Biopsychosocial perspective

3 components of all behavior presentations:
- Biology (genetic endowment)
- Psychology (internal mental processes; monitor and expression of emotions)
- Social (contextual: family, peers, school and community)

Refusing to speak in school (#1)

- 4 ½ yo girl: her preschool teacher reports that Kira is not talking to her teacher or other children at school
- She plays quietly and interactively and she is achieving all early educational goals
- Kira’s mother finds this odd in that Kira talks to her parents and 2 older siblings at home

Refusing to speak in school

- Pre-natal and post natal medical and developmental history is normal. She spoke several words at 12 months and used phrases at 24 months. Her speech is 100% intelligible at home.
- Kira’s parents describe her as shy and prefers to play by herself or with one child at a time.
- Family history of OCD and social anxiety

Specific Anxiety Disorders

- Separation Anxiety Disorder
- School Phobia (School Refusal)
- Selective Mutism
- Simple Phobias
- Social Anxiety Disorder
- Panic Disorder
- Obsessive Compulsive Disorder
- Post Traumatic Stress Disorder
Specific Anxiety Disorders

- Separation Anxiety Disorder
- School Phobia (School Refusal)
- Selective Mutism
- Simple Phobias
- Social Anxiety Disorder
- Panic Disorder
- Obsessive Compulsive Disorder
- Post Traumatic Stress Disorder

Selective Mutism

- Inability to speak in specific social situations despite speaking at home lasting at least 1 month (excluding 1st month of school or new child care)
- An expression of social anxiety in an extremely shy child
- Prevalence 0.1-0.7%

Selective Mutism

- Family history of social phobia (70%) and selective mutism (37%)
- Speech and language delays: 20-30%
- Bilingualism: immigrant children may refuse to speak to strangers in a new environment (to be distinguished from selective mutism); if prolonged, consider selective mutism or a language delay
Selective Mutism: Treatment

- Goal: treat anxiety and improve self-esteem rather than force to speak
- Behavior therapy: positive reinforcement for communication
  Non-verbal (smiling and pointing)
  Verbal (whispering to friends or adults)
- Cognitive-behavioral therapy
- Medication: SSRI (2 RCTs with fluoxetine)

10-month old w/ motor, language and social development delay (#2)

- Normal prenatal and perinatal history
- Unable to sit w/o assistance
- Absent pincer grasp
- No audible language
- Unilateral exotropia
- Microcephaly
- Poor reciprocal social responses but smiles frequently
- PATTERN RECOGNITION

Infant w/ motor, language and social development delay

- Early feeding problems
  (poor suck, reflux)
- Truncal hypotonia
- Tongue thrusting
- Wide mouth
- Prominent chin
- PATTERN RECOGNITION
Angelman Syndrome

- Features with broad diagnostic possibilities
  - Global developmental delay
- Focal features associated with specific dx.
  - Microcephaly
- SMILING in an aphasic, severely developmentally delayed young child

PATTERN RECOGNITION

Angelman Syndrome

- “Happy puppet syndrome” (1965)
- Deletion in the 15q11-q13 region (majority)
  - Absence of maternal gene in this region
- Mental retardation (severe)
- Seizures < 3 years
- Ataxia
- Hand-flapping

15 month old boy with ataxic cerebral palsy, microcephaly and seizures

Angelman syndrome
20-month old with expressive language delay (#3)

- Only 5 words at 20-months
- Normal prenatal and perinatal events
- Normal hearing screen at birth
- Normal motor milestones
- Normal neurological exam
- Head circumference: 25th percentile at birth
  75th percentile at 20mos.

Language Delay
Differential Diagnosis

- Hearing loss
- Psychosocial deprivation
- Autistic Spectrum Disorder
- Global cognitive delay (MR)
- Apraxia
- Expressive vs. Receptive Delay
- Maturational language delay
  Developmental Language Disorder

Language Milestones

Consider Referral If Not Met

- Newborn: turns to soft voice
- 3-months: Cooing sounds (alert, interactive)
- 6-months: Coos/jabbers; turns to new sounds and familiar voices
- 9-months: Babbles “mama, baba;”
  Knows name, turns when called
- 12-months: Points to objects; gives or shows objects;
  One word in addition to “mama/dada;”
  Follows one-step command
Language Milestones
Consider Referral If Not Met

- 18-months: Produces 5 or more words
- Comprehends more than 50 words
- 2-years: Produces >50 words
- 2 words together/follows 2-step command
- Points at picture book
- Uses words to request

ELMS: Early Language Milestone Scale (0-36 mo)
Modern Educational Corp, PO Box 721, Tulsa OK 74140

Challenge to Pediatricians:
Autism presenting as language delay

MCHAT: Modified Checklist for Autism in Toddlers.

www.dbpeds.org (Search MCHAT)
MCHAT
18-24 month Autism Screen
23-question parent form

6 Critical Questions:
1. Does your child take an interest in other children?
2. Does your child ever use his/her index finger to point, to indicate interest in something?
3. Does your child ever bring objects over to you to show you something?
4. Does your child imitate you?
5. Does your child respond to his/her name?
6. If you point to a toy across the room, does your child look at it?

4-year old boy (#4)

- Fatigue for 2 months
- Decreased interest in play

4-year old boy (#4)

- Additional history:

  Review of the medical record revealed that he walked at 18 months with normal language, social and fine motor developmental milestones
4- year old boy w/ fatigue

- PE normal
- CBC and ESR normal
- Urinalysis normal
- Chem panel normal except
  - AST=112
  - ALT=230

6 yo boy: prolonged crying every morning before school (#5)

- Awakens with crying that persists through breakfast and often until he is taken to school
- 1st grade, new school; did well in kindergarten
- Family moved to new city 2 months prior to onset of crying. He left 2 close friends. Father busy with new job while mom is caring for 2 younger children and establishing family in new home without her own social supports
Insidious behavioral changes in some neurological disorders

- Personality change
- Irritability
- Disruptive outbursts
- Sadness
- Social withdrawal
- Drop in school performance

Neurological conditions with early behavior change

- Brain tumor
- Metabolic disorders
  - Wilson’s disease
  - Thyroid dysfunction
- Toxic encephalopathy
  - Lead
  - Carbon monoxide
- CNS infections
  - TB meningitis
  - HIV encephalopathy
- Degenerative disorders
  - Adrenoleukodystrophy
  - Subacute sclerosing panencephalitis
Brainstem glioma: 2 phases of behavioral alterations

1st phase: Withdrawal, apathy, lethargy
Cries easily
Decline in school performance

2nd phase: Hyperactivity
Aggression
Nightmares


Pathologic Laughter and Crying

- PLC due to damage of pathways arising in motor areas of cerebral cortex and descent to brainstem to inhibit a putative center for laughter and crying
- Disinhibition (or release) of the laughter and crying center
- PLC lesions occur in the cerebro-pontine-cerebellar pathways
- Cerebellum modulates laughter or crying in context of situational and cognitive events.


At the end of an uneventful 12 month well child visit: (#6)
“By the way….her left foot turns out.”
The ultimate out-the door statement
Syringomyelia

- Definition - A central cavity in the spinal cord lined with altered glial cells.
- Localized or multi-segmental
- Syringobulbia
- Associated lesions: Intramedullary tumor
  - Spinal cord trauma
  - Arterial insufficiency
  - Developmental anomaly
Recent experiences with look-alike cases of ADHD (#7)

- Obstructive sleep apnea
- Moderate to severe allergic rhinitis: poorly controlled due to under treatment
- Adrenoleukodystrophy
- Marfan Syndrome
- Velocardiofacial syndrome (22 q deletion)

Velocardiofacial syndrome (22 q deletion)

First Visit History (#8)

- 16 y.o. white male
- meds=minocycline, paroxetine
- 2 yr h/o depression/anxiety
- denies substance abuse
- 75% Ht, 40% Wt
- RN BP 140/88, rpt 130/80
- incidental ganglion cyst, hydrocele
**Do We Have a Problem?**

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**Paroxetine Side-Effects**

- Serious Side Effects
  - Withdrawal Syndrome
  - Serotonin Syndrome
  - Mania
  - Seizures
  - Hypoanesthesia
  - Bleeding
  - Extrapyramidal symptoms

- Common Reactions
  - Anxiety
  - Sweating
  - Sexual dysfunction
  - Tremor
  - Constipation
  - Appetite Changes
  - Flatulence
  - Blurred vision
  - Nausea
  - Somnolence
  - Headache
  - Insomnia

**Laboratory Studies**

- Nm1 Electrolytes
  - No renal failure
  - No Hyponatremia
  - No Suggestion of Adrenal DIsorders

- Nm1 Cholesterol
  - No Nephropathy
  - No accelerated atherosclerosis

- Nm1 CBC
  - No polycythemia/thrombocytosis

- Nm1 Thyroid Function
  - No hyperthyroidism
  - Some evidence vs autoimmune proto

- Other tests that could be helpful
  - UA & CX
  - EKG
  - ESR
  - Drugs Abuse Screen
Follow-Up Visit

- RN BP 138/96
- MD BPs 135/80, 135/85
- Home Log ave 132/75
- Increasing Generalized Anxiety
- Episodic “Panic Attacks”
- Paxil increased 2 mos ago

Surgery for Hydrocele

- Hypertension: BP 260/110
- Nml 4 extremity BP
- EKG nml
  - No LVH/Dysrhythmia
- Drug Tox positive for THC only
  - Suggestive of no substance abuse that may cause hypertension

Differential Diagnosis of Hypertension

- Renal
  - Glomerulonephritis
  - Polycystic
  - Scar
  - Any Failure (volume overload)
  - TCC
  - Obstructive Uropathy
  - Renal Artery Stenosis/Thrombosis
- Endocrine
  - Cushing's DZ
  - Congenital Adrenal Hypoplasia
  - Primary Hyperaldosteronism
  - Apparent Mineralocorticoid Excess
- Tumors
  - Phaeochromocytoma
  - Neuroblastoma
  - Wilms

- Cardiovascular
  - Vascular
  - Obstructive

- Ingestions/Drugs
  - Cocaine
  - Amphetamines
  - Inhalants
  - PCE
  - Anabolic Steroids
  - ETOH withdrawal
  - L-dopa, methyldopa
  - Guanfacine
  - Captopril withdrawal
  - OCPs
  - Lead intoxication

- Other
  - Cystinoma
  - BPO
  - Tumor’s extent
  - William’s (ren art sten)
  - LA VSD
CASE PRESENTATION

- 4cm mass at left renal hilum
- 1cm mass to right of aorta

References

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