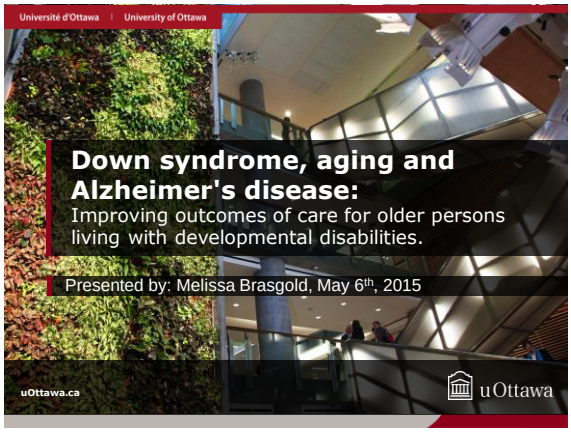

Today's Outline

- Introduction – Who am I?
- Overview of Alzheimer's disease (AD) and its links/prevalence with Down syndrome (DS).
- Symptomology and Diagnosis of AD in DS.
- Treatment and Caretaking practices of AD in DS.
- Discussion, Questions.
- Wrap-up.

Learning Outcomes

By the end of this session you will be able to:

1. Describe the symptoms of AD in a DS individual.
2. Explain key aspects of diagnosis for AD in DS.
3. Identify treatments for AD in DS.
4. Detail and analyze some successful caregiving practices for individuals with AD and DS.

Who am I?

A Gerontology professor and an Educational developer.

PART I

SYMPTOMOLOGY & DIAGNOSIS

Living Longer

- Individuals with DS live much longer and exhibit conditions & characteristics typically associated with older adulthood, as early as in their 30's.

Including:

- Sensory Loss, Hypothyroidism, Sleep-apnea, Osteoarthritis, Osteoporosis, Instability and cervical spine issues, Celiac disease, and Alzheimer's disease.

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Want to know more about the various conditions?

Check out p.7-12 in the following resource:

Link to Information on Various conditions exhibited in aging DS individuals

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Why talk about DS, and Alzheimer's?: Trisomy 21 - Accelerated Aging

- Over 500,000 people with DS live with Alzheimer's in Canada (Alzheimer's Society Canada, 2009).
- Trisomy 21 predisposes DS to AD 3x more.
- More Amyloid protein produced and at a faster rate = deposits seen as plaque in AD brain.
- Prevalence of AD in DS individuals higher and occurs earlier than in general population.

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Why talk about DS, and Alzheimer's?: Trisomy 21 - Accelerated Aging.

Research shows that:

- By the age of 49, as much as 25% of DS have AD.
- By the age of 59, as much as 50% of DS have AD.
- Over the age of 59, as much as 75% have AD.

(Alvarez and Hoffman, 2014 ; Zigman, Schupf, Sersen, & Silverman,1996).

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Alzheimer's Disease

- Most common form of dementia.
- Destruction of brain cells.
- Plaques & Tangles seen in the brain in autopsy.
- Impacts cognitive abilities (memory, learning, judgement, communication).

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Alzheimer's in the General Population

- First signs: depression.
- Then progresses to attack frontal lobes, impacting cognitive abilities.
- Impacts behavior (indifference, unresponsive), personality, emotional function, and judgement and reasoning capabilities.

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Alzheimer's in individuals with DS

- First symptoms: frontal effects of indifference or unresponsiveness (Ball et. al, 2006).
- Memory and other cognitive changes are hard to notice. Need to be dramatic.
- Prevalence: 50-55% of those between the ages of 50-59 show noticeable changes associated with dementia.

(Alvarez and Hoffman, 2014, Clinical Presentation Section).

AD in DS: Early Stage Symptoms

- First symptoms around the age of 50 (range 36-62).
- Diagnosis approximately 2 years later 52 (range 37-62).
- Duration – approximately 10 years after first symptoms (range 47-70).

AD in DS: Early Stage Symptoms

- Confusion.
- Increased time and place disorientation - gets lost.
- Wandering.
- Changes in communication, language and social skills.
- Exaggerated personality traits .
- Changes in sleep or eating habits.
- Inability to make decisions.

AD in DS: Early Stage Symptoms

- Difficulty remembering names or recognizing faces of familiar people or objects.
- Inability to carry out their job duties (Urv, Zigman & Silverman, 2008).
- Visual impairments in perception.
- Impacts on learning ability.

AD in DS: Middle Stage Symptoms

- Advanced deterioration in cognitive and motor skills impacting ADLs.
- Difficulty swallowing & frequent choking.
- Judgement & problem solving skills worsen.
- Further deterioration in memory & communication.
- Problem behavior (e.g. anxiety, suspiciousness, psychosis).

AD in DS: Advanced Stage Symptoms

- Almost comatose and confined to bed.
- Totally dependent of others for care.
- All forms of memory are impacted, and Epileptic seizures are common at this stage.

First Hand Account: Case Study

A Dr.'s personal account of an individual with DS developing Ad. Taken from "Alzheimer Disease in Down Syndrome" by Alvarez & Hoffman, 2013.

Link to description of a DS individual's progression towards AD

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The Top 10 Early Warning Signs

1. Behavioral symptoms (e.g. withdrawal, indifference; Ball et al., 2006).
2. Emotional symptoms (depressive like symptoms).
3. Issues with learning new things.
4. Problems with communication.
5. Issues with decision making and judgement.

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The Top 10 Early Warning Signs

6. Decreases in memory function.
7. Inability to perform even simple tasks.
8. Unable to care for self.
9. Issues with recognizing familiar places or people.
10. Issues with bodily functions.

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Diagnosis: AD in individuals with DS

Diagnosis challenging for a variety of reasons:

- Many conditions common in DS mimic AD (e.g. hypothyroidism and depression). Link to Information on Differential Diagnosis
- Standard diagnostic tests not catered to DS (i.e. limited communication & verbal skills).

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Diagnosing: AD in individuals with DS

- A baseline report completed before the age of 35.
- A detailed medical history.
- A thorough physical and neurological examination.
- Blood and urine tests.
- Neuropsychological testing.
- Neuroimaging tests.
- A mental status test.
- A psychiatric assessment to rule out other disorders.

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Diagnosing: AD in individuals with DS

- Dementia scale for Down syndrome (DSDS).
- Dementia Questionnaire for Mentally Retarded Persons.
- Dementia screening questionnaire for individuals with intellectual disabilities (DSQIID; Deb, Hare, Prior, and Bhaumik, 2007).Link to an overview of the tool

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Tools for Diagnosing AD in the General population

- Check out the following document by the Chronic Care Networks for AD initiative which outlines some tools for assessing AD, but which are not geared towards DS individuals.

Information on tools for early Identification, assessment, and treatment for individuals with AD.

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The Alzheimer Functional Assessment Wrentham Developmental Center, Mass

Via interviews with those close to the individual, the following info is obtained:

- Name
- Activities of daily living
- Description of skills
- Toileting, bathing, personal/oral hygiene, dressing
- Dining
- Walking/motor
- Environmental awareness

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The Alzheimer Functional Assessment Wrentham Developmental Center, Mass

- Walking/motor
- Environmental awareness
- For details on what is looked at in each of the listed categories, please visit **Overview of Alzheimer Functional Assessment Tool**

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Medical Tests: Blood work

- Liver and Renal function tests
- Electrolytes
- Blood glucose
- CBC
- Folic acid
- Vitamin B-12
- Syphilis and HIV
- Thyroid-stimulating hormone (TSH) and thyroxine (T-4) levels
- Amyloid Beta (Abeta) 42

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Neuroimaging: CT, MRI, and PET

- What does the brain of a DS individual with AD look like?
- Can we gain anything from a CT/MRI?

Let's Take a look!
Link to article on CT and MRI in DS with AD
Link to Image of AD vs. Normal Brain
Link to Images or Normal vs. AD, vs. DS brain

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Neuroimaging: CT, MRI, and PET

Observe a reduction in:

- Brain volume.
- White and gray matter.
- Hippocampal volume.
- Frontal and occipital lobe volume.
- Planumtemporale & superior temporal gyrus.

And…

- Atrophy of the corpus callosum.
- Enlargement of the ventricular system.

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In Vivo MRI Support Beacher and colleagues (2009/2010)

Methods:

- 19 DS/AD adults.
- 39 DS only adults.

DS/AD Results:

- Smaller volumes of hippocampus, right amygdala, caudate, and putamen.
- Larger volume of left peripheral CSF.

PART II

TREATMENT & CARETAKING

Treatment of AD in DS

- No cure for Alzheimer's, **but...**
  - Progression may be slowed.
  - Many symptoms (e.g. anxiety, restlessness, depression) can be managed.
- Numerous medications that are too complex to discuss here. To see some with details, consult the following: Treatment of AD in DS

Treatment of AD in DS: Medications

Acetylcholinesterase (AChE) inhibitors

- Tacrine (Cognex)
- Donepezil (Aricept)*
- Rivastigmine (Exelon)
- Galantamine (Reminyl)

N-methyl-D-aspartate (NMDA) blocker

- Memantine (Namenda, Axura)

Antioxidants

Psychotropic medications

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Acetylcholinesterase (AChE) inhibitors

- Only Donepezil and rivastigmine have been investigated for AD in DS.
- Studies (e.g. Boada-Rovira et al., 2005) have shown modest, but non long-term benefits.
- Studies investigating AD without DS have shown modest efficacy for the most part.
- Some industry sponsored studies have shown a 1 yr delay in nursing home placement when AChE used (e.g. Prasher, 2004; Monahan et al., 2009).

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Antioxidants

- No consistent support for the use of antioxidants (e.g. lipoic acid, Vitamin E) in reducing neurogeneration in AD.
- Vitamin B-6, B-12, & folic acid, which metabolize homocysteine do not show any benefit.
- Ginko Biloba has shown some promise in improving social and academic skills in individuals with DS (Don Francesco & Dell'uomo (2004).

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Psychotropic medications

- Benefits associated with using typical & atypical neuroleptics to treat psychosis, agitation, hallucination, and aggression in AD patients.
- But….many adverse side effects noted.
- **Conclusion?** Avoid their usage.

Other drug therapies

- Wide variety of medications have been investigated to treat/prevent/manage AD.

Some examples:

- Anti-inflammatory drugs to reduce Abeta 42.
- Estrogen - protective effect on dementia.
- Antiseizure medications.

Treatment & Caregiving: Support is Key

Important for caretakers to understand:

1. The realities of the disease.
2. That a network of resources/support is needed.
3. The basics of caregiving.

Caregiving: 13 Things to Consider

1. Memory and learning will negatively be impacted.
2. Cognitive abilities & behavior will be negatively impacted.
3. Focus on non-verbal behavior to improve communication & help ensure a supportive, safe and positive environment.
4. Avoid using negative words or tone of voice.
5. Refrain from correcting the individual and be flexible.

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Caregiving: 13 Things to Consider

6. Avoid negotiating or logic.
7. Keep things simple and avoid using long sentences, big words, or lists of things to do.
8. Be specific and avoid too many choices.
9. Repeat things to ensure that the individual got the information.

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Caregiving: 13 Things to Consider

10. Avoid questions; instead, make statements.
11. Learn how to redirect attention to something more positive.
12. Avoid mentioning details that are unnecessary or that will cause uneasiness/anger.
13. Look for behavioural "Triggers".

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Successful Communication

Steps to Successful Communication

GETTING STARTED	FACIAL EXPRESSION	TONE OF VOICE	BODY LANGUAGE
• Approach from the front • Smile • Identify yourself • Use the person's name • If possible, be at eye level	• Establish & maintain eye contact • Be friendly & relaxed • Always remember humor: smiles & laughter go a long way • Be patient and supportive	• Speak slowly & clearly • Use a gentle & relaxed tone of voice • Convey an easy-going manner	• Avoid sudden movement • Be open & relaxed with your stance • Remain calm & confident to provide reassurance • Use gestures such as pointing • Give visual cues

National Down syndrome Society, 2013, p.27.

Be Proactive in Aging: 3 Key Things

1. Ensure the individual remains socially active and connected.
2. Try to maintain Aging in Place by ensuring the living environment meets the medical, physical, social, emotional and safety needs of the individual.
3. Plan for the individual to reduce work and enter retirement earlier.

Person-Centered Coordinated Care

FIGURE 1
Example of person-centered care. The team members surround the center person, and the steps of care coordination are an ongoing and evolving process between all members.

National Down syndrome Society, 2013, p.34.

Team of Coordinate Care

Topics to Consider in Care Coordination

ABILITIES & CAPABILITIES	ENVIRONMENT & ACTIVITIES	CONDITIONS	RESOURCES
• Physical • Psychological / Behavioral • Cognitive • Functional Abilities (activities of daily living) • Sensory (vision, hearing, touch, smell) • Communication	• Living Arrangement • Employment / Retirement • Social Engagement / Activities • Day Program Activity • Daily Routines • Spiritual Support • Likes / Dislikes • Safety Risks	• Nutrition • Oral / Dental • Elimination Pattern • Sleep Pattern • Medical Conditions • Allergies & Intolerances • Medications • Prevention Care • Treatment & Services • Palliative & End of Life Care Needs	• Community Services & Support • Funding • Staffing Requirements • Transportation Needs • Legal

National Down syndrome Society, 2013, p.35.

Some Links to Resources

- Alzheimer Disease in Down Syndrome by Dr. Norberto Alvarez, and Dr. Michael Hoffmann.
- An interesting slideshow that reviews AD and provides diagnostic and caretaking tips.
- A review of AD and DS from Alzheimer's Society

Recommended Links to Resources

- Alzheimer's Australia Overview of AD and DS
- Alzheimer's Australia tip sheets on communication, safety issues, activities, dental care, hygiene, sleeping, eating, etc..
- A review of some cognitive assessment measures

Wrapping things up

- Record a DS's functionality before the age of 35 in order to have a baseline.
- Obtain information from those close to the individual to get the most accurate picture.
- Understand that the individual's cognitive abilities will change and avoid trying to rationalize with the individual.

Wrapping things up

- Make sure to use effective communication to avoid misunderstandings and frustration.
- Stay positive and be flexible when necessary.
- Form a collaborative team that is person centered to help care for the individual.

Anything you wish to add?

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THANK YOU! ☺

Questions? Comments? Email me at: mbrasgol@uottawa.ca

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