

Developmental Issues and Assessment

Application to Organic Acidemia
Patients



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What is Normal Development

- Series of processes
 - Learned skills
 - Language
 - Socialization/Culture
 - Less learned skills
 - More innate (inborn) component
 - Smiling/Responding to faces
 - Walking

Normal Development 2

- Systems
 - Brain
 - Peripheral nervous system
 - Musculoskeletal (muscles, joints and bones)
- Gradual Process
 - Patterns of expected gains
 - Not linear
 - Everyone does not achieve the same level in all areas of development

Areas of development

- Four major areas evaluated in the youngest child
- Gross Motor
- Fine Motor
- Language
 - Expressive = what is produced
 - Receptive = what is understood
- Personal-Social
- Cognition added for the older child
- Achievements are of importance to the individual, family and society

Gross Motor Skills

- Large movements
- Rolling over
- Sitting
- Crawling
- Standing
- Running
- Complex coordinated movements
 - Athletics, dance, swimming, bicycle riding



Fine Motor

- Reach toward object
- Touch/Bat object
- Attain (get)
- Manipulate
- Transfer between hands
- Pincer grasp = opposition of thumb and finger
- Tool/Utensil use
- Writing
- Video game



Expressive Language

- Cries
- Cooing
- Pre-verbal Noises = raspberry/Bronx cheer
- Nonspecific phoneme (single sound)
- Specific phoneme combinations
- Words
- Sentences
- More complex language
 - Plurals, tenses, and abstract concepts

Receptive Language

- Attend to surroundings non-specifically
- Attend specifically
- Respond
- 'No'
- Follow command
 - One step
 - Multistep
- Reading
- Interpretation



Personal-Social

- Smile
- Cry for wants
- Self-feed
- Interactive behavior
- Social compartment
- [toileting]
- Moral/ethical code
- Social integration



Development as a Learned Pattern of Behavior

- Imitation
 - Mimicry
 - Learning to wave
- Language
 - Infants make sounds used in all of world's languages
 - selected down to the sound set for the languages to which they are exposed
- Visual-spatial coordination
 - Hitting a ball
 - Dance
 - Video games
 - Computer use

Who teaches development

- Parents
- Other caregivers
- Teachers
- Therapists
- Any Role Model

Development in Organic Acidemias

- Organic acidemias can affect
 - Muscle and its use
 - Motor functions of the brain
 - Planning functions of the brain
 - Frontal cortex (emotions & social reasoning)
 - Language cortex
 - Visual cortex

Organic Acidemias Can Affect Development

- Muscle
 - Tone can be low = more power needed
 - High tone - interferes with movement
- Brain
 - Motor, accessory motor cortex and other structures
 - Plan movement
 - Initiate movement
 - Smooth movement = feedback process
 - End movement
 - May be at a level under consciousness
 - Frontal brain
 - Emotions and socialization
 - Language Cortex
 - Expressive or Receptive

Re-Learned Development

- Rehabilitation
 - Post – injury
- Not using a skill
 - Not riding a bike for a number of years
- Brain has plasticity
 - To a degree, the brain can recruit other pathways to learn a task
 - Peripheral nervous system has less plasticity
 - Some re-growth of original nerve possible
 - May never be as good as the original ‘wiring’
 - Some recovery after metabolic injury

Assessment of Development

- Informal Comparison
 - Historical
 - What was he doing then vs. now?
 - Contemporary
 - What is she doing compared to?
- Formal Assessment
 - Developmental rating on a test
 - Developmental Quotient
 - IQ = Intelligence Quotient

Why Assess Development?

- **Reassurance** of 'Normality'
 - Meeting expectations
- **Comparison**
 - To peers
 - To siblings
- Recognizing **Strengths and Weaknesses**
- **Planning** for Interventions

Planning for Interventions

- Ready for more complex task?
- Strengths
 - Absolute or relative
- Weaknesses
 - Improvement to a more functional level
 - Improvement to a supernormal level
 - Tutoring/coaching
 - Development of an alternative skill
 - Signing vs. speaking

Methods of Assessment

- History
- Observation during physical examination
- Screening Tests
 - Denver Developmental Screening Test
 - Goodenough-Harris Draw-a-Person
- Non-screening 'normed' tests
 - May require special equipment
 - May be weighted by one area of development
 - May require special expertise to administer or score
 - Appreciation for the variables in the test situation

Psychological Testing

- Cognition
- Personality
- Academic achievement
- Language development
- Motor development
- No single battery of tests fits all
 - Adaptation of test battery based on abilities
 - Valid, reproducible, normalized (standardized)



Testing Children

- Potential problems
 - Activity Level
 - Distractibility
 - Shorter attention span
 - Wariness of strangers
 - Inconsistent performance in a strange environment
- Requires
 - Time
 - Patience
 - Experience

Battelle Developmental Inventory

- Birth to eight years
- Individually administered assessment battery over 90 minutes
- **5 Domains**
 - Cognitive, Adaptive (self-help), Motor, Communication, Personal-social
 - Can be divided into sub-domains
 - Useful for determining eligibility for services
- Administered in three formats
 - Structured format
 - Parental interview
 - Observations

Stanford-Binet: 4th Edition

- 2 years to adult
- 15 sub-tests grouped into four areas
 - **verbal reasoning** (vocabulary comprehension, absurdities, verbal relations)
 - **abstract/visual reasoning** (pattern analysis/block design, form copying; matrices, paper folding and cutting)
 - **quantitative reasoning** (quantitative number series, equation building)
 - **short-term memory** (sentence memory, bead memory, digit memory, object memory)
- Each area yields a Stanford Age Score (SAS)
 - mean of 100 and a standard deviation of 15
- Composite Score (from all 4 areas combined)
 - mean of 100 and a standard deviation of 15.
 - Is equivalent to full scale IQ.

Kaufman-Assessment Battery for Children

- 2 ½ to 12 ½ years
- Multi-subtest battery
- Composite Mental Processing Score
 - calculated from Sequential and Simultaneous areas
 - A non-verbal score can be calculated from appropriate Mental Processing subtests
 - Separate percentile tables are provided for several socio-cultural sub-groups
- Child needs to be able to:
 - imitate the assessor
 - respond to pictures and cards with adult in a "testing" situation

Some Other Assessment Tools

- **Assessment of Motor and Process Skills**
 - Observational tool for goal-directed tasks of domestic and personal activities of daily living.
- **Bruininks-Oseretsky Test of Motor Proficiency**
 - Gross motor, upper limb, and fine motor proficiency in children 4.5 to 14.5 years of age.
- **Canadian Occupational Performance Measure**
 - Interview tool
 - Identify the family's priorities for their child with special needs
 - Aids in developing therapy goals with the child's primary caregiver
- **Childhood Autism Rating Scale**
 - Observational tool
 - distinguish those children with developmental delay from children with autism

- **Developmental Test of Visual Motor Integration - 4th ed.**
 - Identify visual-motor integration deficits in children ages 3 to 18 that can lead to learning and behavior problems
- **Developmental Test of Visual Perception – 2nd Ed.**
 - measures the visual perception and visual-motor integration skills in children 4 to 10 years of age
- **Evaluation Tool of Children's Handwriting**
 - Criterion referenced tool
 - evaluate the manuscript and cursive handwriting skills of children in grades 1 through 6
- **First STEP: Screening Test for Evaluating Preschoolers**
 - Screening tool for children ages 33 to 74 months who need in-depth diagnostic testing in the areas of cognition, communication, and motor development

- **Infant/Toddler Sensory Profile**
 - Judgment-based caregiver questionnaire
 - describe behavioral responses to various everyday sensory experiences in children from birth to 3 years of age.
- **Peabody Developmental Motor Scales – 2nd ed.**
 - Gross and fine motor skills from birth to 5 years of age
- **Pediatric Evaluation of Disability Inventory**
 - Functional abilities (self-care, mobility, social function) in children 6 months to 9 years of age
- **School Function Assessment**
 - Judgment-based questionnaire
 - Measure performance of functional tasks that support a student's participation in the academic and social aspects of an elementary school program
- **Sensory Integration and Praxis Tests**
 - Sensory integration processes that underlie learning and behavior in children 4 to 9 years of age.
 - assess visual perception, somatosensory and vestibular processing, and various types of praxis.

- **Sensory Profile**
 - Caregiver questionnaire
 - Measure the frequency of behaviors relation to sensory processing, modulation, and emotional responsivity to sensory input in children 3 to 12 years of age
- **Test of Visual-Motor Skills – Revised**
 - Measures eye-hand coordination skills needed to copy geometric designs in children 3 to 13 years of age
- **Test of Visual-Perceptual Skills – Non-Motor – Revised**
 - Assesses visual-perceptual skills (discrimination, memory, spatial relations, form constancy, sequential memory, figure-ground, closure) in children 4 to 13 years of age
 - Upper Level: 12 to 17 years of age
- **Toddler and Infant Motor Evaluation**
 - Measure neuromotor changes in children who have atypical development
 - appropriate for use with children who are functioning at or below 3.5 years.



Issues in Organic Acidemias

- Not a static process
 - eg. Progressive movement disorder in MMA
- Developmental perception
 - What do parents, teachers/therapists and physicians see as skills?
- Developmental Surveillance
 - Identify issues
 - Anticipate problems/barriers
 - Coordinate services
- No single point of entry into the medical system

Caregivers' Roles

- Understand current developmental state
- Support the child's effort
 - Positive reinforcement
- Promote success/approval
- Encourage normal/appropriate neurological development
- Collaborative management
 - It takes a team

Parental Worries

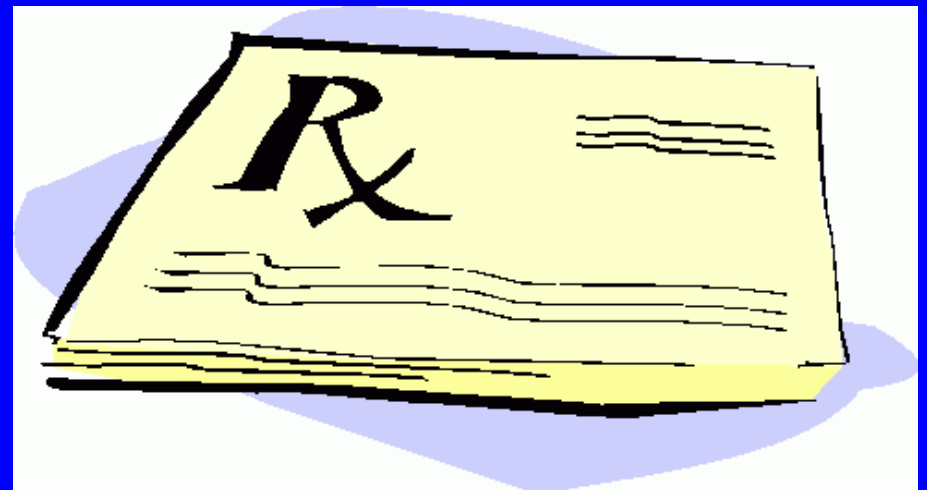
- Many worry about abilities and behaviors
 - Rapid change
 - Patterns may vary and be normal
- Recognition of vulnerability
- Work with the other members of a team
 - Ask questions
 - Find resources

Goals

- Motor
 - Community walking
- Effective communication
 - Needs, desires, emotions
- Societal interaction
 - Self-help skills
 - Monetary skills
- Executive processing = Pre-planning
- Societal acceptance
- Transitioning/increasing responsibilities
- Independence

A therapist's prescription

- Diagnosis
- Precautions
- Type of Therapy
- Frequency of Therapy
- Goals/Objectives
 - Anticipated
 - Realistic
- Duration



Barriers to Progress

- Lack of therapists
- Schools not appropriate for needs
- Money/Insurance
 - Public, 3rd party (commercial), or private (self-pay)
- Time

- Social-cultural issues
 - Acceptance of developmental status
- Environment: chaos or nurturing

The Child - Slowing Progress

- Motivation
- Endurance
- Acceptance of change
- Appropriate socialization

- Appropriate goals
- Appropriate assessment

What about other children?

- Normally developing kids
 - Dislike aggressive playmates
 - Have few positive playmate preferences re developmentally delayed
 - Generally do not automatically reject a developmentally delayed playmate
- Teach other children to accept
 - Bias is learned
 - One role of school rather than home education

Some other issues

- Parents do age
 - Enough years to achieve goals
 - Planning for parental absence
 - Issue especially for the non-independent child
- Involvement of siblings
 - Appropriateness of involvement
 - Ability of sibs to care for a developmentally challenged individual
- Needs of a family as a social unit

Review / Summation

- Development is a series of complex skills
- Learned or Modeled
- Dynamic Process
- Requires Assessment
- Intervention
- Involvement of family, school and medical community
- Goal of highest possible functioning