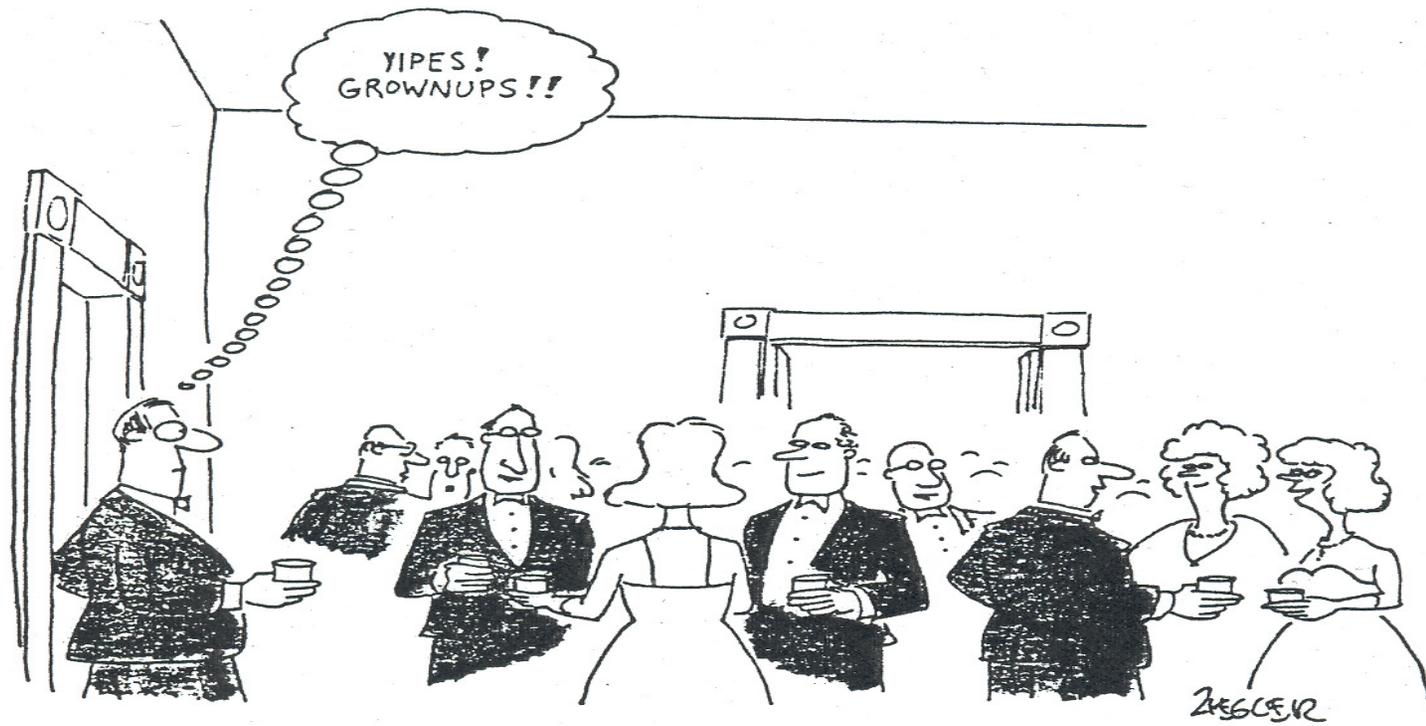


Overview:
Neuropsychological Evaluation of
Autistic Spectrum Disorder
Howard Glidden, Ph.D. FACPN



YIPES!
GROWNUPS!!

ZIEGLER

Neuropsychological Assessment

- Neuropsychology is a subspecialty of psychology that blends the disciplines of psychology and neuroscience.
- APA definition of Pediatric Neuropsychology: “Pediatric neuropsychology is a professional specialty concerned with learning and behavior in relationship to a child’s brain. A pediatric neuropsychologist is a licensed psychologist with expertise in how learning and behavior are associated with the development of brain structures and systems...”

Purpose of Pediatric neuropsychology

- To clarify the diagnosis/differential diagnosis
 - To establish a baseline of functioning
 - To evaluate relative strengths/weaknesses
 - To evaluate change over time
 - To assist in treatment/educational planning
 - To facilitate the therapy itself
-
- Psychological/neuropsychological evaluations differ from medical classification of brain functions which is usually binary (normal/impaired).
 - Neuropsychological assessment can grade function based on a continuum.
 - Neuropsychological assessment allows for increased sensitivity.

General principles of assessment

- No single psychometric test is sufficient or appropriate, by itself, for assessing brain function/dysfunction
- Assessment, in addition to psychometric test administration, takes into account:
 - Developmental history
 - Family history
 - Previous assessments
 - Cultural/language variability
 - Limits of reliability/validity of instruments utilized
 - Test bias
- There may be many reasons why a client performs poorly on a test or group of tests, which affects reliability

All measures, whether they be observation, clinical interview, psychometric or radiologic/lab tests provide estimates of the trait being measure.

- There are no perfect tests
- Tests are predictive of a trait or attribute being considered
- Tests yield scores that indicate the degree to which the trait in question is present
- In general, the higher the score, the greater the amount of the trait being measured

Assessment Guidelines

- Difficulties:
 - wide range of functioning (IQ, language, symptom severity)
 - attention, social, communication, compliance

Assessment Guidelines - continued

- Use the most valid, reliable, and comprehensive measures
- Parental involvement
- Coherent view of the child
- Profile of strengths and weaknesses
- Contact with other professionals

Reliability

Reliability is the degree to which tests scores are consistent, dependable, and repeatable. The greater the error of the test, the lower the reliability.

Validity

Validity is the degree to which a test or measurement accurately measures or reflects what it purports to measure.

Validity refers to the tests usefulness in measuring the question.

There are no valid or invalid tests.

A test can be reliable, but not be valid. A test cannot be valid, if it's unreliable.

Sensitivity/specificity

- Sensitivity is the probability that a test gives a positive diagnosis, given that the individual actually has the condition for which he or she is being tested.
 - Type I error
- Specificity is the probability that a test yields a negative diagnosis, given that the individual does not have the condition for which he or she is being tested.
 - Type II error

Type I/Type II table

Statistical Decision

Actual Condition

ACTUAL CONDITION		
	+	-
Test Result	Present	Absent
Positive +	Condition Present + Positive result = True Positive Hit Valid positive Sensitivity	Condition absent + Positive result = False Positive Type I error Alpha
Negative -	Condition Present + Negative result = False (invalid) Negative Type II error Beta	Condition absent + Negative result = True (accurate) Negative Specificity

Rejecting a null-hypothesis when it should not have been rejected creates a type I error.

Failing to reject a null-hypothesis when it should have been rejected creates a type II error.

In either case, a wrong decision or error in judgement has occurred.

HISTORICAL PERSPECTIVE

- 1801 Itard – “wild boy”
- Mid 1800’s Maudsley – “insanity in children
- Early 1900’s Bleuler – characterized autism as a psychological state associated with “detachment with the outside world”
- 1906 Kraepelin – “dementia praecox”
- DeSantis – “schizophrenia”
- 1943 Kanner – autism
- 1944 Asperger – “autistic psychopathology
- 1949 Rank – “atypical personality development
- 1952 DSM I - “schizophrenic reaction, childhood type”
- 1968 DSM II – “Schizophrenia, childhood type” covered this entire group of disorders
- 1976 Autism Society of America: working definition of Autism
- 1978 Rutter
- 1980 DSM III – Distinguishes Infantile Autism (as a category of Pervasive Developmental Disorders) from Schizophrenia, childhood onset
- 1984 DSM III-R: Highlight Infantile Autism as prototypal Pervasive Developmental Disorder. Provides more specific, developmentally-oriented diagnostic criteria
- 1994 DSM IV: Attempts to define subtypes of autistic disorders, including Rett Syndrome, Asperger Syndrome, Atypical Autism, and Pervasive Developmental Disorder NOS
- DSM V: Stay tuned...

DSM I (1952)

000-x28 Schizophrenic reaction, Childhood Type

- Here will be classified those schizophrenic reactions occurring before puberty. The clinical picture may differ from schizophrenic reactions occurring in other age periods because of the immaturity and plasticity of the patient at the time of onset of the reaction. Psychotic reactions in children, manifesting primarily **autism**, will be classified here.

DSM II (1968)

Autism was not mentioned; the word appears only under the following category

295.8 Schizophrenia , childhood type

- This category is for cases in which schizophrenic symptoms appear before puberty. This condition may be manifested by **autistic**, atypical and withdrawn behavior; failure to develop identity separate from the mother's; and general unevenness, gross immaturity and inadequacy of development. These developmental defects may result in mental retardation, which should also be diagnosed.

DSM III (1980)

- Diagnostic criteria for **Infantile Autism**
 - A. Onset before 30 months of age
 - B. Pervasive lack of responsiveness to other people (autism)
 - C. Gross deficits in language development
 - D. If speech is present, peculiar speech patterns such as immediate and delayed echolalia, metaphorical language, pronoun reversal
 - E. Bizarre responses to various aspects of the environment, e.g. resistance to change, peculiar interest in or attachments to animate or inanimate objects
 - F. Absence of delusions, hallucinations, loosening of associations, and incoherence as in Schizophrenia

DSM III-R (1987)

Diagnostic Criteria for Autistic Disorder

- At least eight of the following sixteen items are present, these to include at least two items from A, one from B, and one from C.
- A. *Qualitative impairment in reciprocal social interaction (the examples within parentheses are arranged so that those first listed are more likely to apply to younger or more disabled, and the later ones, to older or less disabled) as manifested by the following:*
 - 1. Marked lack of awareness of the existence or feelings of others (for example, treats a person as if that person were a piece of furniture; does not notice another person's distress; apparently has no concept of the need of others for privacy);
 - 2. No or abnormal seeking of comfort at times of distress (for example, does not come for comfort even when ill, hurt, or tired; seeks comfort in a stereotyped way, for example, says "cheese, cheese, cheese" whenever hurt);
 - 3. No or impaired imitation (for example, does not wave bye-bye; does not copy parent's domestic activities; mechanical imitation of others' actions out of context);
 - 4. No or abnormal social play (for example, does not actively participate in simple games; refers solitary play activities; involves other children in play only as mechanical aids); and
 - 5. Gross impairment in ability to make peer friendships (for example, no interest in making peer friendships despite interest in making fiends, demonstrates lack of understanding of conventions of social interaction, for example, reads phone book to uninterested peer.

DSM III-R continued

- *B. Qualitative impairment in verbal and nonverbal communication and in imaginative activity, (the numbered items are arranged so that those first listed are more likely to apply to younger or more disabled, and the later ones, to older or less disabled) as manifested by the following:*
- 1. No mode of communication, such as: communicative babbling, facial expression, gesture, mime, or spoken language;
- 2. Markedly abnormal nonverbal communication, as in the use of eye-to-eye gaze, facial expression, body posture, or gestures to initiate or modulate social interaction (for example, does not anticipate being held, stiffens when held, does not look at the person or smile when making a social approach, does not greet parents or visitors, has a fixed stare in social situations);
- 3. Absence of imaginative activity, such as play-acting of adult roles, fantasy character or animals; lack of interest in stories about imaginary events;
- 4. Marked abnormalities in the production of speech, including volume, pitch, stress, rate, rhythm, and intonation (for example, monotonous tone, question-like melody, or high pitch);
- 5. Marked abnormalities in the form or content of speech, including stereotyped and repetitive use of speech (for example, immediate echolalia or mechanical repetition of a television commercial); use of "you" when "I" is meant (for example, using "You want cookie?" to mean "I want a cookie"); idiosyncratic use of words or phrases (for example, "Go on green riding" to mean "I want to go on the swing"); or frequent irrelevant remarks (for example, starts talking about train schedules during a conversation about ports); and
- 6. Marked impairment in the ability to initiate or sustain a conversation with others, despite adequate speech (for example, indulging in lengthy monologues on one subject regardless of interjections from others);

DSM III-R continued

- *C. Markedly restricted repertoire of activities and interests as manifested by the following:*
 - 1. Stereotyped body movements (for example, hand flicking or twisting, spinning, head-banging, complex whole-body movements);
 - 2. Persistent preoccupation with parts of objects (for example, sniffing or smelling objects, repetitive feeling of texture of materials, spinning wheels of toy cars) or attachment to unusual objects (for example, insists on carrying around a piece of string);
 - 3. Marked distress over changes in trivial aspects of environment (for example, when a vase is moved from usual position);
 - 4. Unreasonable insistence on following routines in precise detail (for example, insisting that exactly the same route always be followed when shopping);
 - 5. Markedly restricted range of interests and a preoccupation with one narrow interest, e.g., interested only in lining up objects, in amassing facts about meteorology, or in pretending to be a fantasy character.
- D. Onset during infancy or early childhood
- Specify if childhood onset (after 36 months of age)

DSM-IV-TR Diagnostic Criteria:

- **299.00 Autistic Disorder**
- A total of six (or more) items from (1), (2), and (3), with at least two from (1), and one each from (2) and (3):

DSM-IV-TR Diagnostic Criteria:

- (1) qualitative impairment in social interaction, as manifested by at least two of the following:
 - (a) marked impairment in the use of multiple nonverbal behaviors, such as eye-to-eye gaze, facial expression, body postures, and gestures to regulate social interaction
 - (b) failure to develop peer relationships appropriate to developmental level
 - (c) a lack of spontaneous seeking to share enjoyment, interests, or achievements with other people (e.g., by a lack of showing, bringing, or pointing out objects of interest)
 - (d) lack of social or emotional reciprocity

DSM-IV-TR Diagnostic Criteria:

- (2) qualitative impairments in communication, as manifested by at least one of the following:
 - (a) delay in, or total lack of, the development of spoken language (not accompanied by an attempt to compensate through alternative modes of communication such as gesture or mime)
 - (b) in individuals with adequate speech, marked impairment in the ability to initiate or sustain a conversation with others
 - (c) stereotyped and repetitive use of language or idiosyncratic language
 - (d) lack of varied, spontaneous make-believe play or social imitative play appropriate to developmental level

DSM-IV-TR Diagnostic Criteria:

- (3) restricted, repetitive, and stereotyped patterns of behavior, interests, and activities as manifested by at least one of the following:
 - (a) encompassing preoccupation with one or more stereotyped and restricted patterns of interest that is abnormal either in intensity or focus
 - (b) apparently inflexible adherence to specific, nonfunctional routines or rituals
 - (c) stereotyped and repetitive motor mannerisms (e.g., hand or finger flapping or twisting or complex whole-body movements)
 - (d) persistent preoccupation with parts of objects

DSM-IV-TR Diagnostic Criteria:

- B. Delays or abnormal functioning in at least one of the following areas, with onset prior to age 3 years:
 - (1) social interaction,
 - (2) language as used in social communication, or
 - (3) symbolic or imaginative play.
- C. The disturbance is not better accounted for by Rett's disorder or childhood disintegrative disorder.

Autistic Disorder: DSM-IV-TR

Overview

- Qualitative Impairments in SOCIAL INTERACTION
- Qualitative Impairments in COMMUNICATION
- RESTRICTED REPETITIVE AND STEREOTYPED PATTERNS OF BEHAVIOR, INTERESTS AND ACTIVITIES

DSM IV-TR

Sensitivity = .82

Specificity = .89

- Field Trial for Autistic Disorder in DSM IV (1994)
 - Volkmar, F.R., Klin, A., Siegel, B., Szatmari, P., Lord, C., Campbell, A. Freeman, B.J., Cichetti, D.V. , Ritter, M and Kline, W.

DSM V

- STAY TUNED...

Changes Associated with Diagnosis

- Broader definition of autism
- With the advent of the DSM-IV, questions regarding eligibility of children with PDD-NOS or Asperger's Syndrome for services
- Autism was not recognized as a disability under the IDEA until 1990. Children with autism were typically classified as “other health impaired” or “multihandicapped”

Diagnostic Criteria for Autism

- Impairments in social relatedness
 - Nonverbal communication (e.g. eye contact)
 - Peer interaction
 - Joint attention
 - Emotional reciprocity

Diagnostic Criteria for Autism (cont'd)

- Impairments in communication and play
 - Delay in language development
 - Impaired conversational ability
 - Stereotyped and repetitive language
 - Absent or stereotyped pretend play

Diagnostic Criteria for Autism (cont'd)

- Restricted interests and activities
 - Encompassing preoccupations and interests
 - Adherence to nonfunctional routines or rituals
 - Motor stereotypies
 - Preoccupations with parts of objects



Leo Kanner

Leo Kanner, M.D.

- 1930 first U.S. child psychiatrist
- 1935 first textbook of child psychiatry
- 1943 first description of autism

Leo Kanner's Autism

- Extreme autistic aloneness
- Anxiously obsessive desire for the preservation of sameness
- Excellent rote memory
- Delayed echolalia
- Oversensitivity to stimuli
- Limitation in the variety of spontaneous activity
- Good cognitive potentialities
- Highly intelligent families

Francesca Happe (1994) – Autism: An introduction to psychological theory.

Impairment in Social Interaction

- “inability to relate themselves”
- “extreme autistic aloneness”
- “failure to assume at any time an anticipatory posture”

Impairments in Communication

- 11 case reports (8 boys, 3 girls)
- 3 of 11 “mute”
- Ability to speak delayed in some
- In none has language “served to convey meaning to others”

Restricted & Stereotyped Patterns

- “delayed echolalia”
- “monotonously repetitious”
- “anxiously obsessive desire for the maintenance of sameness”
- “limitation in the variety of spontaneous activity”

Parents

- Intelligent
- Obsessive
- “Very few really warmhearted”

The first case to be described by Dr. Kanner was of a 5-year, 1-month old boy. He wrote: “Before the family’s arrival from their hometown, they sent a 33 page typewritten history that, though filled with much obsessive detail, gave an excellent account of Donald’s background.”

“Autistic Disturbance of Affective Contact”

- “Since 1938, there have come to our attention a number of children whose condition differs so markedly and uniquely from anything reported so far, that each case merits – and I hope, will eventually receive – a detailed consideration of its fascinating peculiarities.”

(p.217)

Wing's Triad of Impairments in Autism

- Qualitative impairment in reciprocal social interaction
- Qualitative impairment in verbal and nonverbal communication and in imaginative activity;
- Markedly restricted repertoire of activities and interests

Primary Features of Autism

- A. Autism is a biological disorder.
- B. Autism is a clinical (behaviorally defined) syndrome.
- C. Autism is a spectrum disorder.
- D. Autism is a developmental diagnosis.
- E. Autism is a retrospective diagnosis.
- F. Autism frequently occurs in association with other syndromes, specific diseases, and developmental disabilities.
- G. While there is no known cure, children with autism improve with proper, early intervention.
- H. Autism is a lifelong condition and requires long-term case management and coordination of services.

Myths of Autism

- Children with autism never make eye contact
- Inside a child with autism is a genius
- Children with autism do not talk
- Progress means that the child doesn't have autism
- Behavior change from maladaptive to adaptive isn't autistic
- Children with autism do not smile at you
- Children with autism do not give or receive physical affection
- People with autism do not notice others and don't pick up cues from peers/adults
- People with autism do not want friends
- Individuals with autism do not relate to peers/adults
- People with autism could talk if they wanted to
- When a child with autism does not respond to a question/direction to which he has previously given a correct response, he is being stubborn/non-compliant/obnoxious
- Autism can be outgrown
- Autism is an emotional disability
- Children with autism cannot learn
- Children with autism will show no imagination
- Bad parenting causes autism
- Autism is rare

Autism is a neurodevelopmental syndrome that is behaviorally defined. It is a retrospective diagnosis with behavioral symptoms that vary over time. Dynamic impairments are qualitative and include deficits in social interaction and communication with restricted, repetitive or stereotyped patterns of behavior.

There are no pathognomic signs:

- Impaired social responsiveness
- Echolalia
- Stereotypies
- Impaired facial recognition
- Eye gaze aversion
- Regression of acquired language and social skills with spared motor skills is a Red Flag

Conditions With Similarities To Autism

- Selective mutism
- Attachment disorder
- Developmental receptive-expressive language disorder
- Hyperkinetic disorder with stereotypies
- Disintegrative disorder
- Intellectual disability
- Rett's syndrome
- Landau-Kleffner syndrome
- Sensory impairment
- Generalized Anxiety Disorder/Social phobia
- Schizotypal personality disorder
- Schizoid personality disorder
- Global developmental delay

If it's not ASD

- Intellectual disability
- Language disorders
- Selective mutism
- Shy, gifted students who are overwhelmed by noisy and chaotic preschool rooms
- Tic disorder or Tourette's (with tics misinterpreted as autistic stereotypies)
- Angelman's syndrome; other genetic/metabolic anomalies
- Fetal alcohol syndrome
- Progressive hearing loss
- Separation anxiety; social anxiety
- Older children: depression; schizoid personality/schizophrenia; bored-to-death gifted child in a stultifying classroom; mild mental retardation; severe dysfluent dyslexia (with good decoding); anxiety disorders

Autism: Components of Cause

Abnormalities in Genetic Code for Brain Development



Abnormal Mechanisms of Brain Development



Structural and Functional Abnormalities



Cognitive and Neurologic Abnormalities



Behavioral Syndrome

- Data from family and twin studies indicate Autism is genetically heterogeneous and several genes are involved in the development of Autism
- The combination of genes varies across the population
- Fatal gene

GENETIC FACTORS IN AUTISM

- A. 3-5% recurrence risk among siblings compared to population frequency of 2-5/10,000 (Bolton et al., 1994).
- B. Twin studies suggest genetic underpinnings of autism (Bailey et al., 1995).
- C. Autism is “complex trait”, i.e., a trait in which multiple genotypes lead to the same phenotype (genetic heterogeneity).
- D. High comorbidity of autism with known genetic conditions, such as Fragile X, Tuberous Sclerosis, and chromosomal abnormalities suggest genetic heterogeneity is present (Ritvo et al., 1990; Rutter et al., 1994).
- E. Family and twin studies suggest a broader phenotype definition of autism which may include social deficits, cognitive and communication disorders, obsessive-compulsive disorder, affective and anxiety disorders (LeContour et al., 1996).
- F. As a result of the Human Genome Project, identification of susceptibility of genes, it should now be possible using linkage analysis on properly diagnosed and collected families.
- G. Once susceptibility genes underlying autism are identified, the role of other risk factors can be studied.
- H. Stoppage factor estimates inheritance among siblings to be closer to 10%.

Rutter (2003) summary of neuropsychological theories

- If specific cognitive deficit causes the syndrome, why are not all severely MR children autistic?
- Brain normally works in a top-down, context-driven, predictive fashion, less in autism, but central coherence is underspecified
- Theory of mind deficits not primary, not universal in autism, not specific, can't explain non-social aspects
- Deficits in executive functions not specific, not primary, can't explain superior performance on some tasks

Neurobiology

Brain abnormalities have been identified throughout the brain

- Structural imaging studies (MRI)
- Neuropathology
- Functional imaging studies (ERPs, fMRI)
- Neuropsychological testing

Neurobiology – continued

- Widespread neurodevelopmental abnormalities
- Autism spectrum disorders affect the development and function of both cortical and subcortical structures with considerable interindividual variation

Neurobiology – continued

- Brain weight is normal in most postmortem (adult) cases of autism, with rare cases of megalencephaly and possible microencephaly.
- Whole brain volume is slightly smaller than normal or normal in the majority of MRI studies of adolescents and adults with autism.
- Skull may be large, despite lower volume

Neurobiological Findings in Autism

- High peripheral levels of serotonin (blood test)
- High rate of seizure disorders
- Persistent primitive reflexes
- Increased head size (toddlers)
- Morphological changes in CNS
- Mini-columns/mirror neurons
- Fusiform gyrus and faces
- Placental abnormalities

-Fred Volkmar, M.D. 2006

Patterns of Brain Growth

- Neonatal head circumference records for 2 to 5 year-olds normal
- 90% of 2 to 4 year-olds with autism had larger than average brain volume
- Pediatric head growth records indicated abnormal growth by 6 to 14 months
 - Courchesne, Carper & Akshoomoff (2003)
 - Evidence of brain overgrowth in the first year of life in autism. JAMA, 290, 337-344.

Abnormal Developmental Trajectory?

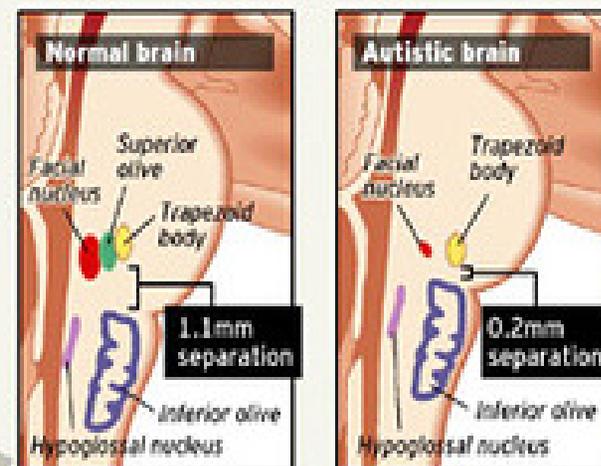
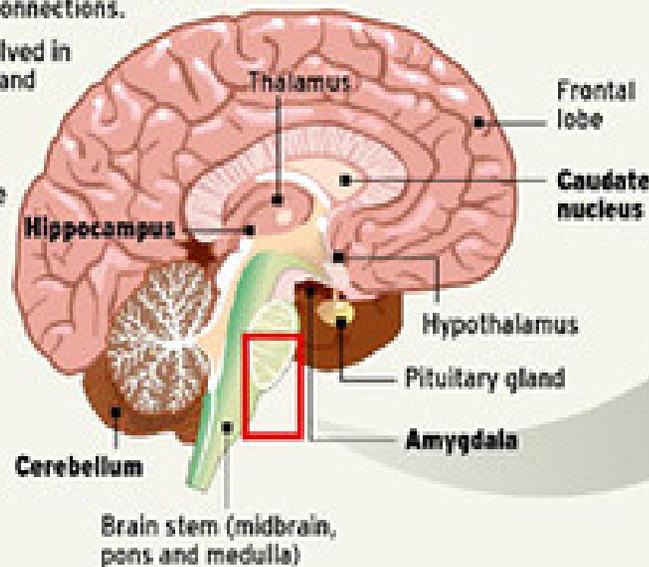
- Macrocephaly infrequent at birth in Autism
- Increased rate of macrocephaly with increasing age
 - Over 1/3rd of subjects showed increased head growth during childhood
- Substantial programmed neuron loss (apoptosis) occurs premorbidly
- Lower glucose metabolism in frontal lobes of autistics on PET scan

Autism and the brain

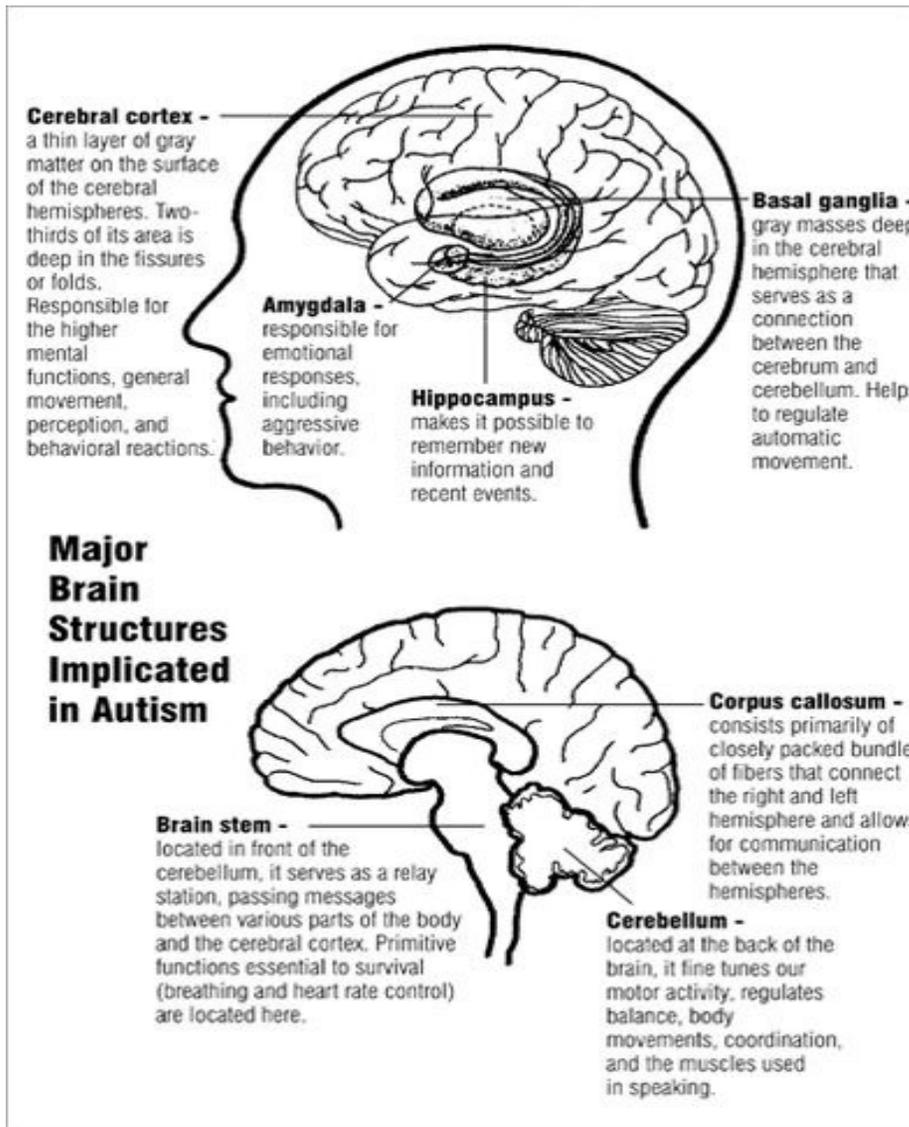
Researchers are finding that the brains of autistic persons are measurably different.

- Cells in certain areas are smaller and more densely packed; others have shorter and less-developed connections.

- The **amygdala**, involved in processing emotion, and the **hippocampus**, involved in memory and learning, are smaller. Similarly, the **cerebellum** and **caudate nucleus**, thought to be involved in shifting attention to different tasks, are significantly reduced.

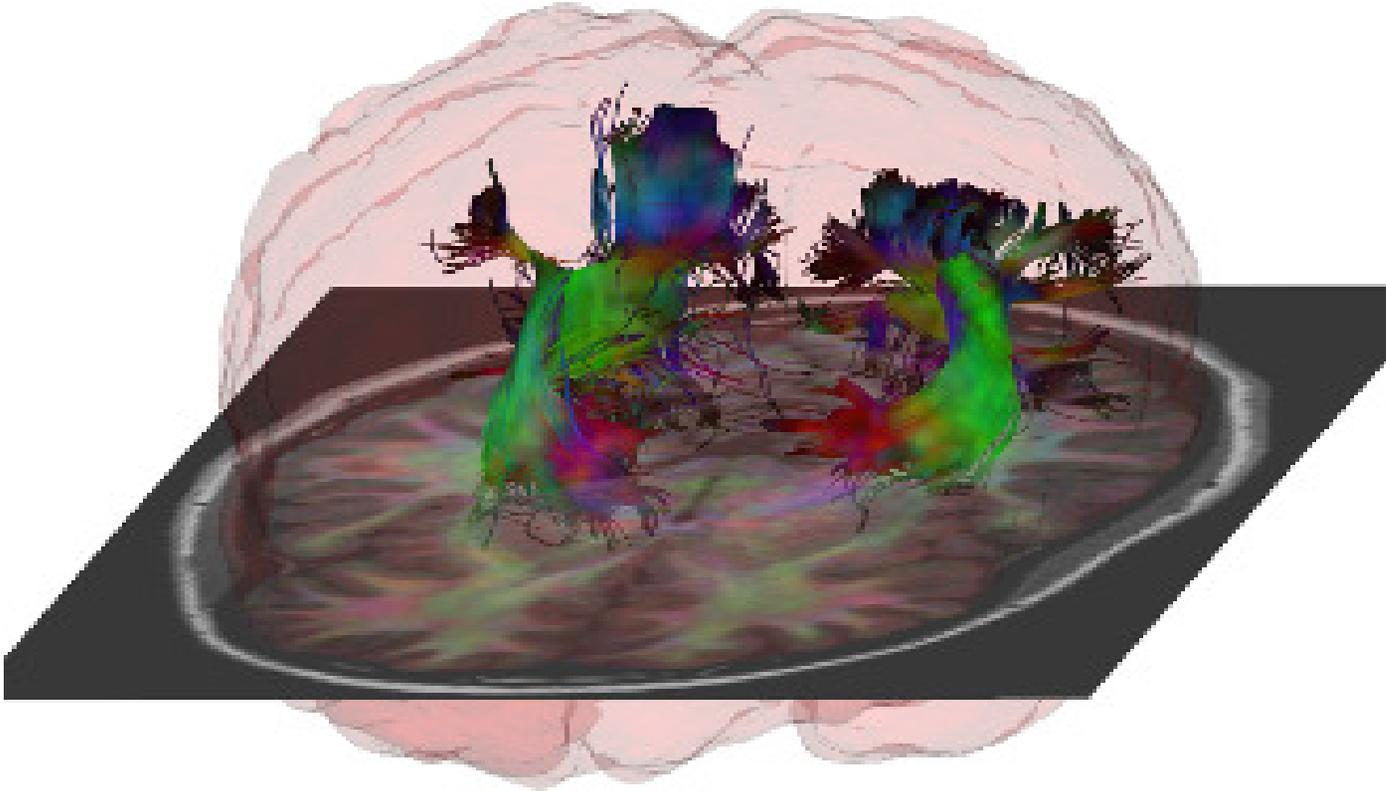


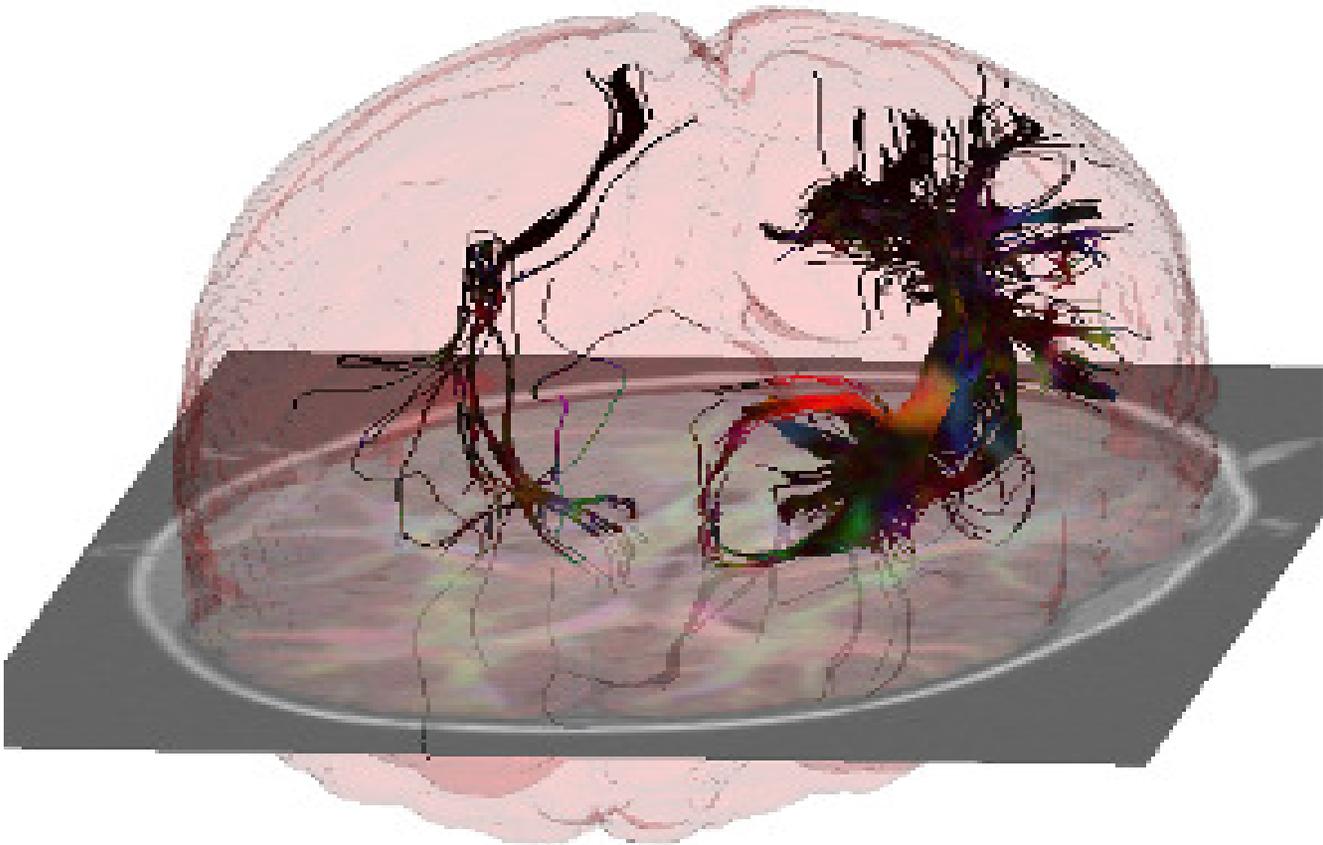
Some research suggests autism may be linked to several physical abnormalities within the brain. Example: A shortening of the brain stem that results in certain structures being bunched closer together, reducing some in size and apparently eliminating others.



Cellular brain development

1. Nerve cell migration
2. Axonal growth
3. Dendritic growth
4. Synaptic formation
5. Apoptosis
6. Myelination





Development of sensory systems

1. Tactile – 7 ½ weeks from exact date of conception; top and back of head are responsive
2. Vestibular – impulses projected to brainstem
3. Auditory – 6 months gestation, structurally equipped
4. Visual – retina and continues to mature by 30 weeks gestation
 - 22 weeks – evoked cortical responses
 - 24-29 weeks – eyelids open and close
 - 6 months – cones developed
 - Infant is blind at birth
 - At 2 years of age – sees 20/20 (like adult)/ visual system is fully myelinated

Normal development

- 3 week post delivery: touch, senses developed, position and taste
- 4 weeks post delivery: brain is divided into functional units

The Broader Phenotype

- Mild behavioral characteristics were described by Kanner (1943) and Asperger (1944)
- Asperger (1944) suggested a genetic mechanism (behavioral difficulties in fathers and sons)

The Broader Phenotype - continued

- Psychological and biological researchers have been preoccupied with seeking explanations for the most peculiar and distinctive aspects of autistic behavior
 - Milder phenotypes seen as “lesser variants”
- The milder phenotype may be seen as variable expression of a genetic susceptibility to autism and possibly other PDDs

Current View

- Happe (IMAR, 2008) argued that endophenotypes should be studied separately, as continua, in relation to imaging, biological, and genetic studies. Waterhouse said this many years ago.

Disorders That May Accompany Autism

- Intellectual disability
- Seizures
- Fragile X
- Tuberous sclerosis

When question #1

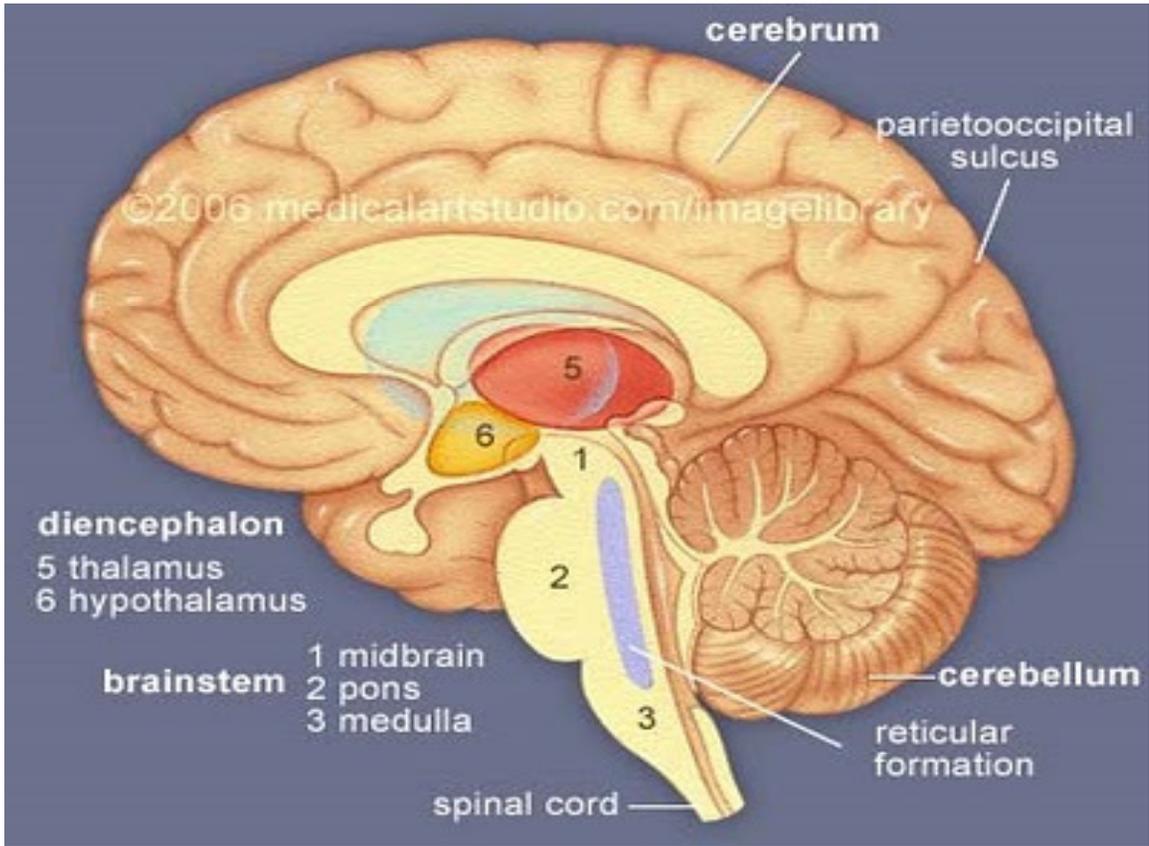
When did the insult occur?

- Lower level functions are more strongly related to the structure of the brain. It is more localized. Relatively less environmentally dependent – academics/learning
- Subclinical vs severe symptoms

Brain Insult

- For children, early injury results in generalized/global impairment in acquisition of developmental skills regardless of location of insult

Lower level functions	Higher level functions
• sensation	• perceptual/ motor
• motor	• language
	• memory
	• executive functions



When question #2 – Why now?

Developmental course of Autism

- **Infancy:** 50% of parents with children diagnosed with Autistic Disorder report they suspected a problem before 1 year of age
- In typically developing children imitation is demonstrated by age 1
- Sensory sensitivity (hyper/hypoarousal) is common
- Neurologically = increased hypotonia, hyporeflexia, stereotypies, increased EEG abnormalities with or without seizures

Abnormalities on the Neurological Examination and EEG in Young Children with Pervasive developmental Disorders,
Akshoomoff, N, et al. (2006).

- May be irritable
- May be excessively quiet
- May have difficulty with transitions

Developmental course of Autism - continued

- **Toddlerhood/early childhood:** children with autistic disorder often become more responsive to their name, but receptive/expressive language impairments become more apparent
- Most are not “aloof”, but have deficits in engaging with others in unstructured settings
- Positive prognostic predictors are: high IQ, language before 5 years
- Imaginative play is limited
- Deficits in social interaction may become more evident upon entry to school
- May exhibit poor sleep hygiene
- May demonstrate food selectivity
- May show an increase in temper tantrums

Developmental course of Autism - continued

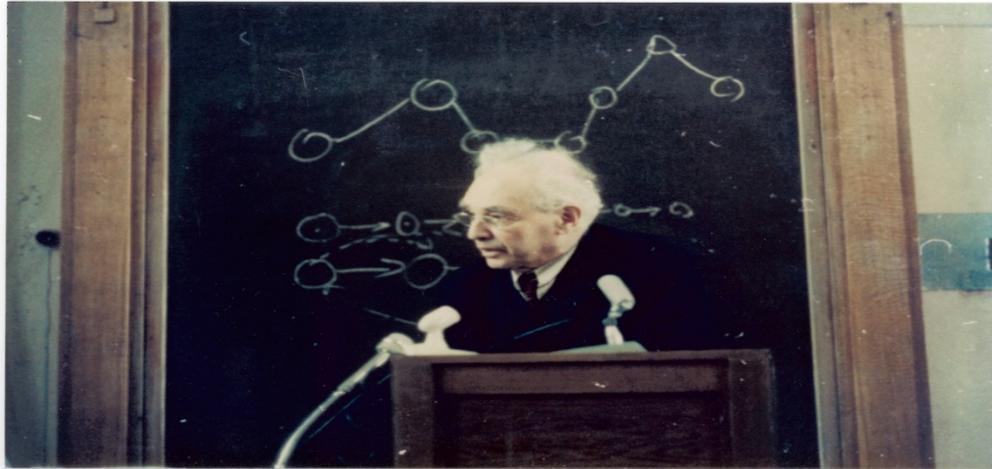
- **Middle to late childhood:** language may improve markedly
 - 33% of children have communicative speech by age 5
 - 50% by age 12
- Pragmatics of language may be impaired: humor, sarcasm, or inferential logic

Developmental course of Autism - continued

- **Adolescence and adulthood:** this is a time of increased social demands and high functioning or Asperger's Disorder may aspire to "fit in", but do not have the requisite skills to do so
- Prone to depression that can be manifested in:
 - Behavioral dysregulation
 - Increased level of obsessive/compulsive behaviors
 - Anxiety
- May be prone to victimization by peers

Arnold Gesell (1880-1961)

- Earned his Ph.D. from Clark University in 1906 and his M.D. from Yale University in 1915.
- Felt the evaluation of impaired children requires a firm foundation in typical development. He was instrumental in establishing the typical trajectory of development.
- Streams of development investigated were:
 - Gross motor
 - Visual motor
 - Language
 - Adaptation
 - Social
- Many of the techniques developed by Gesell were incorporated into developmental measures used today.



Alexander Z. Luria (1902 – 1977)

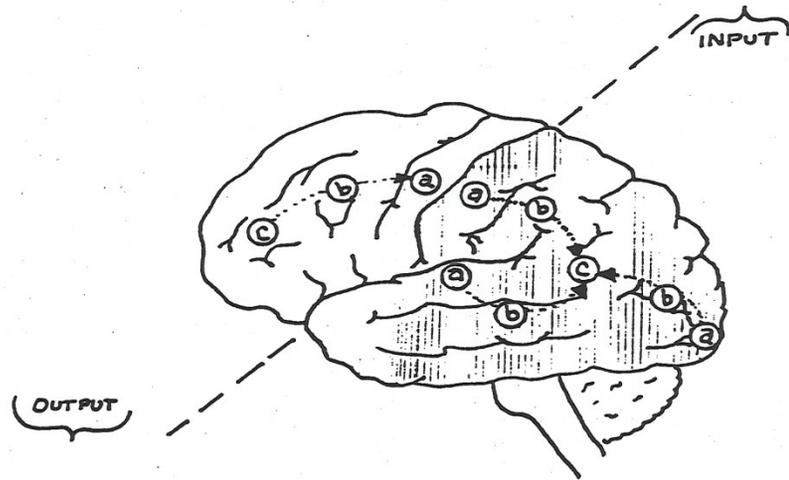
- Russian neuropsychologist/physician developed a system of neuropsychological assessment that aided in diagnosis and rehabilitation of brain damage, as well as in work-up with children
- Neuropsychological assessment evaluated information processing

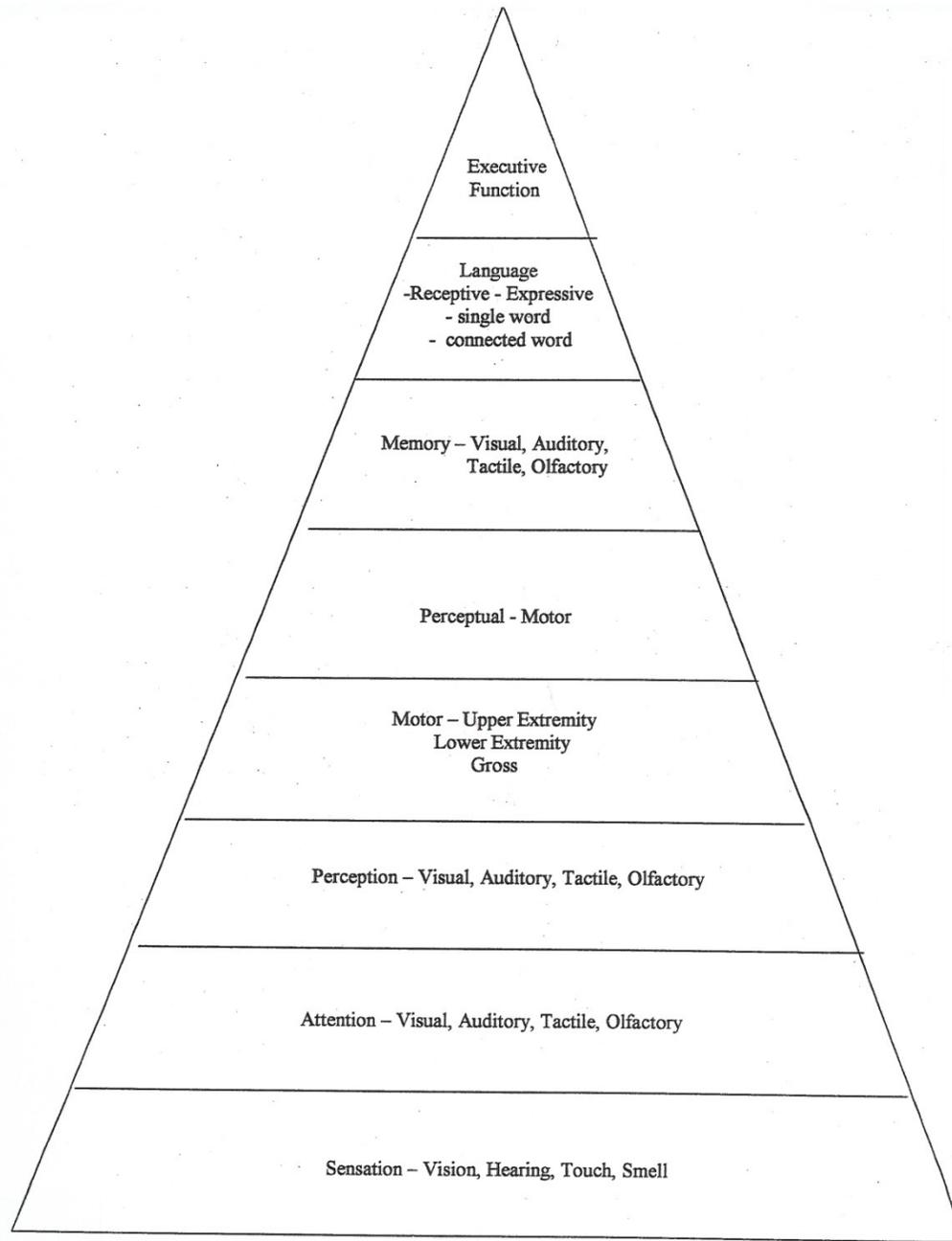
Luria systems

Arousal	Birth – 12 months
Primary motor and sensory areas	Birth – 12 months
Secondary motor and sensory areas	Birth – 5 years
Tertiary sensory input areas	5 – 8 years
Tertiary output/planning	12 – 24 years

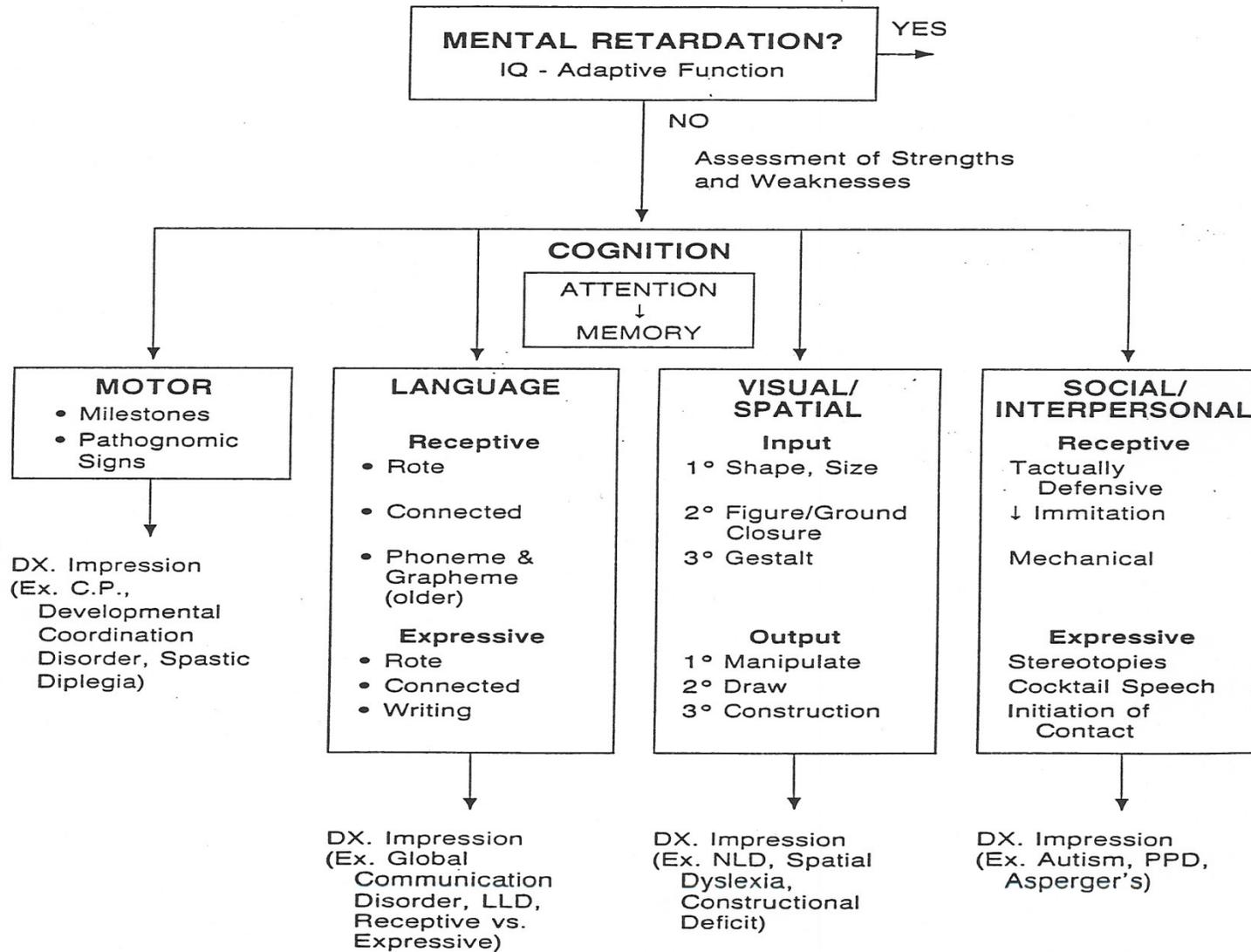
3 units of processing:

- 1) **Arousal (orientation) unit** - Attention
 - a. Brainstem-Reticular-Frontal-Thalamic (diffuse)
 - b. Filters/ selects sensory processing inputs (survival needs)
 - c. induces/suppresses arousal
 - d. augments/lowers functional arousal (social needs)
- 2) **Sensory input units** – modality specific, but composed of primary, secondary, and tertiary stages of refinement and integration. Posterior cortex.
 - a. Primary – reception, registration of elemental sensory data, some of may already be integrated (i.e. eye)
 - b. Secondary – integration, elaboration and perception of incoming input from primary centers. Checks memory for recognition of familiar.
 - c. Tertiary – cross modal integration located primarily at the juncture of the parietal, occipital and temporal lobes (PTO). This is where auditory, visual and tactile information is fixed and analyzed simultaneously.
- 3) **Output/planning unit** – the executive function responsible for initiating and modulating behavioral responses. Anterior cortex.
 - a. Primary - the motor output area where commands are sent from to engage specific muscles/muscle groups to perform any behavior
 - b. Secondary – organizes the sequences of motor acts, plans, and rewrite all necessary elements. Checks for recognition of familiar motor plans.
 - c. Tertiary - primarily the prefrontal lobes, responsible for decision-making, evaluation of ones actions, impulse control, focusing attention, flexibility, and emotional integration/control.





MODEL FOR PEDIATRIC NEUROPSYCHOLOGICAL ASSESSMENT



“No child is untestable”

Isabel Rapin, M.D.

- Need to know how to test skills at different levels of development
- Knowledge of cognitive development, social development, language development, etc.
- Validity and reliability
- Observation
- Patience is a virtue

ASSESSMENT OF THE SYNDROME

A. Historical Information

1. Early Developmental History
2. Nature/Type of Onset
3. Associated Medical Problems, Family History

ASSESSMENT OF THE SYNDROME - continued

B. Psychological/Communication Assessments

1. Verbal and Nonverbal Skills must be assessed independently
2. Assessment of Social-Adaptive and Social-Emotional Behaviors
3. Communication Evaluation (not just speech)

ASSESSMENT OF THE SYNDROME - continued

C. Behavioral Assessment

1. Nature of Social Relatedness
2. Behavioral Features
3. Play Skills
4. Family/Living Conditions

ASSESSMENT OF THE SYNDROME - continued

D. Medical Evaluations/Laboratory Studies

1. Associated Conditions
2. Genetic Screening
3. Auditory Testing
4. EEG/MRI/Neurological/other consultations as needed

Global Developmental Delay

- Definition – delay in 2 or more of:
 - Gross/fine motor activity
 - Speech/language performance
 - Cognitive ability
 - Social/personal interaction
 - Activities of daily living

Global Developmental Delay - continued

- Term used for children less than 5 years
 - (Varies by state)
- Thereafter, when I.Q. testing is reliable, the term is mental retardation
- Age specific deficits in adaptation and learning skills
- Not synonymous, nor always predictive of Intellectual disability

Global Developmental Delay - continued

- Incorrect diagnosis leads to inappropriate counseling, stigmatization, etc.
- Better to defer diagnosis than to arrive at an incorrect diagnosis.
- This does not imply corrective therapies should be delayed.

Intelligence

- Approximately 70% in Intellectually Disabled range
 - 30% Full Scale IQ = 51-69
 - 40% Full Scale IQ = < 50
- Often peaks in performance
 1. Block Design (for middle and older children WISC-IV)
 2. Digit Span
 3. Object Assembly (for younger children WPPSI-III)
 4. Matrix Reasoning
- Poor performance on
 1. Comprehension
 2. Picture arrangement
- VIQ < PIQ – difference tends to reduce in school age
- Intelligence is a significant factor in level of impairment

Sensation

- Low reaction to sounds/ intolerance for specific sounds
- Hypersensitivity to texture
- Selective food preferences
- High tolerance for pain
- Lines up objects
- Peripheral gaze

Attention

- Overfocused for preferred activities
- Impaired for non-preferred activities
- Shifting attention impaired
- Sentence memory < Digit Span (greater semantic salience)

Perception

Visual perception

- Insensitivity to complex motion
- Poor face processing
- Visual-spatial ability typically a strength
- Visual scanning approach is often disorganized

Auditory perception

- Comprehension of speech more impaired than in individuals with intellectual disability

Motor

- Hypotonicity resolves by early childhood
- Praxis-motor planning/ motor imitation often poor
- May have poor coordination
- Simple motor speed (tapping) intact
- Complex motor coordination (pegs) impaired

Somatomotor

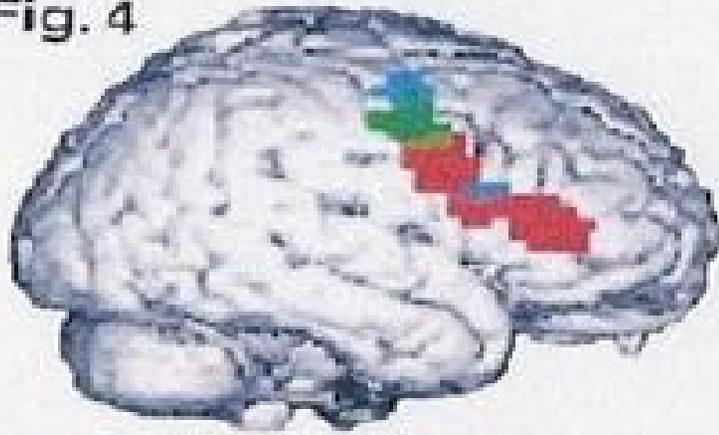
- Mirror cells
- Joint attention
- Imitation
- Emotional face processing



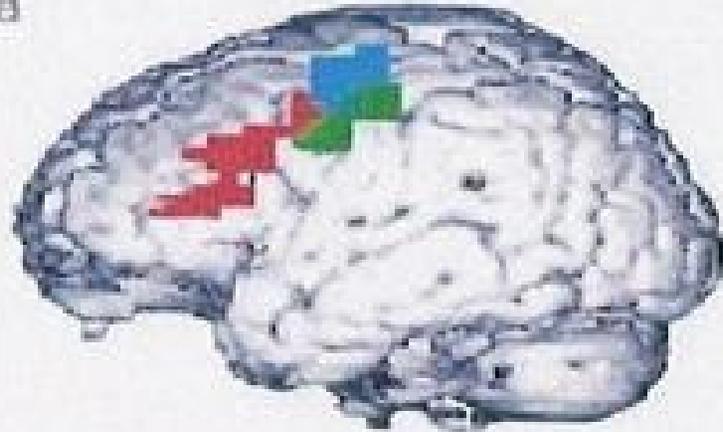
Viewer With Autism

Normal Comparison Viewer

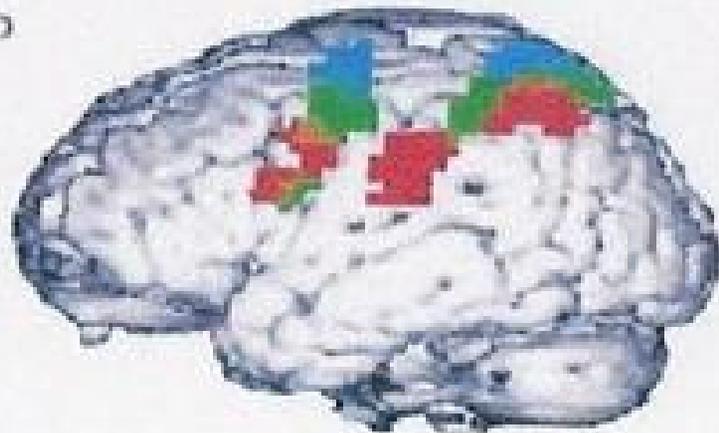
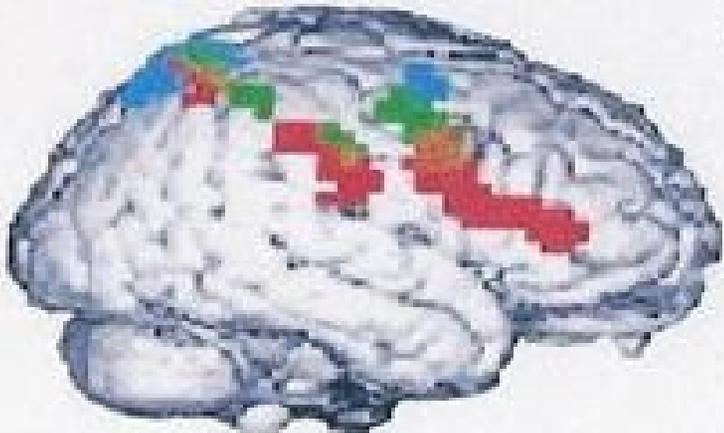
Fig. 4



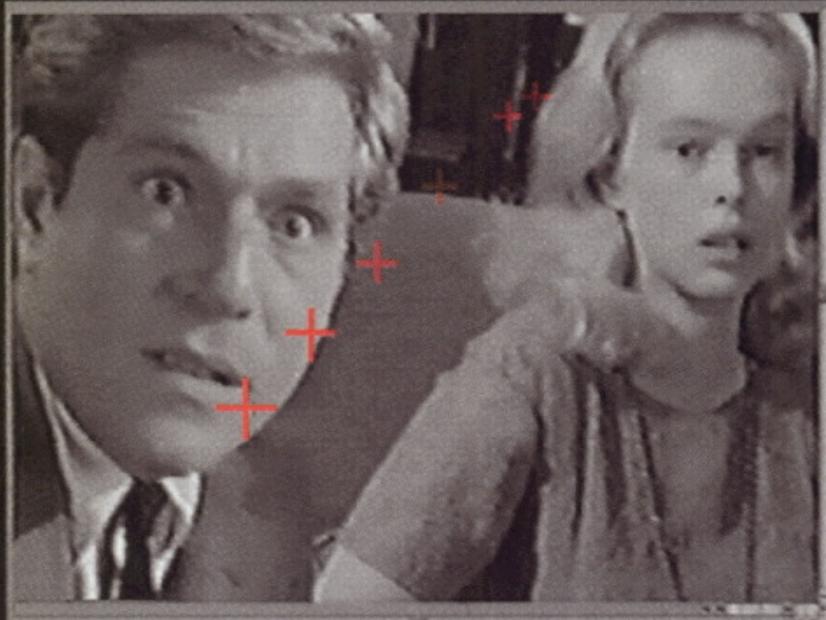
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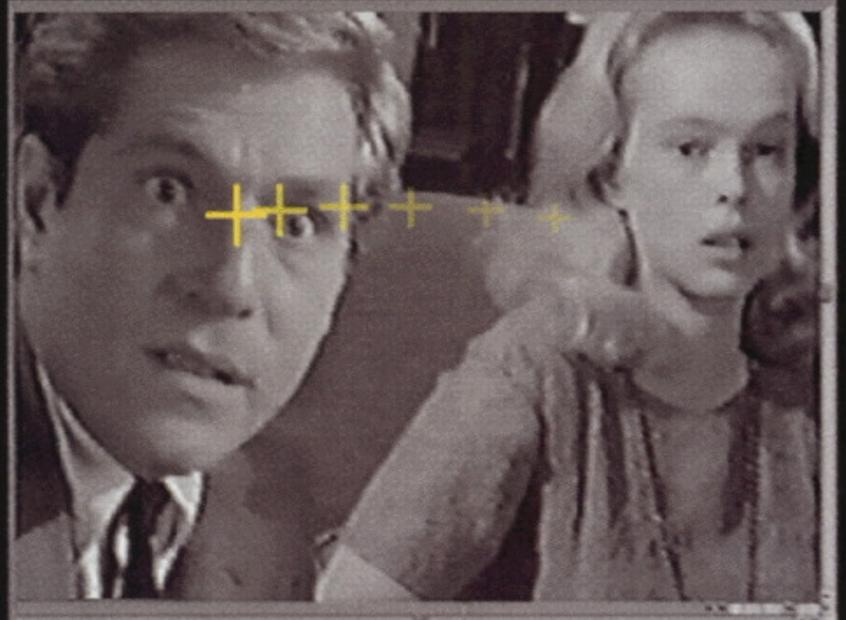
b



Viewer With Autism



Normal Comparison Viewer



Language

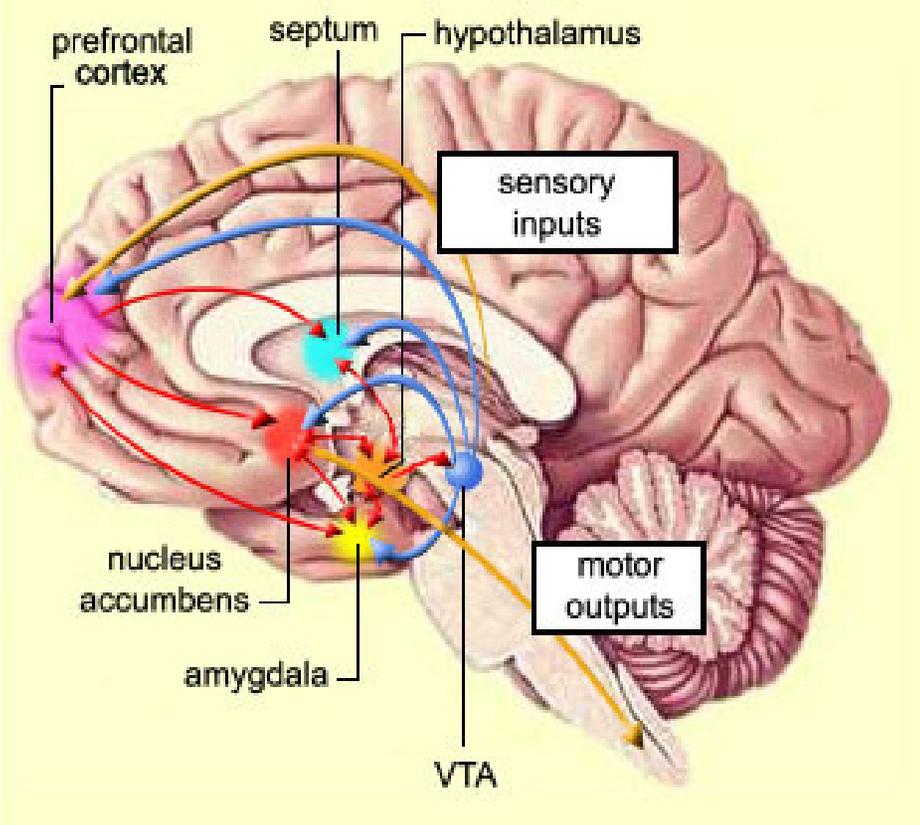
- 50% nonfunctional
- Low interest/attention to language
- Poor nonverbal communication; gesture
- Poor comprehension
- Echolalia persists – immediate/delayed
- Reduced vocalization
- Prosody impaired – flat, exaggerated, rate and volume may be abnormal
- Intact reading decoding/impaired reading comprehension
- Impaired pragmatics
 - Reduced eye contact
 - May intrude physical space
 - Insensitive to conversation partner's reaction
 - Impaired humor
 - Impaired appreciation of sarcasm
- Unusual word choice or overly formal language

Memory

- Encoding, storage and retrieval are relatively consistent with overall cognitive functioning
- Nonverbal memory strength/verbal memory poor
- Memory for events may be good
- Memory for emotional events poor
- Reduced semantic clustering
- Poor source memory

Executive function

- Impaired “top down” processing - frontal-striatal (cortical-subcortical) circuitry
- Limited planning/organization efficiency (Tower of Hanoi)
- Reduced mental flexibility – perseveration
- Reduced set shifting
- Impaired working memory
- Hypothesis testing often impaired
- Perseveration covaries with language functioning
- Disinhibition
- Stimulus bound behavior
- Poor generalization of learning



Social reciprocity

Characteristics include:

- Interpersonal relatedness – the capacity to coordinate effective perspective of others
- Imitation – skills needed to reproduce the actions of others
- Joint attention – the ability to coordinate one's attention with another person to an object or event
 - Felt to impact the ability to develop Theory of Mind (ToM) – the cognitive capacity to make inferences regarding the mental state of others

Academics

- Reading decoding usually intact
- Reading comprehension impaired - especially abstract, inferential
- Spelling often good
- Math highly variable
- Writing usually impaired, especially reports requiring organization

Social Cognition

- Impairment in:
 - Understanding subtle emotions
 - Matching affects to situations
 - Following another's gaze
 - Self and other face recognition
 - Abnormalities in face processing on fMRI
 - Recall of emotional stories
 - Generating scripts for familiar routines
 - Theory of mind
 - Humor/absurdities

