Aging in the intellectually and developmentally delayed community: comorbid I/DD + dementia.

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Abstract

Humankind gains of greater longevity increase greater numbers of aging people worldwide, including those with Intellectual and Developmental Disorders (I/DD) (Alvarez, 2015). When an I/DD is present in an individual, and age-related disease (such as dementia) is also diagnosed, it is often referred to as a comorbid diagnosis. Comorbid diagnosis simply means that there is more than a single disease or condition present in one body at the same time. Diagnosis of a comorbid I/DD + dementia can be complicated and needs to be based on previously existing levels of cognitive disabilities in the individual with I/DD. Establishing an individualized record of peak performance baseline functioning at age 25-35 builds a threshold for measuring future age-related decline in these individuals with I/DD. Baseline function can differentiate if a newly appearing symptom in an individual with I/DD is a normal aging-associated process, or a disease-related-decline.

Through a literature review, informed by research based scholarship, program based publications, and professional sources (including professional websites) on topics of normal aging, I/DD, AD and dementia, this project will 1) describe prevalence/incidence studies of comorbid I/DD + dementia diagnosis in locations around the world, including challenges and conflicts of the research, 2) explore *non-disease-related, age-associated patterns* common in the I/DD community, 3) examine common indications of *disease-related cognitive decline* of individuals with I/DD + dementia, 4) discuss support frameworks available for comorbid (I/DD + dementia) individuals and caregivers, 5) suggest where to look for support networks that can assist caregivers and family members, post-diagnosis, in best practices for the comorbid individual, and themselves, as caregivers

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To all of the professors I have had throughout my time at K-State I say thank you. Learning is what keeps a mind active alive.

Dedication

This Masters Report is dedicated to my family. Without their patience and willingness to “take a back seat” I would have never gotten it done. Special dedication to Tom, my husband for supporting, encouraging, and helping me to know that I could do it.

I would also like to dedicate this project to a few of my beloved I/DD friends who have "gone on ahead of me”. Their life experiences, which were shared with me, have been the inspiration and the motivation to research the growing concern of aging in the Intellectually and Developmentally Delayed community.

Preface

From the dawn of time, humankind has coped with conditions that impaired general functioning of affected individuals within their communities. These conditions often affected cognitive abilities and/or physical abilities. Culturally, individuals with these disabilities were often stigmatized. Once termed mental retardation, in modern times this condition is termed Intellectually and/or Developmentally Delayed (I/DD). The use of the term I/DD is currently considered more respectful to the individual by the professionals who assist them.

Figure 1 (Appendix C page 62) is a historical timeline showing how the life of an individual with Down Syndrome has changed over time. Down Syndrome (DS) falls under the diagnostic umbrella of Intellectual and Developmental Delay. During the early parts of this timeline (in the 1700s), people with DS, and many other I/DDs, were incarcerated. Those people who were not incarcerated were kept in their homes and never publicly discussed or viewed. Many individuals born with an I/DD disability, including DS, did not live longer than 9-12 years and were rarely seen outside of a small area within their homes (Down Syndrome Timeline, 2018).

In some individuals, disease-related cognitive decline presents itself at the end of the lifespan. In the 1700s, Philippe Pinel named this condition 'dementia'. A colleague of Pinel's, Jean Esquirol, termed the condition 'Des Maladies Mentales' and wrote a book about it. Esquirol claimed in *Des Maladies Mentales* that this condition was caused by age, syphilis, mercury, wine, masturbation, and menstrual disorders (Boller & Forbes, 1998; Daly, n.d.). The people who developed this condition were often incarcerated or institutionalized into workhouses or poorhouses. Other families with respectful attitudes towards the elderly, supported and cared for their elders in their family home (Anderson, 2015).

Dementia has been found in recorded history as 'memory loss related to aging' as far back as 2000 BC in Egypt (Boller & Forbes, 1998; Daly, n.d.). I/DD and dementia were part of society but had no common meeting point until the Industrial Revolution began. With the onset of the Industrial Revolution medical science began to improve personal health, find cures for diseases, and increase life expectancy (Bagosklonny, 2010). Through the progress of modern medicine, humanity's average life span has steadily increased (Bagosklonny, 2010).

Seventy-five percent of people in London in the year 1662 in Graunt's life table (as cited in Bagloskonny, 2010) were expected to die before they reached the age of 26. In the year 2010 Blagosklonny predicted that with the civilization's continuing progress in medicine and human services, human life expectancy was increasing at 2.5 years per 10 years of life. Much of this extension has occurred in the last century.

Alvarez (2015) states that the lifespan of a person with Down Syndrome (DS) in the 1920s averaged around 9 years of age. By 1960, it was 30 years of age, rising to 55 years of age by 1993. At Wrentham Developmental Center in Massachusetts, in 1999, the average human lifespan for patients with DS was 61-years of age (Alvarez, 2015). The average lifespan of a person with Down's Syndrome, as of 2019, is still at 60 years (ndss, 2019). This is compared to the estimated life expectancy of the general population in the United States, which is 78.8 years (CDC, 2018).

Alvarez (2015) reports that over half a million people with I/DD live in the United States alone. What does this mean for people with I/DD? In older ages they are coping with normal aging-associated conditions and/or aging disease-related declines, that have never before been addressed. These declines are compounded with pre-existing cognitive delays. Because of this,

there is a pressing need to research aging-associated disease-related declines in the I/DD community and best practices to cope with them.

Chapter 1: Introduction

The first topic I will address is the prevalence and incidence of reported comorbid I/DD + dementia diagnosis in communities across the globe. Historically, the most prevalent comorbid combination is Down's Syndrome (DS) and Alzheimer's Disease (AD).

Before discussing prevalence, or incidence, let's clarify which disease(s) we may be talking about associated with each taxonomic umbrella. What diagnoses fit under the Intellectually and Developmentally Delayed umbrella? What conditions, or diseases fit under

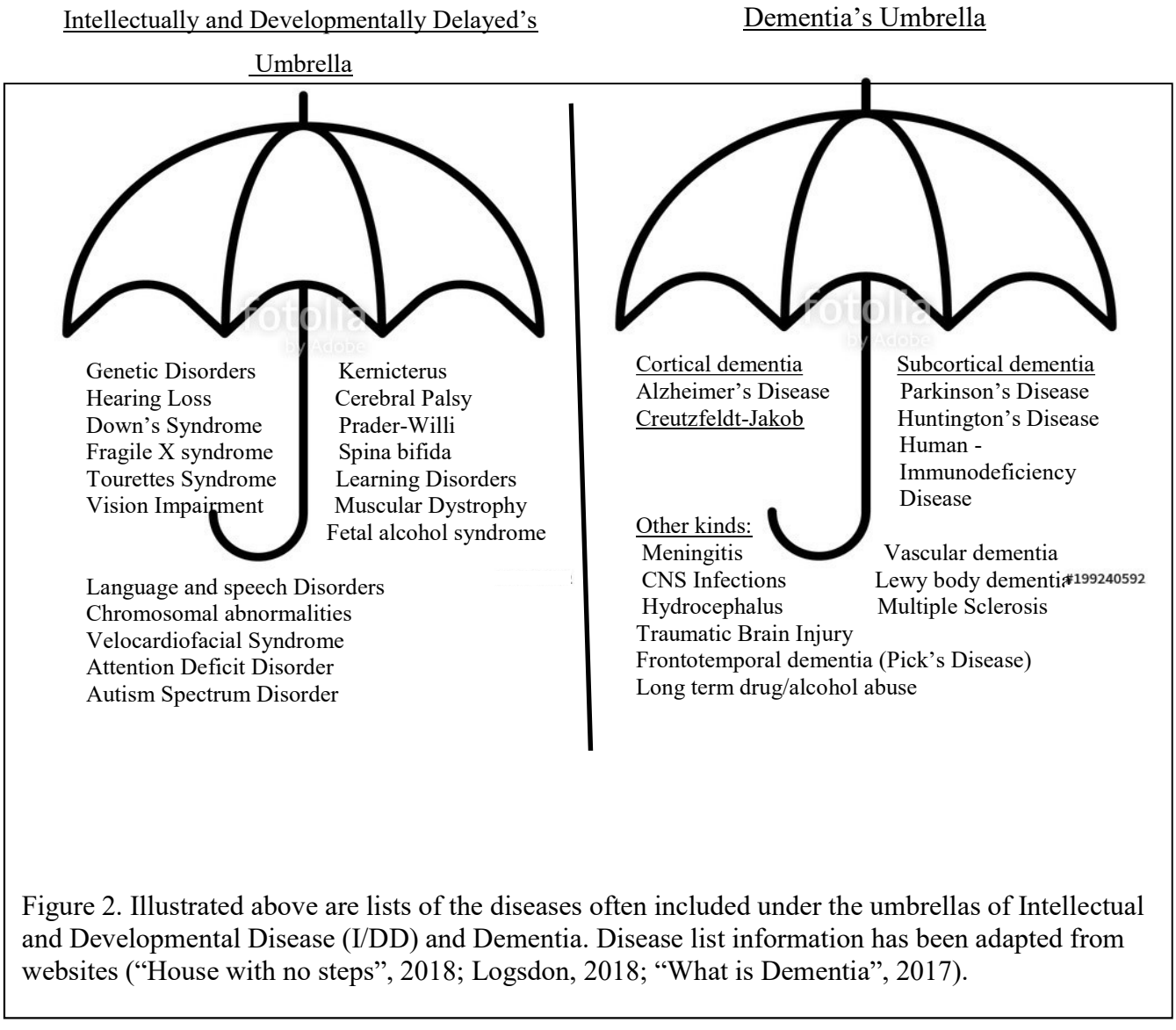

Figure 2. Illustrated above are lists of the diseases often included under the umbrellas of Intellectual and Developmental Disease (I/DD) and Dementia. Disease list information has been adapted from websites ("House with no steps", 2018; Logsdon, 2018; "What is Dementia", 2017).

the dementia umbrella? Figure 2 lists different diseases and disorders that are associated with these separate conditions. Not included in Fig. 2, under the I/DD umbrella, is the rare case where the intellectual or developmental delay cannot be explained by another disorder or disease. In contrast, many people when they hear the term dementia, automatically think AD, but dementia is caused by other diseases or disorders as well. As can be seen in Figure 2, there are many happenings in a person's life that can lead to the development of dementia. A young person may have an I/DD diagnosis of Autism Spectrum Disease from birth and belong to a family with several individuals who have Parkinson's Disease. Parkinson's (a progressive disorder) can result in related dementia as part of its course ("How Parkinson's Disease Progresses", 2018). If the Parkinson's associated dementia is diagnosed in the person with a pre-existing I/DD diagnosis, the two diseases together can create a comorbid I/DD + dementia. The difference in symptoms and needs of this person with ASD and Parkinson's related dementia, in contrast to the symptoms and needs of a person with comorbid DS + AD, offer an example of how a single set of guidelines for diagnosing a comorbid I/DD + dementia cannot be sufficient. Therefore, an individual baseline of cognitive function must be created for each person. I will discuss this in a later chapter. Let's get back to prevalence and incidence.

With extended life expectancy, worldwide, are those with I/DD living long enough to develop aging-related dementia? Are there enough epidemiological studies from different areas of the world to determine the extent of the malady? Economic, cultural, and social patterns affect research on prevalence and incidence data in different areas of the world. This paper will discuss how and why these patterns affect research. Standardization of epidemiological data collection and diagnostic protocols worldwide will help to improve data analysis and remove discrepancies among studies.

What are baseline cognitive functioning levels for average middle age individuals with I/DD? What tests and procedures are used to measure an individual with I/DD's cognitive levels of performance and at what stage in life should this be accomplished? What are normative aging signs in people with I/DD? 'Normal aging' for those individuals not affected by I/DD can appear much different than it does for someone who has pre-existing cognitive delays. In addition to describing the methods of determining a baseline functioning level, I will also compare age-related decline due to primary aging in a person with I/DD to those declines due to disease-related decline. With this comparison, I hope to offer caregivers of individuals with I/DD a method to recognize the most common indications of disease-related decline. An adult peak functional baseline for the cognitively delayed individual with I/DD must be created to be able to recognize most common indications of disease-related increasing cognitive decline.

Once the baseline functioning levels and most common indications of disease-related cognitive decline have been determined and compared, I will discuss support frameworks (with an emphasis on the USA) that are available for comorbid individuals and their caregivers when the signs of increasing cognitive decline might signal dementia has been clearly identified. What is available to help people with a new diagnosis of co-morbid I/DD and dementia? Do caregiver(s) need any special support? As a part of the framework, I will also briefly suggest best routine post-diagnostic practices that can assist caregivers and family members to create the best level of lifestyle in their environment.

What does all this information mean? Once prevalence, incidence, a baseline, normal vs. disease-related factors, support framework, and best practices have been discussed, I will discuss how to apply this information. How can the person with I/DD and their caregiver benefit from

this information? As well, what are future directions of research that might benefit both people with comorbid I/DD + dementia and their caregiver(s) to live more fulfilling lives?

Chapter 2: Prevalence & Incidence

Determining a prevalence of co-morbid I/DD and dementia worldwide

It is easy to view the world with a sort of tunnel vision based on one's surroundings. Living in a part of the world that values higher education and is more socially advanced in the treatment of all citizens can skew our perception of topics such as disability and dementia. In more developed countries, research on dementia in individuals with I/DD is a topic of interest and there is much data that is available. However, this is not the case worldwide. Ferri et al. (2005) point out in their opening summary on the global prevalence of dementia that "evidence from well-planned, representative epidemiological surveys is scarce in many regions" (p. 2112). Therefore, a consensus approach must often be used to estimate an overall result. There are two main consensus methods or approaches used for consensus. These are the Delphi and the nominal group technique. The Delphi is more commonly used in research related to medical science. When there is inadequate or conflicting published information on a topic, insights of appropriate experts are gathered, and a median of the answers of those experts is forecast as the result of the research question. "The theory behind the Delphi method is that the interaction of experts may lead to a reduction in individual bias" (Cam, McNight, & Doctor, 2002, p. 990). Jones and Hunter (1995) point out that although the consensus approach may often be more accurate than actual data, it is an estimate or a forecast of experts. From this author's viewpoint, a forecast of experts could easily be considered a good point for creating a hypothesis for research, but it should not be considered as final data. Data collections can sometimes prove a research hypothesis to be wrong. Could a Delphi Consensus Approach also find a null hypothesis?

According to Lemmi et al. (2015) in A Systematic Review for International Initiative for Impact Evaluation, there are an estimated one billion people globally with disabilities, including long term physical, mental, intellectual, and sensory impairment(s). Of this one billion, 80% reside in low and middle-income countries. If 80% of the world's disabled reside in low and middle-income countries, it would be of higher accuracy if Ferri et al.'s (2005) statement was changed to read: "evidence [of dementia in persons with I/DD] from well-planned, representative epidemiological surveys is scarce in many regions [that are low and middle-income]" (p. 2112). Lemmi et al.'s (2005) research is a 4th systematic review for Community-based Rehabilitation. This summary is published by International Initiative for Impact Evaluation (3ie). 3ie publishes information that is used by WHO and the UN for information, so it should be considered trustworthy. Changing the wording of their statement would make the literature closer to an accurate representation. This literature does not, however, mention a comorbidity of disabilities. It does, however, point out the lack of data that exists on the topic of disabilities (comorbid or not) in large areas of the globe. This is important because developing a prevalence value for comorbid dementia + I/DD depends on first identifying the underlying disability and its prevalence in different locations.

Third world country examples: Africa

At the beginning of this section, the ease of falling into tunnel vision was mentioned. One possible reason for needing a consensus approach in many areas is that life is viewed by people all around the world through different cultural lenses. Each lens is restricted by its own mode of0 tunnel vision. Disabilities do not respect race, wealth, education, or culture, and appear everywhere that there are human beings. In some third world countries culture dictates a totally different outlook at disabilities, including dementia. With increasing media attention on

economics and populations in many of these third world countries, disabilities are beginning to catch the attention of researchers in places like Afica and Middle Asia. Research on the topic of I/DD seems to be increasing rapidly in several African countries. Literature from African studies is easier to access. For this reason, I have chosen Africa as an example of third world countries and research studies in progress.

In 2006, the United Nations (UN), through the Convention on the Rights of Persons with Disabilities, created the Committee on the Rights of Persons with Disabilities (UN DESA, 2016). In Africa, the Committee on the Rights of Persons with Disabilities (CPRD) is tasked with erasing the stigmatization of disabled persons, examining root causes of stigmatization and teaching the consequences of harmful behaviors perpetrated towards the disabled. Another task of the CPRD is raising awareness of disabilities through activities and measures that combat harmful practices in these areas. In 2016 the UN created A Toolkit on Disability for Africa (TDA) to train workers who would be going to work in regions such as Cameroon, Ethiopia, Senegal, Uganda, and Zambia. The TDA was intended to help these workers understand the different cultural outlooks of each region on individuals who have I/DD; or dementia (UN DESA, 2016). It is highly informational on tribal beliefs and traditions concerning the disabled.

In other research, not related to the previous UN study, Eskay, Onu, Igbo, Obiyo, and Ugwanyi (2012) point out that even in this modern era, children in Nigerian schools who fall behind in their educational pursuits are usually excommunicated from their tribe. Interestingly, this study states that the disabled person who is excommunicated has most basic necessities still met, but they are shunned, ignored, not spoken to, or acknowledged. Occasionally the whole family of the disabled individual may be excommunicated, often because it is believed that the disability is a punishment for a wrong-doing by the mother, a curse, or possibly possession of an

evil spirit. In cultures with these beliefs, the individual with I/DD is ignored and sometimes terribly abused (TDA, 2016). There is little local interest in studying or researching the actual causes, symptoms, or treatments of the disability. Instead of researching the I/DD, it is stigmatized or shunned. This is a common cultural occurrence; throughout several of the tribes in Africa (TDA, 2016).

Autism Spectrum Disorder (ASD) falls under the umbrella of I/DD and seems to draw more research attention than other diagnoses. Zeliadt (2017) points out in an article about ASD awareness in Africa that until currently, parents were without assistance when coping with a child with ASD. Often the children are kept in small rooms or tied to chairs; in an attempt to control them simply because parents have no clue about how to care for them. In Ethiopia, two different centers for ASD children have recently been opened. These two centers offer help to 400 students and there is a waiting list. The creator of one of these centers, Zemi Yenus, a mother of a son with ASD, has become a familiar face around Africa as she attempts to educate everyone about intellectual disorders, such as ASD, via television and the internet. She emphasizes the rights of persons with disabilities and attempts to lower the false beliefs that these disorders are curses or possession of evil spirits. Her efforts have caught the attention of researchers from other areas who were interested in I/DD.

In another part of Zeliadt's (2012) ASD awareness article she tells of a Dr. Petris De Vries who arrived in Capetown, South Africa to continue the research on prevalence of ASD in the African population. He soon learned that determining the prevalence of ASD in Africa made no difference if there were no solutions to the problem of how to diagnose and help those who have ASD. In 2015, there were only 50 child and adolescent psychiatrists for 1 billion population in Sub Saharan Africa. Across the African continent, there are 2,000 different

languages spoken. Testing and treatment materials were in English, not a common language of Sub Saharan tribes. Language hurdles were slowing progress and education. De Vries has translated the Autism Diagnostic Observation Schedule (ADOS) and the Autism Diagnostic Interview-Revised (ADI-R) to Afrikaans, one of the 11 main languages in Africa. Another research fellow at Kenya Medical Research Institute in Kilifi began helping and has translated these same diagnostic tools into Kiswahili, another of the 11 main languages. With the information in native languages, these doctors hope to motivate better education among the indigenous people (Zeliadt, 2017). Activities such as this translation are preparatory steps to collecting information on the prevalence of comorbid I/DD + dementia. If indigenous people can educate themselves about I/DDs and types of dementia in a familiar language, they will be more likely to assist in data collection for research studies.

The stigmatization also surrounds those people who develop dementia. In the abstract of their article, George-Carey et al. (2012) state that "Information on dementia prevalence in Africa is [also] very limited" (p. 1). Paula Barbarino, Chief Executive Officer for Alzheimer's Disease International (2017) in her forward written for *Dementia in sub-Sahara Africa Challenges and Opportunities*, a study on aging in Africa, states "older people are seen as not contributing to economies, and their voices and rights are routinely excluded and ignored. Inequality is also a huge issue.... [in another paragraph of the forward she writes] at policy making level, it is clear to me that mother and child health issues and infectious diseases are the top priority in this region and that non-communicable diseases (NCDs) and older people's issues [such as dementia] are not really on the radar of many sub-Saharan countries" (p. 3). Multiple studies (George-Carey et al., 2012; Guerchet et al., 2017) suggest that older people with dementia are treated in much the same way as individuals with I/DD. Dementia is viewed as

"just part of aging" and no attention has been given to finding a cause or how to better treat it. Alzheimer's Disease International (ADI) has taken an interest in dementia in this region. Hopefully, research will begin to move forward for persons with dementia also.

The continent of Africa, having many low to middle-income countries inside of its boundaries, is one of the "many regions" lacking "evidence from well-planned, representative epidemiological surveys" referred to by Ferri et al. (2005 p. 2112). However, it is in its infancy regarding research. The future promises to hold much more information on I/DD and dementia and its comorbid relationship in that area of our world.

Research in more developed nations

I/DD affects individuals in all races across the planet. Dementia is also becoming a condition that is recognized worldwide. Countries like the UK, Ireland, Australia, and the USA are actively researching comorbid I/DD and dementia. Because of the "scarcity of epidemiological survey" data in less developed countries (Ferri et al., 2005), a consensus approach (see description page 5) appears to be needed to determine the prevalence of dementia in individuals with I/DD worldwide. A description of the consensus approach needed to cover the "scarcity of study" data in this case would be too much to cover in this literature review. For present purposes, we will continue with data from areas where I/DD + dementia comorbidity is actively being studied. Because of the difficulty of recognizing and diagnosing furthering cognitive decline in someone who already has cognitive delays, the topic is of considerable interest. There is a greater amount of data on this phenomenon in more developed nations that is available for review.

So far, examples of initial steps of research in third world countries, as exampled by Sub Saharan Africa, have been discussed. Several countries that are not classified as lower social

economic status also have limited published literature available about I/DD + dementia comorbidity. Australia, Canada, and Japan all admit to scarcity of documented literature in their countries. The Netherlands is known for its comprehensive research and care of its elderly and I/DD communities. However, database searches resulted in only one empirical article on the comorbidity of dementia + I/DD in the Netherlands. Main empirical sources of information about aging in the I/DD community used in this report are based in Ireland, the United Kingdom (UK), and the United States (USA). Most of the literature is from ongoing longitudinal studies. Some of the studies are still in progress in 2019. The focus of this report is on individuals with I/DD without DS. However, due to available data and the close relationship between DS and AD, DS is often included in the research samples of available literature. In an attempt to fully cover many different areas of the globe, some studies including the criterion of DS will need to be included in my report.

One of the limitations of published literature is the difficulty of choosing a representative sample. Articles mentioned in the next few paragraphs are interesting reads about conditions that are comorbid with I/DD, however, the only purpose that they serve for this review is to stress the scarcity of "representative epidemiological surveys" (Ferri et al., 2005).

Australia's, White, Chant, Edwards, Townsend & Waghorn (2005) eliminated most remote areas and penal system prisoners from their sample. The focus of this study was more on anxiety and depressive type mental health issues with a lack of coding for dementia. There was no coding for dementia.

Canada's Shooshtarl, Martens, Burchill, Dik, & Naghipur (2011) was limited to only Manitoba, Canada. This sample did not include individuals with I/DD who were institutionalized or already receiving care of some sort. A positive factor of the study is that the

research findings showed a significantly higher prevalence of dementia and depression in the I/DD group who lived in the community.

Japan's Sekijima et al. (1998) focuses on dementia in the individual with I/DD related to people with DS who are living in Nagano Prefecture. The lack of individuals with I/DD without DS in the sample was a large limitation.

Although, the Netherland's Van Schrojenstein Lantman-de Valk, den Akken, Maaskant, Haveman, Urlings, Kessels, and Creblder (1997) study was much more comprehensive, the sample was limited to 146 individuals with I/DD, living in institutions and group homes. Individuals with I/DD living in the community were not included. This study suggested that often symptoms of mental health issues, such as depression, manifest similarly to dementia, so diagnostic testing should be done with extreme caution. The concept of diagnosing dementia from an individual, pre-existing baseline was strongly recommended. As in other literature sources, I/DD individuals with DS were included in the sample percentages of Van Schrojenstein Lantman-de Valk et al.'s (1997) study. These are the countries with a scarcity of published literature. Now we will discuss literature from countries with larger amounts of research.

Ireland has focused more attention than many countries on the research of dementia in individuals with I/DD. Much of its data is based on DS in I/DD *only* and does not contain individuals with I/DD without DS. Mary McCarron leads an ongoing study of 609 people with I/DD (The Irish Longitudinal Study of Ageing (TILDA), Intellectually Disabled Supplement (IDS), 2017). It was begun in 1996, throughout Ireland, to determine the incidence and prevalence of dementia in I/DD over time. TILDA is designed to consistently continue updating data with the most current information. Another Irish study, Tyrell et al. (2001), completed a cross-sectional study of 289 DS-affected participants listed with service providers as having DS.

Tyrell et al. (2001) found that dementia in DS individuals who lived in an institutional setting was more likely (20.1%) than in those who lived in the community (7.5%). This was attributed to the fact that the institutional settings housed more older residents, and dementia is closely related to age. The Tyrell et al. (2001) study attempted to predict a baseline from which to determine the functional decline in dementia. At the time of Tyrell et al. (2001) cross-sectional study, the prevalence of dementia among the DS individuals studied was 13.3%. This sample was reported to be followed for at least 4 years from the time of the original study with plans to expand. No follow-up research literature has been located suggesting any associated future studies. In yet another Irish study, McCarron, McCallion, Reilly, Dunne Carroll, & Mulryan (2017) found that 97.4% of a sample of 37 subjects, all women with DS, over the age of 35, followed over a 20-year period (1996-2015) from an Irish memory clinic developed dementia. At age 50, there were 23% with dementia, which increased as the group aged to 80% at age 65. Again, most Irish epidemiological studies targeting the prevalence rates of dementia are conducted on large numbers of DS in the I/DD sample. Since DS is closely related to dementia prevalence/incidence rates appear high in this country.

Andre Strydom, a professor at Kings College in London, UK, appears to be one of the foremost researchers of dementia in the I/DD community worldwide. Strydom, Chan, King, Hassiotis & Livingston (2013) will be the main research focus for the present literature review. Strydom et al. (2013) used a follow-up sample from the Becoming Older with Learning Disabilities (BOLD) cohort. This cohort was used for samples for several research projects. It is described by Strydom as one hundred forty-two people with I/DD without DS from 5 inner-city and suburban boroughs in London. All participants of the sample for Strydom et al. (2013) were aged 60 or older. Two-hundred twenty-two people participated in the baseline study. All

participants had I/DD without DS. The study sample was chosen from a balance of community and institutional settings. In a longitudinal design, this sample was reassessed, and new data were collected 2.9 years after the first collection (second collection is labeled T2). This study found a significant incidence of dementia in adults aged 65 and older, five times higher than the general population. Incidence peak was at 70-74 years of age for the I/DD individuals compared to 90 years of age in the general population. There were no significant differences associated with gender. Please note that these data are for incidence rates, not prevalence. Strydom et al. (2013) believe that prevalence rates are highly influenced by mortality rates which are associated with old age, as is dementia. They prefer to focus on incidence over prevalence, as shown in the statement: "Prevalence rates may, therefore, underestimate a population's risk for dementia" (pg. 1882).

In another UK study, Cooper's (1997) research findings agree more closely to Strydom's data than other literature. Cooper (1997) studied the entire "learning-disabled" (Cooper's terminology, pg. 610) community 65 years and older (N=134) of Leicestershire, UK. Dementia was diagnosed in 21.06% of the group, higher than the 5.7% she had predicted. The percent of diagnosis of dementia increased as the group's age increased (i.e. age 65-84=23.5%, age 85-94=70%). In this study there was a higher *incidence* rate of comorbid dementia and I/DD in females, and they were more likely to live in health services institutions.

Consistency in Study Data

There is no scarcity of published research on comorbid I/DD + dementia from the US, UK and Ireland. However, the published literatures tend to have discrepancies. Standardization of epidemiological design would assist data collection and analysis to more closely resemble other research. Strydom et al. (2009) offer an example of this when they state, "Although the

UK estimates were based on samples representative of service users, the diagnoses were based on a single evaluation rather than longitudinal assessments, and this may have over-estimated dementia. The American study may have under-estimated dementia by not including all dementia subtypes, by using more restrictive criteria, or by being less representative than the UK studies...." (p. 10). Standardization of representative sampling method, when comparing studies, would include regulations on sample size, inclusion and exclusion criteria (ex: presence of DS), recruitment network (ex: institutional or community), bias in sampling methods, and country/culture of research. In the literature reviewed for this report, all of the studies, except Janicki and Dalton (2000) (n=794) were close in sample size (n=101-n=142). Inclusion criteria could have caused some discrepancy in the comparison for this literature review. UK studies used samples combining individuals with and without DS. Ireland's samples, in both studies, used only individuals with I/DD with DS. The US studies used samples of individuals without DS. DS is closely related to AD on the 21st chromosome. This causes higher percentages of prevalence in the individuals with DS and affects age of onset. Recruitment networks varied in all of the research. This could have been a cause for discrepancy in comparisons also. There can be a large variation of characteristics between individuals who live in institutions compared to those who reside in the community. As was emphasized in this report's section on Africa, culture can also cause large differences in sample composition and, therefore, research findings. The bottom line is that sampling methods are of high importance for international standardization for determining prevalence worldwide of comorbid I/DD + dementia.

Another ideal in standardized epidemiological design is fit of design. As a progressive disease, comorbid I/DD + dementia needs to be observed over time. This suggests that a longitudinal (taken over time) design is the best for researching comorbid I/DD + dementia. A

cross-sectional sample, taken at a single point in time, might work for prevalence percentages, but is not a good fit for incidence. Zigman et al. (2004) describe their study as a cross sectional design with 18-month intervals carried out over 4.5 years. My understanding of this would be that a new sample is studied every 18 months, then all data is analyzed. A longitudinal design would study the same sample group at each interval. TILDA, in Ireland, is a longitudinal design study that has been ongoing for twenty-four years. TILDA can offer very valuable data for researching how I/DD and dementia progress over time. However, Diesfeldt, van Houte, and Moerken (1986) report that the usual duration of dementia from onset to death is 7.2 years +/- 4.1 years. I would think that TILDA, having run for 24 years, would have had to collect new sample participants for study. Would collecting new sample members make it a cross-sectional with intervals as described by Zigman et al. (2004)? Possibly, the TILDA can offer an example of study design that would be appropriate for international standardization of study design.

Another important factor that needs standardization for international purposes is criteria of analysis. An example from the literature reviewed for this report is that Strydom et al. (2009) falsely suggested that Zigman et al. (2004) had limited dementia criteria to only the AD type in his analysis. Zigman et al. (2004) may have classified some mild dementia as Mild Cognitive Impairment (MCI), underestimating the dementia risk, while Strydom et al. (2009) did not even mention MCI. MCI does not usually lead to dementia. However, inclusion of MCI is highly important when considering the diagnosis of dementia in individuals with I/DD because of pre-existing cognitive delays. Strydom, Chan, Fenton, Livingston, & Hassiotis (2013) re-examined the original sample of 142 people with I/DD without DS from Strydom et al. (2009) three years after the original study. The 2013 study included MCI classification, the 2009 did not. Re-examination results found that 33% of the original sample were no longer demented, and 48% of

the original sample redefined operationally as having MCI. Silverman, Zigman, Krinsky-Hale, Ryan, & Schupf (2014), who compared Zigman et al. (2004) to Strydom et al. (2009), stated "results clearly illustrate that subtle differences in case definition can have substantial impacts on estimates of incidence/prevalence" (p. 6). To reiterate the point on discrepancy in studies at the beginning of this section, I would like to point out that Strydom is a researcher from the UK and Zigman is a researcher from the US. Standardizing criteria used in analysis, worldwide, could prevent situations such as the MCI inclusion confusion. If individuals with I/DD without DS are showing no significant difference for risk of dementia compared to the general population in repeated studies, maybe classification of data, as in the difference between MCI and mild dementia should be looked at closer?

If standardization of 1) samples for collection of data, 2) design of study, and 3) criteria for data analysis were to happen, results would be more consistent. Consistency would increase the ability to find appropriate treatments and supports for individuals with I/DD.

What does it all mean?

Studies show conflicting data on the prevalence of dementia in individuals with I/DD without DS. In American literature, there appears no significant increase of dementia in individuals with I/DD not related to DS when compared to the general population. Here lies the conflict. If individuals with I/DD without DS do have a higher risk of developing dementia, do they need any special treatment or support? Individuals with I/DD with DS have a higher risk of developing AD type dementia at an earlier onset age because of the DS to AD relationship (mentioned earlier). There appears to be a debate amongst American researchers on specific criteria needed during analysis. There is a need for consistency in study design and analysis criterion.

In the UK, there was a significant increase in dementia in individuals with I/DD. Most of the samples from the UK included individuals with DS, affecting prevalence rates. Susan Cooper (1997) was the only study that showed a significant difference between genders. Results from the UK studies tend to be closer in design, analysis, and have more in common with each other. UK versus other countries' studies vary in sample composition, study design and criteria for analysis.

Most studies from Ireland included DS in their research. As a quick reminder, DS falls under the I/DD umbrella. The data including DS will always have larger prevalence rates because of DS's close relationship to AD on the 21st chromosome and related dementia.

Countries other than the USA, UK, and Ireland have too few data to determine a reliable prevalence of comorbid I/DD + dementia. Their statistics are interesting to look at, but they do not offer much in the concept of overall worldly prevalence of dementia in aging individuals with I/DD without DS. It is doubtful that the region of the world where the studies are being done, or the culture of the area have significant influence to change prevalence values, or risk of the individual developing comorbid I/DD + dementia in data analysis. Variance of method, such as prevalence vs. incidence, longitudinal vs cross-sectional, sample selection differences {institutional residence vs. community residence}, and the criteria used for analysis appear to be a larger influence(s) on analysis results. Analysis results determine prevalence of comorbid I/DD + dementia, and if individuals with I/DD without DS might be at higher risk to develop a dementia.

Standardization of sample selection, data analysis, and method of collection of data is needed for consistency across the globe to create non-conflicting, beneficial data from varied locations. Until the time that these research guides are created, it will be difficult to determine if

there is a higher prevalence of dementia among individuals with I/DD without DS in any area of the world. With prevalence data, it might be possible to determine the impact of factors, such as differences in culture, religious beliefs, and climate, of areas less affected by comorbid dementia and I/DD. Information like this will be of benefit in assisting regions that are more affected by these factors to overcome their barriers and move forward.

Chapter 3: Baseline, Normal, & Disease-related Functioning

What is Considered Normal?

Due to, often lifelong, lower cognitive functioning in a majority of individuals with I/DD, their daily functioning cannot be compared to what is considered 'normal functioning' in the general population. Through an Intelligence Quotient (IQ), the mental capacity of each individual with I/DD is measured during childhood. Mental capacity usually affects the individual's capacity to function in life skills (Pederson, 2018). According to the DSM-5, a person with an IQ of 19 and under is considered 'profound', 20-34 is considered 'severe', 35-49 is considered 'moderate', 50-70 is considered 'mild' (Gluck, 2018; Thorpe, 2006). The dissertation of Canadian Lillian Thorpe (2006) reports that about "85% of people with mental retardation fall into the mild category, 10% in the moderate, 3-4% in the severe and 1-2% are in the profound category" (p. 9).

Some studies concerning the relationship of DAT (Dementia of the Alzheimer's Type-early onset) and DS have determined that lower measures of IQ at an early age are related to higher levels of aging-related decline. This is Cognitive Reserve Theory. Cognitive Reserve Theory claims that higher reserves (higher levels of cognitive ability, education and literacy) should be protective against, and delay, the onset of dementia (Zigman et al., 2004). Other studies contradict this (Thorpe, 2006; Bush & Beail, 2004). In a literature review hypothesizing why the contradictions about the relationship of IQ and aging-diseases might exist, Bush & Beail (2004, p. 88) concluded

> A number of methodological problems are likely to be contributing to the variability in the findings reported [on the relationship between DAT and IQ]. These need to be addressed if the relative influence of different potential risk

factors is to be established. The major methodological limitations to be considered include cohort bias through inadequate sampling, problems in diagnosing DAT, and inadequate measurements of change in cognitive functioning.

What Does Normative Aging Look Like in an Individual with I/DD?

The variability of these "pre-existing conditions" makes it difficult to detect specific group-related extended memory impairment or other cognitive changes. What, then, can be considered usual changes in primary aging in the I/DD community?

As will be discussed later in this report, it is important to determine cognitive abilities of an individual at or around age 30. This is the peak functioning level of most people's adult lives, for those with I/DD or without (Marill, 2018). Many parts of aging are caused by simply living. The aging person's hair loses some of its original pigment. Their eyes, ears, muscles, and blood vessels develop wear and tear. Eye tissue may develop cataracts, and glaucoma and/or become more rigid due to long exposure to light. Internal ear tissues get stiffer from use and build up inside causing hearing to decline. Blood vessels become tougher and blood pressure may rise. Weight gain, trouble sleeping at night, stiffer muscles, and heart problems (muscle) may occur. Small episodes of memory loss, such as forgetting the name of someone the person has not seen in a long time, or the place where keys were left are normal in everyone, individuals with I/DD included. The individual loses a small amount of ability to do activities of daily living (ADLs), like toileting, tooth brushing, and combing hair with normative aging. When the individual develops an inability to do ADLs, caregivers should seek guidance.

Emotions, such as frustration or anger, can also be common for persons with I/DD as they experience the effects of aging. It may be difficult for them to express that they cannot see as well, or that they are exhausted because of higher blood pressure or diabetes. This failure to

adequately express themselves can be converted to anger or frustration, difficult for both caregiver as well as patient. Some memory loss is even expected. When problems with memory become inability to use language, recognize objects, or remember what you ate for lunch that day, if that was not already a lowered function in the person with I/DD, become noticeable, it is time to seek guidance (Marill, 2018). It may be that dementia is creating more serious declines.

Finding Baseline Functioning in Individuals

Before diagnosing an individual with I/DD (with or without DS) with comorbid dementia, a baseline of cognitive abilities needs to be established. In Assessment and Diagnosis of Dementia in Individuals with Intellectual Disability: A Toolkit for Clinicians and Caseworkers Gregory Prichett (2017) advises how to create baselines and diagnosis of dementia. Pritchett (2017) states, "This toolkit was developed for clinicians and caseworkers....[who lack training in the needs of individuals with intellectual disability]....It compares and contrasts the incidence, prevalence, and clinical features of dementia...in adults with ID...with that of the general population and then outlines the modification to diagnostic approaches that are needed to improve diagnostic accuracy for those ID adults potentially affected by dementia" (p. 4).

Nearly all individuals with I/DD with DS develop neuritic plaques, neurofibrillary tangles, and other characteristic brain lesions of AD by the age of 40, even though they may not develop signs of dementia (Aylward, Burt, Thorpe, Lai & Dalton, 1997). Developing baselines for cognitive functions needs to be accomplished on personal levels, prior to suspicions of demented behavior, but at the same time related to a standardized criterion. Because of early development of plaques and tangle, in a person I/DD caused by DS, this would be prior to age 40. Is that age a reference point for all individuals with I/DD? As stated above, a baseline of what might be considered 'normal' for the individual with I/DD needs to be developed at an age

before dementia would commonly begin. For this purpose, the Working Group for the Establishment of Criteria for the Diagnosis of Dementia in Individuals with Intellectual Disability was founded (Aylward, Burt, Thorpe, Lai & Dalton, 1995). It is comprised of experts in the fields of aging and mental retardation. This working group of experts recommends assessing for baseline functioning before the age of 36 in individuals with I/DD with DS and age 50 in individuals with I/DD without DS (Pritchett, 2017). Note that the Working Group's recommendation of age 36 is very close to Marill's (2018) peak functioning age of 30 in all adults. The age of 35 is also offered as a perfect point to develop baseline cognitive function with annual updates (Burt & Aylward, 2000; Nieuwanhauis-Mark, Schalk, & de Graaf, 2009). This age recommendation allows the person with I/DD with DS to be assessed before the customary brain lesions appear. The early age baseline measurement is not needed in an individual with I/DD without DS because the same growth of characteristic brain lesions does not develop early in most brains that are not affected by DS (Pritchett, 2017; Aylward et al, 1995; Thorpe, 2006).

The National Task Group on Intellectual Disabilities and Dementia Practices (NTG) recommends regular screening be continued at yearly intervals (Pritchett, 2017). By screening regularly, conditions that closely resemble dementia, like vitamin B12 deficiency, depression, thyroid imbalance, delirium, medication imbalances, normal pressure hydrocephalus (NPH) or tumor, can be identified, properly diagnosed and treated in less time. To sustain good health practices, early detection of any condition is important.

As of the printing of Pritchett's Handbook (2017), there was no one specific diagnostic criterion used to diagnose dementia in the I/DD community. The previously referenced Working Group (Aylward et al, 1995) recommends use of the ICD-10 as the most appropriate test for diagnosing dementia for this group. For a copy of the full ICD-10, see World Health

Organization link in reference section. This criterion places more of an emphasis on the non-cognitive aspects of dementia, which are often better indicators of early decline in the individual with I/DD. Indicators of decline in the individual with I/DD will be discussed later in this paper. The ICD-10 also offers subtyping, which allows evaluators the ability to link AD to DS in cases where they might connect. The two-step process, which allows subtyping, also helps to control bias of clinical situations by providing greater scientific certainty (Pritchett, 2017; World Health Organization, 2003).

The NTG recommends evaluations for managing and diagnosing dementia in the individual with I/DD. Components of a standard workup should include: 1) detailed medical history, 2) thorough physical history, 3) thorough neurological exam, 4) psychiatric assessment, 5) routine lab tests, including blood and urine, 6) chest x-ray, 7) electrography, and any other specialized tests related to an individual's personal health issues. (Pritchett, 2017; National Task Group, 2013). As mentioned earlier, it is recommended that evaluations and re-screening be done annually (Pritchett, 2017; National Task Group, 2013). All possible conditions having similar symptoms to dementia must be ruled out before proceeding with diagnosis.

How to Determine What is Disease-Related Decline

With so many conditions and diseases having symptoms that resemble dementia, it can be problematic to recognize the difference between age-associated decline due to disease with primary aging. Prasher, Chung and Hague (1998) caution that assessment of an individual with I/DD suspected of having dementia should be longitudinal and extend over a three-year period. Adaptive, cognitive and behavioral features should be observed for at least two years before interpreting the cause of the decline. A basic definition can be found in the DSM-IV instrument for mental disorders.

> Dementia is defined in the DSM-IV…as progressive cognitive…decline that has reached the point of significant impairment in social or occupational functioning. It must represent a significant decline from a previous level of functioning, which differentiates it from early life cognitive disabilities such as ID. (Thorpe, 2006, p 23)

For diagnostic purposes, the DSM-IV also requires deficits in cognitive functioning such as aphasia (loss of ability to understand speech), apraxia (inability to perform a task when asked to), agnosia (inability to identify objects), and loss of executive function (Thorpe, 2006).

Table 1 Signs of Dementia per stage in the I/DD Community

Stage	Signs observed in stage
First Phase	Mild memory impairment Reduced verbal output Apathy Inattention Spatial disorientation
Second Phase	Loss of self-help ADLs Gait changes Seizures
Final Phase	Non-ambulatory Bedridden Flexed postures Sphincter incontinence Pathological reflexes Aspiration Pneumonia Other re-occurring infections

Table created using literature from Lai, F. & Williams, R. (1989).

Dementia is a progressive disease. Memory impairment is not the first sign of dementia in individuals with I/DD. As shown in Table 1, the original signs of an age-related disease in the individual with I/DD are reduced verbal output, apathy, inattention, spatial disorientation, and decreased social interaction. As decline continues into what Lai & Williams (1989) classify as Phase 2, there may be losses in self-help ADLs, a change of gait, and possible seizures (which can be pharmacologically controlled).

As the dementia-related decline progresses towards the final stage, the individual with I/DD can become non-ambulatory, bedridden, in continual flexed postures, have sphincter incontinence, pathological (ex: upgoing toe; extensor response) reflexes, aspiration pneumonia, and infections that are difficult or impossible to treat (Thorpe, 2006).

There are alternate frameworks for dealing with phases or stages of dementia. Table 2 shares a listing of the seven stages of AD as they typically affect the general population (Thorpe 2006). The purpose of these tables is to compare signs of disease-related decline in the general population (Table 2, below) to those of the I/DD community, in Table 1(above). Some of the symptoms observed in Table 2 may exist as pre-existing cognitive delays within a person with I/DD who has had a baseline assessment (Thorpe, 2006). The preexisting symptoms could add difficulty to creating and following an individual baseline for observing and diagnosing aging people with I/DD. This comparison illustrates the importance of creating a baseline cognitive function for individuals with I/DD at their peak adult functioning age (Marill, 2018).

Different from the general population, the original signs of disease-related decline often manifest as personality changes in an individual with I/DD. According to most of the clinical literature, the initial signs of dementia are not a large change in memory, contrary to public

Table 2 Signs of Alzheimer's Disease per stage in General Population

Stage	Signs observed in stage
First stage	No subjective memory problems No abnormalities observed during clinical interview
Second stage	Some subjective memory problems No subjective memory problems during clinical interview
Third stage	Mild changes in: a) orientation, b) vocational performance, c) word finding, d) memory, and e) concentration.
Fourth stage	Clear cut deficits in: a) knowledge of current & recent personal events, b) memory of current or recent personal events, c) concentration (easily distracted), d) orientation, and e) ability to handle finances.
Fifth stage	No longer able to live alone. Deficits in: a) recalling address & phone number, b) recalling family names, c) recall of schools attended.
Sixth stage	Deficits in memory of spouse's name Unaware of current or recent personal events Need help with basic ADL's such as bathing, and dressing. Incontinent Cannot travel Changes in: a) personality, and b) behavior. Development of: a) delusions, b) repetitive actions, c) anxiety, e) agitation, f) aggression, and g) apathy.
Seventh stage	Verbal skills are lost. 24-hour care is required. Complete loss of ADL's Deficits in motor function Extrapyramidal side effects.

Table created using literature from Thorpe, L. (2006)

belief. Individuals with I/DD often live with short term memory delays every day. Memory decline would not be as visible in a person who already has it. Therefore, memory loss is more obvious in the general population. As mentioned earlier, the first signs in the individual with

I/DD will be more social. Apathy and personality changes are not listed for people in the general population until much later in the progression of the dementia.

In Phase 2 (Table 1), it is mentioned that the individual with I/DD will have a change of gait. Gait is not directly approached in the general population literature. A lack of orientation could cause an older person with or without a disability, to move differently and be less confident in their steps, causing a gait change.

Incontinence is mentioned in both the general population and I/DD community at a similar point of progression of the dementia. Many of the physical conditions, like inability to travel and need of 24-hour care, also compare at similar points in the progression of both general populations and I/DD communities. Twenty-four-hour care, however, could be pre-existing in the individual with I/DD and already part of a baseline assessment. Seizures are another symptom that could commonly already exist in an individual with I/DD. Possibility of 24-hour care and seizures already existing are examples of why baseline assessments are very important when determining decline and/or changes in the individual with I/DD.

Information on the individual with I/DD's signs or symptoms is sometimes hard to collect. Many individuals with I/DD cannot answer questions about their own spatial orientation, especially if they have reduced verbal output or a lower IQ. Caregivers would need to report observations in these cases. In many situations there is not a staff member or caregiver who has been connected to the individual with I/DD for a long enough space of time to be able to accurately present information to a medical professional. Often there is no training or education offered to the residential staff to recognize a problem or sign if it is happening. Written records are a necessity in residential living situations with higher staff turnover, for consistency in care. This will be discussed later in this paper.

Universal "Guideline" Criteria for Disease-Related Decline

Dementia signs in an individual with I/DD can be very subtle. Loss of memory is a good example of this. As stated earlier, the individual with I/DD may have always had a problem with short term memory. If disease-related decline in this area is present, it can easily be overlooked because it is not a dramatic change in the individual with I/DD's behavior. Somewhere in the individual with I/DD's medical history (hopefully in a baseline cognitive assessment) there should be mention of their short-term memory level of functioning. This measure can be compared to a new measure to determine if there is a large decline occurring. A caregiver's observations of the individual's behavior must be vigilant.

Useful and easily accessible tools have been created to aid the caregiver in assessing their ward's signs of decline. A useful instrument available online is The National Task Group Early Detection Screen for Dementia (NTG-EDSD). It is a valuable tool designed to be used by family caregivers or residential staff in an annual review, or when the individual with I/DD appears to be experiencing further cognitive decline. The NTG-EDSD format is user-friendly. It assesses general conditions that can indicate dementia or rule out other diseases or conditions. This screening tool is a good way to use a universal guideline criterion to help diagnose dementia while also collecting specifics for the individual with I/DD. Further information on the NTG-EDSD can be found at: https://aadmd.org/sites/default/files/NTG-EDSD-Final.pdf.

Nieuwenhuis-Mark et al. (2009) recommends a few brief cognitive screens when trying to diagnose dementia in an individual with I/DD. For initial screenings the DemTract (Kalbe et al. (2004) takes about 8-10 minutes and it is very easy to administer. The DemTract measures 1) word list, 2) number transcoding, 3) word fluency, 4) digit span reverse, and 5) delayed recall of word list. The 7-Minute Screen (Solomon et al., 1998) consists of screening in 1) enhanced cue

recall, 2) temporal orientation, 3) verbal fluency, and 4) clock drawing. As this test's name suggests, it takes about 7 minutes to administer the total screen and is very easy to administer. Neiuwenhuis-Mark et al. (2009) suggests that both screening instruments are useful in the first baseline cognition level assessment as well as longitudinally throughout the annual screenings.

The Barthel Index (BI) is also a good tool for use in the first screening and continuing annual screening. The Barthel Index (Mahoney & Barthel, 1965) measures ADLs and iADLs in the individual with I/DD. The BI measures cognitive abilities, especially in executive functioning, at the age 35 mark. It can then be used to compare annual screenings to the pre-morbid baseline screen. This helps to recognize what is a slow and steady decline that can be associated with normal aging, or a disease-related decline, which might indicate early aging disease signs.

Other instruments mentioned in diagnosis of comorbid I/DD + dementia are the Camdex, Camdex-R (adapted for retardation), Camdex-DS (adapted to fit Downs Syndrome), Dementia Scale in Down's Syndrome (DSDS), Dementia Questionnaire for Person with MR (DMR), and the Mini Mental State Exam (MMSE). An instrument used for a diagnosis of dementia should be administered after the individual with I/DD has shown first signs of dementia, has had some screening, and their needs have been assessed. The instrument used should be chosen with care, selected to best match the individual with I/DD's symptoms. Accompanying the test above, a psychological assessment should also assess depression, delirium, and other mood changes. Observations screenings of these behavioral changes should be completed by caregivers who know the individual with I/DD well. Neiuwenhuis-Mark et al. (2009) suggest that the caregiver should have been in a relationship of close capacity for at least six- months. Observations should also be longitudinal in design. Physical exams (as listed on page 25 of this document) should

also accompany these assessments. Neiuwenhuis-Mark et al. (2009) claims that no one test is rated more efficient than another.

To sum up this section on diagnosis testing, it is difficult to make a clear-cut diagnosis of comorbid dementia within individuals with I/DD because of their varied personal and individual pre-morbid needs. A single method of testing to define a diagnosis of dementia in every individual tested is not possible. There are different tests, however, that are available that contain questions that are disease specific but also can be tailor made to fit the varied individual. Although is appears contradictory, in the end, it IS possible to have a general basic set of questions that can be individually modified or adjusted to use for comorbid diagnosis of I/DD + dementia.

Chapter 4: Support Frameworks and Best Practices

Who Should be Involved in the Reporting of Decline?

An individual with I/DD is going to have a relationship in some capacity with a caregiver. If their condition is categorized as mild, it may only be a family member who visits now and then. Generally, they will live in a family home, in a residential (community) home, or an institution and will have caregivers. The caregiver is commonly the person who will see the first signs of normative age-related decline, or disease-related decline.

The NTG offers steps of evaluation to prepare for diagnosis, Step 1: Gather a pertinent medical and psychiatric history that includes cardiovascular, cerebrovascular, neurological structure abnormalities, head injuries, sleep disorder, thyroid function, vitamin B12 deficiency, and metabolic syndrome. Step 2: Obtain historical description of baseline function. Supplemental Table 1 at http://www.mayoclinicproceedings.org offers exact information needed for a baseline function. Step 3: Obtain a description of current function and compare to baseline. Does the observed behavior interfere with functioning? Does it represent a decline? Step 4: Perform a focus review of systems. What are the common issues seen with increasing age? Supplement Table 2 at http://www.mayoclinicproceedings.org can offer guidance in this step. Step 5: Review medication lists thoroughly for signs of polypharmacy like drug to drug interactions or adverse effects. Also, review multiple doctors prescribing medications. If multiple doctors are prescribing medications, without full medication review, the medications could be the cause of observed symptoms by reacting against each other. Step 6: Obtain pertinent family history. Look for signs of cerebrovascular events, stroke, diabetes, heart disease rheumatoid arthritis and lupus in immediate family. Step 7: Assess for other psychosocial issue or changes. Look for triggering events. Assess for co-existing mood disturbance or psychiatric

disease. Step 8: Review social history like living environment and level of support. Check out safety concerns and fit of residential environment. Step 9: Synthesize the information that has been gathered. Cross reference all information to determine which direction the signs are guiding the diagnosis. Simultaneously, as the caregiver is evaluating the above steps, the individual with I/DD should have a physical and a cognitive examination performed by a health professional. All information that cannot be provided by the individual with I/DD should be provided by a trusted source who has known the individual with I/DD closely for at least six months. Once all these steps have been completed, then the individual should be evaluated by a professional health care provider who is familiar with dementia. The team involved should be caregivers, medical records, doctors for physical and cognitive exams, and finally a medical professional with an understanding of dementia.

Supports are Available for the Patient with Disease-Related Decline

The main goal of most supports for an individual with comorbid I/DD + dementia is to be able to remain in familiar settings, customarily the family home or a community residential living home. To assist in this situation, the National Task Group (Jokinen, Janicki, Keller, McCallion & Force, 2013, p. 2) has created these guidelines for developing a support system:

- Promote quality of life.
- Use a person-centered approach.
- Affirm individual strengths, capabilities, skills, and wishes.
- Involve the individual, their family, and other close supportive persons.
- Access and make available appropriate diagnostic, assessment, and service resources.
- Plan and provide services that effectively support the individual to remain in their chosen home and community.

• Provide access to services and supports that are available to other persons in the general population affected by dementia.

• Undertake proactive strategic planning across policy, provider, and advocacy groups.

Jokinen et al. (2013) estimate that in the United States 75% of older adult individuals with I/DD reside with their parents. Since dementia is a progressive disease, it must be considered that symptoms will become more complex with time. Support will be needed in larger amounts. Parental caregivers will also be aging. There is a possibility that a parent caregiver will become unable to provide the entire amount of support needed for the individual as the dementia progresses. Extra support will be necessary. Immediately after being diagnosed, while the individual with I/DD + dementia is still at the best capability of making decisions for themselves, a support plan should be created, approved by both individual with I/DD + dementia and caregiver, and recorded. This will help to ease transitions in care that will be needed as the dementia progresses.

Sources of informal support for family caregiving are a) extended family members, b) neighbors, c) friends, and d) members of faith communities (Jokinen et al., 2013). In family caregiving there is also the possibility of adding paid, formal caregivers when needed. Sources for formal caregivers include 1) Alzheimer's Association chapters, 2) Aging and Disability Resource Centers, 3) Hospital Discharge Planning, 4) Area Agencies on Aging, 5) Caregiver Support Programs, 5) Community Agencies, and 6) National Down Syndrome Services. Many of these organizations have websites that can lead to specific needs. To view an example of a website with this capacity, visit http://www.mindandmemory.org/ .

In community residential homes, multiple paid caregivers attend the individual with I/DD + dementia every day. Residential staff tend to have a fast turnover of employees due to the high

demands, physical and emotional, of the job. For suitable care it is of great importance in this setting that caregivers keep accurate and timely records. In the community residential setting it is also important to have a support plan in place soon after diagnosis that caregivers are aware of and can periodically review. The afore mentioned agencies are also available for use in formal caregiving. An added benefit of community residential living is that multiple people offer the care for one individual so there is less "burn out" or feelings of being overwhelmed with the burden. Having multiple caregivers also adds different perspectives to the individual with I/DD + dementia's care. With different perspectives (sometimes called "fresh eyes"), problems can sometimes be caught in earlier stages resulting in better care. The opposite can also be true. Multiple caregivers and an over worked staff can lead to insensitive treatment for the individual with I/DD + dementia. Insensitivity can result in abuse and neglect. In community residential settings there should be frequent, regular evaluations of care. The full description of care of individuals with I/DD + dementia is beyond the scope of this report.

Supports Available for the Caregiver of an I/DD Individual with Dementia

As mentioned above, being a caregiver for an individual with I/DD + dementia is very demanding, physically and emotionally. In informal family caregiving, often there are no chances for a break. Caregivers are "on duty" twenty-four hours a day, seven days a week. This can create negative feelings between the informal caregiver and the individual with I/DD, which may lead to abuse and neglect. It is important to have frequent, regular evaluations of care in this setting, just as in the community residential home. Support groups are available in most locations to offer these evaluations and give the caregiver someone to talk to and get ideas from. Information about them may be available online or listed through medical offices providing care for the individual with I/DD + dementia that they work with.

Another source of care assistance is respite care. Local agencies usually offer listings of qualified people who can assist the caregiver and allow for breaks or that sometimes much needed nap. The Alzheimer's Society offers respite care. There is also an American Alzheimer's Association (AAA) caregiver program in many localities. For more information on the AAA caregiver program visit https://www.alz.org/help-support/caregiving/care-options/respite-care . For those situations where finances are limited, respite care is also covered by Medicare. See https://medicare.com/caregiver-resources/respite-care-tips-caregivers for more information on this service.

Adult Day Care provides another source of support for informal caregiver. This works best for the milder stages of dementia and for most levels of I/DD. Adult Day Care facilities are generally open from the hours of 7:00 a.m., to 6:00 p.m. Some offer evening hours for caregivers who works later hours or have special events. Adult Day Care offers more autonomy and socialization than an institution. There are different types of Adult Day Care. Transitional is usually offered for a post-hospital stay. Transitional Adult Day Care provides a place for the individuals to go during the day because they are not fully ready to fully care for themselves after their short hospital stay. Congregational Adult Day Care is hosted by a religious organization. An example is the Catholic Social Service Adult Day Care Services. Active Adult Day Care is for elders who do not have extensive physical or medical issues. It offers socialization, exercise and a sense of community. Palliative Adult Day Care's goal is to improve health and wellness. It is not meant to be an "end of life" place, but a "let's get better" place. Military Veteran Adult Day Care helps veterans with ADLs, provides meals, and general help with daily needs. Memory Care Adult Day Care is just what its name says. It is designed for elders who have AD and other forms of memory disorders. Memory care is designed to be a safe

place. Memory care also offers cognitive and memory training therapies. Most insurance, Medicaid, Medicare, and VA benefits all help to pay Adult Day Care. Costs can run from $25 to $125/day depending on supervision needs. Adult Day Care is a positive way to provide respite care (Seniorliving.com, 2019).

Best Practices for the Care of a Newly Diagnosed Co-Morbid Individuals

Determining best practices for care of an individual with I/DD + dementia is a controversial issue. Some professionals believe that institutional placement increases mortality for an aging person with I/DD. Others believe that "those with multiple complex medical conditions may receive inadequate care in the community unless intensive supports are available, whereas those with mild disabilities may improve in the community because of decreases in institutional mediated infections and improved biopsychosocial well-being." (Thorpe, 2006, p.13). Institutional placement offers caregivers with knowledge of multiple complex medical conditions. Knowledge of these medical conditions is a benefit; however, in an institutional environment, caregivers/staff have several patients to care for in a determined time frame or shift. Individual supports given can be much less effective. Treatments can be missed, or expedited, causing lowered benefits. The individual with I/DD + dementia's biopsychosocial well-being can be affected by this less personal posture of their caregiver. From a physical perspective, as mentioned above by Thorpe (2006), infections are more likely to occur in institutions due to higher exposure to contagions from multiple sources. When considering best practices, caregivers in an institutional setting's higher knowledge of existing conditions must be measured in comparison to less time for personal attention. For best practices, placement of an individual with I/DD + dementia needs to be person-centered. Instead of following a cookie cutter plan of care, each person's unique conditions need to be addressed.

Another concept to be considered when determining where to "place" an individual with I/DD + dementia, concerning best practices, is the "aging in place" concept. This concept can be completed through the individual with I/DD + dementia remaining at home and receiving support from informal family members. In 2011, President Barrack Obama signed a National Alzheimer's Project Act (NAPA). In connection to this act the NTG created guidelines for best practices of care of the individuals with I/DD from the point of suspicion of comorbid dementia through to their death (Jokinen et al., 2013). Informal, family and community care settings were a large focus of these guidelines. Best practice models for each stage of progression of dementia, from suspicion of presence of the disease to death of the individual are presented in Jokinen, et al. (2013). To view full information on best practice models please visit https://onlinelibrary-wiley-com.er.lib.k-state.edu/doi/pdf/10.1111/jppi.12016.

Summing Up

Several forms of literature comprise this review. These research and scholarly sources reveal that global prevalence of comorbid I/DD and dementia is difficult to predict due to "evidence from well-planned, representative epidemiological surveys [being] scarce in many regions" (Ferri et al., 2005, p. 2112). There is a continuing need to standardize sample selection, collection of data, and analysis of data to provide non-conflicting, beneficial data analysis from varied locations. Consistency in data collection and analysis of data across the globe should help to improve future data comparisons. The professional literature also recommends that a baseline should be established during the mid-30's for people with I/DD to serve as a guide for later diagnosis of dementia, if needed. Guidelines for baseline measurement and recording have been determined. Supports for both individuals who are comorbid

and their caregivers have been discussed. Best practices have been partially discussed for the care of the individual with comorbid I/DD + dementia diagnosis and their caregiver.

Chapter 5: Discussion of Applying Information

Application of This Literature Review in Family Relationships

This literature review has helped me to understand the prevalence and incidence of comorbid I/DD + dementia better. Its intent is to help the reader to understand precautions that should be taken during an individual with I/DD + dementia's early adult life, in order to create a baseline cognitive measure for use when/if signs of disease-related decline begin. Support systems have been identified. How can we apply this information to our own real lives?

For me, this is an important question. I have two biological children, a son and a daughter, now adults, who have I/DD. The main diagnosis, for both of them, is a Porencephalic Cyst (very rare), which is genetic. They both have other comorbid diagnoses related to their I/DD. Both of my children are considered mildly mentally retarded. Both have epilepsy. My son also has diagnoses of Attention Deficit Disorder (ADD), and Borderline Personality Disorder (BPD). In the past, he has received diagnoses of Obsessive-Compulsive Disorder (OCD) and Schizoaffective Disorder (SOA). For a temporary time, as a teenager, his psychiatrist was terming the Schizoaffective Disorder as Schizophrenia. As an adult, OCD and SOA were singly dropped from his diagnoses and then grouped into the BPD diagnosis to qualify him for services at Pawnee Mental Health Services. My daughter also copes with Depression and Language/Speech Delays. My daughter is high functioning and, as an adult, does not appear to need many I/DD services, except counseling sessions with Pawnee Mental Health for her depression. My son is another story.

My son and I moved to Kansas in 1997, while he was still in High School. He qualified for a Special Education Cooperative covering Clay and Washington Counties and was bussed to Clay County Cooperative High School (CCCHS) every day for school. His Individual Education

Plan (IEP) had him scheduled to continue to the closest Community-based Developmental Disability Organization (CDDO) for adults, Big Lakes Development Center (BLDC), upon graduation. After a visit to his father's newly combined family in Idaho (his father had re-married), he chose to return to Idaho to live in the summer of 1998, which removed him from the Special Education Program in Clay Center. Leaving CCCHS removed the IEP that had grandfathered him into BLDC. The IEP had offered him a direct line into the CDDO without having to apply and go through a qualification process. Within 6 months, my son realized that he had made a mistake and started begging to return to Kansas. In the interim, my son had his 18th birthday. His father and step mother, for protection reasons, had taken adult guardianship/conservatorship of him and, for personal reasons, they were not allowing him to return. It was 2002 before the situation in Idaho allowed for him to return to Kansas. He had substantial I/DD and Mental Health symptoms and was in serious need of services. Since he had been in Developmental Services in Idaho and had at one time been on track to attend BLDC, our first step to attain help was to apply to Big Lakes for services. The application process was definitely a big learning experience for all involved.

To be admitted into a Community-based Developmental Disability Organization (CDDO) for adults with I/DD in the United States, an individual must have an overall IQ, an average of performance and verbal skill, of 70 or under (personal experience, 2002) unless they have been 'grandfathered in' through Special Education Program IEP. There are many individuals with I/DD in the world, who are slightly intellectually delayed, who have IQ's slightly over this "standard measure' criteria, but are still considered mentally retarded (MR). My son with I/DD has a performance IQ of 68 with a verbal IQ of 80. When they are averaged for an overall IQ, his overall IQ is a 74, four points above the national cut off level. This makes him ineligible for

CDDO services. If his performance IQ alone was used to qualify for service, he would qualify (personal experience, 2002). He has been labeled as, "mildly mentally retarded," but cannot get services. He has been through Vocational Rehabilitation Services and is considered unemployable because of his mild retardation + comorbid mental health diagnoses (mentioned earlier). Many of his daily life skills, which he needs help with, fall under the performance IQ umbrella (as listed above, his is 68). He attended Pawnee Mental Health Day Support Services for 1-2 years but discontinued attendance. To this day, he is followed by a case manager who meets with him periodically. That is the extent of his disability services.

Many mildly retarded people in the US, like my son, fall through the cracks, if they don't have an advocate working for them. These people who fall through the cracks need to have professional and/or medical caregivers that understand the importance of creating a baseline of their cognitive skills at a younger age. Because my son has a genetic cyst in his brain, has had epileptic brain surgery, and has a family history of dementia, he is also considered at higher risk for a comorbid diagnosis of I/DD without DS + dementia in the future. This is true for many individuals with I/DD without DS, who are falling through the cracks. When these people with I/DD reach the critical age of developing comorbid dementia, their advocates, or caregivers, might not be available to assist them any longer. It is important to have a baseline measure of cognitive functionality recorded in a place that can be easily reached by any caregiver or health care professional. My son has a neurologist, psychiatrist, orthopedist, general physician, and is a patient at the Epilepsy Center in University of Kansas Medical Center. If he were to have a baseline cognitive functioning measure done, the test results would need to be, not only in his own, private, personal medical files, but also saved at Pawnee Mental Health Services or the Epilepsy Center because he does not qualify for services that most individuals with I/DD receive

(allowing easier access to records). A medical professional trying to access a baseline measure for someone who has fallen through the cracks, and has no current advocate, must know where to go to find this baseline. This is something that a caregiver, or advocate needs to be aware of, when the individual with I/DD is younger, and record it. This literature review can be usefully applied to our family by defining what measures need to be followed and at which stages of our individual with I/DD's life it should be accomplished. Since life expectancy is increasing, even in individuals with I/DD, there are great chances that I, as advocate, will not be around to help when my children reach the age of developing possible dementia. Planning needs to be done now to advise later caregivers what actions need to be taken for proper care of my children, if they develop comorbid dementia with their existing I/DD.

Applying this Literature Review to Community Residential Relationships

The work that I do in our community offers exposure to many situations in the I/DD community. Several years ago, there was a man with DS who developed pneumonia. Towards the end of his life, he would be in the hospital for a length of time during which the pneumonia would get treated and improve, then he would return to his residential care home and become worse again. This happened several times. I wondered often what it was that the staff at his home were doing that was making it so that he could not get fully well. I even went so far as to think possibly they were neglecting him. He was 65 years old. From this literature review I have learned that because this man had DS, he had a high risk of having comorbid AD. Looking back, I can remember signs of disease-related decline in his behavior. I have also learned from this literature review that pneumonia is part of the progression of dementia. If the residential staff at this man's home had had training in best practices for care of older individuals with co-

morbid I/DD + dementia, or even a source to go to in order to understand what was happening to this man, his end of life could have been a better experience for all of them.

Community Residential staff needs to be trained on what to look for in the behavior of residents at their home. They need to have access to personal records to see if a particular behavior change might be a sign of disease-related decline or is the pre-existing functioning level of the individual with I/DD. Training in what is normal aging behavior in individuals with I/DD compared with what may be disease-related decline should be offered to all residential staff that are hired in all levels of housing. Reading materials should also be kept at each home as reference when/if a staff has questions.

Applying this Literature Review to Institutional Relationships

As mentioned earlier, the current inclination of individuals in the general population who are aging is to "age in place". This means that a person who is aging will remain in the place that they are familiar with as they grow older. Many communities, called Continuing Care Retirement Communities (CCRC), are being built that allow an aging person to move into an "independent living" apartment, then into an "assisted living" situation and finally to a "memory" (dementia) or "acute care" living all while staying in one facility's location. This allows the person to receive the care he/she needs as the decline happens, reducing the stress of constant adjustment to new surroundings.

Care of I/DD is developing a similar outlook. Individuals with I/DD are part of the 'Inclusion Revolution". They are often housed in neighborhood houses in regular communities with 24-hour staff to help them as needed. Finding housing that fits for the adult with I/DD is a goal of most parents. Adult individuals with severe or profound mental retardation also live in community residential housing, which I have personally observed. The residential trend is

moving away from institutional setting. When the comorbid individual with I/DD + dementia moves into the final stage and needs more medical assistance, they are at that time moved into institutions.

When the Usual Caregiver can No Longer Care for the Individual

As mentioned earlier, sometimes, if the caregiver is the parent, the caregiver themselves might move into disease-related decline and not be capable of caring for the individual with I/DD + dementia. This is the time that it is very important that plans have been made, early in the dementia stages, for support. If the caregiver and individual with I/DD + dementia have laid out a plan, while both were in their right minds support might continue with little adjusting. Sometimes the support that is provided for the individual with I/DD + dementia can also benefit the past caregiver. Occasionally the caregiver will be mentally capable but not able to physically care for the individual with I/DD + dementia any longer. This is another occurrence that, for smoother transitions for all, should be addressed when dementia stages are early, while planning for later support.

When the individual who has been informally cared for, moves into the final stage of dementia and is non-ambulatory, bedridden, has flexed postures, sphincter incontinence, pathological reflexes, aspiration, pneumonia, and other re-occurring infections (Lai & Williams, 1989) they may need to be moved to an institution that can provide 24-hour medical care. Informal caregivers will find themselves lost for a while during this adjustment period. They may visit the individual several times a day, or they may be thankful to not have the burden of being a caregiver any longer and involve themselves in many activities. Different personalities cope with change and adjustment in different ways. In the case of the individual with I/DD + dementia, there is a chance that the caregiver has been in the role of caregiver for a very long

time, possibly, since birth. The last transition might be a larger event for the caregiver, than it is for the individual. Caring for the individual with I/DD + dementia can be a large project for the caregiver. The final stages of the disease are a difficult adjustment time for all involved.

Chapter 6: Future Directions

Getting Education to Caregivers

My overall conclusion from my review of literature is that much research needs to be conducted not only on comorbid I/DD + dementia but also on the "stand alone" single conditions. Education of the conditions and diseases under the I/DD and Dementia umbrellas need to be readily available EVERYWHERE and to EVERYONE. Third world countries and low SES areas need to be included. In this technology savvy modern age, there should be no places in the world that still believe that an I/DD or a dementia is a curse or a punishment. Care guidelines and provisions need to be available to all people. To provide for the needs of all people, a proper measure of prevalence must exist. Assistance, both monetarily and bodily, should be offered to current researchers from organizations like WHO. When areas of higher prevalence are found in certain areas, providing education materials should be a higher priority to those areas. Attention needs to be drawn to research on all disabilities all over the world.

Brochures, Pamphlets, Booklets

Existing brochures, pamphlets, booklets, and diagnostic instruments need to be translated into more world languages. As in the example of Petris De Vries noted earlier in this document, availability of these items in multiple languages needs to be shared with all medical doctors. Medical doctors should also be educated on medical, epidemiological, and treatment/support literature that is available for their patients who may be coping with an I/DD, dementia, or comorbidity.

Information on brochures and other materials should include topics such as building a baseline during the peak of adult functioning. Pamphlets continuing information on support systems should be offered in not only physicians' offices, but also in mental health facilities,

hospitals, health clinics, any offices of assistance to lower social economic status (SES) groups, special education programs, and anywhere that someone with an I/DD might attend.

It would be of great value to also place education materials into community residential homes for staff to have available for review and reference. Informal family caregivers should also be provided with classes to train them on what to expect. There can never be enough information available on disabilities and comorbidity of them.

Programs for Training of Caregivers

Through this literature review and planned research to come, my goal is to create a training program to be administered to all people who will be stepping into the shoes of a caregiver. This training program would be mandatory for all community residential services and institutions who offer I/DD + dementia care in the USA. It should also be made mandatory for informal family caregivers of individuals with I/DD + dementia. It should be translated into other languages to help other countrys' caregivers.

Home based caregivers (informal) would benefit from this program by learning about comorbid I/DD + dementia. They would learn how to recognize signs of disease-related decline in their ward. Included would be techniques for handling decline-related behaviors and references for building a support network. Recommendations on where to record important information for possible later caregivers and support would also be offered.

Residential caregivers (formal, paid) would receive training, upon hire, about recognizing disease-related decline in the individuals with I/DD that they will work with. Information about techniques for handling decline-related behaviors and references for building a support network would be included. Recommendations on how to document records in the community residential system that they work in would also be included in the training.

A training program that covers all the little factors of caring for an individual with comorbid I/DD + dementia will need to include comprehensive research on anything that might affect that condition. For example, a program should be able to teach the people participating in it, how to differentiate symptoms of dementia compared to MCI; or other diseases with symptoms resembling dementia. Multiple people with comorbid I/DD + dementia need to be observed to determine which residential placement, treatment, and support are "best practices". The NTG has a good start and I will be following their existing guidelines to create this program. This literature review has been a good step in beginning the walk that will make a difference in the aging of future I/DD elderly.

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Appendix A: List of Abbreviations

AD	Alzheimer's Disease
ADC	Adult Day Care
ADD	Attention Deficit Disorder
ADHD	Attention Deficit Hyperactive Disorder
ADI	Alzheimer's Disease International
ADI-R	Autism Diagnostic Interview- Revised
ADL	Average Daily Living Skills
iADLs	Individual Average Daily Living Skills
ADOS	Autism Diagnostic Observation Schedule
ASD	Autism Spectrum Disorder
BI	Barthel Index
BLDC	Big Lakes Developmental Center
BOLD	Bold Cohort (Strydom literature)
BPD	Borderline Personality Disorder
CCCHS	Clay County Cooperative High School
CCRC	Continuing Care Retirement Community
CDC	Center for Disease Control
CDDO	Community based Developmental Disabilities Organization
CRPD	Convention on the Rights of Persons with Disabilities
CSP	Community Service Provider
DAT	Dementia of the Alzheimer's Type
DMR	Dementia Questionnaire for persons with MR

DS	Down's Syndrome
DSDS	Dementia Scale in Down Syndrome
DSM-IV	Diagnostic and Statistical Manual of Mental Disorders-5th edition
ID-10	International Statistical Classification of Disease-related Health Problems
IQ	Intelligence Quotient
I/DD	Intellectually and/or Developmentally Disabled (or Delayed)
IDS	Intellectually Disabled Supplement
IEP	Individual Education Plan
MCI	Mild Cognitive Impairment
MMSE	Mini Mental State Exam
MR	Mentally Retarded
NAPA	National American Protection Act
NCD	Non-communicable Disease
NDSS	National Down Syndrome Society
NTG	National Task Group
NTG-EDSD	National Task Group-Early Detection Screen for Dementia
OCD	Obsessive Compulsive Disorder
RCSP	Residential Service Provider
SES	Social Economic Status
SOA	Schizoaffective Disorder
TDA	Toolkit of Disability for Africa
TILDA	The Irish Longitudinal Study of Aging
UN	United Nations

UN DESA	United Nations-Convention on Rights of Persons with Disabilities
UK	United Kingdom
USA	United States of America
WHO	World Health Organization
W/O	Without

Appendix B: Sources of Supplemental Data

Forms of Supplemental Data

ICD-10:

https://apps.who.int/iris/bitstream/handle/10665/37108/9241544554.pdf?sequence=1&isAllowed=y

DSM-V:

https://goo.gl/images/xhZqzf

NTG Guidelines:

http://aadmd.org/sites/default/files/NTG-communitycareguidelines-Final.pdf

Appendix C: Figures and Tables

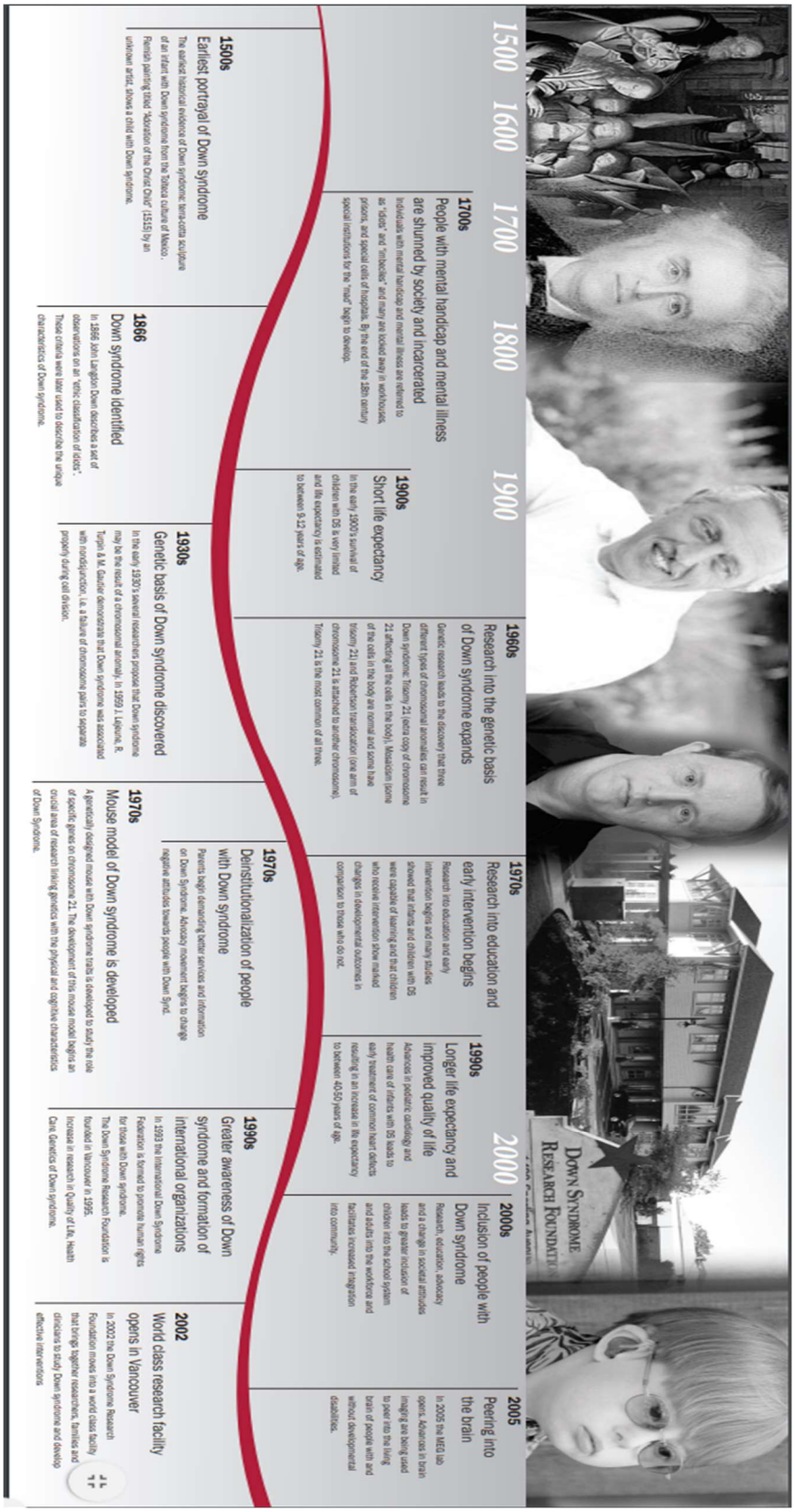

Figure 1: Historical to present day growth in the awareness and treatment of Down's Syndrome, classified as an Intellectual and Developmental Delay. Retrieved from https://www.dsrf.org/media/timeline.pdf.