Developmental Delay

Recognition, Assessment, and Management

Introduction

■Children are not *"miniature adults"*

■They are dynamic organisms changing constantly not only in terms of physical growth but also in the other areas of development

■We expect them to make progress in their development according to schedule

■However for some, this does not happen and delays are observed

Introduction

■Developmental delay is a problem not infrequently encountered

▪1%-3% of all children are globally developmentally delayed

▪16% of all children have learning difficulties, and other developmental and behavioral problems

▪Only 20%-30% are detected prior to school entry

■Thus, routine screening is recommended as part of health maintenance visits

Introduction

■Low-frequency, high-morbidity disabilities such as CP, MR, sensory impairment and autism are likely to be identified in the preschool years

■Whereas high-frequency, low-morbidity disabilities like LD and ADHD are likely to be more recognized in school-age children

Introduction

■The acquisition of various developmental and intellectual functions occurs gradually

■This indicates neuromaturational processes occurring in the CNS

■An individual is conceived with a predetermined number of neurons (approximately 100 billions)

■Neuronal migration and organization are almost complete at birth

Introduction

■Brain growth in humans is largely a postnatal event, brought about by myelination, increase in glia, and elaboration of dendritic and axonal processes

■Normal neuromaturation requires an "ideal" environment free from noxious or adverse effects that may seriously affect the ultimate structure of the CNS

■Various areas in the CNS are endowed with specific functions, but with considerable overlap

Introduction

■The acquisition of the various milestones (i.e., gross and fine motor, personal and social, and language) occurs hand in hand

■With time, more complex functions are increasingly acquired

■Deviation on development especially delay should be taken seriously

CNS Plasticity

■Potential for recovery in children is significantly greater than the mature brain
■An innate property of the brain, not compensatory
■Brain injury that happens prior to the acquisition of function leads to no deficit, but in time delays are seen
■Brain injury that happens after the acquisition of function leads to a deficit similar to the adult, but is generally transient (<10 years)

Early Identification

■Early identification of developmental problems is important because of the potential for improvement through education and habilitative services
■Mode of presentation of developmental delay is age-dependent
■Most common presentation: Failure to achieve age-appropriate skills

Mode of Presentation

■First months: Poor suck, hypotonia or hypertonia, lack of visual or auditory response
■Through 18 months: Motor delays (gross and fine/adaptive)
■≥2 years: Speech delay and aberrant behavior
■School-age: Academic underachievement and ADHD symptomatology

Normal Motor Development
Adapted from DDST
Cephalad-caudad Progression

■Head control
▪Prone up to 45^{o} 2-2.5 months
▪Prone up to 90^{o} 2.5-3 months
▪Sits, head steady 3.5-4 months
■Sits without support 6.5-8 months
■Stands/walks alone 13-15 months
■Walk up steps 21-22 months

Milestone Guideline
Gross Motor

■Function	Expected Age	Referral
■Head control	2 mos	4 mos
■Sitting	6 mos	8 mos
■Crawling	9 mos	12 mos
■Walking	12 mos	18 mos

Delayed Motor Milestones
Causes

■Systemic diseases (acute and chronic)
■Cerebral palsy
■Spinal defects
■Muscle disorders
■Non-specific global delay (mental retardation)

Delayed Motor Milestones
Causes
- Familial
- Environmental (lack of stimulation)
- Non-neurologic disorders of joints, ligaments and bones
- Sensory deprivation (e.g., blindness)
- "Benign" hypotonia – (not entirely benign)
- Unknown

Milestone Guideline
Fine Motor/Social

Function	Expected Age	Referral
■ Smiling	6 weeks	3-4 mos
■ Reaching	4 mos	5-6 mos
■ Transfer	6 mos	8 mos
■ Pincer	8 mos	>12 mos

Common Patterns of Behavior Necessitating Referral if Persistent
- Hand regard
  - Emerges: 12 wks
  - Disappears: 6 mos
  - Referral: 6 mos
- Mouthing
  - Emerges: Birth
  - Disappears: 13 mos
  - Referral: 18 mos
- Casting
  - Emerges: 12 mos
  - Disappears: 18 mos
  - Referral: 20 mos

Language Development
- Two most important requisites for the acquisition of language
  - Intact hearing (***must*** be documented through hearing testing)
  - Sufficient intelligence

Language Development
Characteristics
- Normal
  - Single most important intellectual accomplishment <5 y
  - Only public form of cognition in the early years; window to the developing mind
  - Best predictor for cognitive functioning
- Delay
  - Predictor of intellectual retardation
  - Strong predictor of later learning disabilities
  - May persist through the school years

Milestone Guideline
Language

Average age (mos)	Observed behavior
6-10	Canonical babble (dada, mama)
9-10	Word comprehension
12-13	Word production
18	Vocabulary burst
20	Word combination
24-36	Grammatization (>3 word phrases)

Early Signs of Impairment
Language Development

Average age (mos)	Observed behavior
■ 11-12	No giving, showing or pointing
■ 18	Comprehends <60 words; produces <5 words
■ 24	Produces no 2-word phrase

Language Delay
Causes
■Mental retardation
■Hearing loss
■Developmental "aphasia"
■Autism
■Psychosocial deprivation (lack of stimulation)

Academic Underachievement
Causes
■Mental retardation
■Specific learning disabilities
■Below Average IQ
■Neurobehavioral disorders (e.g., ADHD)
■Sensory deprivation (e.g., blindness, deafness)
■Psychosocial factors (emotional, dysfunctional families)

Reading Problems
Causes
■Mental retardation
■Specific learning disabilities
■Psychosocial factors (emotional, dysfunctional families)
■Poor teaching methods
■Visual, auditory and spatial difficulties

Mental Retardation
Definition
■Subaverage intellectual functioning, the deficit has resulted from an injury, disease, or an abnormality in the brain that existed before age 18 years, together with difficulty adapting to the environment
■IQ <75
■Static progress (distinguish from dementia, which is progressive intellectual deterioration)

Mental Retardation
Definition
■Definition insists that the brain dysfunction has occurred during the pediatric age group
■Intelligence is inferior to peers, but will show progress with age and training
■Progress may come to a "halt" when the patient has reached maximum potential – repeat psychometrics may show lowering of IQ; not necessarily indicating deterioration

Mental Retardation
Causes
■Most frustrating for MDs to undertake
■Mild MR are particularly difficult; some are familial, and may be influenced by low

SES
■Severe MR – greater chance in identifying a cause
▪Chromosomal aberration – most common
▪CNS injury (pre, peri, and postnatal)
▪CNS anomalies
▪Dysmorphic syndromes
▪Endocrine and other metabolic problems

Pseudo Retardation
Causes
■Inadequate exposure to a culture for which the test is formulated (e.g., language, migration)
■Environmental understimulation (e.g., low SES, lack of schooling, illiteracy, etc)
■Emotional disturbance
■Chronic illness (academic underexposure)
■Seizures
■Motor handicaps (e.g., CP)
■Sensory deprivation (blindness, deafness)

Developmental Assessment
Indications
■When the parents express anxiety about the child's progress or behavior (parents' complaints **MUST** not be ignored)
■When the clinician notices some delay or deviation of development, or persistence of a finding expected to have disappeared

■When the child has a known disability

Developmental Assessment
Plan
■Detailed history (provides ample data to suspect previous insult to the CNS)
■Clinical tests for hearing and vision
■Physical and neurological examination
■Developmental assessment (use any of the developmental screening tests)
■Laboratory and radiographic investigations
■Psychometric evaluation

Developmental Assessment
Screening Tests
■Identifies 70% of abnormal children
■Identifies 90% of normal children
■Caveat
■10% of normal children are initial labeled abnormal
■30% of abnormal children are labeled normal

Developmental History
■Delayed or uneven acquisition of milestones (cognition, speech and language, gross and fine motor, adaptive skills) calls for further evaluation
■Parental assessment is extremely important as parents are the best monitors
■Utilize immunization booklets with various developmental milestones
■Parental education starts before the baby's discharge from the nursery
■Track and review progress during well-baby visits

Physical Examination
■Growth abnormalities (height, weight, head circumference)
■Major and minor congenital anomalies (dysmorphism)
■Skin findings (neurocutaneous signs)
■Organomegaly
■Eye findings (cataracts, corneal opacities, etc)
■Others

Hearing Screening
■Hearing should be tested especially when there is no response to auditory stimuli and/or the child is speech delayed; also very important in children with neonatal hyperbilirubinemia, sepsis (ototoxic antibiotics) and post-meningitis
■BAER and/or Behavioral Audiometry

Vision Screening
■Visual alertness, tracking, visual fields, picking small objects
■Referral to a Pediatric Ophthalmologist for children at high risk for visual problems (e.g., prematures, CP, genetic syndromes, etc)
Neurologic Examination

■Standard exam: normal for most children but identifies gross neurologic impairment (e.g., CP)
■Primitive reflexes disappear <6 months
■Postural equilibrium reflexes and protective responses emerge >6 months
■Asymmetry of tone & function, persistence of primitive & non-emergence of postural reflexes suggest CP
■Review of the progression of motor milestones is a useful tool to help distinguish delay from disability

Developmental Screening
■<6 years:
▪Denver II
▪Goodenough Draw-A-Person
■>6 years:
▪WRAT
▪Goodenough Draw-A-Person
▪Wechsler's Verbal items (General Information, Similarities, Vocabulary)
▪Bender-Gestalt or Beery-Buktenika
▪Peabody picture vocabulary

Developmental Screening
■Highly influenced by the well-being of the child:
▪Defer assessment in a sick child
■Highly affected by environmental factors (e.g., prolonged hospitalizations, dysfunctional families, etc)
■Significant factor in developing countries: low socio-economic status
Developmental Screening
■Calculate "DQ" in each area
■DQ = DA/CA x 100.
▪Should approximate 100
▪Not an IQ score
▪If there is a significant difference in scores (i.e., >10), the depressed score is usually the primary deficit

Correction for Prematurity
■No exact guidelines
■Up to 1 year: Correct by subtracting the number of months of prematurity from chronologic age
▪e.g., 7 months CA, 2 months premature
7 mos – 2 mos = 5 months (corrected age)
■Between 1 and 2 years: Correct by 50%
▪e.g., 18 mos CA, 2 months premature
18 mos – 1 mo (50%) = 17 months (corrected age)
■After 2 years: No correction

Ancillary Testing
■Not all developmentally delayed children need to have costly laboratory and radiographic examinations
■Tests often requested: Chromosomal analysis, metabolic screening, EEG, CCT and head MRI
■Nothing compares with a thorough history, physical and neurologic examinations and developmental screening

Chromosomal Analysis
■Children with major and multiple minor anomalies
■Not necessary in Down syndrome unless the mother is young
■Remember! Not all cells manifest the abnormality (>20 cells should be studied)
■Newer and more modern procedures may not be available

Electroencephalography
■Epilepsy in general population: 0.5%
■Epilepsy in developmental disabilities: 12% to 58%
■All types of seizures are reported in DD
■Specific syndromes like Lenox-Gastaut and West syndromes are associated with MR/DD
■Unless seizures are present or strongly suspected, EEG is of little value and should not be routinely requested

Cranial CT Scan
■CT provides excellent resolution of intracranial contents with the ability to image bony structures and intracranial calcifications
■Less expensive
■But –
▪Entails radiation
▪Not ideal for lesions in posterior fossa
▪Crude image

Cranial MRI
■Delineates more delicately the intracranial structures (demarcates GM and WM)
■Obtained in many planes without radiation
■Eliminates bone, and ideal for posterior fossa structures
■Inferior to CT in intracranial calcification
■Very costly

Cranial CT and MRI
Who Needs Them?
■No definite answer
■Indications not precise
■When there is a need for diagnosis, genetic counseling and possible treatment
▪Congenital infections, neurocutaneous syndromes, craniofacial dysmorphism, macrocephaly, HIE, ICH, postmeningitis

▪Sudden change in behavior and cognition, head size, motor function and new onset seizures

Metabolic Screening
■Far from ideal as only a few disorders are tested
■Negative results gives false sense of security
■Very low yield
■Should not be routine
■Specific testing should be considered in -
▪Deterioration in clinical status
▪Myoclonic seizures
▪Seizures refractory to medical treatment
▪Family history of similar problems or early demise

Overall Management
■Cornerstone for any DD is the therapeutic preschool
■Early identification means early intervention
■Management depends on the specific delay
▪Gross motor delay with abnormalities of tone: PT & OT
▪Fine motor/adaptive delay: OT
▪Speech delay: Speech & language therapy
▪Learning disability: Appropriate class placement
▪Behavioral problems: Counseling, behavioral modification
▪Global delay: Multidisciplinary approach

Medical Management
■AED for seizures
■Stimulants for ADHD
■Replacement therapy for specific deficits (e.g., thyroid hormone)
■Dietary regimen for specific disorders (e.g., PKU)
■Psychopharmacology for severe behaviors (may need assistance of psychiatrist)

Conclusions
■Developmental screening should be performed as part of health maintenance visits
■Early identification and parental acceptance are crucial to the management
■With early implementation of appropriate intervention, changes are bound to happen for the better, and may make these children functional and responsible adults!