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An introduction to the identification and care of individuals with Down syndrome complicated by Alzheimer's disease

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Running head: ALZHEIMER'S DISEASE IN DOWN SYNDROME

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Table of Contents

Appendix

Chapter One

Introduction

The average life span for an individual with Down syndrome (DS) has increased from less than 10 years in 1920, to over 60 years in 2000 (Watchman, 2003). While the correlation between Down syndrome and Alzheimer's disease (AD) was recognized as early as 1948 (Lott & Head, 2001), the awareness of the impact of this finding has not had clinical relevance until the recent past, as individuals live longer and thus manifest the dual disease process in much greater numbers. Mainstreaming of the Down syndrome population from institutional settings to community-based homes places a greater responsibility on community-based direct care providers to be cognizant of and responsive to the health care needs of this vulnerable group of individuals.

In the general population, Alzheimer's disease is often heralded by the onset of memory loss. In contrast, an individual with Down syndrome in early stages of Alzheimer's disease is much more apt to display changes in personality and behavior (Ball, Holland, Hon, Huppert, Treppner & Watson, 2006). Failure to recognize warning symptoms can result in failure to provide early intervention; deleterious in this group as Alzheimer's disease already occurs at a much earlier age and with greater incidence as compared to the general population. The course of AD in DS has been described in an article by Janicki et al. (2002) as "...compressed, the duration short, and the decline precipitous." It is widely acknowledged that individuals with Down syndrome possess the neuropathological changes necessary for a diagnosis of Alzheimer's disease by age 40 (Visser et al., 1997; Schupf, 2002; Teipel & Hampel, 2006; Head & Lott, 2004), however few studies have been done to assess the efficacy of various forms of treatment, including the use of acetylcholinesterase inhibitors. While detecting cognitive decline in

individuals with Down syndrome is challenging, devising methods to provide early diagnosis and prompt treatment/interventions is essential if we are to help these individuals, as best they are able, to lead full and meaningful lives.

A major difficulty in diagnosing Alzheimer's disease in Down syndrome results from the cognitive impairment these individuals already display. While numerous tools have been designed for the purpose of dementia diagnosis, most are not applicable to an individual with preexisting cognitive impairment. To this end, tools have been designed in an attempt to capture the more subtle indicators of this population. As with all individuals, the diagnosis of Alzheimer's disease is one of exclusion, and all possible differential diagnosis must first be excluded before a dementia diagnosis can be made.

Another problem inherent to this population is the general lack of awareness of the potential for someone with Down syndrome to acquire Alzheimer's disease, the increased risk, earlier manifestation and more precipitous course. Although 60 years have passed since the first description of the correlation between Alzheimer's disease and Down syndrome, individuals with Down syndrome are now living much longer, allowing those responsible for their care to become cognizant of the co-morbid condition and the need for appropriate care.

The purpose of this project is to generate awareness of the propensity for individuals with Down syndrome to succumb to a disease process that severely limits their potential to experience a satisfactory quality of life. Awareness brings the potential for providing means of earlier diagnosis and appropriate care. Many individuals with Down syndrome are now living in the community in group homes, family settings, or in individual apartments. The onset of Alzheimer's disease necessitates a major revision in plan of care concerning housing, work, medication usage, social life and more. Determining care that best meets the needs of individuals

with continued cognitive decline is an essential responsibility of those for whom their care is entrusted. End-of-life issues will also be addressed.

An assumption for this project is that many individuals with Down syndrome possess only a mild to moderate degree of cognitive impairment. This, in fact, is not always the case. Many individuals with Down syndrome suffer profound retardation; the ability to diagnose a co-morbid condition in those with less impaired cognition can be extremely difficult, and to recognize the changes in those with profound retardation can be daunting.

A factor limiting the treatment of these individuals is a failure to include this group of individuals in trials designed to test medications used to treat Alzheimer's disease, including such agents as donezepril. Lack of knowledge regarding safety and efficacy impacts a practitioner's decision to use treatment modalities, and requires a type of trial and error approach to therapy. Donezepril has been used with varying results, however further dedicated trials are needed.

Conceptual/Theoretical Framework

The theoretical model chose for this project is The Elements of Nursing: A Model for Nursing Based on a Model of Living designed by Nancy Roper, Winifred W. Logan and Alison J. Tierney. While their works have undergone several revisions, the basic tenets of the framework have persisted. First published in 1980 as The Elements of Nursing, the framework was last revised in 2000 as a monograph to preserve the conceptual model while allowing the individual practitioner to adapt the model to individual circumstances (Tomey & Alligood, 2002).

In this framework, individuality in living is assessed through 12 dimensions of activities of living as elucidated below:

- Maintaining a safe environment
- Communicating
- Breathing
- Eating and drinking
- Eliminating
- Personal cleansing and dressing
- Controlling body temperature
- Mobilizing
- Working and playing
- Expressing sexuality
- Sleeping
- Dying

These factors are further influenced by biological, psychological, sociocultural, environmental and politicoeconomic factors, and are further defined on a continuum of dependence and independence which can in some circumstances be seen as a changing value.

Individuality in nursing is seen as addressing all of the same issues as those attendant to individuality in living, with the addition of provision for providing individualized nursing care through appropriate assessment, planning, implementing and evaluating.

This theoretical model is appropriate for this topic because it fully recognizes and addresses the needs of individuals within multiple settings, recognizing the value of people at all stages of life regardless of psychological or sociocultural circumstances. It addresses the need for

preventative care in an effort to maintain health and optimal functioning, but it also recognizes that not all problems can be solved, and that coping with death and dying in a positive way is a means of furthering optimal individuality in living.

Attending to activities of daily living is seen as one way that healthcare providers can attend to the changing needs of a person with Down syndrome complicated by Alzheimer's disease. In fact, alteration in activities of daily living may well provide the means of prompting caregivers to examine subtle changes that may well serve as early indicators of early cognitive decline.

Definitions

Alzheimer's Disease – Defined by Tabers (20th Ed) as "A chronic, progressive, degenerative disorder that accounts for more than 60% of the dementias."

Amniocentesis - In suspected Down syndrome, the retrieval of an amniotic fluid sample to allow for genetic testing.

Beta-amyloid plaque – A substance formed from the cleavage of amyloid precursor protein (APP) which is deposited in neuritic plaques; referred to as "the hallmark of AD pathology" by Sun et al. (2006).

Brachydactyly – Abnormal shortness of fingers and toes.

Chorionic villus sampling – Obtaining a sample of the membranes surrounding a fetus, generally performed at 8 – 12 weeks, to determine the presence of chromosomal abnormality. Can provide a means of prenatal diagnosis of Down syndrome.

Clinodactyly – Hypoplasia of the middle phalanx of one or more fingers which results in an inward curving of the affected finger. Often noted in the small fingers of a person with DS.

Down syndrome – Defined by Tabers (20th Ed) as "the clinical consequence of having three copies of chromosome 21."

Intellectual disability – Below-average intelligence as measured on scores of ability, a term used to describe life-long mental, physical or combined mental/physical limitations.

Intelligence Quotient (IQ) – A measure of intelligence based on an individual's response to a set of arbitrarily chosen questions; may not provide an accurate picture of skill or potential.

Self-determination – In health care, the ability of the individual to control and manage decisions regarding their own health care

Simean crease – An apparent fusion of the two flexion creases found on the palm of the hand, forming a single transverse crease as noted in a variety of developmental abnormalities including Down syndrome.. The term simian refers to a type of monkey, from whom the description is obtained.

Significance of the Project

According to the National Down Syndrome Society, there are approximately 350,000 individuals with Down syndrome living in the United States today, and another 5000 children are born each year. The average lifespan of an individual with Down syndrome is approaching 60 years, and occurrence of Alzheimer's disease in this age group is estimated at 30 – 75%. The face of Down syndrome may change over time with improved genetic counseling/screening and advancements in medicine, but current numbers point to a need for aggressive intervention for the current population.

In 2001, The Arc, a group of people devoted to improved support and services for people with intellectual and developmental disabilities, estimated that there were 9000 adults with

mental retardation affected by dementia, and that the number was expected to triple over the next 20 years. These figures are supported by Janicki et al. (2002). This is perhaps a conservative estimate, as tracking of individuals with Down syndrome and Alzheimer's disease is difficult. It would be anticipated that the numbers would continue to grow with increasing longevity due to better health care.

Deinstitutionalization of the target population places the care and responsibility of these individuals in the hands of general practitioners, unlike earlier times when care was dependent upon a select few. It is likely that the enhanced health care these individuals have received has been a major contributing factor to their markedly increased life span.

Chapter Two

Review and Critique of Related Studies

Down syndrome, as defined by Tabers (20th Ed) is "the clinical consequence of having three copies of chromosome 21." This nondisjunction of chromosome 21, accounting for the syndrome known as Trisomy 21, is present in 95% of cases. Other causes include Robertsonian translocation of chromosome 21 to chromosome 14 (4% of cases) and mosaicism. The importance of genetic counseling to determine the specific cause of Down syndrome is most important when considering another pregnancy, as the risk of having another child with Down syndrome is altered significantly by the mechanism involved (Tyler & Edman, 2004).

Tyler & Edman (2004) assert that Down syndrome "is the most common chromosomal cause of mental retardation", and that Down syndrome generally results in a person having mild to moderate mental retardation with an IQ ranging from 30 to 70. The average IQ of the general population has arbitrarily been set at 100.

The ability to diagnose Down syndrome antenatally is achieved by use of the triple screen, consisting of maternal serum alpha-fetoprotein (AFP), unconjugated estradiol and human chorionic gonadotropin (HCG) (Roizen & Patterson, 2003). In Down syndrome, levels of maternal serum alpha-fetoprotein and unconjugated estradiol are reduced, while levels of maternal serum HCG are doubled in number. Measurement of the triple screen in the first trimester of pregnancy allows for a 69% detection rate with a 5% false-positive rate ((Roizen & Patterson, 2003). Use of ultrasound to detect nuchal thickening increases the detection rate to 80-85%, with a 5% false-positive rate (Roizen & Patterson, 2003). Other measures of antenatal detection include chorionic villus sampling and amniocentesis. Diagnosis of Down syndrome after birth is by physical appearance (Tyler & Edman, 2004). Children with Down syndrome

(DS) can exhibit over 100 anomalies including the characteristic epicanthal folds, oblique palpebral fissures, brachycephaly, brachydactyly, Simean crease, flat nasal bridge, short stature, protruding tongue, hypotonia, excess nuchal skin, hyperflexibility, short 5th finger with clinodactyly, and short broad hands (Roizen & Patterson, 2003; Tyler & Edman, 2004; Bosch, 2003). Borth (2003) stresses the importance of confirming a DS diagnosis with karyotyping to exclude other possible genetic etiologies, and for genetic counseling for families. Caring for individuals with DS requires knowledge of less visible manifestations as well, including sensory deficits, possible cardiac involvement and duodenal atresia (Borth, 2003). This project is limited to the manifestation of Alzheimer's disease in DS.

Alzheimer's disease, as defined by Tabers (20th Ed) is "a chronic, progressive, degenerative disorder that accounts for more than 60% of the dementias. Birth numbers of individuals with DS are declining, with estimates of pregnancy termination rates as high as 92% of Down-afflicted fetuses (Tyler & Edman, 2004). Nevertheless, with a life-span now approaching 60 years and beyond (Watchman, 2003), the increased propensity for developing Alzheimer's disease in this population has become increasingly evident. Whereas Jervis first identified the connection between Down syndrome and Alzheimer's disease in 1948 (Lott & Head, 2001)) the impact of his findings have become more relevant with increased longevity. Deinstitutionalization of this population into community settings has also raised awareness of the need for a plan of care if these individuals are to be assisted in living productive and meaningful lives.

The neuropathological changes of Alzheimer's disease are the result of beta-amyloid plaques and tangles, resulting in neuron cell death (Temple et al., 2001; Lott & Head, 2004; Lana et al., 2007; Sun et al, 2006). The pathophysiology is complex and incompletely understood,

involving numerous factors such as BACE 1, BACE 2, SOD 1, and more (Bush & Beail, 2004; Sun et al., 2006). A review of the pathological processes involved in the development of AD in DS is beyond the scope of this paper. What is important to note is that people with DS possess the neuropathological changes necessary for a diagnosis of AD by age 40 (Allen et al., 1999; Head et al., 2007; Lott & Head, 2005; Teipel & Hample, 2006; Temple et al., 2001). Although the median age of onset of dementia in individuals with DS is 50 years (Margallo-Lana et al., 2004), the characteristic plaques and tangles have been noted in a DS individual as young as eight years (Head et al., 2003). By the age of 65, an estimated 30 – 75% of the DS population will have AD, compared with 13% of the general population (AARP, 2007).

Diagnosing the onset of dementia in individuals with DS is difficult because of the preexisting cognitive deficits. If coupled with sensory impairments such as hearing and vision loss, detecting subtle indicators of decline can be a formidable task. In addition, most exams that are utilized to detect dementia are geared towards a population of normal intellect, such as the Mini-Mental State Exam. Tests that have been specifically devised to address the needs of the DS population include the Dementia Screening Questionnaire for Individuals with Intellectual Disabilities (DSQIID) by Shoumitro et. al. (2007), the Dementia Scale for Down Syndrome by Geyde (1995), the Dementia Questionnaire for Persons with Mental Retardation (Evenhuis, 1996) and the Spatial Recognition Span (Moss et al., 1986). The administration, scoring and interpretation of these exams should be done by a clinical psychologist or others with experience in testing people with learning disabilities. Several authors point to the need to administer a test battery to determine baseline cognitive function by age 30, which could be reviewed periodically as a means to provide recognition of early decline, at a time when medical intervention might be most therapeutic (Burt et al., 2000; Stanton & Coetzee, 2004; Shoumitro et al., 2007; Pyo et al.,

2007). Another option in detecting early changes of a dementia process is proposed by the evaluation of an individual's ability to complete activities of daily living (ADLs), an example being the Alzheimer Functional Assessment Tool, a copy of which is provided at the end of this paper (see Appendix). Categorically identifying and then systematically reviewing an individual's ability to complete ADLs over time is an inexpensive and noninvasive means of obtaining data that might assist in raising the index of suspicion and securing appropriate intervention for an individual exhibiting decline in self-care. For best results, observation and completion of an ADL inventory should be accomplished by a family member or direct caregiver with extensive knowledge of the person in question.

Suspicion for a beginning dementia process requires first that alternative diagnosis be excluded. In the case of DS, several conditions may mimic AD including hypothyroidism, depression or psychological condition, normal aging, obstructive sleep apnea, sensory loss (hearing & vision), change in environment, delirium, chronic hepatitis, medication effect (i.e. anticholinergics), abuse, other forms of dementia, neoplasms and metabolic abnormalities (Van Allen et al, 1999; Stanton & Coetzee, 2004; Bush & Beail, 2004). These same authors propose a test battery for the exclusion of these and other disorders including a CBC, Comprehensive Panel 12, ESR & CRP (inflammatory markers), thyroid function studies, folate & B12, CT/MRI of the brain, EEG, plasma levels of drugs such as digitalis and anticonvulsants, urinalysis, vision and hearing screening and more. Intended as a reference only, the proposed studies would be completed at the discretion of the party involved as well as his/her significant others and the primary provider. 'It's Your Move,' a publication from Down Syndrome Scotland provides a comprehensive visual aide to assist a caregiver in determining treatable conditions that might mimic AD, and is included in the Appendix to serve as a reference.

Unlike the general population for whom loss of memory provides the first indication of declining cognition, a person with DS most frequently presents with alteration of personality and behavior, changes consistent with a frontal lobe dementia (Ball et al., 2006). Symptoms mentioned by Ball include social withdrawal and apathy, excessive stubbornness, depressed mood, restlessness, disturbed sleep and uncooperativeness. These findings, as with those of declining ADL ability, require a review of the differential diagnosis to provide an appropriate diagnosis, thus maximizing the treatment potential.

The goal of care in DS complicated by AD can be assessed via the ECEPS approach (Janicki et el., 2002).

1. Early screening and diagnosis – periodic assessments with early referral for differential diagnosis

2. Clinical support – assistance of trained consultants to provide assessment and intervention. These persons provide support for direct caregivers and assist with developing programs and care management, as well as training for families and support staff.

3. Environmental modifications – modification of the existing environment to allow "aging in place" if possible, including such things as door alarm devices, wandering paths, enhanced lighting, modification of bathroom facilities, and more.

4. Program adaptations – restructuring the environment to reduce stimulation while retaining the level of functioning and independence as best a person is able.

5. Specialized care – mid-stage and end-stage interventions to meet the increasing needs of someone with a progressive disease.

For an individual with DS/AD, the decision to remain in their usual care setting might be ideal, but perhaps not feasible. Safety concerns or health care needs may dictate alternate

placement, such as a long-term care facility. If individuals with DS and AD are to remain in their usual care setting, financial reimbursement needs to be increased to provide for the additional resources needed to maintain care, including additional staffing and remodeling of the existing environment. Persons with DS complicated by AD require a significant increase in caregiver time as noted by McCarron et al. (2002). This study notes that in mid-stage dementia, the number of hours of care climbed from 2.53 per 24 hours per person in the non-demented population, to 9 hours per 24 hours per person in the mid-stage demented population, related primarily to the need to attend to management of behavioral problems. At end-stage, the average number of hours was 8.86 hours per 24 hours per person, with more time being devoted to meeting physical needs. Additional areas of concern are caregiver strain related to DS/AD, the impact of the diagnosis on other parties that might share a group home, and behavioral issues that are not easily controlled. Because the issue of AD in DS is an evolving one, the best answers to these and other questions regarding care remain unanswered.

End of care issues in DS/AD are not unlike those in the general population. As a person's condition deteriorates, decisions need to be made whether to pursue such options as feeding tube placement to maintain nutrition and hydration, how to maintain skin integrity in the face of severely limited mobility and incontinence issues, pain control issues, and whether or not to treat frequent infections, among others. The ethics of care are complicated with no easy answers for involved parties. This is perhaps a time when professional and non-family caregivers, with greater awareness of disease progression, can guide a family in making difficult choices when their loved one is no longer capable of any semblance of self-determination.

Service (2002) describes how sensory experiences supplant cognitive experiences in the late stages of intellectual disability (ID) complicated by AD. She notes the usefulness of

activities such as "gentle massage, quiet music, scented hand lotion, flavored lip glosses and talking to the individual even when he/she is unresponsive…" She quotes Stephen Post (1995) in stating that "the first task of dementia ethics is to secure the underpinnings of care; that is, the appreciation and respect for compassion. In a culture such as in the U.S., that is hypersensitive and values productivity, it is easy to think that people with dementia lack any moral significance." His position is perhaps magnified when the person in question is also afflicted by mental retardation.

Perspectives vary in what type of care, how much care and by whom it should be given for individuals with DS complicated by AD. Service (2002) reminds us that the amygdala, the portion of the brain which receives and attends to emotions, is "one of the last parts of the brain affected by this disease". She stresses that emotions are accessible to carers until very late in the disease process; the implications for care are self-evident. And finally, Stuart Todd (2002) reminds us that "if the lives of people with intellectual disabilities are to count as significant, then surely their deaths must also have some consequence."

Chapter Three

Methodology/Procedure

The goal of my project, as defined by the title, was to prepare a presentation that would assist caregivers in identifying the correlation between Down syndrome and Alzheimer's disease, including age of onset, presenting signs, and the need for early diagnosis and intervention. The program was supported and offered by the Collegial Discipline Team of the Grafton Developmental Center, and all staff and consumers of the North Dakota Developmental Center were encouraged to attend. The Collegial Discipline Team provided campus-wide advertising, and scheduled the program at a time that was likely to allow participation. With more than one presenter present that day, individuals were able to sign up for topics of personal/work-related interest. Ten people chose to attend the presentation on DS with AD representing multiple disciplines across the health care delivery continuum, including clinical psychologists, nurses, occupational therapists, and one consumer. Two of the participants had family members with Down syndrome. Participants were asked to complete a 10 point multiple choice and true/false quiz prior to the presentation (see Appendix) dealing with items such as expected life span, age of AD onset, symptom presentation and more. Questions and participant input were solicited before, during and following the presentation. At completion, the participants were asked to complete the same 10 point multiple choice and true/false quiz so that evidence of learning could be assessed. The average score on the pre-presentation quiz was 54.44; average score on the post-presentation quiz was 84.44, representing a 30 point increase in test results. Correct answers are provided in bold in the appendix.

The first test item asked what the current life span was for an individual with Down syndrome, with answers ranging from 40 to 70 years of age. Eight persons answered this

incorrectly on the initial quiz, with the vast majority believing the life span to be 40-50 years of age. Only one person answered incorrectly on the post exam. This item was intended to provide the learner with an awareness of current life span.

Item number two asked the median age of onset of Alzheimer's disease in Down syndrome, with answers again ranging from 40 to 70 years. Seven persons answered this incorrectly on the pre-exam, with most respondents replying that the average age of inset is 40 years old. On the post test, only two persons provided an incorrect response.

Item number three asked at what age individuals with Down syndrome display the characteristic brain lesions necessary to provide the diagnosis of Alzheimer's disease. On both pre and post-exam, only one person provided a wrong response. This was surprising, in that I felt this piece of information would not be a well-known fact.

With information provided by the North Dakota Department of Health, item number four asked the average number of individuals born with Down syndrome per year from 1996 to 2006 (no data from 2004 were available). This item was included solely to provide an awareness of the incidence of Down syndrome in North Dakota; learners were cautioned that no statistically relevant information could be gleaned, but that the record provided a way of looking at trends since 1978. One-half of the respondents answered incorrectly in assuming a number larger than the 6.11 average births/year in the 10 years for which data was available. No one got the item wrong on the post test.

Items 5-10 were true/false statements.

Item number five stated that neurological changes in Down syndrome occur at the same time as they do for the general population. In essence, this was a rewording of test item number three, and in this case no one answered incorrectly on either quiz. The respondents of this

particular group seemed well-versed in the early onset of neuropathological changes in the individual with Down syndrome.

Item number six addressed the neuropathological changes of AD as being related to beta-amyloid deposition. While the intent of the lecture was not to provide a pathophysiological background, the item was included to provide a baseline level of understanding of the pathological process. The item was scored correctly 100% on both pre and post-test, again supporting the level of learner understanding of the process of AD.

Item number seven stated that changes in memory provided the first evidence of AD in DS. Four answered incorrectly on the pre-quiz, two answered incorrectly on the post-quiz. My expectation was that this would have been a lesser understood concept, with more respondents believing that AD would manifest with memory impairment in DS, as is typical in the general population.

Item number eight stated that the loss of cognitive ability was diagnostic for Alzheimer's disease. Four people answered this incorrectly on the pre-test and the post-test quiz did not show an improvement. This was disappointing, as one of the key points of the presentation was to make the audience aware that loss of cognitive ability was a reason to pursue a differential diagnosis before assuming that a decline in actual cognitive status was occurring. It is possible that the wording of the test item was suggestive or contrived, or that there was failure to convey the importance of ruling out alternative causes for apparent cognitive decline. This would be an area that would require review and revision on a subsequent presentation, as it is key to the concept of early, appropriate diagnosis.

Item number nine stated that individuals with DS/AD are less apt to feel and exhibit emotion as Alzheimer's disease progresses. While this was meant to be a false statement, three

responders believed it to be true on pre-test, and the number increased to four on post-test. This item was addressed as an end-of-life issue, and my intent was to make carers aware that loss of the ability to feel and express emotion is one of the last abilities to be lost. I believe that wording of the item was problematic, in that while individuals with DS/AD do feel and exhibit emotion, there is undoubtedly some decline in that ability. It is also possible that learners were operating from their own personal work experiences or worldview. The lone attending consumer, whose quiz results were not included in the quiz averages, wrote only on her test that "we have feelings too."

The final item stated that symptoms of AD progress at the same rate in individuals with DS as they do for the general population. While this is false, four respondents answered incorrectly on the pre-test. There were no incorrect answers on the post test.

The primary objectives of the presentation were to increase the learner's awareness of the prevalence of AD in DS, the difference in age of onset, presentation and disease progression. Test items were worded to address these primary concerns.

This power point presentation was provided to a group of learned individuals with expertise in the care of individuals with developmental disabilities, including Down syndrome. A limitation of the presentation was that participants were moving from lecture to lecture, providing little time for interactive discussion and questions. Including space and inviting participants to provide suggestions for improving the format and/or content would have provided useful feedback for the instructor as well. Having a clear idea of the target audience prior to delivery would also help the instructor to target the information to the needs of the learner.

Implications for Nursing

Practice: As has previously been noted, deinstitutionalization necessitates that care of individuals with DS and AD no longer falls into the hands of a select few. These people are receiving their health care in community clinics, hospitals, long-term care and group-home settings. Many health care goals are the same for individuals with and without disability, such as the need for disease prevention, safety awareness, immunization practices and education, however individuals with DS, especially if complicated by AD present with a host of special concerns, of which nursing needs to be aware. Impaired cognition requires a honing of skills to extrapolate key information to provide an accurate diagnosis and plan of care. Involving care partners and other disciplines is mandatory to facilitate comprehensive care.

Research: AD in DS requires extensive research in this specific population to determine the mechanism of action for the co-morbid condition. In the interim, research needs to be done to determine what types of testing are apt to yield the best results concerning onset of AD and what treatment measures are most effective. Specifically, studies must be performed to determine the safety and efficacy of pharmaceutical interventions, placement and care options, use of non-pharmacological agents to enhance quality of life and how best to care for these individuals when end-of-life approaches. Research must address how to meet the needs of individuals in the most therapeutic yet cost-effective manner.

Education: Because care of individuals with DS has traditionally happened in a sheltered type of setting, the mainstreaming of individuals, especially when their health deteriorates, has been at times unnerving and problematic for carers not familiar with this population. Curriculum of study for any type of health care provider needs to include information on the health care needs

of these individuals. Knowledge enhances a provider's sense of capability, and will result in improved understanding, care and compassion.

Summary/Assessment

The concept of Alzheimer's disease in Down syndrome is not new. What seems most apparent is lack of awareness of the co-morbid conditions in the general population, as well as knowing how to intervene through early recognition and prompt intervention. The issue is a decidedly difficult one, as recognizing AD in a person with a pre-existing cognitive deficit is not easy. Financial constraints, staffing, lack of education and lack of evidenced-based studies determining best care practices contribute to the potential for delayed recognition and inadequate care measures. Care of these individuals has fallen to community-based organizations dedicated and wholly-committed to the well-being of DS/AD individuals. Care centers are coping with the dual diagnosis as best they can, generally without additional reimbursement and always with awareness of the effect of the increased workload on staff, other clients and the afflicted individual. As prevalence rates continue to increase, research will need to devise ways and means to enhance quality of life for the person with DS/AD, while lightening the load of care providers.

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APPENDIX

Alzheimer's Disease in Down Syndrome

Jody Sharp

What is Down Syndrome?

- "The clinical consequence of having three copies of chromosome 21" - Tabers, 20th Ed.
  - Nondisjunction
  - Translocation
  - Mosaicism
- Most common chromosomal cause of mental retardation (MR)
- Results in mild to moderate cognitive impairment
  - Average IQ 50; range 30-70

Physical Characteristics

- Short head
- Short fingers and toes
- Broad hands with a single palmar crease (Simean crease)
- Epicanthal folds
- Oblique palpebral fissures
- Inward curving of the 5th finger

Physical Characteristics, cont.

- Flat nasal bridge
- Open mouth with protruding tongue
- Short stature
- Wide gap between great and 2nd toe
- Excess nuchal skin
- Hypotonia
- Lax ligaments with hyperflexibility
- Congenital cardiac defects
- Duodenal atresia

Diagnosing Down Syndrome

- Prenatally:
  - Triple Screen
    - Decreased levels of maternal serum alpha-fetoprotein & unconjugated estradiol
    - Two fold increase in maternal serum HCG
  - Ultrasound
    increased nuchal fold thickness due to edema
  - Amniocentesis
  - Chorionic villus sampling

Diagnosing Down syndrome, cont.

- Using triple screen with ultrasound results in 80-85% detection rate, with 5% false positive
- Postnatally – Diagnosis is by physical appearance

DS Items of Interest:

- Pregnancy termination rates:
  - Estimated at 27 - 92%
- Number of Down syndrome births has declined from 1:700 to 1:1000
- Life span
  - From <10 years in 1920 to >60 years in 2000
- Review of North Dakota statistics
  - ~ 6 births of individuals with DS per year over past 10 years (see Appendix)

DS Items of Interest, cont

- Current estimates of 350,000 individuals with Down syndrome in the United States
- In 2002, estimates of 9000 individuals with intellectual disability and dementia, with numbers expected to triple in the next 20 years

What is Alzheimer's Disease?

- "A chronic, progressive, degenerative disorder that accounts for more than 60% of the dementias" -Tabers
- Discovered by Alois Alzheimer, a German neuropathologist and psychiatrist
  - 51 year old female case history (1901)

Stages of AD-General Population

- Stage 1
  - Mild memory loss
- Stage 2
  - Deterioration of intellectual functioning
  - Personality changes
  - Speech/language problems
- Stage 3
  - Dependent on others for ADLs
  - Minimal to no communication
  - Immobile

Neuropathology of AD in DS

- Chromosome 21 implicated in development of DAT
- Neuropathological changes:

  1. Deposition of extracellular beta-amyloid in neuritic plaques, due to cleavage of APP; increased four to five-fold due to chromosome triplication

  "Deposition of beta-amyloid in the brain is the hallmark of Alzheimer's Disease pathology"

  Sun, et al., (2006)

  2. Intracellular neurofibrillary tangles

Neuropathology, cont.

- Other factors:
  - BACE1 - contributes to pathogenesis
  - BACE 2 - noncontributory
  - SOD1
  - Estrogen - neuroprotective
  - Less brain weight/cognitive reserve
  - Statin use - neuroprotective
  - Environmental factors
    - Schooling, employment, living conditions

Results of neuropathology:

- Deposits of beta-amyloid present in children with DS as young as 8 years old
- By age 40, virtually all individuals with DS have the brain lesions to meet criteria for a diagnosis of AD
- Neurological changes in DS occur 10 - 30 years ahead of individuals without DS
  - median age of onset is 50
  - average age at onset 50-55
- At age 65, 30 -75% of individuals with DS have AD; compared with 13% of non-DS population

Diagnosing AD in DS

- Requires evidence of clinically significant decline in cognitive ability from previously attained level of performance
- Requires that all other possible sources of decline have been eliminated from the differential diagnosis

What Mimics AD?

- Hypothyroidism
- Depression, anxiety, phobia
- Normal aging
- Obstructive sleep apnea
- Sensory loss (vision and hearing)
- Change in environment
- Delirium

What Mimics AD, cont:

- Chronic hepatitis
- Medication effect (anticholinergics)
- Abuse
- Other types of dementia
- Neoplasm
- Folic acid abnormalities (for those on anticonvulsant medications)

Testing for Alzheimer's Disease

- Should be performed by a clinical psychologist with experience in testing people with learning disabilities
- Examples of tests:
  - Test for Severe Impairment (Albert & Cohen, 1992)
  - Spatial Recognition Span (Moss et al., 1986)
  - Dementia Scale for Down Syndrome (Gedye, 1995)
  - Dementia Questionnaire for Persons with Mental Retardation (Evenhuis, 1996)

Tests for AD, cont.

- EEG - slowing of background activity is consistent with late onset seizure activity and cognitive decline (requires serial testing)

Establish a baseline of cognitive function in early adulthood; be alert for sensory impairment while testing

Functional Assessment

- Caregivers as first line of defense:
  - Alzheimer's functional assessment tool (see appendix)
  - Social Skills Inventory
  - Early Signs of Dementia Checklist
  - Dementia Screening Questionnaire for Individuals with Intellectual Disabilities

Clinical Investigation: Differential Diagnosis

- CBC
- Comprehensive Panel 12 (liver, renal, glucose, electrolytes)
- ESR & CRP (inflammatory markers)
- Thyroid function
- Folate & B12
- CT/MRI brain
- CXR
- EEG

Clinical Investigation: Differential Diagnosis, cont

- Plasma levels of drugs such as digitalis and anticonvulsants
- Urinalysis
- Vision and hearing screening

How Does AD Manifest in DS?

- Evidence of frontal lobe dysfunction
  - Changes in personality & behavior
    - Social withdrawal and apathy
    - Unusual, excessive stubbornness/uncooperativeness
    - Increased maladaptive behaviors
    - Depressed mood
    - Restlessness
    - Disturbed sleep
    - Diminished communication

Manifestations of AD in DS, cont.

- Change in functional status
- Confusion
- Late signs
  - Seizure activity (up to 80% of AD/DS individuals)
  - Immobility
  - Lung troubles
  - Feeding disorders
  - Dyspraxia

Relevance of Findings in AD/DS

- Personality and behavioral changes occur before memory decline is apparent
- When these changes are apparent, or when caregiver reports decline in functional ability, proceed with differential diagnosis list to provide early diagnosis and maximize treatment potential

Treatment Goals of AD in DS

- Psychosocial intervention to maximize cognitive function. Encourage participation in as many activities as individual will allow
- Provide stability in the environment.
- Provide safety
- Aging in place, in-place progression of care

Treatment Goals, cont

- Minimize sensory deficit
- Continued medical assessment
- Medications:
  - Aricept, Namenda
  - Antipsychotics
  - Antidepressants
  - Benzodiazepines,
  - Carbamazepines

Treatment goals:

- ACEPS Approach
  - Early screening & diagnosis
  - Clinical support
  - Environmental modification
  - Program adaptation
  - Specialized care

Ethics of Care

- Dealing with families
- Mobility
- Feeding/hydration (tube feedings)
- CPR
- Finances
- Caregiver strain
- Aggressive care vs palliation

End of Life Issues

- Capacity to feel and exhibit emotion persists, and is one of the last parts of the brain affected by AD
- Provide comfort, reduce anxiety and promote security

Key Points

- Alzheimer's disease in Down syndrome:
  - Presents early
  - Often manifests in changes of personality and behavior
  - Can progress rapidly, with compressed course and precipitous decline

Resources

- It's Your Move and other information from Down Syndrome Scotland
- Functional Assessment Tool
- Dementia Scale for Down syndrome
- NDDH Vital Records data, used with permission from NDDH

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ALZHEIMER'S DISEASE IN DOWN SYNDROME QUIZ

1. What is the current life span of an individual with Down syndrome?

   A. 40 years
   B. 50 years
   C. 60 years
   D. 70 years

2. What is the median age of onset of Alzheimer's disease in Down syndrome?

   A. 40 years
   B. 50 years
   C. 60 years
   D. 70 years

3. At what point do most individuals with Down syndrome possess the brain lesions necessary to meet the criteria necessary for a diagnosis of Alzheimer's disease?

   A. 40 years
   B. 50 years
   C. 60 years
   D. 70 years

4. In the past ten years, what is the approximate average number of individuals born in North Dakota per year with Down syndrome?

   A. 4
   B. 6
   C. 8
   D. 10

True/False Questions:

5. Neurological changes in Down syndrome occur at the same time as they do for the general population.
   T **F**

6. Neuropathological changes in AD are related to beta-amyloid deposition.
   T F

7. Changes in memory provide the first evidence of AD in DS.
   T **F**

8. Loss of cognitive ability is diagnostic for Alzheimer's disease.
   T **F**

9. Persons with DS/AD are less apt to feel and exhibit emotion as Alzheimer's disease progresses.
   T **F**

10. Symptoms of Alzheimer's disease progress at the same rate in individuals with Down Syndrome as they do for the general population.
    T **F**

1978-2006 ND RESIDENT BIRTHS WITH DOWN SYNDROME REPORTED ON THE NORTH DAKOTA CERTIFICATE OF LIVE BIRTH

YEAR	NUMBER OF RESIDENT BIRTHS
1978	8
1979	6
1980	7
1981	7
1982	7
1983	12
1984	8
1985	13
1986	11
1987	8
1988	11
1989	17
1990	6
1991	8
1992	11
1993	5
1994	11
1995	9
1996	8
1997	5
1998	9
1999	3
2000	7
2001	7
2002	7
2003	3
2005	7
2006	6

SOURCE: VITAL RECORDS, NORTH DAKOTA DEPARTMENT OF HEALTH

Alzheimer's Functional Assessment Tool

This is a summary of scoring for the Alzheimer's Functional Assessment Tool. This tool can be used to document the progression of the symptoms, and it can also be helpful to evaluate the usefulness of any drug treatment or behavioral interventions. It is not intended to make the diagnosis of Alzheimer's Disease.

Toileting

1. Can use bathroom in familiar and unfamiliar environments independently
2. Goes to the toilet independently or asks for assistance; may need reminders to use toilet paper and wash hands
3. Has occasional toileting accidents; needs verbal reminders
4. Needs assistance going to the bathroom on a schedule (does not go to the bathroom independently); remains continent 90% of the time
5. Needs assistance going to the bathroom on a schedule (does not go to the bathroom independently); remains continent 50% of the time or less
6. No bowel or bladder control; may require frequent changing or special clothing (for example, pads, diapers)

Dining

1. Can prepare simple food (for example, sandwich, toast), can set table and clean up after meal, uses knife and fork to cut food, may or may not use adaptive equipment to eat independently
2. Can use fork and spoon to eat independently but needs food to be cut
3. Eats independently with the help of adaptive equipment
4. Can use fork and spoon to eat independently but may need occasional prompts to start or continue eating, may finger feed, needs food to be cut
5. Needs physical assistance to complete the meal
6. Develops swallowing problems, needs change in consistency of food or thick drinks
7. Completely dependent on assistance, may need specialized feeding program

Walking/motor

1. Independent walking (ambulation), able to walk steadily, able to start - stop - and change direction without falling, able to walk fast or run, able to ascend and descend stairs, capable of leaving premises without assistance
2. Independent ambulation for short distances, walks up and down the stairs one step at a time by holding rails, able to leave premises without assistance
3. Independent but cannot go up or down stairs, unable to leave premises without assistance

4. Can walk without support but requires supervision, may be unsteady, requires supportive measures at times

5. Needs assistance (for example, another person to hold, walker) to walk, "cruises" around using structures such as furniture and walls as support, unable to leave premises independently

6. Needs wheelchair but can move independently

7. Needs an adapted wheelchair and cannot move independently, needs to be pushed

Bathing

1. Can independently carry out an appropriate bathing routine (disrobing, washing, drying, and dressing)

2. Can carry out an appropriate bathing routine with occasional reminders to do a step or wash more thoroughly

3. Needs verbal prompts to initiate and/or complete some steps in the bathing process (because of low-level confusion and/or fear), continuous staff supervision at shower time not necessary, may use toiletries inappropriately

4. Requires continuous staff supervision at shower time to ensure complete bathing and safety (for example, problems due to confusion and/or fear), hand-over-hand assistance may be necessary at times, alternatives to showering or a specialized program may be recommended because of fear of showering, safe use of hot and cold water needs monitoring

5. Primarily passive during bathing, requires some form of assistance for all steps, may be able to stand and move a body part when given a verbal or touch cue, fear of water may be present

6. Physically and cognitively unable to participate actively in bathing process, may respond to stimulation during bathing with vocalizations or changes in facial expressions

Dressing (skills and appropriate dress)

1. Dresses independently or with physical assistance due to handicap, can choose appropriate clothing (for weather or activity of the day) and cares for own clothing (for example, places dirty clothes in hamper, hangs clothing, stores properly)

2. Occasionally needs reminders to dress appropriately ("It's cold out today") and to care for clothes ("Remember where your dirty socks go?")

3. Dresses with minimal assistance or verbal prompts

4. Dresses inappropriately for weather (layers clothing and/or puts clothing on inappropriately), may undress at an inappropriate time and/or place, may benefit from adaptive clothing to retain dressing skills; makes no attempt to care for own clothing

5. Needs assistance in dressing (50% or more of task) and may be resistive; may assist when compliant (for example, puts arm through sleeve)

6. Lies passively during [illegible] does not respond to dressing or undressing

Personal/oral hygiene (hair brushing, teeth brushing, sanitary pad, shaving)

1. Able to perform all personal hygiene tasks
2. Able to perform all personal hygiene tasks within regular routines, may show difficulty in performing tasks if routine is changed (for example, hospitalized, moved)
3. Able to perform all personal hygiene tasks but requires occasional reminders from staff to complete the task
4. Able to perform personal hygiene tasks but requires frequent reminders from staff to complete the task, may need staff guidance (verbal and point cues) in some parts of some tasks (for example, may forget steps), may still be proficient in one area and lose ability in another area
5. Requires staff supervision (verbal and point cues) to complete some personal hygiene tasks and staff assistance (light, moderate physical cues) to complete others
6. May still be able to perform some steps of some personal hygiene tasks with staff assistance but depends on staff to meet other personal hygiene needs
7. Depends on staff to meet all personal hygiene needs

Environmental awareness

1. Aware (cognizant) and responsive, in a relevant way, to familiar and unfamiliar people and other environmental stimuli
2. Generally responsive to familiar and unfamiliar people and situations but seems self-absorbed and/or confused most of the time
3. Cognizant and responsive in a relevant way to familiar people and situations but shows a delayed or inappropriate response to unfamiliar people and situations
4. Cognizant and responsive to stimuli, but response is often inappropriate, even in familiar situations
5. Mostly awake but seems self-involved, showing little or inconsistent response to the environment
6. Sometimes awake but shows little interest in surroundings, sleeps at other times
7. Sleeps most of the day, needs to be aroused repeatedly to maintain interaction

http://www.emedicinehealth.com/alzheimer_disease_in_individuals_with_down_syndro/page6_em.htm

DEMENTIA SCALE FOR DOWN SYNDROME

NAME: (Surname) _____ , _____ ; SEX: M F # _____

BIRTHDATE: (Mo) _____ (Da) _____ (Yr) _____ ; ____ Trisomy 21; ____ Mosaic; ____ Translocation; _____ Not DS

Congenital Heart Defects ___ Yes, ___ No; Cataract(s) ___Yes, ___ No; Hearing Loss _____No, or R Ear _____ L Ear____

Hypothyroidism _____Yes (year treated _____) ____ No; Seizures ____Yes, ____ No; OTHER: _____

_____

MENTAL LEVEL: (Highest level as an adult):_____ Borderline Range or higher; ___ Mild; ____ Moderate; ____ Severe; ____ Profound

Highest Mental Age as an Adult: _____ years (e.g., 7-8, 5, 3 1/2, below 2), Social Age: _____

RESIDENCE: ____ Group Home; ____ Family Care Home; ___ Natural Home; ___ Large Facility (Name:_____)

Address: _____

_____

OTHER: Surgery (e.g., hip, cataract) or major traumas in recent years such as head injury, loss of close friend/relative, etc.

Date & Type: _____ Date & Type: _____

Date & Type: _____ Date & Type: _____

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