MODULARITY

• Williams Syndrome (WS):
  – (more or less) general cognitive retardation except in language
  – Results from the deletion of the long arm of chromosome 7.

• Specific Language Impairment (SLI):
  – Language problems without any obvious cause.
  – Fifty to seventy percent of children with SLI have at least one other family member with the disorder

➢ If the brain is a general purpose problem solver, one’s general intelligence should be predictive of abilities such as language.

➢ For people affected by WS or SLI that’s simply not true.
A genetic marker for WMS is the **deletion of one copy of a small set of genes on chromosome 7**, band 7q11.23, shown in the ideogram. This region is expanded to the right to illustrate genes that are missing one copy in WMS, including the gene for elastin. The regions involving the common breakpoints in WMS are also illustrated (see Korenberg et al., this volume). Thus, WMS is characterized genetically by deletion of one copy of a small set of genes on chromosome 7, including the gene for elastin (illustration based on Korenberg et al., 1998).
Total volume of the brain is reduced in WS but frontal lobe, neocerebellum, temporal limbic areas and superior temporal areas are preserved or even enlarged.
Neocerebellum

- Involved in movement.
- Among the brain's newest parts, appearing in human ancestors about the same time as the enlargement of the frontal cortex, which controls much of rational thought and reasoning.
- The neocerebellum is significantly smaller in people with autism, who are generally antisocial and poor at language, the reverse of people with Williams.

Limbic system

- memory, emotions
Neurocognitive predictions

- Impairment on right hemisphere functions.
- Relative sparing of left hemisphere function.
Williams Syndrome (WS) medical features

*Neurological*

- average IQ 55 (range 40-90)
- poor coordination
- hypersensitivity to sound
- hoarse voice

  - enhanced musical ability
  - relatively spared language
  - overly friendly personality
Williams Syndrome (WS) medical features

Facial Features

- full prominent lips
- stellate iris pattern
- prominent ear lobes
- wide mouth
- small, widely spaced teeth
- medial eyebrow flare
- flat nasal bridge
- short nose/anteverted nares
Williams Syndrome (WS) medical features

Cardiovascular

- supravalvular aortic stenosis
- peripheral pulmonary artery stenosis
- pulmonic valvular stenosis
- ventricular/atrial-septal defects
Williams Syndrome (WS) medical features

Other

- elastin deletion probe (FISH)
- transient infantile hypercalcemia
- developmental delay (infants height and weight < 5th percentile)
Persons with WS are bad at:

• Spatial cognition tasks
Model:

Williams
Age 11;1

Williams
Age 9;1

Control
Age 6;1

From: http://www.ling.udel.edu/colin/courses/ling101_f98/lecture1.html
Persons with WS are bad at:

- Spatial cognition tasks
  - But they know that their models are not correct.
- Contrast between visuo-spatial and language abilities:
Persons with WS are bad at:

- Spatial cognition tasks
- Distinct modes of failure in WS and Down:

**Task:** REPRODUCE IMAGE

**Williams subjects**

**Down subjects**
Persons with WS are bad at:

• Piagetian conservation tasks
  – Ability to recognize that number, mass or volume does not alter when physical appearance alters.
  – E.g. ■ = ◆

• Math and number concepts
• Folkbiology (Johnson & Carey, 1998)
Persons with WS are good at:

• Music.
  – Individuals with WS generally have a higher rate of musicality.
  – WS persons tend to have an “affinity” for music -- this, however, does not necessarily entail musical talent.
  – The percentage of individuals with Williams syndrome who have perfect pitch is higher than the percentage among individuals in the general population.
Persons with WS are good at:

• Auditory short term memory.
  – Recall: This is precisely what’s *impaired* in dyslexia and SLI
Persons with WS are good at:

• Face processing.

Figure 13. The strengths and weaknesses in visuospatial processing in WMS show an unusual profile. The results are shown from two tasks that are both visuo-perceptual tasks, sensitive to right-hemisphere damage, where the correct answer requires only pointing to a picture without any constructional component. Note that the same subjects with WMS perform very differently on the two tasks. The contrast in performance on line orientation (Benton Judgment of Line Orientation, mean percent correct = 36.67) and face discrimination (Benton Face Recognition, mean percent correct = 92.59) is shown for 16 subjects with WMS ($t = 18.69; p < .0001$). On the Line-Orientations task, several individuals with WMS could not even pass the warm up items. In great contrast, exactly the same subjects with WMS perform remarkably well on a very difficult face discrimination task that involves recognizing the same individual under different conditions of lighting, shadow, and orientation. In both tasks, performance of normal individuals is indicated by the broken lines.
Persons with WS are good at:

• Theory of mind
Persons with WS are good at:

• **Language**
  – But there is a debate about whether language is really spared.

  For excellent resources on this debate, see Andrea Zukowski’s UMD website:
  

  – For example, it’s been shown that WS kids perform badly on embedded relative clauses.
    
  (Karmiloff-Smith et al, 1997; Volterra et al., 1996; Mervis et al., 1999)
What does the test look like?

“The circle the star is in is red”
• Hardly surprising if a person with general mental retardation does not do so well on this task.
• Challenge: to devise tasks that tap onto language without being taxing for other cognitive resources.
Example
(from Zukowski’s work)

Method: Elicited production technique
(Hamburger and Crain, 1982)

Children are asked to tell a parent which of two similar characters a change happened to.
Which cow is Max looking at?
“The cow who um the boy’s pointing to.”

(12-year-old Williams Syndrome male, IQ = 56)
(12-year-old Williams Syndrome male, IQ = 56)

"The cow who um the boy’s pointing to."
Which boy turned blue and which boy turned purple?
(16-year-old Williams Syndrome male, IQ = 40)

“The boy that’s pointing to his arm turned purple, and the boy that turned blue is pointing to his finger”
(16-year-old Williams Syndrome male, IQ = 40)

Subject Gap Relatives

“The boy that’s pointing to his arm turned purple, and the boy that turned blue is pointing to his finger”
(16-year-old Williams Syndrome male, IQ = 40)

"The boy that’s pointing to his arm turned purple, and the boy that turned blue is pointing to his finger"
What was each WS individual capable of producing?

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<th>OG Relative</th>
<th>CE Relative</th>
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# Control Children

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Every WS child except one was capable of producing examples of each of the components of embedded relative clauses
• WS kids produce complex syntactic structures as long as the task demands are otherwise low.
Individuals with Down’s and Williams Syndromes have similar IQs
Figure 4. Individuals with WMS (12 years and older) perform significantly better than those with DNS (age- and Full Scale IQ-matched to WMS subjects) on syntactic processing tasks (e.g., conditional sentences) on both grammar and content. Examples of responses by subjects with WMS and DNS are shown. Normal control levels are shown by a line.
Figure 6. (a) The figure shows stimulus examples of homonyms task with primary and secondary meanings (e.g., 'bank'). (b) Subjects with DNS and normal mental age controls provide more primary meanings than secondary meanings of the homonyms, as would be expected. Subjects with WMS, in contrast, provide an equal number of primary and secondary meanings, suggesting anomalous semantic organization. (c) Sample responses show that WMS subjects are able to access both the primary and secondary meanings of homonyms while DNS subjects access only one meaning, sometimes with two responses. For the meaning of 'club,' a DNS individual said “Go to a club” and then “I'm in the key club” whereas a WMS individual said: “A secret kind of club, and a club with spurs . . . .”
WS and irregular vs. regular morphology

• Overregularization of irregulars in WS (Clahsen et Almazan; Clahsen et al 2003).
  – “rules” are intact; lexical memory is impaired. Taken as evidence for dual mechanism theories morphology.
Zukowski (2001): WS regular vs. irregular morphology

(1) What’s this?
A FOOT!
A RAT!

(2) And here are a bunch of…?
FEET!
RATS!

(3) What would you call someone who eats…?
FEET-EATER!
RAT-EATER!
*RATS-EATER!

• Gordon (1985): kids as young as 3 years never produce rats-eater.
Zukowski (2001): WS regular vs. irregular morphology
Zukowski (2001): WS regular vs. irregular morphology

- What about WS kids? Producing regulars is very easy for them while producing irregulars is hard. Would WS kids produce *rats-eater*?
  - No, never.