

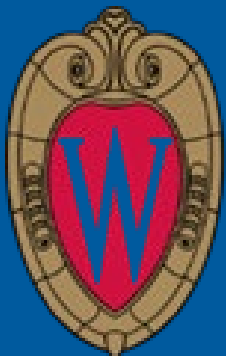
Cognitive and Behavioral Profiles in Down Syndrome Across the Life Course

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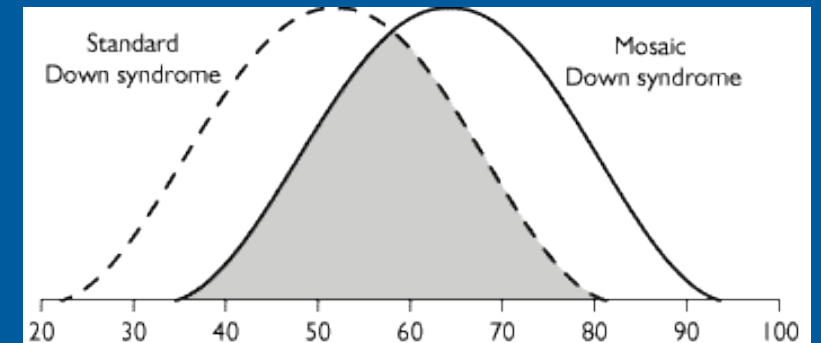


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Cognitive and Behavioral Functioning in Down syndrome (DS)

- Overall, score on IQ measures in mild (IQ: 50-69) to moderate (IQ: 35-49) range (Hamburg et al., 2019)
 - 10-15% severe to profound (IQ < 35) range
 - >5% borderline (IQ: 70-85) or average (IQ: 85+) range
 - full trisomy (mean IQ: 50-55) vs, mosaic (mean IQ: 63-68)
 - Support and education



- At risk for maladaptive behaviors and mental health problems
 - Up to 2/3rds of adults with DS do not qualify for a mental health disorder (Mallador ete al., 2014; Mantry et al., 2008)
- Unique profile of strengths and challenges across the life course

Cognition: Infancy and Toddlerhood

Challenges

- Reduced/delayed vocal reactivity and responsiveness
- Delayed development of inhibitory processes
- Delays in canonical babbling and first words
- Expressive language
- Social cognition

Strengths

- Interest in face-to-face interactions
- Joint attention
- Gestural communication
- Language comprehension
- Pragmatic aspects of language
- Visual learning



Univ. College London website

Maximizing Learning

Understand more than they can say

Allow time to react/respond

Clear, simple phrases and visual cues/modeling

Multiple ways to communicate

Leverage positive interactions

Cognition: Childhood and Adolescence

- **Challenges**

- Auditory memory
- Expressive language (esp. syntax, phonological processing and intelligibility)
- Phonic learning
- Executive Functions

- **Strengths**

- Comprehension
- Social skills
- Visual learning
- Visual memory
- Sight vocabulary



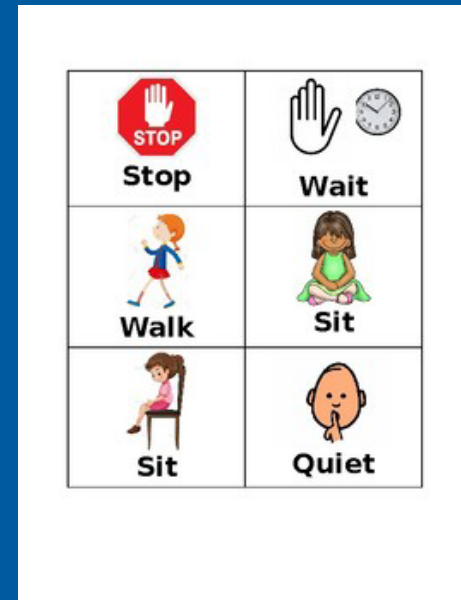
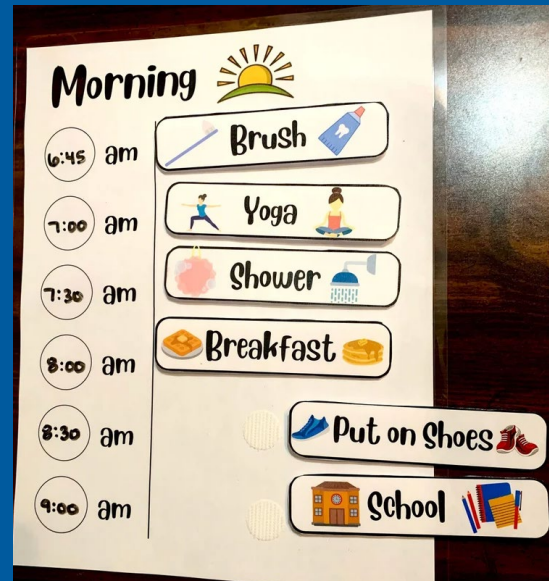
Center for Disability Studies;
<https://www.cfdsny.org/dsahrc>



<https://www.down-syndrome.org/en-gb/resources/teaching/rli>

Strength in Visual Learning and Memory

- Strong visuospatial abilities relative to verbal processing skills (Costanzo et al., 2013; Jarrold et al., 1999; Klein & Mervis, 1999; Fidler et al., 2006)
- Aspects of visuospatial processing especially strong (Fidler, 2005)
 - visual memory
 - visual-motor integration
 - visual imitation



Challenge in Memory & Executive Functions



Maximizing Learning

Direct, clear and short phrases

Visual demonstrations, pictures, charts

Learning by doing

Repetition

Break tasks into small steps, simplify

Reduce distraction

Lots of breaks between periods of learning

Explicit directions and cues to regulate behavior

Cognition: Adulthood

Challenges

- Auditory memory
- Expressive language skills relative to other adaptive behavior skill areas
- Intelligibility problems and sound production errors
- Executive Functions

Strengths

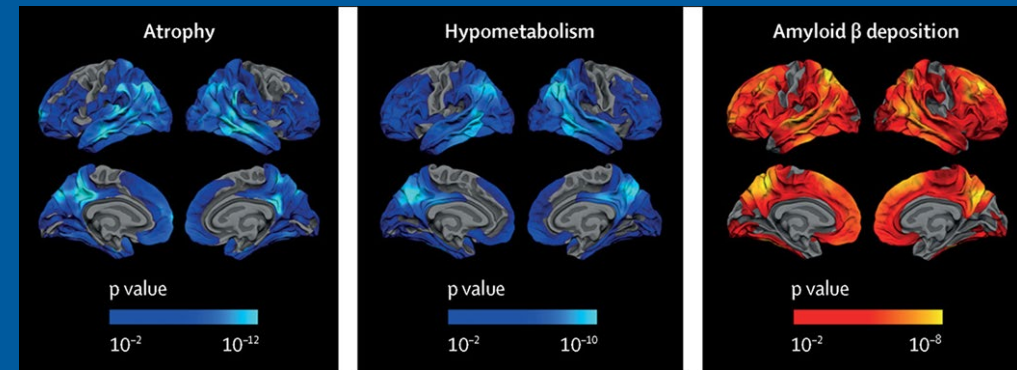
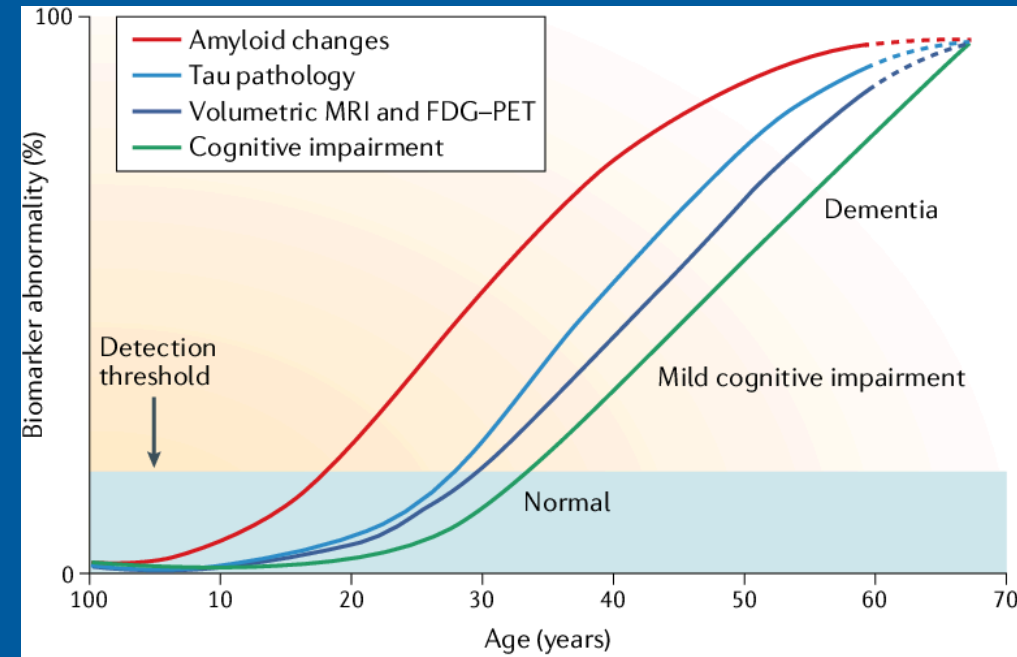
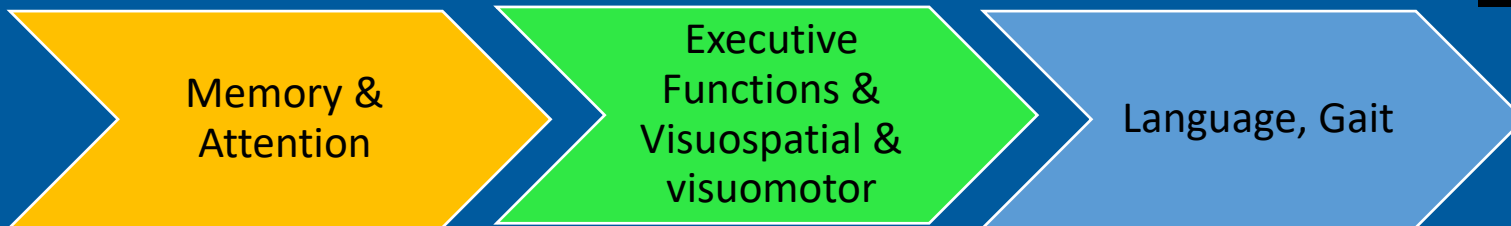
- Comprehension skills
- Activities of daily living and employment skills
- Visuospatial abilities
- Visual memory and learning

Cognition: Aging in DS

- Alzheimer's disease

- Brain pathology - amyloid-beta ($A\beta$)- present by age 40 years (Fortea et al., 2021; Head et al., 2012; Lao et al., 2017)
- Over half of adults with DS exhibit Alzheimer's dementia by age 55 years (Rubenstein et al., 2020)
 - Variability; genes such as APOE (Fortea et al., 2020), health conditions (Lao et al., 2022), and lifestyle (e.g., Mihaila et al., 2020)

- Sequence of cognitive decline



Fortea et al., 2020; Lao et al., 2017; Zammit et al., 2020; Head & Lott, 2019

Hartley et al., 2020; Fleming et al., in press

Alzheimer's Biomarker Consortium - Down Syndrome



This study is for:

- People with Down syndrome
- 25 years of age or older
- Accompanied by study partner (e.g. parent, caregiver, legal guardian)



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What does this study involve?

- Up to 4 visits over five years
- Study Partner completes questionnaires
- Participants completes physical exam, blood draw, and tests of thinking
- Participants complete an MRI & PET scans, and an optional lumbar puncture
- Invited to report on lifestyle



Ways to Preserve Cognitive Functioning with aging in DS

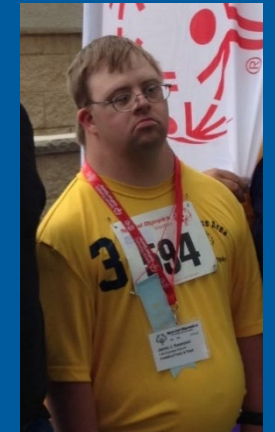
- Sleep

- Sleep disruptions and sleep disordered breathing associated with executive functioning and memory (Cody et al., 2020; Fleming et al., 2021)
- Regular sleep screenings and compliance with treatments and recommendations



- Physical Activity

- More sedentary behavior and too little physical activity associated with poorer executive functioning and memory (Fleming et al., 2021)
- Build activity into a routine and make social



- Cognitive Stimulation

- Identify leisure or employment/day program/other activities that make you think (Mihaila et al., 2019)

“...you know the memories might go away, but what will never go away – your family.”

-Danny Chafetz, Edgewood College Cutting Edge Program

Profile of Maladaptive Behaviors - Childhood/Adolescence

Challenges- Toddlers & Children

- Wondering
- Disruptive and impulsive and hyperactive
- Oppositional and disruptive behavior
- Anxious, inflexible and stuck behaviors

Challenges- Adolescence

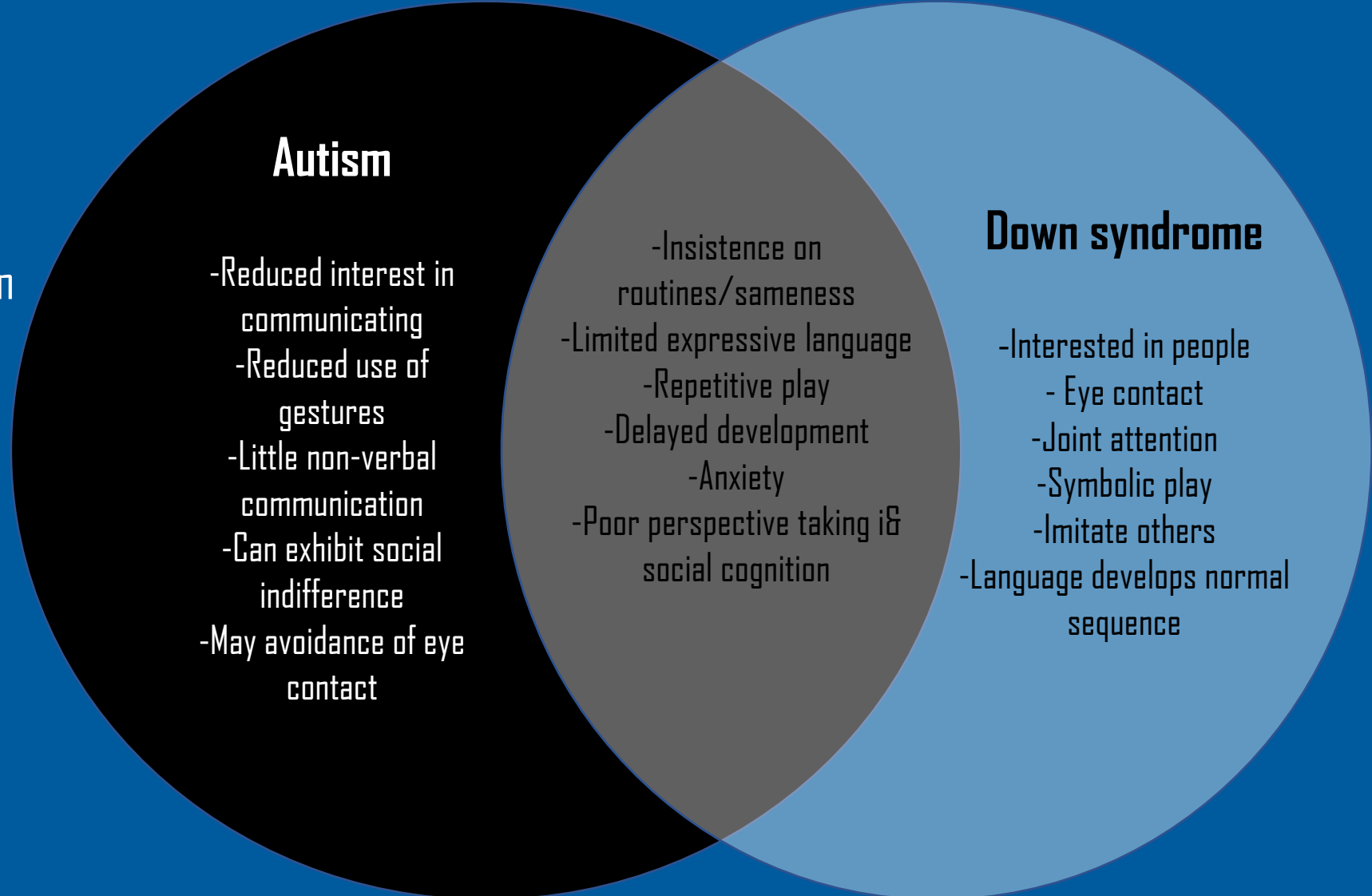
- Depression, social withdraw
- Generalized anxiety
- Obsessive compulsive behaviors

Strengths

- Social motivation
- Routines
- Social skills

Autism

- 5-16% children with DS have diagnosis of autism spectrum disorder (Richards et al., 2018)
- Frequency and severity of autism symptoms negatively associated with IQ and adaptive functioning (cite)



Profiles of Maladaptive Behavior - Adults

Challenges

- Regression
- Anxiety and depression
- Obsessive compulsive behavior
- Self-Talk

Strengths

- Employment and daily living skills
- Routines
- Social skills

Self-Talk

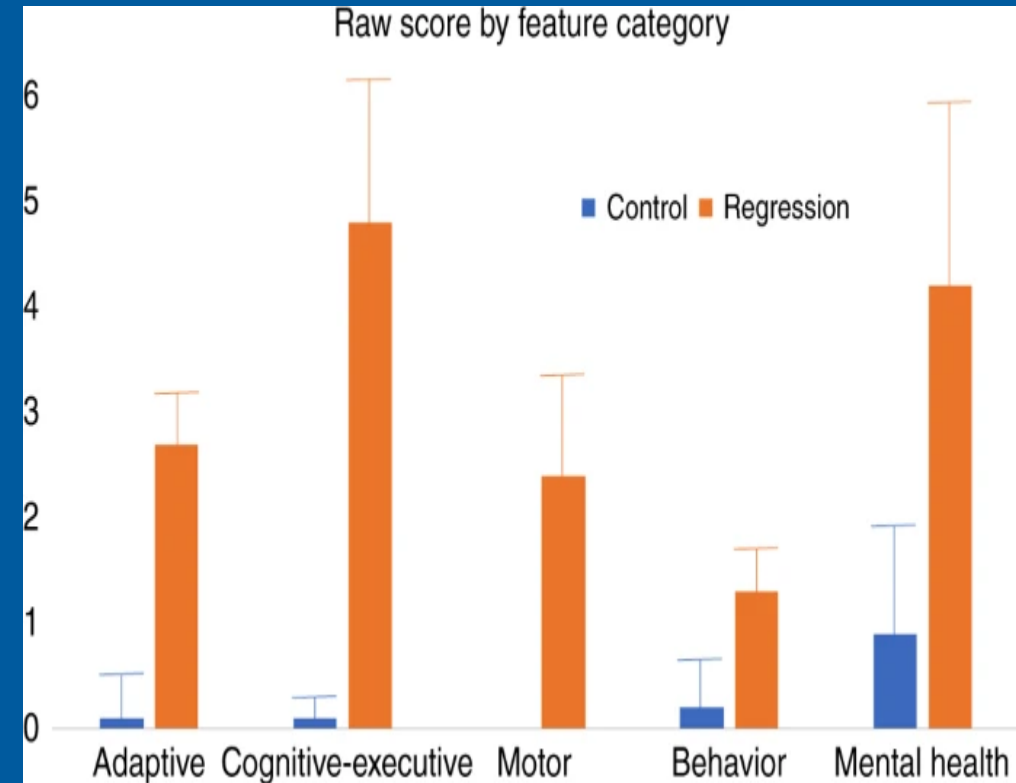
91% of children and adults engage in self-talk
(Patti, Andiloro, & Gavin, 2008)

Employment

- Janitorial Work
- Clerical work
- Speaking/Advocacy
- Food industry
- Bagging/shelving
- hosting
- assembly
- College

Regression

- Sudden or progressive 'regression'
- 1-17%; late adolescence to early adulthood
- Range of symptoms – changes in social skills, adaptive behavior, attention, and internalizing behavior most common (see Rosso et al., 2019; Walpert et al., 2021)
- Treatment often anti-depressants or anti-psychotics
 - Typically partial recovery of abilities (Walpert et al., 2021)
- Not Alzheimer's disease, but could share etiological mechanisms and/or alter timing (i.e., tau and NfL) (Handen et al., 2021)



Santoro et al., 2019

Summary

- Individuals with DS have a unique profile of strengths and challenges in cognition and behavior
- Leverage strengths and reduce impact of challenges to promote learning and enhance quality of life
- Profiles presented on today are based on average patterns; there is a TON of variable among children and adults with DS. Many individuals with DS will have additional or alternative strengths or challenges



Artwork by Dan Campbell



Photo by Clark Kibbe, Wiseman Communications

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World Down Syndrome Day – March 21st 2022

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