this study when I want to. ☐

I do not have to give a reason.

I understand that all information I give during this study will be kept safe and private. ☐

I will not be named in any reports.

I understand that there are no known risks with this study. ☐

YOUR CONSENT

Your name: _____
Your phone number: _____
Your address: _____
_____

_____

Please sign your name:
_____
Date: _____

THE PERSON SUPPORTING YOU

I have supported the person named above to fill out this form. I believe they understand the information and have freely agreed to take part in this study.
Print name: _____
Relationship to the person named above: _____
Phone number:_____
Signature: _____
Date:_____
Please return this consent form to the field researcher before the interview commences.

IDS-TILDA, The University of Dublin, Trinity College, School of Nursing & Midwifery, 24 D'Olier Street, Dublin 2
Tel: +353 1 8963186/8963187 Fax: +353 1 8693001 Email: idstilda@tcd.ie

Contract of Agreement and Data Access Request Form
The Intellectual Disability Supplement to The Irish Longitudinal Study on Ageing (IDS-TILDA)
August 2018

Intellectual Disability Supplement to TILDA
Trinity College Dublin, The University of Dublin
School of Nursing & Midwifery
2 Clare St

Dublin 2
Ireland

Telephone: (01) 896 3186 / (01) 896 3187
Fax: (01) 896 3001
Email: idstilda@tcd.ie

Applicant Contact Details:	
Name:	
Address:	
Phone:	
Email:	

Applicant Contact Details:	
Name:	
Address:	
Phone:	
Email:	

Supervisor (if PhD/MD student):	
Name:	
Address:	
Phone:	
Email:	

IDS-TILDA Collaborators[1] supporting this application:	
Name of any additional IDS-TILDA Collaborators:	
Address:	
Phone:	
Email:	

[1] *In addition to the PIs Prof McCarron and Prof McCallion*

Description of Proposal (300 words maximum)

Background:

Aim(s):

Methodology & Study Design: (please include a planned completion date)

Which IDS-TILDA modules do you intend to use: (please highlight as many as are relevant)	
☐ Socio-Demographic Characteristics of Adults with an Intellectual Disability Ageing in Ireland	☐ Adults with an Intellectual Disability Ageing in Ireland as Members of their Families and Communities
☐ Physical and Behavioural Health of Adults with an Intellectual Disability Ageing in Ireland	☐ Mental Health and Cognitive Function of Older Irish Adults with an Intellectual Disability
☐ Health and Social Care Utilisation of Adults with an Intellectual Disability in Ireland	☐ Employment, Retirement, Day Services and Lifelong Learning
☐ Personal Choice, Planning for Daily Life, Personal Wellbeing and Beliefs About Ageing	☐ Objective Measurers
☐ Other (Give details)	

What this study will contribute to IDS-TILDA's data cleaning and /or variable construction:

Are you applying for funding to use IDS-TILDA data (please tick)?			
Yes		**No**	
Funder:			
Timescale:			

Signatures

By signing below I am confirming that I have read and agree to abide by the conditions as set out in this document and the following:

☐ IDS-TILDA Data Use Protocol and Procedure (please tick)

☐ TCAID Data Protection Manual (please tick)

☐ IDS-TILDA Data Management Guidelines (please tick)

_____ _____

Applicant Date

_____ _____

Supervisor (if applicant is MD/PhD student) Date

Please complete this form and send to IDS-TILDA Project Manager Margaret Haigh (haighm@tcd.ie) and the IDS-TILDA Data Manager Dr Rachael Carrol (carrolr5@tcd.ie). Thank you.

IDS-TILDA Approval

_____ _____

Name Date

Additional requirements (if any)

_____

Appendix 13: IDS-TILDA data use protocol

Data Use Protocol and Procedure of the Intellectual Disability Supplement of The Irish Longitudinal Study on Ageing (IDS-TILDA)
August 2018

Intellectual Disability Supplement to TILDA
Trinity College Dublin, The University of Dublin
School of Nursing & Midwifery
2 Clare St
Dublin 2
Ireland

Telephone: (01) 896 3186 / (01) 896 3187
Fax: (01) 896 3001
Email: idstilda@tcd.ie

Developed and reviewed by	Review Date
Prof. Mary McCarron, Prof. Philip McCallion, Eilish Burke	24/10/2011
Prof. Mary McCarron, Prof. Philip McCallion, Dr Rachael Carroll, Eilish Burke	12/03/2014
Prof. Mary McCarron, Prof. Philip McCallion, Dr Rachael Carroll, Dr Máire O'Dwyer, Eilish Burke	16/07/2015
Prof. Mary McCarron, Prof. Philip McCallion, Dr Rachael Carroll, Dr Mary-Ann O'Donovan, Dr. Eilish Burke	09/05/2017
Dr Rachael Carroll, Margaret Haigh	15/02/2018
Michael Foley	27/08/2018

Contents

1.0 Protocol Statement

This policy will provide guidance on the use of IDS-TILDA research project data findings and results for the IDS-TILDA research team itself, for PhD/MD Masters students and any person with whom the PIs may collaborate in the development of peer reviewed articles, presentations or any electronic or other dissemination.
In support of fundamental research ethical principles requiring the safeguarding of participant confidentiality and strict adherence to fundamental precepts of research integrity, all access to, use of and publications from IDS-TILDA data will be managed and approved by the IDS-TILDA PIs, supported by the Project Manager and the Data Manager.

2.0 Background

Currently, the IDS-TILDA dataset is only available with the permission of the PIs. With suitable protection, the PIs will consider access to the dataset as they recognise the criticalness that publications from IDS-TILDA data be developed including by students and collaborators to:

(1) increase knowledge about the ageing of people with intellectual disability across a range of disciplines
(2) support the training of researchers
(3) influence consideration of key policy concerns
(4) offer data reports likely to encourage support for subsequent waves of data collection for the benefit of people with ID

3.0 Definition of Terms

IDS-TILDA: The Intellectual disability supplement to the Irish longitudinal study on ageing in Ireland hereinafter referred to as the project.
Investigators or PIs: Refers to the Principal Investigators Professor Mary McCarron and Professor Philip McCallion.
Research team member: All staff employed in the IDS-TILDA research study project either full time, part-time or under a contract basis as well as PhD/MD/Masters students supervised by the PIs.
Collaborators: All persons invited to contribute, write and work in a joint intellectual capacity with the IDS-TILDA team and investigators.
Public Use Dataset: At a future time, the IDS-TILDA team will make the IDS-TILDA dataset available in an appropriate data archive. The specific protocol for the use of such a dataset will be posted there. In the interim, there is no public use dataset.

4.0 Scope of the Protocol

The protocol applies to all uses of the data other than by the PIs and research team staff they designate. The scope of this document may be amended in light of any future developments.

5.0 Purpose of the Protocol

The purpose of this policy is to provide all IDS-TILDA research team members and collaborators clear guidance regarding the use of project data to include all analyses,

publications, oral presentations, electronic dissemination and/or other dissemination of the data.

6.0 Data User Roles and Responsibilities

All persons wishing to have access to data shall complete an IDS-TILDA Contract of Agreement and Data Access Request Form and furnish this to the PIs (hardcopy and email).

Until such time as data files are posted as a public use dataset, the IDS-TILDA Contract of Agreement and Data Access Request Form must include:

(1) specification of a time period to access the data
(2) an explanation of the research question and related analyses
(3) intended type of publication/dissemination
(4) a signed agreement that all PI-stated restrictions on use will be adhered to and that a copy of the final data analysis file will be provided to the PIs for the IDS-TILDA data archive
(5) acknowledgement in all products that IDS-TILDA is the source of the data, using the language provided by the PIs
(6) acknowledgement that the PIs will be the final arbitrator for all publication and data access-related decisions

6.1 Prerequisites for Access to Data

Conducting research carries the responsibility to protect the confidentiality and privacy of participants. Access to data will therefore be carefully managed and granted to appropriate persons on the sole discretion and permission of the PIs. All persons wishing to access data shall do so in adherence to the IDS-TILDA Contract of Agreement and Access to Data Procedure and in line with the Data Protection Manual.

6.2 Data Access and Use

In providing access to the data, the PIs reserve the right to directly write articles themselves, support PhD/MD/Masters students they supervise in the development of approved theses, and work with research team members and approved collaborators on the development of additional articles.

(1) After receiving signed approval for the project proposal (see Contract of Agreement and Data Access Request Form) the research team member or collaborator will request the specific variables to be used in the proposed analysis.
(2) Data will be accessed on secure computers located in the IDS-TILDA office[2]. All efforts will be made to protect these computers from a data breach, e.g. strong passwords used, screens locked when not in use, avoidance of unsecure websites.

[2] *Any alternative arrangement to data access will only be made in exceptional circumstances and only with the written agreement of both PIs. In all cases, data should always be on a secure, encrypted laptop belonging to IDS-TILDA. 'Exceptional circumstances' will ONLY refer to qualitative data access.*

(3) At all times, access to the main IDS-TILDA quantitative dataset will occur at the IDS-TILDA office level ONLY.
(4) Permission to access will ONLY be for the approved variables and timeframe.
(5) No data can be removed, copied or accessed outside the IDS-TILDA office. No laptops will be removed.
(6) The research team member or collaborator will participate in an exit process at the end of their research, as outlined in the Data Protection Manual
(7) Agreement to the terms of this protocol and procedure is understood on signature of the Contract of Agreement and Data Access Request Form.

Access to the data is granted with the additional understanding that the PIs will have the opportunity to be among the authors on all publications, determine the scope of each article, authorise order of authorship and establish the timelines for publication. These requirements are further specified in 6.3 and 6.4.

6.3 Authorship

For research team members, the PIs will authorise all aspects of the proposed publications. This includes the content of the paper, early discussion of publication and authorship practice for the work, the appropriate authorship, the place of publication, the protection of intellectual property rights and any release of results on the Internet.
An approved thesis will be the work of the PhD/MD/Masters student and will be completed within a timeline agreed with the PIs.
Articles by research team members and collaborators are encouraged by the PIs and the scope, content, timeline for completion and order of authorship of articles to be completed will be negotiated in advance with the PIs. In particular, the PIs reserve the right to take back control of an article if the agreed timelines are not met. This may include changing the originally agreed order of authorship.
The PIs will be the final arbitrators of all related decisions.

6.4 Criteria for authorship

To be recognised as an author, a research team member or collaborator shall:

- Contribute substantially to the IDS-TILDA creative process within any of the following areas: generation of hypotheses, data collection, analysis or interpretation of data.
- Contribute substantially to the preparation of the article to be published either through preparation of drafts or through critical revision.
- Accept, in writing, the final draft and be prepared to take public responsibility for the content.
- Meet the specific requirements for authorship required by any journal considered for publication.
- Within reasonable limits, accept responsibility for the contents of the publication as being based on honest research.

For members of the research team or collaborators, the PIs must have the opportunity to review and approve the final version of any article/presentation/report to be submitted for publication or other type of dissemination.

7. Data Access Procedure for Non-Research Team Members or Investigator supervised PhD/MD students

After receiving signed approval from the PIs for the project proposal (see Contract of Agreement and Data Access Request Form and Data Protection Manual):

(1) Researcher or PhD/MD student (with a co-sign by their supervisor) will complete a variable request form (approved list will be available on confirmation of proposal) and submit the form to the Project Manager. Should the researcher seek variables beyond the approved list (e.g. identifiable variables such as open questions), a separate request shall be made to the PIs, cc the Project Manager and Data Manager.

(2) The Project Manager will obtain related access permissions to the Drive on the secure IDS-TILDA server (this will require at least one week after receipt of data access approval).

(3) On receipt of access permission, the Data Manager will compile the required list of variables on the Drive.

(4) The dataset is then available to access on a computer available in the IDS-TILDA office; the procedure for doing this will be demonstrated on the first day at the office.

(5) All analysis shall be undertaken in the IDS-TILDA office. To that end, approved researchers and PhD/MD students must arrange dates for 'hot desk' access (this can be arranged when requesting folder setup) by liaising with the Executive Officer or through the Project Manager or Data Manager.

(6) No data shall be removed, copied or accessed except within the assigned folder on the secure Drive.

(7) No data shall be removed, copied or accessed outside the IDS-TILDA office.

(8) Agreement to the terms of this protocol and procedure is understood on signature of the Contract of Agreement and Data Access Request Form.

The investigators will be the final arbitrators of all related decisions.

References

An Bord Altranais (2007) *Guidance to Nurses and Midwives regarding Ethical Conduct of Nursing and Midwifery Research.* An Bord Altranais: Dublin.

Trinity College Dublin (2009) Policy on Good Research Conduct accessed on https://www.tcd.ie/research/dean/TCDGoodResearchPractice.pdf 24.10.2011.

Appendix 14 IDS-TILDA data protection regulations

IDS-TILDA GDPR Regulations

Data Statement for IDS-TILDA

IDS-TILDA is hosted by the Trinity Centre for Ageing and Intellectual Disability (TCAID). The aim of TCAID is to address the inequalities and health disparities that occur for people with intellectual disability as they age.

We do this by conducing rigorous engaged research on the physical and social determinants of health and wellbeing and translation of findings to inform models of policy and practice. To do this work, we gather, store and use 'personal data' such as names, email addresses, photographs and telephone numbers. All data are important to us and we always aim to treat them in a clear, secure manner as a way of respecting the people who gave us the data about themselves.

IDS-TILDA Data

While we already provide information in our cover letter, our information leaflets and on our consent forms, we want to ensure that we can keep everyone informed on all aspects of how we work. In December 2018, the Department of Health issued a document called 'Guidance on Information Principles for informed consent for the processing of personal data for health research'. Prompted by this guidance, we are happy to share the following information:

1. General

Title and Purpose of the Research

IDS-TILDA is the Intellectual Disability Supplement to the Irish Longitudinal Study on Ageing.

It is a longitudinal study researching ageing in Ireland among people with an intellectual disability aged 40 and over. The aim of the study is to identify the principal influences on successful ageing in persons with an intellectual disability, and then determine if they are the same or different from the influences for the general population.

Personal Data Collected and Used

Name; address; phone number; age; date of birth; living arrangements; friendship and family; quality of relationships; social activities; physical, mental and cognitive health; health care utilisation; medication, occupation and functional limitations. In addition, personal physical health measures and biological samples will be collected to include - Activity patterns; Blood drop sample; Blood pressure; Bone density; Breath; Calf circumference; Cognitive functioning; Foot health; Grip strength; Hair sample; Height; Mobility and falls risk; Nutritional health; Oral health; Waist and hip measurement; Weight.

Using Pseudonymised Data

IDS-TILDA removes information like name, address, contact details, etc. from the collected data. This information is kept apart from the other data so that the person cannot be identified. This process is called pseudonymisation. Pseudonymised data is used in IDS-TILDA because the researchers need to go back to the same people during the next wave of data collection and must have a way of keeping in touch with them.

Benefits of IDS-TILDA

The purpose of IDS-TILDA is to understand the ageing process for people with an intellectual disability, what health conditions they have and how this relates to the wider

determinants of health and wellbeing such as social contact, living arrangements, etc. This research endeavours to inform policy and practice in the area of intellectual disability so that both the State and services can be better equipped to support healthy ageing among this population.

Finding Out about Health

Anyone who takes part in the IDS-TILDA Health Fair is provided with a report card on data gathered. This information may lead them to take action about any of these results. People also have a choice about whether they give samples of blood, hair or breath. If they agree, they can request the results. If, after analysis, the sample shows something unexpected, the person will be informed.

Giving Consent

IDS-TILDA seeks consent on an ongoing basis throughout the data gathering process – we call this 'process consent'. We check that consent is given freely and voluntarily. If someone decides not to consent, it will have no adverse consequences. People can also withdraw their consent by contacting the Project Manager, Margaret Haigh at the contact details below – by letter, by email or by phone. Personal information will be deleted from the dataset, apart from any data that has already been analysed or in the process of being published.

Complaints

If someone involved in IDS-TILDA would like to make a complaint, they can do so by contacting the Project Manager, Margaret Haigh at the contact details below – by letter, by email or by phone.

2. Details on who is conducting the research

Data Controller

The data controller for this research is Trinity College Dublin and the Data Protection Officer:

Data Protection Officer
Secretary's Office, Trinity College Dublin
Dublin 2, Ireland.

Principal Investigators

IDS-TILDA has two Principal Investigators for the study. Prof Mary McCarron is Professor of Ageing & Intellectual Disability in the School of Nursing and Midwifery and is the Director of the Trinity Centre for Ageing and Intellectual Disability. Prof Philip McCallion is Director of the School of Social Work in Temple University, Philadelphia. He is a visiting Professor at Trinity College Dublin.

Prof Mary McCarron
Trinity Centre for Ageing and Intellectual Disability
2 Clare Street
Dublin 2, D02 CK80

Prof Philip McCallion
c/o Office of Admissions
College of Public Health
1101 W. Montgomery Avenue
Suite 370, Philadelphia

Data Processors

A data processor is used to hold and process data which has been gathered by researchers in the field. One of the data processors is engaged to design the CAPI (Computer Assisted Personal Interviewing) data collection tool which will be used in face-to-face interviews. It holds the data securely until such time as it is transferred to the appropriate secure servers within Trinity College Dublin.

This data processor for Wave 4 is Behaviour & Attitudes, Milltown House, Mount St Annes, Milltown, Dublin 6, D06 Y822. This company acted as data processor in Wave 3 of the study and has the skills and expertise needed to fulfil this role. Behaviour & Attitudes subcontract work to two data management companies: IT Force, 27 Fitzwilliam Street Upper, Dublin 2, D02 TP23; and Kefron, 53 Park W Rd, Cherry Orchard, Dublin, D12 F8RK.
A number of other data processors are used in order to hold and process data which has been collected during the Health Fair. This is due to the fact that the study itself does not have the capacity to analyse the samples provided.
The data processors for Health Fair data in Wave 4 are: for blood samples, MedLab Pathology, Unit 3, Sandyford Business Centre, Sandyford Business Park, Dublin 18; for breath samples, Biochemistry Department, Laboratory Medicine Directorate, St. James's Hospital, James's Street, Dublin 8; for dietary information Open Lab, Floor 1, Urban Sciences Building, 1 Science Square, Science Central, Newcastle Upon Tyne, NE4 5TG, United Kingdom; for hair follicle samples, Dresden LabService GmbH, Tatzberg 47-49, D-01307 Dresden, Germany.
Physical activity / sedentary behaviour information will also be gathered using devices and software created by PAL Technologies Ltd (50 Richmond Street, Glasgow G1 1XP, Scotland, UK) but they will not have any access to the data gathered using their devices or software.
In addition, interviews carried out as part of the End-of-Life part of IDS-TILDA research will be transcribed by Audiotrans, 25 Pinelawn, Oldbawn, Tallaght, Co. Dublin, D24 KX4P, a company specialising in transcription of research material.
IDS-TILDA also uses couriers to ensure that secure information is delivered straight to the IDS-TILDA offices. For international packages, IDS-TILDA will use FDS Worldwide and for Ireland-wide deliveries, IDS-TILDA will use Cyclone.

Confidentiality

All researchers involved in the gathering or analysis of IDS-TILDA are bound by the rules and regulations that protect the identity of anyone involved in the research.

Funding

IDS-TILDA is funded by the Health Research Board and the Department of Health. These funders do not have access to any of the data provided for the study.

Data Protection Training

All staff in the Trinity Centre for Ageing and Intellectual Disability and researchers using IDS-TILDA data must complete the online module on GDPR provided by Trinity College Dublin. All field workers are also provided with a half-day workshop on the influence of GDPR and research integrity on IDS-TILDA as part of their comprehensive training to participate in IDS-TILDA.

3. Obtaining, use, storage and disclosure of personal data

Data from Healthcare Providers

Principal data will be collected, with consent, from those involved in the study, i.e. people with an intellectual disability, their family members or carers. In certain cases, key workers employed by disability support services may support the person to participate in IDS-TILDA.

Processing Promise

Personal data will be processed only as is necessary to achieve the objectives of IDS-TILDA and will not be processed in a way that damage or distress will be caused to the data subject.

Duration of Data Retention

As this is a longitudinal study, all data will be retained for the duration of the study plus a

further five years, in a pseudonymised format. By its nature, this study compares data over different waves of the study and the data from early waves must remain available to researchers in order to compare with data from later waves.

At the End of the Study

While IDS-TILDA is in progress, the data will remain psudonomysed and available to TCAID / IDS-TILDA researchers for analysis. Any personal contact information will be available to core IDS-TILDA staff only for the purposes of the logistics of the study. On completion of the study, the data will be fully anonymised and a review will take place of what data could be archived in a public repository which would not identify any living participants of the study.

Disclosure of Data

Any relevant contact data is disclosed to a data processor (currently Behaviour & Attitudes) for supply to an IDS-TILDA field researcher in order to identify interviewees for the study. All data collected by the IDS-TILDA field researchers is also handled by that data processor for the sole purpose of supply to IDS-TILDA.

Once the data is gathered, it is no longer retained by the data processor or any field researcher. The data is then only disclosed to researchers working with the IDS-TILDA team and is not disclosed to anyone external to that group.

For the health fair information, pseudonymised samples are created. These are then supplied to data processors for analysis, without any other personal data. Once analysis by the processor is complete, samples are destroyed and results are returned to IDS-TILDA. In any case where a data processor wishes to retain data, it will be done with the explicit consent of the person who supplied the sample.

Non-Commercial Purposes

No data will be used or disclosed for commercial purposes. In the event of any commercial use being proposed, re-consent would be sought.

Data Security

All paper copies of data are held in locked filing cabinets in secure, alarmed offices in the Trinity Centre for Ageing and Intellectual Disability.

All electronic copies of data are held in secure folders on the TCAID server only accessible by a small number of TCAID staff.

Risk Assessment

A Data Protection Impact Assessment, which identifies whether there are any risks involved in research such as IDS-TILDA, is being carried out.

Data Leaving the State

None of the data from the main study is intended to leave the State. Data gathered from the Health Fair will be sent to data processors in the UK and in Germany for analysis. This data will be pseudonymised prior to sending.

4. Research Ethics Committee

Ethical Approval was granted in December 2018 by:
Health Sciences Ethics Committee
Health Sciences' Faculty Office
Ground Floor, Chemistry Building
Trinity College Dublin, Dublin 2
Email ethicscommittee@tcd.ie

The Principal Investigator was the Dean of the Faculty of Health Sciences at the time of the application. All TCAID staff who work on IDS-TILDA are employees of Trinity College Dublin.

All approved research requires six-monthly Monitoring reports and an End of Project

report.
No conditions were attached to the research by the Research Ethics Committee

5. Lawful basis for the research

Under Article 6 and Article 9 of the General Data Protection Regulation, the legal basis for the processing of data is that the person has given their explicit consent for the use of their data in pursuit of the aim of IDS-TILDA.

6. Follow-up contact

As this is a longitudinal study, all participants will be followed up during future waves of the research. IDS-TILDA also has a 'keep in touch' strategy where it distributes a newsletter to participants and greeting cards at Easter and Christmas.

7. Project Manager

For further information about these or any other aspect of the IDS-TILDA study, please contact:

Margaret Haigh
Project Manager, IDS-TILDA
Trinity Centre for Ageing and Intellectual Disability
2 Clare Street
Dublin 2 D02 CK8