



Sheba - Academic Medical Center Hospital

Validation of the Computerized Neurocognitive Battery in individuals with 22q11.2 deletion syndrome (22q11DS)

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Big Thanks

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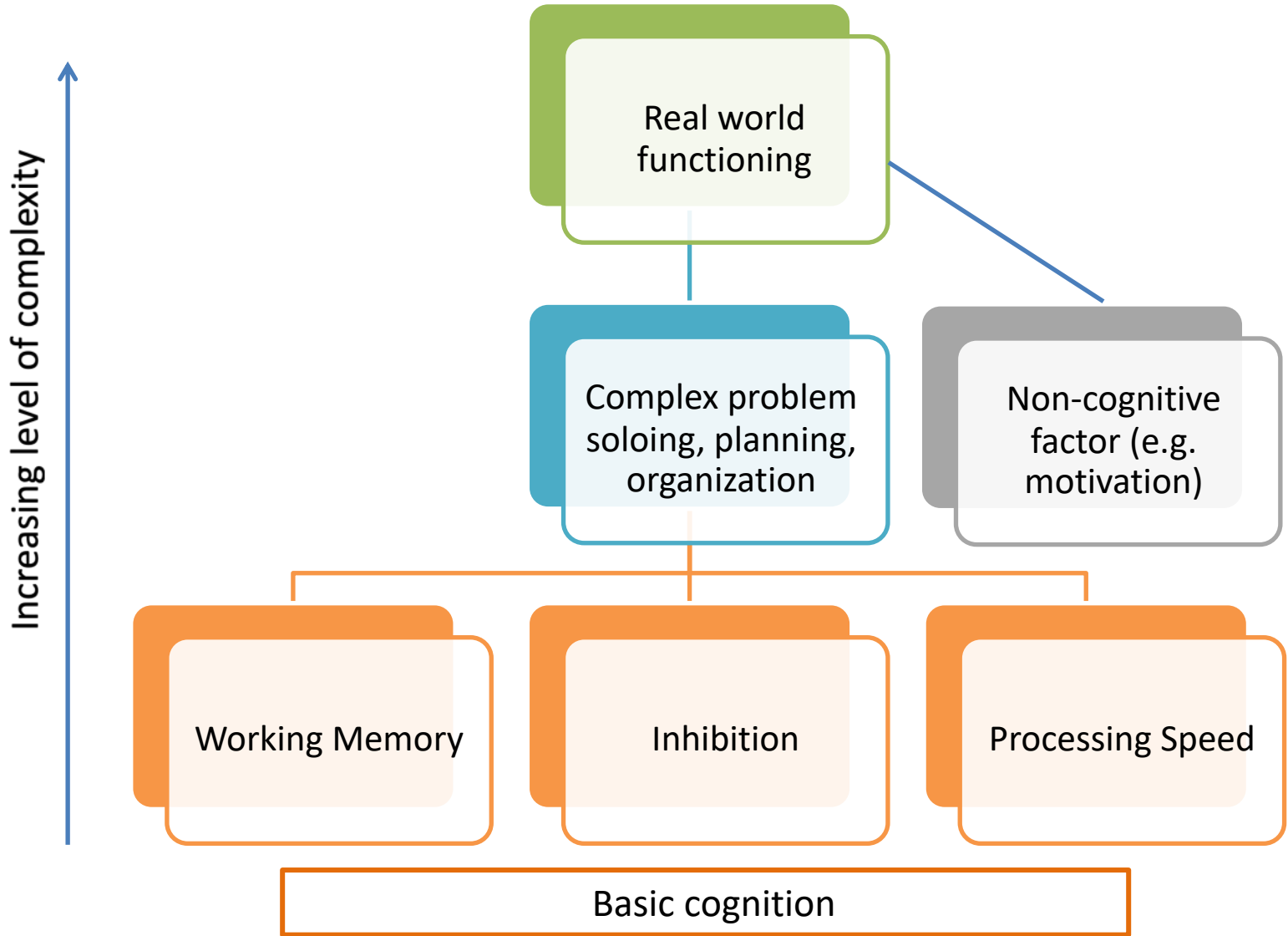
Children's Hospital of Philadelphia,
Penn University

Main Ideas

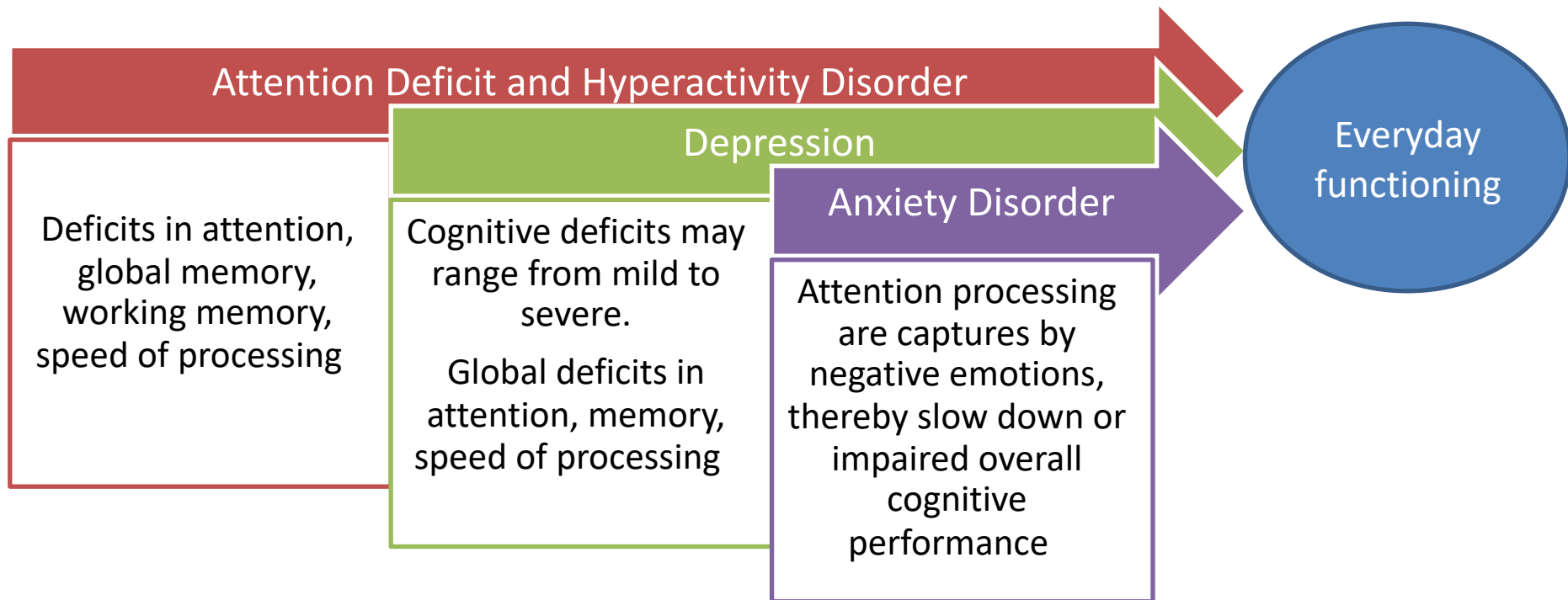
- The importance of cognition in everyday functioning
- Results from cognitive studies with individuals with 22q11DS
- The Computerized Neurocognitive Battery (CNB)

Most of individuals with 22q11DS cope with learning disabilities

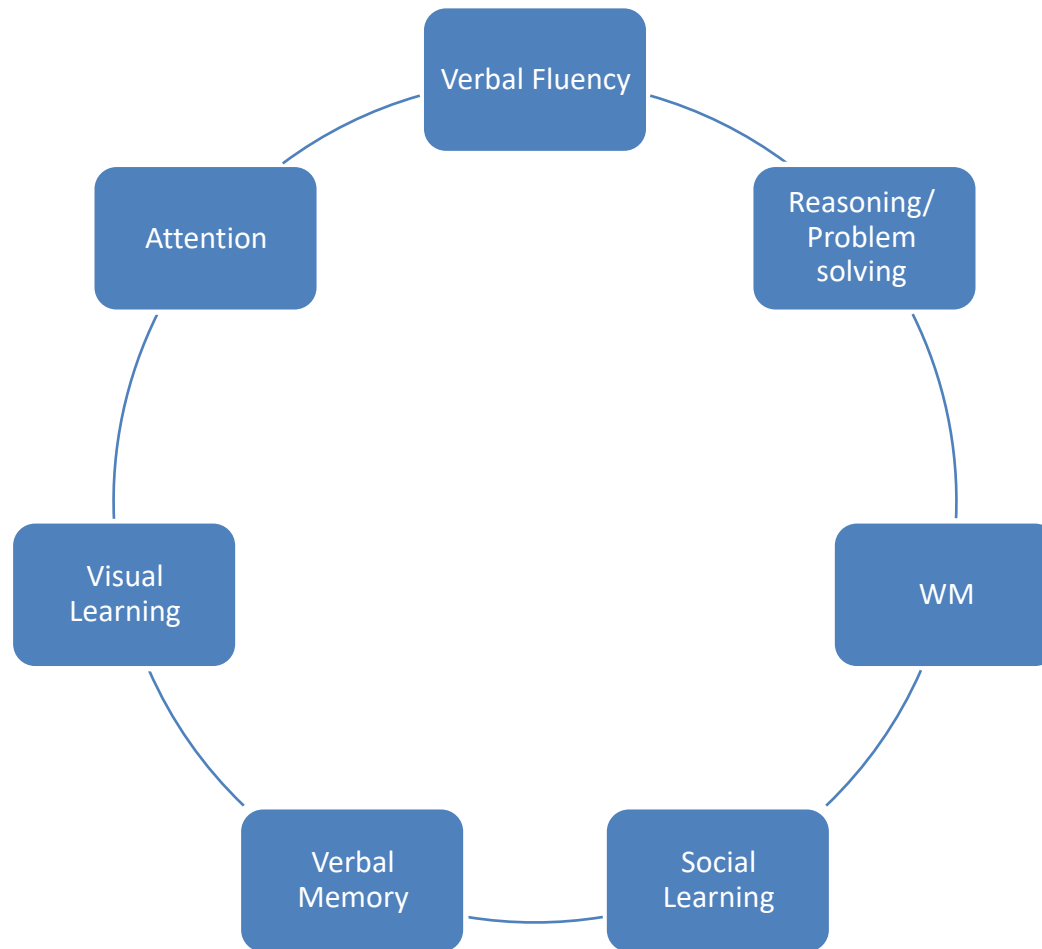




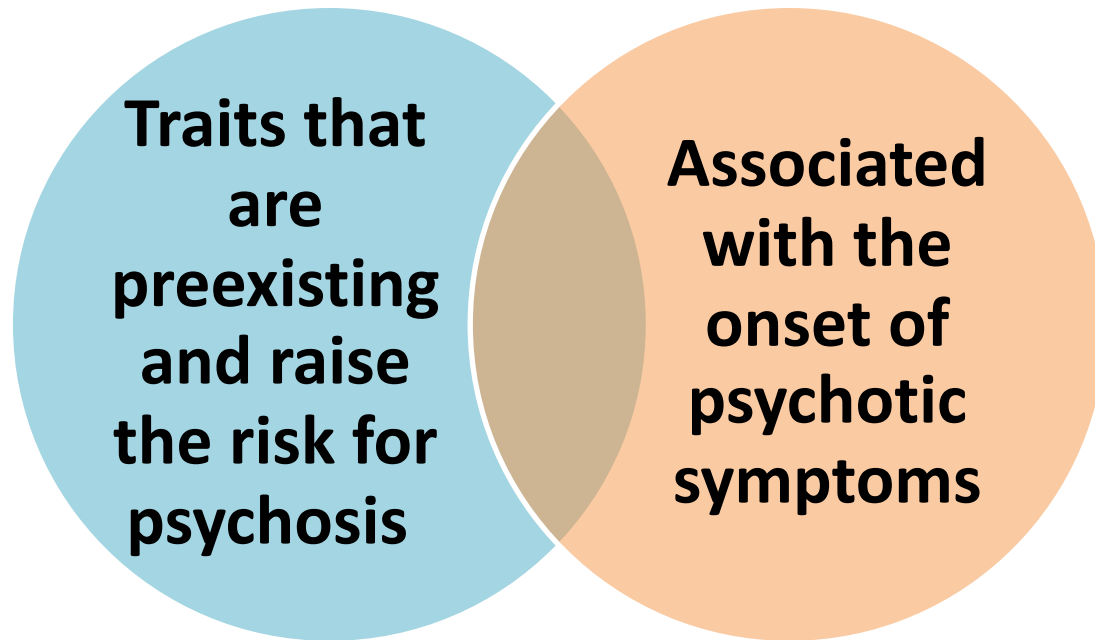
The Cognitive Profile of Select Psychiatric Disorders



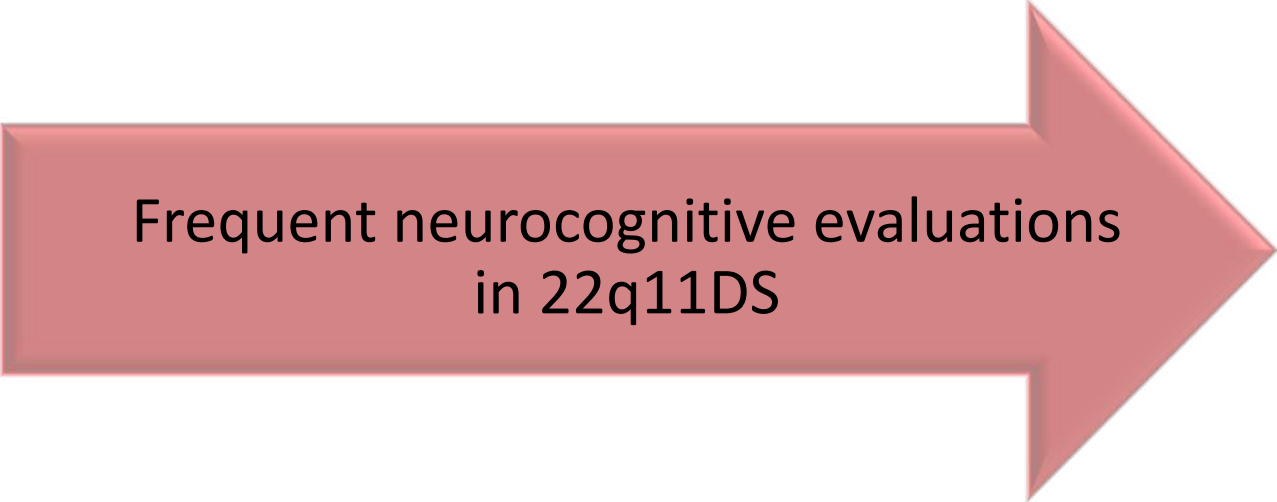
Neuro- Cognitive Deficits in Schizophrenia



Cognitive Deficits in Psychosis



Studies recommendation



Frequent neurocognitive evaluations
in 22q11DS

Bassett, A. S., et al (2011). Practical guardedness for managing patients with 22q11.2 deletion syndrome.

Limitations of Previous Studies

- IQ test is essential for later diagnosis- **BUT** it has limitations
- Little used a comprehensive computerized battery adapted for use in 22q11DS

The field should place emphasis on early identification and intervention

Neurocognitive evaluation should be...

Reliable

Thorough

Comprehensive

Efficient

Can be used
periodically

Can be used
frequent if
needed

The Penn Computerized Neurocognitive Battery (CNB)

- Develop in Penn university
- Previous used in schizophrenia studies, and was adapted for 22q11DS research
- It takes ~1 hour to administer, by trained staff
- CNB performance is compared to chronological age, based on typically developed controls

Computerized Neurocognitive Battery

Executive Function

- Continuous Performance Test
- Conditional Exclusion
- Letter-N-back

Complex Cognition

- Line Orientation Test
- Matrix Analysis

Social Cognition

- 5 Emotion Recognition
- Pair of faces, which one is older
- Pair of faces with emotions, which one is more intense

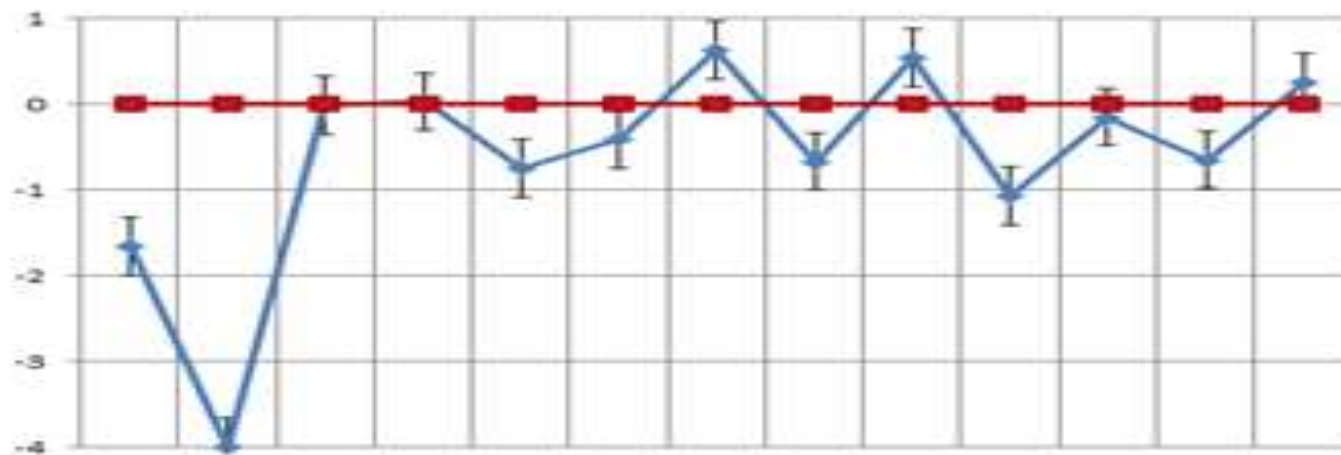
Episodic Memory

- Words Recall
- Faces Recall
- Visual Object learning

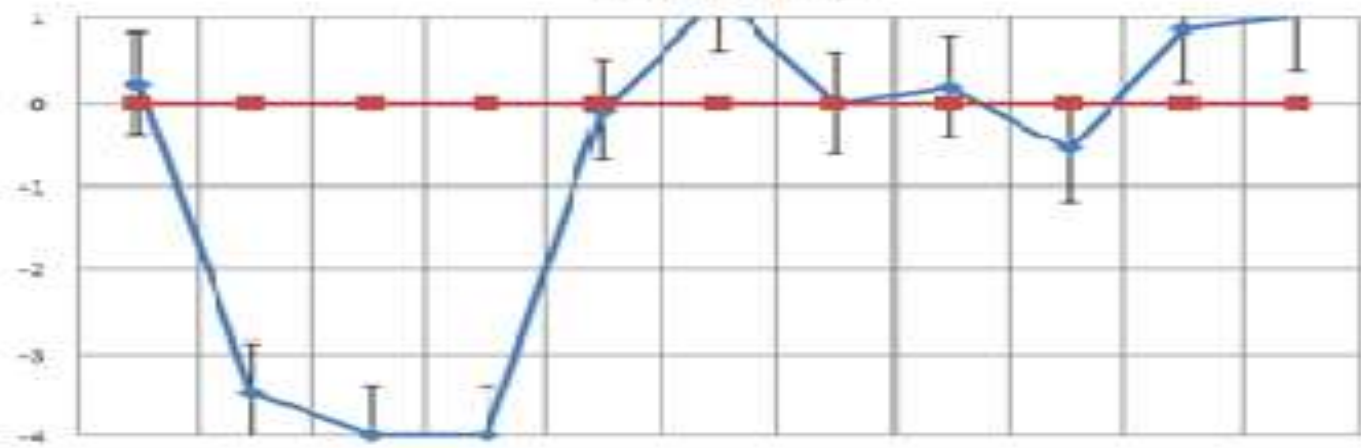
Praxis Speed

- Click a moving green square that gets smaller
- Spacebar tapping

Reaction Time



Accuracy



ABF ATT TVM VEME FMEM SMEM DVR SPA EM EMQ AGD

Examples for clinical recommendations

Problems with following new rules

- Ensure frequent repetition and reinforcement of information

Problems in social cognition

- Escorting John through interpersonal relationships and in complex social situations

Assess memory

- Visual memory may be a relative strength

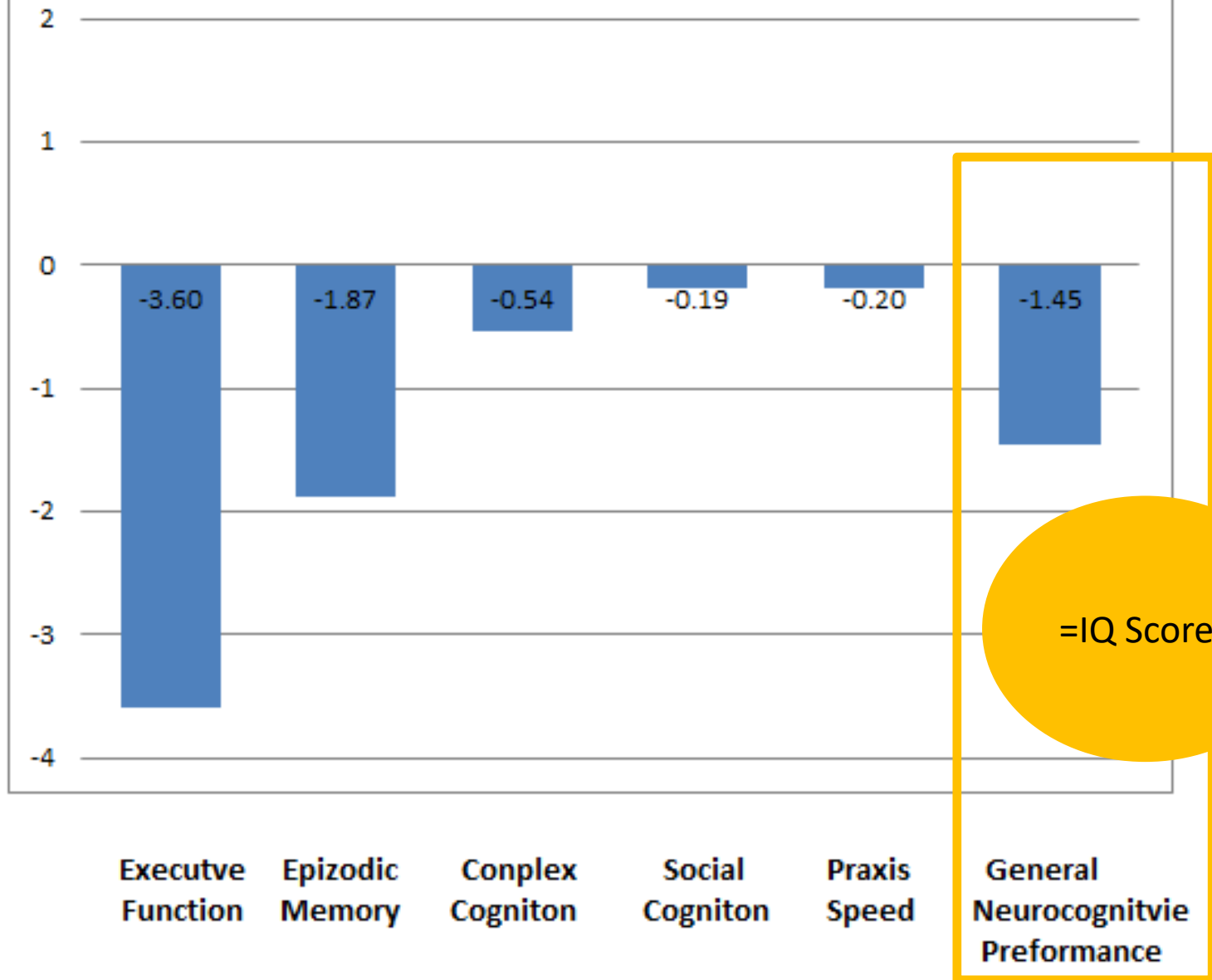
Problems in attention

- To monitor the attention functions

Problems in abstract tasks

- John seems to perform better when the instruction is clear and familiar

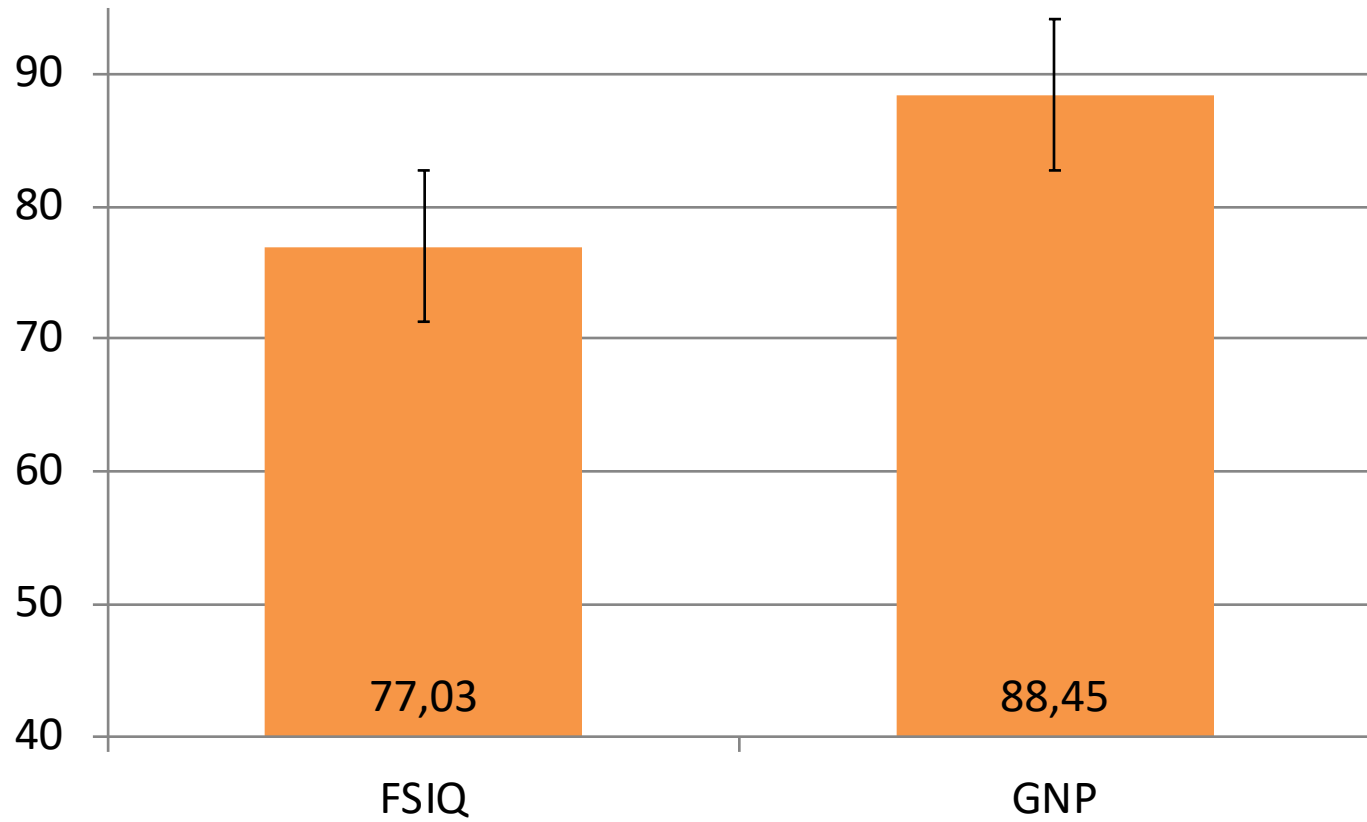
Computerized Neurocognitive Battery

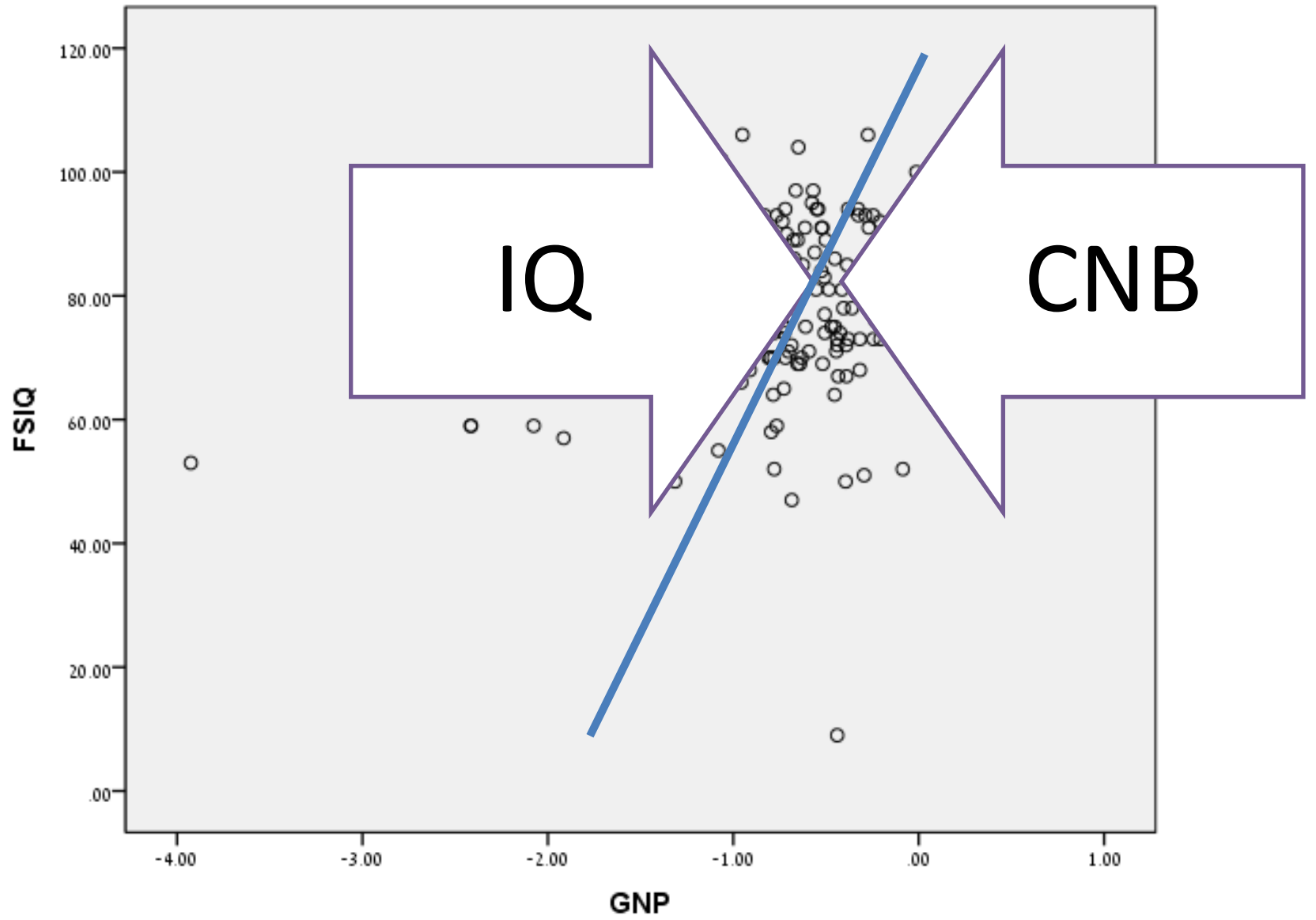


Demographic Characteristics

n	356
Age (mean)	22.6±8.3
Age range	8.06-52.44
Male / Female (%)	51/49
FSIQ	77.03±12.05

CNB - IQ





Neurocognitive evaluation should be...

Reliable ✓

Thorough ✓

Comprehensive ✓

Efficient ✓

Can be used
periodically ✓

Can be used
frequent if
needed ✓

Conclusions

- The CNB can be used to assess **efficiently** neurocognitive performance in individuals with 22q11DS
- The CNB can obtain a reliable **proxy of general intelligence**

SOPS SUMMARY SHEET

Positive Symptom Scale

0 Never, Absent	1 Questionably Present	2 Mild	3 Moderate	4 Moderately Severe	5 Severe but Not Psychotic	6 Severe and Psychotic
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Positive Symptoms								Totals
A1. Unusual Thought Content/Delusional Ideas	0	1	2	3	4	5	6	
A2. Suspiciousness/Persecutory Ideas	0	1	2	3	4	5	6	
A3. Grandiosity	0	1	2	3	4	5	6	
A4. Perceptual Abnormalities/Hallucinations	0	1	2	3	4	5	6	
A5. Conceptual Disorganization	0	1	2	3	4	5	6	_____

Negative, Disorganized, General Symptom Scale

0 Absent	1 Questionably Present	2 Mild	3 Moderate	4 Moderately Severe	5 Severe	6 Extreme
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Negative Symptoms								Totals
B1. Social Isolation and Withdrawal	0	1	2	3	4	5	6	
B2. Avolition	0	1	2	3	4	5	6	
B3. Decreased Expression of Emotion	0	1	2	3	4	5	6	
B4. Decreased Experience of Emotions and Self	0	1	2	3	4	5	6	
B5. Decreased Ideational Richness	0	1	2	3	4	5	6	
B6. Deterioration in Role Functioning	0	1	2	3	4	5	6	_____

Disorganization Symptoms								Totals
C1. Odd Behavior or Appearance	0	1	2	3	4	5	6	
C2. Bizarre Thinking	0	1	2	3	4	5	6	
C3. Trouble with Focus and Attention	0	1	2	3	4	5	6	
C4. Personal Hygiene/Social Attentiveness	0	1	2	3	4	5	6	_____

Risk for subthreshold psychotic symptoms by association to neurocognitive abilities

Adjusted probability for the emergence of baseline negative subthreshold psychotic syndrome

	22q11DS	WS	p
GNP (general neurocognitive performance)	0.74	0.26	.008
Executive Function	0.70	0.37	.040
Social Cognition	0.67	0.38	.050

Neurocognitive trajectories and effect of medication in 22q11DS

- Deficits in **GNP** predicted **negative subthreshold psychotic syndrome** in 22q11DS
- There was a **significant progress for individuals with 22q11DS under psychiatric medication** Also, medication treatments is a significant predictor for GNP change.

Conclusions

- The CNB is a potentially a useful tool for identifying cognitive deficits associated with psychosis disorders in 22q11DS.