

Early Recognition & Screening of Alzheimer's Disease in Persons with Down Syndrome

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Objectives - Part 1: Alzheimer's & Down Syndrome

- Different types of dementia
- What is Alzheimer's prevalence in general population & in Down Syndrome
- DM-ID diagnostic criteria of Alzheimer's
- What conditions can be mistaken for Alzheimer's
- Tips & challenges in recognition in Down Syndrome

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Objectives - Part 2: "Effective" Practices

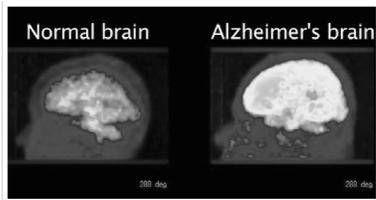
- Early screening for Alzheimer's in Down Syndrome
- Establishing "Baseline" Skills & Abilities
 - Achievement, Cognitive, Activities of Daily Living measure
- Monitoring for changes
 - Compare Current Abilities to Baseline Abilities
 - Screen for Behavioural indicators of dementia
- Determine Need for Referral
 - Patterns, severity of change
 - Medical, physical, and Mental Health Conditions

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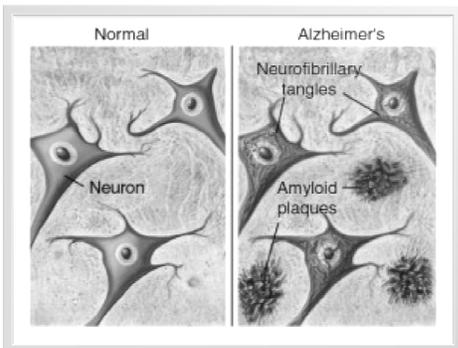
Dementia

- Progressive, usually gradual loss of mental abilities such as thinking, planning, remembering, judgment & reasoning
- Types of dementia
 - Alzheimer's
 - Vascular
 - Dementia due to other medical conditions
 - Substance induced dementia
 - Mixed dementia (d/t multiple causes)

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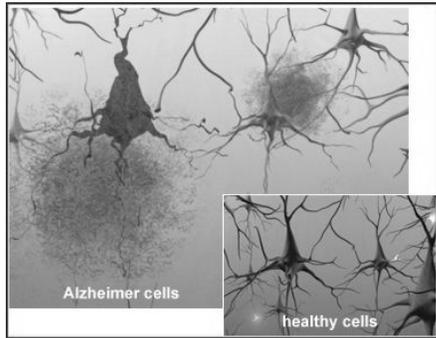


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Alzheimer's – Healthy vs. Diseased Brain



(from Alzheimer's Association)

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Vascular Dementia



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Prevalence of Dementia Subtypes

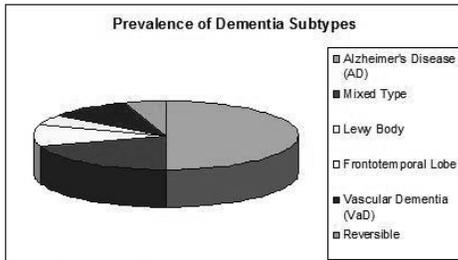
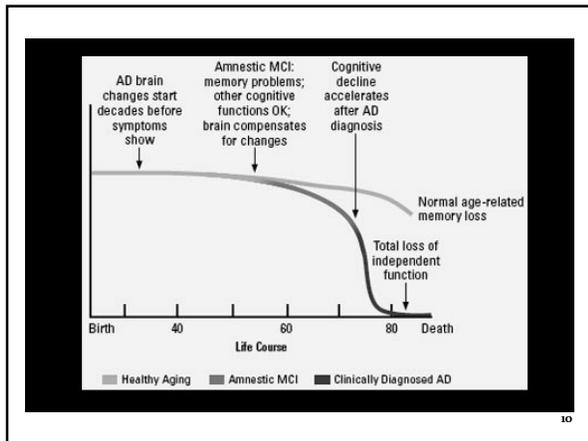
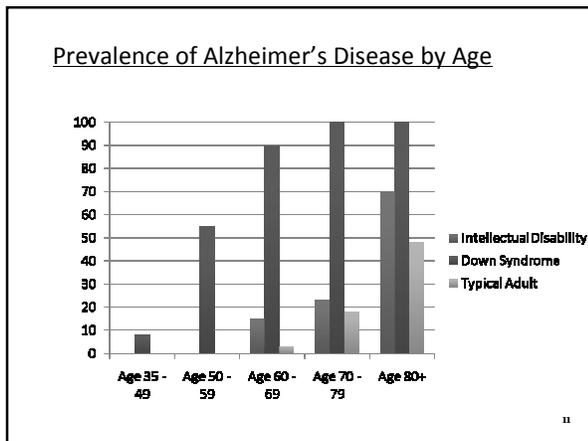
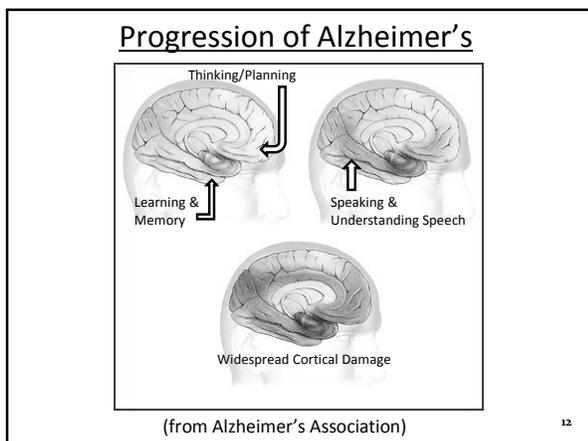


Figure 3: Prevalence of Dementia Subtypes [4]

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Diagnostic Criteria for Alzheimer's (DM-ID)

A. Development of multiple cognitive deficits manifested by both:

1. Memory impairment for learning new information or recalling previously learned information

1. 1 or more of:
 1. Disturbance of language (aphasia)
 2. Impairment in performing motor activities despite intact motor function (apraxia)
 3. Failure to recognize objects despite intact sensory function (agnosia)
 4. Disturbance in executive function (planning, organizing, sequencing, abstracting)

Diagnostic Criteria for Alzheimer's (DM-ID)

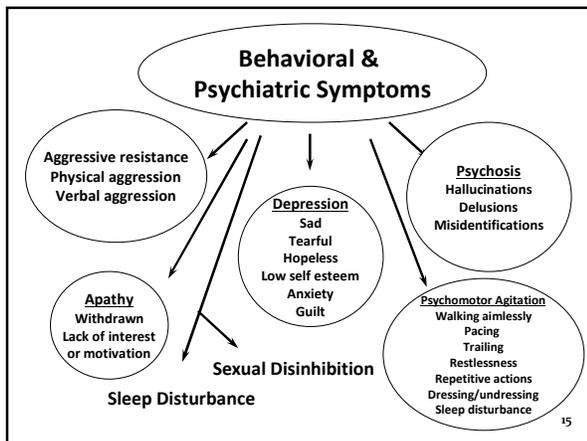
B. These deficits cause significant functional impairment and represent significant decline from previous functional level

C. The course is of gradual onset & continuing cognitive decline

D. Deficits are not caused by any other central nervous system or systemic condition which causes dementia, nor are induced by substances

E. Deficits do not exclusively occur during delirium

F. Deficits not better accounted for by another Axis I disorder – e.g. Major Depression



Differential Diagnosis

(What can be mistaken for Alzheimer's)

- Mild Cognitive Impairment (MCI)
- Other types of dementia
- Medication Side Effects
- Major Depression
- Delirium
- Endocrine disorders – e.g. Hypothyroidism
- Sensory loss – vision, hearing
- Other neurological disorders
- Tumour & head injuries

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Normal Forgetfulness

- Recognize people & places, even if cannot recall their names
- Remember the day & time, might forget the date
- Forget details of a recent experience, but not the experience itself
- Forget items, but will often remember later



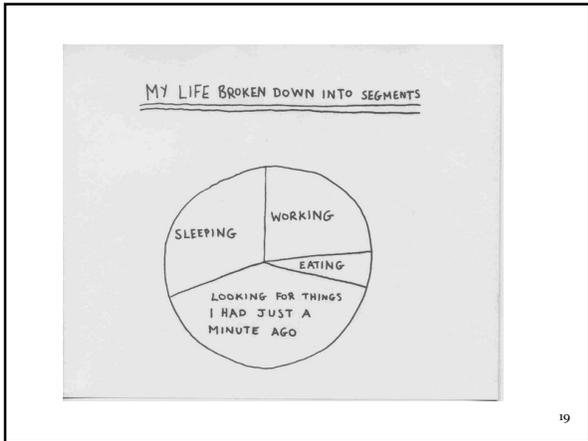
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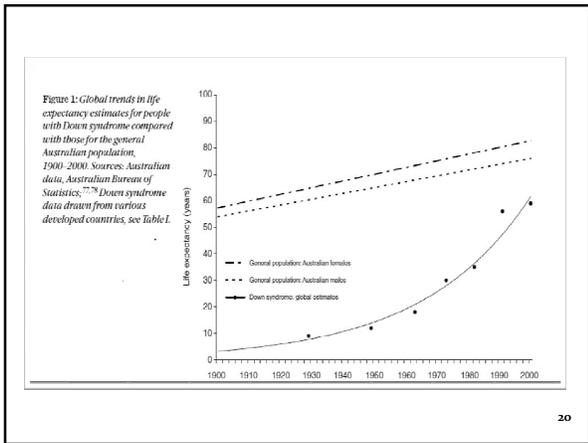
The difference between Alzheimer's and typical age-related changes

(From the Alzheimer's Association)

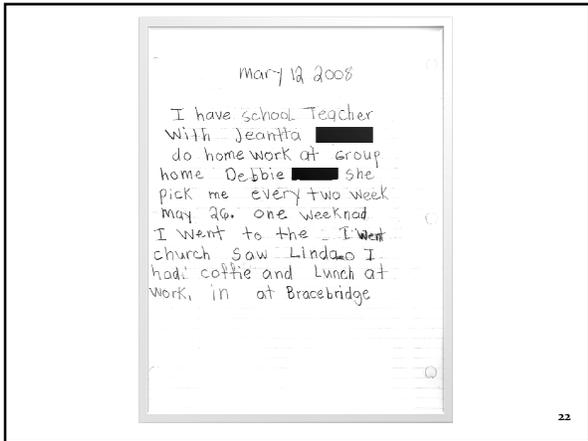
Signs of Alzheimer's	Typical age-related changes
Poor judgment and decision making	Making a bad decision once in a while
Inability to manage a budget	Missing a monthly payment
Losing track of the date or the season	Forgetting which day it is and remembering later
Difficulty having a conversation	Sometimes forgetting which word to use
Misplacing things and being unable to retrace steps to find them	Losing things from time to time

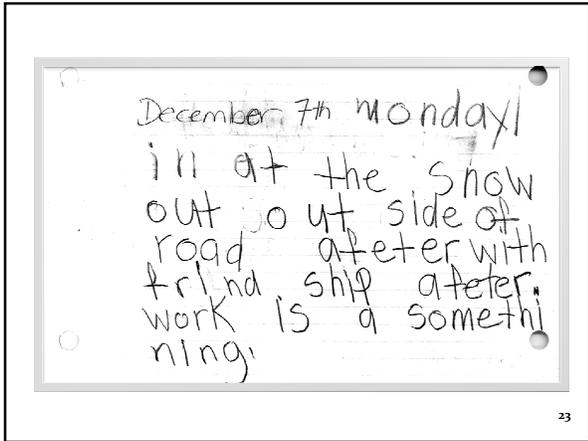
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- ### When to Suspect Alzheimer's
- Loss of memory which impairs daily function
 - Difficulty performing familiar tasks / daily living skills despite intact sensory & motor function
 - Increased difficulty learning new tasks
 - Reduced language skills
 - Decrease in judgment
 - Misplacing items consistently
 - Loss of orientation
 - Loss of initiative / apathy
 - Deterioration in adaptive social skills
 - Onset of seizures for the first time
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Challenges in Recognizing Alzheimer's in Persons with ID

- Conditions which can mimic Alzheimer's must first be ruled out
- Lack of definitive biological tests or markers for AD
- Cognitive screening tests for general population usually invalid in persons with ID
- Lack of baseline functional information for comparison over time
- Insufficient expectations of person to ascertain whether his/her capacity is declining
- Caregiver issues
 - Carers may be unaware of insidious decline
 - Carers notice Sx which impinge on them (aggression etc.) & not necessarily key diagnostic Sx of Alzheimer's
 - Carers change frequently

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Resources

- *Diagnosing Alzheimer's Dementia in Down Syndrome: Problems and Possible Solutions*; Research in Dev Disabilities 30 (2009) 827-38
- Alzheimer's Association: <http://www.alz.org/>

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Early Recognition & Screening of Alzheimer's Disease in People with Down Syndrome

Part Two:

Effective Practices



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Early Screening

- Alzheimer's disease is a diagnosis of exclusion in persons with Down syndrome, just as it is in the general population.
- Clinicians rely largely on informant reports
 - May paradoxically be more difficult for those who see them daily, than those who see them weekly or monthly
 - "Diagnostic Overshadowing"
 - "Baseline Exaggeration"

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'Diagnostic Overshadowing'

(Reiss *et al*, 1982)

- Attribution of changes in behaviour or ability to the intellectual disability or syndrome.
- Can result in late or no referral to specialists.
- Can reduce the significance attached to abnormal behaviour

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'Baseline Exaggeration'

- The exacerbation or worsening of cognitive deficits and maladaptive behaviours that predated the dementia.
- The person's reduced ability in abstract thought and clear communication can result in limited subjective symptoms – e.g. they can't tell you what's happening.
- Changes in presentation might be related to social or emotional difficulties.

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Differences in Presentation

- General Population –
 - Hippocampus atrophies first
 - Episodic memory & orientation problems generally first signs
- Down Syndrome –
 - "Prefrontal Lobe symptoms" may be more common (Azizeh *et al*, 2000);
 - Difference may be due to earlier amyloid deposits in the frontal lobes of DS
 - Frontal lobes underdeveloped in DS
 - "Reserve Capacity" model
 - Alzheimer's dementia more common in mild to moderate ID
 - Frontal lobe dementia appears to be more prevalent in those with more severe ID

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Differences in Presentation

- Personality & behavioural changes preceded declines in memory & orientation
 - Indifference, uncooperativeness, apathy,
 - Depression
 - Socially deficient communication,
 - Impaired adaptive functioning
- Decline in "Executive Functioning" (Das & Mishra, 1995) - verbal fluency, planning, attention span, clock drawing, abstract thinking
- Level of pre-existing cognitive function:
 - Closely associated with the rate of decline
 - Not the risk of developing dementia (Temple *et al*, 2001).

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Changes Over Time

- Schapiro (1988) suggested that cognitive decline in Down Syndrome occurred in two stages, separated by 20 years or less
 - Decline in processing skills
 - Decline in "over-learned" skills in social, occupational, domestic, adaptive areas

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Other Differences

- **Females with DS typically**
 - Better cognitive abilities
 - More developed speech
 - Less challenging behaviour than men
- **Medical & Physical**
 - Medication
 - Sensory (hearing & vision)
 - Endocrine
 - Nutritional
 - Emotional
 - Other neurological or physical conditions

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Rationale for Early Screening and Evaluation

- Diagnosis of AD by exclusion
- Mental health disorders often present differently in persons with Down syndrome and other disabilities because of cognitive and expressive language limitations in this population.
- Traditional testing less helpful for assessing, making diagnosis, and monitoring declining abilities in persons with an intellectual disability
- Several authors have suggested routine baseline screening for people with Down's syndrome in early adulthood
 - Ensures optimum level of achievement attained
 - Easier to identify decline later in life.

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"Effective" Practice Guidelines: Screening for AD in People with Down Syndrome

- Longitudinal follow-up
 - Initial baseline at age 25 years
 - Annual screening for DS >35 to 40 years
- Focus should be in recognizing change & decline in relation to a person's premorbid baseline
 - Step 1 – Medical, Physical & Mental Health Conditions
 - Step 2 – Baseline "Optimum" Skills & Abilities
 - Achievement,
 - Cognitive
 - Activity of Daily Living measure
 - Both general (ADL)
 - Instrumental (IADL)
 - Step 3 – Monitor for Decline in Abilities and Daily Skills
 - Compare Current Abilities to Baseline Abilities and
 - Screen for Behavioural indicators of dementia
 - Step 4 – Determine Need for Referral

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A WORD OF CAUTION

- Just a reminder that this screening package is NOT designed to diagnose, nor is it intended to compare one individual to another, or to a normative group. It is designed to be administered by non-specialist raters, who have been trained in the application and requirements of the scale. Please note that the scale will require specific training in test administration to ensure consistency and reliability. The training and subsequent screening with the individuals would be most "Effective" if 2 or more staff were designated as being responsible for ensuring consistent monitoring and application of the baseline & dementia measures. To ensure reliability and validity over time, it is critical that a trained person be available to administer the measure, as well as being as consistent as possible over time. If the conditions of consistency and training are maintained, a change in total scores over a period of time, in a co-operative person, may be indicative of dementia if there are no other physical or mental factors affecting that person's performance. If you are wanting additional training in the screening package, please contact someone in your agency who can then contact one of the professionals in your region who are familiar with test protocols.

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“Effective” Practice Guidelines: Screening for AD in People with Down Syndrome

STEP 1: Establishing a Medical Baseline

1. Differential Medical Examination: to avoid missing potentially reversible causes of decline.
 - Down Syndrome Health Watch - Adults (B. Sullivan, 2009)
 - Medical history and physical exam
 - Regular Vision and Hearing evaluations
 - Blood work:
 - Thyroid function, vitamin B₁₂, folic acid,
 - Complete blood count (CBC), and chemistry profile.
 - Mental Health Evaluation
 - Depression, Anxiety

“Effective” Practice Guidelines: Screening for AD in People with Down Syndrome

STEP 2: Establishing a Functional Baseline

1. Determining the Person’s Best Level of Functioning: Collect information about the person’s...
 - i. Personal Achievement History:
 - Academic history – levels;
 - Employment / Volunteer history;
 - Living Arrangements
 - Personal Interests & Community Involvement
 - ii. Optimal cognitive level of functioning
 - iii. “Best of Ability” – Adaptive / Independent living skills

STEP 2: Establishing a Functional Baseline

i) Personal Achievement History

- Academic History
 - Grade Achieved
 - Type of Special Education Programming
 - Life Skills Program
 - Vocational Placements
- Day Program Involvement:
 - Recreational, Vocational
- Volunteer and/or Employment History
 - Type of position held
 - Duration of position
 - Reason for leaving

STEP 2: Establishing a Functional Baseline

i) Personal Achievement History

- Living Arrangements:
 - Parent's Home
 - Supported Independent (hours of support)
 - Shared
 - Independent
 - Supported Living – Group Home
- Personal Interests
- Community Involvement

STEP 2: Establishing a Functional Baseline

ii) Optimal Cognitive Development Level

BASIC COGNITIVE ABILITIES SCALE

Orientation

- Time
- Place
- Person

Recall

- Immediate recall
- Delayed recall

STEP 2: Establishing a Functional Baseline

ii) Optimal Cognitive Development Level

BASIC COGNITIVE ABILITIES SCALE

Language

- Written Expression: printing/writing
- Expressive Vocabulary: naming common objects & colours;
- Expressive Information: naming the days of the week;
- Expressive Description: describing a picture
- Receptive Comprehension: following simple requests

STEP 2: Establishing a Functional Baseline

ii) Optimal Cognitive Development Level

BASIC COGNITIVE ABILITIES SCALE

Motor Skills

- Copying and Drawing: copy
- Object Imitation: copy shapes with sticks
- Object Function: demonstrates the action
- Self-Help – demonstrates simple dressing skills
- Simple Gestures: wave, point, and whistle

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STEP 2: Establishing a Functional Baseline

ii) Optimal Cognitive Development Level

BASIC COGNITIVE ABILITIES SCALE

Calculation

- Rote Counting: up to 10
- Counting Objects: count items up to 10
- Money Recognition: label common coins
- Money Counting: adding combinations coins

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BASIC COGNITIVE ABILITIES SCALE (Example)

LANGUAGE			
Expression: Naming Objects			
Show the following objects one by one and ask, "What is this?":			
NOTE: If the person is non-verbal, follow the instructions below:	Paper-Clip	0	1
Non-Verbal Instructions:	Tissue	0	1
- Lay 5 items on the table at a time	Comb	0	1
Ask the person to "Touch the (item)"	Fork	0	1
Mark as correct if the item is touched or held, or given to you	Toothbrush	0	1
After requesting 2 items, vary the items that are displayed	Bar of Soap	0	1
On the table, and ask for the next two items	Scissors	0	1
Continue this procedure until all items have been requested	Knife	0	1
Subtotal Labels			

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STEP 2: Establishing a Functional Baseline

iii) "Best of Ability" Adaptive / I.L. Scale

- Demographic Information
- Staff information – e.g. Length of time knowing person being rated
- Psychiatric Diagnosis
- Physical Concerns
- Medications

STEP 2: Establishing a Functional Baseline

iii) "Best of Ability" Adaptive / I.L. Scale

- *Daily Living Skills*
 - Dressing
 - Bathing
 - Grooming
 - Toileting
 - Physical Mobility
- *Communication*
 - Receptive (Comprehension)
 - Expressive

STEP 2: Establishing a Functional Baseline

iii) "Best of Ability" Adaptive /I.L. Scale

- *Orientation*
 - Person
 - Place
 - Time
- *Memory*
 - Recent
 - Past

STEP 2: Establishing a Functional Baseline

iii) "Best of Ability" Adaptive / I.L. Scale

- *Social*
 - Initiating
 - Responding
 - Maintaining
- *Interests*
 - Day-to-day
 - Events
 - Self

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"Best of Ability" Adaptive Scale (Example)

1.	DAILY LIVING SKILLS - In the past WEEK, the person:
0	Could not live independently, and needed a lot of help with all activities of daily living
1	Could not live independently, but needed only minor help with activities of daily living
2	Could live in a supported independent living situation - but needed help with some activities of daily living
3	Could live independently with only minor help
2.	DRESSING - In the past WEEK, the person:
0	Was either totally dressed by caregiver or remained in bedclothes
1	Person was able to dress but needed frequent assistance.
2	Dressed with only minor supervision. (For example, has clothes laid out or had to be reminded to dress).
3	Initiated and completed dressing without assistance

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STEP 3: Monitor for Decline in Abilities and Daily Skills

- Repeat Baseline Measures
 - Basic Cognitive Abilities Scale & "Best of Abilities" Adaptive Scale
 - At age 35, then 40, then every 2 to 5 years (depending on decline)
- Complete "Early Dementia Screening For Individuals With Intellectual Disabilities" (EDSIID)
 - Age 35, then 40, unless person is scoring 3 or 4 on over 25% of items
 - Then annually

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STEP 3: Monitor for Decline in Abilities and Daily Skills

Early Dementia Screening For Individuals With Intellectual Disabilities” (EDSIID)

- Interview with parents or caregivers who have known person for at least 6 months, preferably before changes were noted:
 - Personality & Mood
 - Cognitive & Communication
 - Motor Skills
 - Memory
 - Orientation
 - Personal Skills

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STEP 3: Monitor for Decline in Abilities and Daily Skills

Evaluate Cognitive and Functional Decline:

in the areas known to be affected by dementia in individuals with Down Syndrome

Criteria A

- Personality & Behaviour
 - Emotional reactions – irritable, crying, flat
 - Apathy – activities, hobbies, other people
 - Social interactions – initiate, maintain, participate

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STEP 3: Monitor for Decline in Abilities and Daily Skills

Criteria B

- Other cognitive skills –
 - General mental functioning
 - Language (expressive & receptive)
 - Visual perception & recognition
 - Voluntary Motor Movements (carry out familiar actions and skills)
 - Executive functioning (planning, decision making, problem solving)

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STEP 3: Monitor for Decline in Abilities and Daily Skills

Criteria C

- *Memory*
 - Short Term, events
 - Word finding
 - People
- *Orientation*
 - Time
 - Place
 - Person

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STEP 3: Monitor for Decline in Abilities and Daily Skills

Criteria D

- Loss of adaptive skills
 - “Everyday” Life Skills
 - Money skills
 - Domestic skills
 - Community safety
 - Self-care
 - Eating, feeding, dressing, grooming, bathing
 - Continence

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(EDSID – Example Item)

EARLY DEMENTIA SCREENING FOR INDIVIDUALS WITH INTELLECTUAL DISABILITIES	Does not apply to this person	Typical – always been the case	Always present, but not worse or Symptom occasionally seen	New Symptom – previously didn't have concern
EVERYDAY ADAPTIVE SKILLS				
1. Decline or loss of ability to independently bath/wash.	0	1	2	3
Comment/Description:				
1. Loss of usual dressing skills. (e.g. A person, who could formerly dress when handed items one-at-a-time, starts putting clothing on front-to-back or otherwise incorrectly.)	0	1	2	3
Comment/Description:				
TOTAL ADAPTIVE SKILL SCORE =				
TOTAL PERSONAL & SOCIAL SKILL SCORE =				
TOTAL COGNITIVE & COMMUNICATION SKILLS =				
TOTAL MEMORY SKILL SCORE =				
TOTAL TIME/ORIENTATION SKILL SCORE =				
TOTAL MOTOR SKILL SCORE =				
TOTAL DEMENTIA SCREENING SCORE =				
Comments on Score:				

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“Effective” Practice Guidelines: Screening for AD in People with Down Syndrome

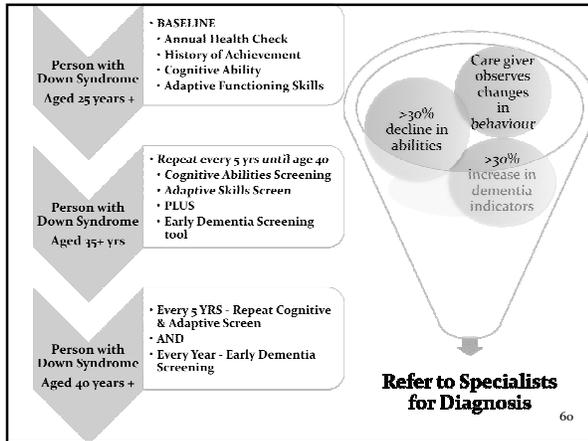
STEP 4 – Determining the need for referral to Specialists for diagnosis

Duration & Pattern of Changes

- Gradual loss
 - Otherwise suspect mental health, medical, or physical causes
- Longer than 6 months
 - Usually more evident in home settings
 - May maintain skills longer in more structured programs
- Decline of >30% on Ability Scales
 - Cognitive ability
 - Adaptive Skills
- Increase of 30% or more on “Dementia Screening Scale”
 - Number of behavioural indicators scored as 3 or 4

Essential Components of a Dementia Responsive Program

- Establishing “Optimum” Ability levels
- Long-term Monitoring
- Early & Regular Screening
- Diagnostic Follow-up
- Person Centered CARE Plan



Essential Components of a Dementia Responsive Program

- Person Centred Care Plans (PCCP)
 - Environmental Modifications
 - Program Adaptations
 - Modified Interaction Techniques- less stimulation, additional visual cues
 - Specialized Activities – less challenge, more familiar, daily activities
 - Intervention for Behavioural / Mood Changes – monitor antecedents
 - Specialized Care Needs
 - Identification of stages & adaptations needed
 - Early stage – similar to those with more limited cognitive ability
 - Middle stage – more individualized
 - Late stage – nursing, long-term care

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- Lisa R. Stanton & Rikus H. Coetzee Down's syndrome and dementia, *Advances in Psychiatric Treatment (2004), vol. 10, 50–58*
- PRUDHOE COGNITIVE FUNCTION TEST (PCFT) developed by: Kay D.W.K., Margallo-Lana, M.L., Moore P.B., Department of Psychiatry, University of Newcastle upon Tyne, Newcastle upon Tyne, UK; and Tyrer S.P., Prudhoe Hospital, Prudhoe, Northumberland, UK; Fletcher R., Berney T.P. & Vithayathil E.

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Questions?

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