1 Common neurological problems in children and how they may affect learning and behavior

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2 Disclosures, and background

- No industry speaker's bureaus, consulting, or other financial relationships are relevant to this talk
- As part of my work I do clinical trials with new medications for which my institution is reimbursed. I won't be discussing any of those however.
- I do occasionally review cases relating to alleged vaccine injury. I donate the proceeds for medical research

My own educational background

- I attended both public and parochial primary and secondary schools in Michigan
- My wife attended public schools in Michigan
- Our children have attended both public and private schools in Ohio and currently attend Walnut Hills HS
- My wife has a PhD in clinical psychology and works in the public schools in Ohio as a school psychologist
- I taught Math in a private high school before going to medical school

4 My current clinical work

- I completed my training in 1998 in Pediatrics and Child Neurology
- I currently spend approximately 25% of my time in direct patient care activities evaluating and treating children with neurological problems
- My subspecialty expertise is Movement Disorders
- I supervise a resident clinic and do inpatient and consult work at Cincinnati Children's which provides broad clinical experience in the other areas we will be discussing today

My other responsibilities

- Child neurology residents train for 5 years after medical school. I am the CN residency program director
- I also teach and do research
- My research involves understanding the causes and "wiring changes" in Tourette Syndrome, ADHD, and a few other conditions. I also do genetics research

6 Outline

- Brain Doctors who are we?
- A Brain and development primer
- Specific Diagnoses: Science, Latest Treatments, Roles ("me" "You")
- Communication
- Self Education tips

7 Outline – specific diagnoses

• Specific Diagnoses: Science, Latest Treatments, Roles ("me" "You"), how might these affect learning?

- ADHD
- Hyperkinetic Movement Disorders (Tics, Stereotypies)
- Medication-Induced Problems
- Headaches/Migraines
- Epilepsy
- Staring and Dissociation
- Functional Neurological Symptom Disorder/Conversion Disorder
- Mental Retardation
- Weakness

8 Brain Doctors (for infants, children, adolescents)

- Neurologists
- Psychiatrists
- Physiatrists
- Neurosurgeons
- Developmental Pediatricians
- Psychologists you know better than I!

9 Training, BE/BC

- Medical School → MD or DO
- Residency: Pediatric Subspecialist Medical Doctors Train in General Pediatrics first
- Subspecialty Training comes second
- Fellowship training may come third
- Completion of ≥ 3 years residency training after medical school necessary to be <u>Board</u> Eligible
- Passing Examinations required to be **Board Certified**
- BC is an important metric

10 As a diagnostician

- I like the data from the deficit model
- I read your reports and use your scores in helping to think about patients I see

11 Primary Care Physicians

- The Go-To, front line doctors: Start Here
- Pediatricians, Family Practitioners, affiliated NPs
- Diagnose common problems like ADHD (Am Acad Pediatrics: ADHD Toolkit)

12 Adult Neurologists

- Goal 1: <u>Diagnosis.</u> Ideally <u>Specific</u>. Should have a biological (cellular, genetic, molecular) basis, based on careful history and physical examination as well as from diagnostic testing: blood/urine/spinal fluid; MRI, Neurophysiology/EEG
- Goal 2: Rational, Evidence Based Intervention
- Often work in teams
- · Prescribe medications
- Typically receive 3 months of pediatric neurology training

13 Adult Neurologists

- Perspective: DSM-5 is a book full of made-up labels of conditions with no biological markers (therefore are not real diagnoses)
- Work with social workers around occupational disability and accommodation issues but not necessarily around educational ones

 Note – in many communities the most accessible neurologist for a child to see is an adult neurologist

14 Child Neurologists

- Goal 1: <u>Diagnosis</u>. Ideally <u>Specific</u>. Should have a biological (cellular, genetic, molecular) basis, based on careful history and physical examination as well as from diagnostic testing: blood/urine/spinal fluid; MRI, Neurophysiology/EEG
- Goal 2: Rational, Evidence Based Intervention
- Often work in teams. We do not see ourselves as "pediatricians first" but we are still full medical doctors
- Prescribe medications
- Typically receive 12 months of adult neurology training

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15 Child Neurologists

- Perspective: Children are not small adults
- We are actively involved on the genetics frontier
- We understand that the diseases we see affect education but our training does not necessarily emphasize this in helpful ways
- The majority of us work in academic centers, not in private practice or in more rural areas

16 Child Neurologists

- Common referrals
 - Spells: Seizure, Epilepsy, other paroxysmal events
 - Headache, Migraine
 - Tic Disorders
 - Developmental delay, cerebral palsy
 - Behavioral problems
 - Traumatic or other acquired brain injuries

17 Child Neurologists

- Less common
 - Neuromuscular diseases, e.g. Muscular Dystrophy
 - Neurodegenerative diseases genetic, metabolic, etc
 - Other neurogenetic conditions
- · Really rare
 - There are a lot of really rare diseases we see
 - -Some of these are partly managed in a few specialized centers around the country

18 Child Neurologists

- Most of what we treat is not curable
- There are increasing numbers of treatments but there are still chronic conditions for which we can provide little directly beneficial treatment

19 Neurosurgeons

- Goal 1: discern when and when not to drill or saw through the skull
- Goal 2: Fix Problems
- Goal 3: If 2 not possible, relieve suffering and limit resulting deficits
- Referral source: primary clinician, neurologist, radiologist
- Pediatric medicine training: None/minimal

20 Pediatric Neurosurgeons – rop referral diagnoses

- Hydrocephalus
- · Skull shape abnormalities
- Brain Tumors or vascular diseases and malformations
- Spinal cord diseases/ structural anomalies

21 Pediatric Physiatrists "PM&R"

- Goal: rehabilitation and adaptation to diseases and injuries of the brain
- Diagnosis has already been made, typically
- Multi-disciplinary work with physical, occupational, speech therapists to improve function, independence, well being

22 Pediatric Physiatrists - referrals

- Traumatic Brain and Spine Injury
- Cerebral Palsy particularly to oversee medical aspects of interventions, e.g. medications and Botulinum Toxin for spasticity
- · Work closely with pediatric orthopedics

23 **Developmental Pediatricians**

- Goal: manage the medical and psychosocial aspects of children's and adolescents' developmental and behavioral problems
- Emphasis more on disordered or atypical development, less on disease or brain injury
- Like physiatrists work collaboratively with therapy teams
- Very interested in schools

24 Psychiatrists

- Medication Treatment of DSM5 Diagnoses
- Work with a "med check" model for follow ups
- Interest/skill in therapy variable
- Can do a one-year fellowship in child psychiatry

25 Child Psychiatrists

- Medication Treatment of DSM5 Diagnoses
- Work with a "med check" model for follow ups
- May take particular interest in neurological condition that has high psychiatric comorbidity
- Interest/skill in therapy variable
- Badly Outnumbered because they are treating society, not just disease

26 **Development**

- The "substrate" for developing skills is the brain and its connections to the body
- The trajectory of skill development requires changes in white matter and gray matter

27 White matter

- White matter is myelin
- Myelination progresses through childhood
- Myelin coats the axons the connections of the nerves
- A primary purpose is to enhance the speed of signaling speed increases as myelin develops

28 White Matter Diseases

• Abnormal oxygen and blood flow before birth, at birth, or related to premature birth can affect the cells that make myelin, and the myelination process.

• Premature Infants often have Periventricular Leukomalacia

29 White Matter Diseases

• In inflammatory diseases, myelin may be attacked by the immune system (Acute Disseminated Encephalomyelitis, Multiple Sclerosis)

30 White Matter Diseases

• Many genetic diseases also affect myelin formation. Some of these prevent formation of normal myelin. Others allow toxic metabolites to accumulate which damage myelin

31 Effects of White Matter Disease on Movement

- Can be severe
- Child is/ becomes very TIGHT "spastic" due to loss of speedy input from the brain or spinal cord to the nerves and muscles

32 Effects of White Matter Disease on learning

• These effects are often profound. Patchy diseases may produce patchy deficits. The slowing of signal transmission slows performance of all brain activities including thinking, behavioral regulation. Eventually the demyelinated neurons may also die off, accelerating the loss of skills. Some of these conditions are ultimately fatal.

33 Gray matter

- Gray Matter is the neuron cell bodies
- A major part of gray matter development involves "pruning" of uneccessary cells and connections
- Learning requires neuroplasticity connections between neurons are strengthened or weakened

34 Gray Matter and Neuroplasticity

- Long Term Potentiation (LTP) connections / synapses are strengthened
- Long Term Depression (LTD) connections / synapses are weakened
- These processes underlie learning but also recovery from injury

35 Gray Matter Diseases

- A huge variety of acquired and genetic conditions can alter the performance of neurons themselves or the LTP / LTD processes necessary to make new connections
- Many environmental and genetic factors can also results in neuron cell degeneration and death

36 Gray Matter Diseases

- Loss of gray matter, or dysfunction of gray matter, leads to cognitive impairment or dementia. It can also results in epilepsy (seizures)
- Progressive gray matter diseases result eventually in death

37 **Delay"**

- Delay can occur because of slower gray or white matter maturation
- Delay can also occur because of injuries to gray or white matter in early life, which permit some if not full recovery over time
- Particularly before the age of 6 years, we may not be sure if the child can "catch up"

38 Derailed"

- We don't tell parents that a child's development is "derailed" but this can be a more accurate way for us to think about certain children
- We can in more severe cases tell parents that the long-term possibilities do not include

normal motor, cognitive, or emotional function

39 Delayed" vs. "Derailed"

- "Delay" implies capacity for significant improvement or even to "catch up"
- May also indicate uncertainty about cognitive potential in a younger child
- Some children will never catch up or will experience only miniscule improvements despite all available therapies
- In many cases, the Doctor knows this very early but may not have communicated this
 effectively to the family

40 **Degenerating**

- Degenerative diseases are rare in children but really tragic
- The school community can play a wonderful role for families affected by these rare conditions, enhancing the quality of life of the child and easing the emotional burden on the parents

41 Definitions are imprecise

- Disorder the brain's structure is intact. The primary problem is with signalling
- Disease cellular or larger scale brain-cell death is involved. Includes progressive, denegenerative
- Conditions nonspecific, global

42 ADHD: is that really the diagnosis?

- The scales are subjective
- Maybe it is just bad parenting?
- "If that mom were raising me, I would have ADHD too!"

43 ADHD is more common in:

- Tourette Syndrome
 - Turner Syndrome
 - Neurofibromatosis
 - Fragile X
 - High Functioning Autism
 - Childhood epilepsies particularly absence
- Traumatic Brain Injury
 - Very Low Birthweight Infants/Prematurity
 - Hypoxic/Ischemic Injury

44 Large Study of kids' brains

- 223 ADHD kids, 223 typical kids
- 824 MRI scans
- Thickness measured in 40,000 locations

45 RESULTS

- Maturation progressed in a similar manner regionally in both children with and without ADHD.
- However, there was a marked, 3 year delay in ADHD in attaining peak thickness throughout most of the cerebrum:
- The delay was most prominent in prefrontal regions important for control of cognitive processes including attention and motor planning.

46 Non-medical Treatments for ADHD

- WIRE UP THOSE FRONTAL LOBES
 - Activities the promote cognitive and motor control, sustained attention, impulse inhibition
- MUSIC
- ART
- Strategy Games
- SPORTS
- Not video games

47 MTA Study

- NIMH Collaborative Multisite Multimodal Treatment Study of Children with ADHD
- · Randomized Controlled Trial
- 579 Children Diagnosed ADHD, ages 7.0 to 9.9 years old
- 4 treatment groups

48 MTA 14 month treatment

- Med Mgmt expert treatment with stimulants
- Behavioral Tx- see next slide
- BOTH combined
- Usual "Community Care"

49 MTA Behavioral Interventions for kids and parents

- 27 session group parent training plus 8 individual parent sessions
- 8 week summer treatment program for kids
- 12 weeks classroom administered behavioral therapy with a half-time aide
- 10 teacher consultations

50 MTA Study <u>14-month</u> <u>outcomes</u>

- Medication and Combined Treatment were similar for many core ADHD symptoms
- Behavioral therapy *alone* (i.e. no meds) and community care were significantly <u>less</u> effective

51 MTA study <u>3-year outcomes</u>

- Improvements in most children were maintained 3 groups
 - Large initial improvement (52%)
 - More married parents, higher IQ, lower behavior problem scores
 - Smaller initial improvement, but continued gains out to 3 years (34%)
 - Transient benefit, then return to pre-treatment levels (14%)

52 MTA study 3 year follow up

- Advantages of initial 14 month intensive medical treatment gone groups converged by 3 years
- All treatment groups have higher levels of substance abuse (17%) and delinquency (27%) than Community Controls (~7%)
 - Molina et al, Journal of the American Academy of Child and Adolescent Psychiatry 2007

53 MTA at 8 years

 Molina BSG, Hinshaw SP, Swanson JM, Arnold LE, Vitiello B, Jensen PS, Epstein JN, Hoza B, Hechtman L, Abikoff HB, Elliott GR, Greenhill LL, Newcorn JH, Wells KC, Wigal T, Severe JB, Gibbons RD, Hur K, Houck PR, and the MTA Cooperative Group. *The MTA at 8 years:* Prospective follow-up of children treated for combined type ADHD in the multisite study.
 Journal of the American Academy of Child and Adolescent Psychiatry. Online ahead of print March 2009.

54 ADHD: What the MDs Do

- Pediatricians diagnose using ADHD structured, validated interviews with Parent/Teacher ratings
- No medical tests needed in most cases
- Refer some proportion of children to psychology, developmental pediatrics, psychiatry, (neurology)

55 ADHD: What you can do

- Encourage teaching staff to fill out (thoughtfully) and return (promptly) rating scale forms
- Talk to parents about keeping an ongoing flow of information from them and the school to the prescribing physician

56 ADHD: What you can do

- Accurately identify children with IQ<70
 - Re-calibrate expectations
- Talk with parents about sharing your evaluations and in-class observations with treating physicians

57 ADHD: What you can do

- Know the science behind the pharmacology:
- ADHD stimulant medications work with the brain's own neurotransmitters dopamine and norepinephrine
- They have been prescribed for decades
- They do not cause brain damage

58 ADHD: What you can do

- Alert parents about possible side effects
- Insomnia
- Appetite suppression and weight loss
- Increased picking or other repetitive behaviors
- Personality change e.g. zombie effect

59 ADHD: What you can do

- Watch out for neuro-quackery
- Diagnosis should conform to standard practice
- Treatment should be evidence-based

60 ADHD: What you can do

- Understand limitations of current treatment
- This is orchard work, not an annual garden
- Meds are like fertilizer, lots of tending is still needed for growth

61 Tics - 1

- Patterned, countable (usually)
- Repetitive but Non-rhythmic
- Increase with stress, decreased with focused activity

62 Tics - 2

- Semi-voluntary/ Semi-suppressible
- Sensory Urge may accompany
- Onset usually after age 3

63 Stereotypies-1

- Patterned but Complex bilateral movements
- Younger children unaware; older children may enjoy
- Onset usually before age 3 years

64 Stereotypies-2

- Occur multiple times daily, last for years
- Often noted in certain situations, eg excited by new toy, bored

65 Stereotypy management

- Educate
- Communicate note for school, other adults
- Empathize/ sympathize try to put yourself in the parents' shoes

66 Tic Diagnoses (From DSM-5)

- Provisional Tic Disorder
- Tourette Syndrome
- Chronic Motor/Vocal Tic Disorder

67 Provisional tic disorder

- One or a few tics
- Tics with less than one year duration may resolve

68 Clinical Criteria for diagnosis of Tourette Syndrome (DSM-5)

- 2 or more motor tics, 1 or more vocal tics
- Duration > 1 year
- Anatomic location, number, frequency, complexity, severity changes over time
- Onset before age 21
- Not iatrogenic or a symptom of another Diagnosis

69 Four Tic Problems which may be indications for intervention

- 1. Pain
- 2. Social Interference
- 3. Disruptions common with vocal tics
- 4. Functional Interference

70 And – a *related non-tic issue* for kids with tics

- About 80% of kids who present for medical attention for tics have at least one other difficulty: ADHD, OCD or other anxiety disorders, Anger problems/low frustration tolerance/ immaturity, etc.
- The other problems may be priority for treatment interventions

71 **1. Tic Pain**

- Direct in the ticcing area, e.g. neck
- Indirect e.g. headaches worse from blinking
- Most dramatic severe strong painful tics in kids are brief
- Guide: If analgesic use is excessive, treat the tics

72 2. Social interference from tics

• Frequently perceived, not always accurately, by child or parent

- Problem: Peak age for tics is 8-15 years
- Male predominant
- Teasing, bullying are significant issues for some kids
- It's not always the tics

73 How do you talk to your peers about tics?

- Be cool: Jokes and T Shirts
- Fit in: be good at something
- Just like anything else "you have asthma, I have tics"
- TSA-USA resources: DVD "I have Tourette's but it Doesn't Have Me"

74 3. Special Considerations for Vocal Tic Disruptions

- Untimed tests, tests in separate room
- Tic breaks
- Standardized tests in another room write letters for SAT/ACT etc.
- Read "Front of the Class" by Brad Cohen

75 4. Functional interference from tics

- Not common for skilled or brief focused activities
- Some interference with sleep initiation
- · Worsening of pre-existing inattention may occur
- Social inhibition may contribute too worry/ attempts to suppress tics cause social or academic interference

76 Treatment principles for children with tics

- Consider all diagnoses and their associated impairment
- Rank and target
- Facilitate realistic expectations
- Start only one medication at a time, then reassess
- Start low, go slow

Elements of Behavior Therapy for Tics

- Function Based Assessment: Assess and improve the Environment
 - Remove reinforcers
 - Reduce stress
- Habit Reversal Training
 - Increase self awareness monitor tics, tic-urges
 - Train in Competing Responses

78 Treatment approach implications

- No one approach helps all problems
- No one medication improves everything in all persons with Tourette Syndrome (or ADHD or OCD...)
- Every child or adult is unique in terms of their pattern of involvement and responses to treatments

79 Medical options for treatment of ADHD in kids with tics

- Stimulants
- Alpha-2 adrenergic agonists
- Selective Norepinephrine Reuptake Inhibitors

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80 TACT Study

- 4 groups: clonidine, methylphenidate, BOTH, or placebo, n=136, 16 weeks
- ADHD results: all groups improved. Combined group did the best.
- Tic results: All groups but placebo improved. Even MPH group alone had fewer tics

81 Take home point:

For many kids with tics or Tourette Syndrome, stimulant treatment for ADHD does not unmask or exacerbate tics

82 Tics, Tourettes, Stereotypies: What Docs Do

- Accurate Diagnosis
- Assess for ADHD, Anxiety/OCD
- Education, Counseling, Anticipatory Guidance, Advocacy Resources
- Minimal or no diagnostic testing needed
- Treatment based on interference or impairment medication/behavioral

83 Tics, Tourettes, Stereotypies: What You can Do

- When you see a child with Tics or OCD, let parents know (calmly)
- Recognize one or both parents may have tics, ADHD, OCD/Anxiety
- Be aware child may also have ADHD, Anxiety/OCD, LD if school failure
- Work with child on anger, flexibility (getting unstuck), impulse control, anxiety as needed

84 TS: Disclosure and Ownership

- Prior to about 4th grade child may not have maturity to discuss tics with adults, peers
- By abut 4th grade aspire to have the child feel comfortable discussing it with peers better to be accepting and open of self than to reject self, waste cognitive reserve on tic suppression

85 Tics, Tourettes, Stereotypies: What You can Do

- Educate child: tics are OK, not a disease
- Educate peers
- Consider accommodations for vocal tics, OCD
- Use behavior plans correctly
- Support accessing appropriate resources, e.g. TSA-USA; TSA-Ohio

86 Another repetitive behavior

- This is one you may see in preschool aged girls or sometimes older girls
- It can be upsetting to parents.

87 Infantile Masturbation (Gratification) Syndrome

- Onset 3-6 months in girls
- Normal examination
- Behavior is interruptible
- Mean duration 5.5 years (Rodoo, Hellberg. Acta Paed, 2013)
- Normal developmental outcomes

88 Tremor

• Rhythmic back-and-forth movement about a joint, relatively symmetric

89 Developmental Coordination Disorder, Developmental Tremor

Clumsy kids

Donald L. Gilbert MD MS Cincinnati Children's OSPA 2014

- · Tremor with fine motor skills
- Non-specific indicator that other problems may be present with learning, attention, emotional regulation
- OT sometimes helpful

90 Teen Tremor

- Essential Tremor
- Drug Induced Tremor
- Psychogenic Tremor

91 Tremor – what docs do

- Try to determine cause/etiology
- · Rate level of interference
- Discuss treatment options
- Collaborate with psychiatrist if psych meds are causing tremor

92 Tremor – what you can do

- Handwriting practice may not work
- Tolerate slow, messy writing
- Reduce writing work, timed tests
- Encourage art, music
- OT
- Strategies

93 Headache Causes

- Headache disorders-such as migraine
- · Infections e.g. sinusitis
- Head injury
- Stress, anxiety, or depression
- Environmental triggers
- Other health conditions, e.g. high blood pressure

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94 Bad, Secondary Headaches

- New, different, unremitting
- Worse pain when horizontal
- Headache with double vision
- Vomiting, headache relieved after vomiting
- Confusion, personality change with headache
- Paralysis, seizure with headache

95 Primary Headaches – mostly migraines

- Migraine headaches, or "sick headaches," are recurring episodes of intense, pounding, nauseating head pain.
- The pain lasts for 30 minutes to up to three days.
- Migraine headaches are common in children and adolescents.
 - 3 percent of preschool children
 - -4 to 11 percent of elementary school children
 - up to 23 percent of teenagers have migraines.
- As children reach their teenage years, the number of headaches may increase, particularly in females.

96 Migraine Treatment in kids

- Lifestyle, lifestyle, lifestyle
- Abortive medications usually acetaminophen or ibuprofen, sometimes caffeine agents, ergots, "triptans"
- Preventative medications taken daily to reduce headache frequency and severity

97 Migraines, Chronic Daily Headache and the school

- Hydration is important
- Quick treatment, within the first ½ hour, is best
- Try NOT to send the child home in most cases
- Be supportive: Validate the pain; but don't reinforce Illness Behavior/ Sick Role
- Reinforce Well Behavior and Coping

98 Chronic Headache – other issues

- Headache/pain personalities passive children who let their parents speak for them
- Social/School avoidant children AND parents
- Pain as a symptom of untreated depression or anxiety
- · Pain as a form of rebellion or control

99 Headache/Migraine What Docs DO

- Accurate diagnosis, with judicious use of tests (often not needed)
- Assess impairment
- Education, non-medical symptom management
- Identify comorbid psychiatric disorders
- Prophylactic, abortive treatment

100 Headache/Migraine What You can do

- Have a compassionate, headache treatment plan with family including access to fluids and abortive medications
- Recognize these kids and parents are often anxious
- Work on stress management with child
- Validate the pain, but don't enable the sick role
- Keep the child in school
- Alert parents if you suspect anxiety, depression

101 Therapy for Migraine

- Doctors are not just researching drugs and pushing pills
- Giving kids tools to enable them to gain control is beneficial
- Therapies can be studied rigorously

102 What are seizures?

- Seizures result from episodic (paroxysmal) alterations of neurologic function
- So do:
 - -Tics
 - Myoclonus
 - Migraines
 - Even syncope (though the brain is downstream)

103 **Definitions: Seizure**

A paroxysmal discharge of cortical (brain surface) neurons resulting in altered neurologic

function

- motor, sensory, cognitive, autonomic
- Crude, non purposeful, and often rhythmic "output" occurs during cortical discharges, such as jerking, or automatic behaviors

104 Differential Diagnosis of <u>Spells</u>

- SEIZURE
- NOT SEIZURE
- Many other types of spells and brief bits of behavior: tantrums, tics, stereotypies, breathholding spells, syncope, dissociating from environment due to anxiety/ obsession/ psychosis

105 Seizure/Spell plan

- Place child on side on floor, away from furniture
- Observe and time episode
- Child will not swallow tongue so stay out of mouth
- Call out firmly, try to stop the movements by firmly grasping (as appropriate)

106 Fainting

- The brain is an innocent bystander
- May occur at predictable times
- Needs medical evaluation, usually not neurological evaluation
- Seizure-like jerking may occur briefly with the loss of consciousness but recovery is much quicker, with at most minimal confusion

107 The Epilepsies

- Diagnosis based on history a clinical diagnosis (usually Doc doesn't see the seizures occur)
- Requires two or more seizures
 - One seizure ≠ Epilepsy (may never recur)
 - Two seizures usually = "Epilepsy"
- Seizures occur without provoking factor (trauma, hypoglycemia, fever...)
- Usually, medical treatment is initiated after the second seizure

108 The Epilepsies

- Two or more unprovoked seizures is an indicator that the patient's brain has a tendency to have seizures
- Epilepsy is classified into many types based on a variety of clinical factors
- Most pediatric epilepsies do not result from structural lesions visible on MRI

109 Feared consequences of non-treatment of seizures

- Brain damage extremely rare
- Refractory epilepsy irrelevant
- Status epilepticus
- Sudden death extremely rare but does occur

110 Realistic consequences of non-treatment of seizures

- Trauma water, wheels
- Psycho-social consequences
- Status epilepticus
- Family/life disruption
- ER visits, hospitalizations, intensive care unit

111 Febrile seizures

- Most common cause of acute seizures
- Children who have these probably have a very mild form of epilepsy, although not always referred to in medical literature this way
- Seizure that occur between ages of 6 months and 6 years, in context of febrile illness, sometimes right at illness onset prior to fever detection

112 Status Epilepticus

- Status epilepticus (SE) is a seizure, or series of seizures, that lasts more than 30 minutes. SE is a life-threatening emergency. It needs to be evaluated and treated in a hospital.
- In the United States, SE affects more then 30,000 children under age 18 each year.
- It is most common in infants and toddlers.

113 Staring Spells

- There are many reasons to stare
- Most children stare off because they are bored are or thinking of something else
- This occurs at predictable times and doesn't interfere with the child's activities
- Some children stare because they are having seizures

114 Staring Seizures - Absence

- <u>Absence Epilepsy</u> is a *generalized epilepsy*, meaning the whole surface of the brain is involved
- Usually diagnosed in early elementary school years
- These seizures occur many times per day, without warning, at unpredictable times
- They are brief, may last just a few seconds
- Lights off (stare); lights on
- Interrupt normal activities and reduce ability to learn because child constantly misses brief bits of what is being said

115 Staring Seizures – Complex Partial

- Epilepsy with complex partial seizures usually comes from the memory lobe of the brain the temporal lobe
- It can begin at any time in life
- These seizures usually do not occur many times per day, and there is sometimes a warning or aura before them
- They may last seconds to minutes
- Lights off (stare; some automatic behavior such as chewing may occur), <u>then confusion</u>, <u>recovery</u>, then lights back on

116 Primary Epilepsies

- In children with normal, near normal intelligence (but who may have subtle learning differences)
- Normal brain structure (per MRI scan)
- Genetic contribution suspected, usually unknown
- Medication Treatment successful in ~70%
- Maturational component: Child may outgrow!

117 Secondary and Refractory Epilepsies

- Brain less likely to appear normal may have a variety of structural lesions
- More learning, behavior problems
- Once child has failed 2-3 medication trials, seizure freedom is unlikely

• Polypharmacy, high doses, side effects all more common

118 Seizures and Epilepsy – What Docs Do

- Accurate Diagnosis
- Education, Prognosis, Anticipatory Guidance
- Prescribe and Monitor Medications
- Refer for advocacy services
- Treat for 2 or more years
- If seizure free, consider stopping meds

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119 Seizures and Epilepsy – What you can do

- Provide accurate description of a first witnessed seizure
- Try to stop unclear spells (e.g. bored staring)
- Be aware children with epilepsy are more likely to have other brain-based problems; therefore consider interventions early
- Other Health Impaired
- Have a seizure safety plan with the family, possibly including use of intranasal or rectal medication
- · Be vigilant for absence seizures

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120 Seizures and Epilepsy – What you can do

- Family support
- Reassure other children
- Educate other children, de-stigmatize
- Discuss safety with parent water, wheels, (womb)
- Advise parent if you think seizure frequency increasing

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121 Cerebral Palsy

- Cerebral palsy (CP) is a common childhood neurological disorder.
- It is a static disorder of movement and posture present in the first year of life.
- May be due to a variety of possible causes which result in permanent structural damage to the brain.

122 Cerebral Palsy

- Brain MRI usually shows the location of the pathology involved in a child's CP
- CP is a brain disorder that typically does not worsen as time passes. In fact, most children with CP improve to some degree as they become older.
- Some children with CP will develop worsening dystonia or scoliosis over time

123 Types of Cerebral Palsy

- Spastic Diplegia most common in premature infants
- Hemiplegia one side, arm and leg
- Quadriplegia all four limbs, usually cognition impaired as well
- "Choreoathetoid CP" rare
- "Ataxic CP" rare

124 Barriers to communicating really bad news

 It makes us feel sad and uncomfortable, therefore it is easier to sugar-coat, or just to avoid discussing • When you give really bad news, it feels hopeless, or like giving up and abandoning the child and parent

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125 Effective Communication about Really Bad Brain Diseases

- Is compassionate
- Employs realism, not sugar coating
- Employs direct, understandable terms:
 - -"This child will never walk or talk (again)"
 - "Some problems are beyond repair"
- The doctor needs to state this (when true) and then school personnel should be aware and reinforce this reality

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126 Survival in Children with Profound Neurological Impairments

- In vegetative or minimally aware children, lifespan is significantly shortened despite the best of parental and medical care
- Complications ultimately develop, usually in the lungs, that naturally bring the child's life to an end over a period of years

127 Cerebral Palsy, severe neurologic impairment – What Docs Do

- Search for cause
- Clarify severity
- Discuss prognosis
- Direct medical/motor therapies
- Medical interventions cannot speed up development or enhance cognition
- Medical interventions cannot reverse damage if present

128 Cerebral Palsy – What You Can Do

- Try to have an accurate estimate of the child's intellectual and motor potential
- Provide therapies
- Maximize participation if possible
- Steward resources if there is NO potential for meaningful gains, emphasize compassion and parental respite
- Don't feed into unrealistic beliefs

129 Progressive Weakness!

- Difficulty running and jumping
- Difficulty climbing steps
- Frequent falls
- Large calf muscles
- Difficulty getting up from a lying or sitting position

130 Muscular Dystrophy

- Duchenne muscular dystrophy (DMD) is the most common form of muscular dystrophy in children.
- DMD is a genetic—or inherited—disorder. It is linked to the X-chromosome. DMD mainly affects boys.
- An estimated one in 3,500 males worldwide has DMD. Around 400 boys in the US are born with DMD each year. Females are rarely affected.
- There are other forms of muscular dystrophies that affect both boys and girls.

131 Progressive Weakness – What docs do

- Identify accurate diagnosis
- · Offer treatment if possible, e.g. steroids for muscular dystrophy
- Clinical trials
- Multi-disciplinary care
- Advocacy, support

132 Progressive Weakness – what you can do

- Support families
- · Provide counseling to child
- Educate peers
- Provide services to reduce contractures, maintain mobility
- Be aware of significant stress on family
- Note muscle diseases often are linked to lower IQ

133 Functional Disorders

- Conversion Disorders
- Functional Disorders
- Psychogenic Disorders
- Psychosomatic Disorders
- "Pseudo- " e.g. Pseudoseizure
- Not the same as malingering or factitious disorders

134 Prior Conception: Somatization disorders in pediatric neurology

- Symptoms develop which resemble those of a neurologic/medical illness but are inconsistent or the neurological diagnosis is "ruled out"
- Emotional trauma converted to physical symptoms
- Reversible there is no actual brain damage

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135 Somatization disorders in pediatric neurology

- Examples: Nonepileptic spells which resemble seizures,
- Weakness/paralysis despite normal brain/spinal cord/nerve function,
- Shaking/tremor-like movements or other abnormal movements
- Symptoms can be very dramatic
- · Long term, even life-long disability may occur

136 **Case 1**

 A previously healthy 14 year old develops abnormal arm movements which start and stop abruptly. She has not been able to attend school for 4 days. She has been to the ER twice but has no diagnosis. Her neurology appointment is not for 3 months and she now presents to your office.

137 Case 2

 A 16 year old adolescent female has had 4 convulsive episodes characterized by thrashing her head back and forth, flailing her arms, bicycling with her legs, and rocking her body.
 Her eyes are reportedly clenched tightly shut during these episodes. These have been witnessed by physicians in the emergency room. The mother is angry that the physician suggested she was faking seizures.

138 Functional Disorders - Outline

- I. Definitions and Language
- II. Examples
- III. Diagnostic process
- IV. Treatment/Management of two cases

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139 DSM – 5 Somatic Symptom and Related Disorders

Conversion Disorder (Functional Neurological Symptom Disorder)

Criteria for conversion disorder (FNSD) are modified to emphasize the essential importance of the <u>neurological examination</u>, and in recognition that relevant <u>psychological</u> factors may <u>not</u> be <u>demonstrable at the time</u> of diagnosis.

140 Language

- Offensive?
- ² "Hysterical"
 - Faking
 - Wacky
 - Conversion
 - Pseudo
 - Psycho-anything
 - · Stress-induced

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- 3 Acceptable?
- Real / Organic
 - Software Problem
 - Something incomprehensible
 - Stress-induced
 - Functional Disorder

141 Examples of Functional Neurological Symptom Disorder

- 1. Non-epileptic "spells" / "pseudoseizures" → "psychogenic seizures"
- 2. Functional/Psychogenic Movement Disorders (Tremor, Tics, Dystonia, Ataxia/Astasia-Abasia)
- 3. Functional/Psychogenic weakness / sensory loss

142 III. Diagnosis – *Positive,* Incongruous Findings

- Symptoms and Signs Incongruous with disease:
 - Motor; Sensory neuro-anatomy
 - Movement Phenomenology
- Seizures: Non-seizure semiology clinically recognizable / verifiable with ictal EEG

143 Refer early and quickly

Positive prognostic factors:

Short duration

Early diagnosis

Satisfaction with care

144 III. Diagnosis – Psychological Factors

Is a MAJOR Stress being CONVERTED?

145 Stress and psychogenic illness:

Not always

- The "magnitude" of the stress may be small (Camel-straw)
- Child/family may be unable to interpret any physical symptoms as being produced by stress, may not recognize emotions

146 Stress and psychogenic illness

- Small stresses can seem big if:
 - Feelings about the stress are unresolved
 - An underlying mood disorder is present
 - Note this may not be identified by the psychologist or psychiatrist initially
 - The child has not developed coping skills
 - The primary role model/parent has personality disorder

147 III. Diagnosis: faking?

- Typically not faked
- Be vigilant for malingering and Munchausen's but this should be infrequent

148 IV. Treatment

Communication about the diagnosis is part of the treatment

- Some families are open to the idea that an illness could be stress-induced or psychogenic (perhaps parent volunteers this information)
- "One of the possible causes of the symptoms your child is having is a brain disorder which *causes* TREMOR, *resembles* PARKINSONS, but is actually psychogenic stress or emotions cause the brain to develop these symptoms"
- Other families are opposed to the idea that an illness could be stress induced or psychogenic

Psych = "offensive"

"One of the possible causes of the symptoms your child is having is a brain disorder which *causes* TREMOR, *resembles* PARKINSONS, but is actually *functional* – the brain is mis-firing ("software problem") and producing these symptoms"

153 Your approach can start with:

"What is your understanding of the cause of your child's problem?"
"What diagnosis did the doctor give?"

154 The parent may say

"The doctor didn't give a diagnosis"

155 🔲 ?

- Parent may feel uncomfortable disclosing this
- The parent may outright reject a psychogenic diagnosis and not want to hear any more about it

• Physician may have tiptoed around the diagnosis and not educated the family about it

156 Your attitude may influence the outcome

- "It sure sounds like *Brittney* has a serious problem."
- "We will do all we can to keep her in school despite this problem. As it turns out there are many children who attend school full time despite serious medical or neurological diagnoses and we are here for her"

157 Your attitude may influence the outcome

- "My understanding is that part of the treatment for these kinds of problems is rehabilitation, and part of the rehabilitation is going to school full time.
- "From what I have read, staying home from school makes this kind of condition worse."

158 Case 2

- A 16 year old adolescent female has had 4 convulsive episodes characterized by thrashing her head back and forth, flailing her arms, bicycling with her legs, and rocking her body. Her eyes are reportedly clenched tightly shut during these episodes. These have been witnessed by physicians in the emergency room. The mother is angry that the physician suggested she was faking seizures.
- "I have read that some kids can develop NONEPILEPTIC seizures. The brain is mis-firing ("software problem") and sending signals out to the body make the body jerk very similar to how it jerks in an epileptic seizure."

160 Effective Treatment

- A collaboration between the physician/ physician team, behavioral health, the parents, and school personnel
- Resume normal well behavior
- Hospitalization is sometimes needed: rehab; psychiatry
- Psychology for stress identification, coping, cognitive behavioral treatment

161 Functional/psychogenic illness: What you can do

- 1. Support, reinforce, normalize the diagnosis
- 2. Reinforce the good news about diagnoses they don't have ("This is not epilepsy, MS, etc")
- 3. Empathize: This is stressful, it IS A REAL PROBLEM

162 Functional/psychogenic illness: What you can do

- Reinforce Well-Behavior, Limit attention to Ill-Behavior
- · Limit nurse office visits
- Ask for doctor note if you need one legally to keep child in school
- Sending the child home enables the illness behavior and prolongs the illness, potentially indefinitely

163 Functional/psychogenic illness: What you can do

- Do not suggest/offer crutches or wheel chair
- Do offer PT/OT in some situations
- Do not say "it's in your head" or "you are faking" that's not the same thing
- Recognize child/parent may deny stressors, may not be able to accept psychogenic diagnosis readily

164 **Acceptance**

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• Early acceptance of diagnosis increases the likelihood of a positive treatment response

- Gelauff J et al, JNNP, 2013
- Some children, no matter what we do as a team, may have long-term disability

165 Iatrogenic Movement Disorders in Kids – The Big Four

- Stimulants
- Dopamine Receptor Blockers
- Selective Serotonin Receptor Inhibitors
- Anti-seizure meds

166 Anti-seizure RX

- Sedation
- Lethargy
- Dizziness
- Personality Change
- · Worsening seizures

167 Kids at higher risk for stimulant side effects?

<u>Autism, OCD/Anxiety, Tic Disorders, Non-autistic stereotypy</u> kids may have increased vulnerability to stimulant side effects

168 Vulnerability of the individual to manifest certain movement symptoms

- Kids on the autism spectrum:
 - Prefer/need sameness, routines
 - Manifest/are gratified by many repetitive behaviors
 - ruminate/obsess or perseverate on fears, special interests
- Pre-RX repetitive behaviors susceptible to Iatrogenic Repetitive Behaviors

169 Stimulant side effects

- Weight Loss
- Insomnia
- Personality changes: zombie, loss of spontaneity
- Psychosis

170 Examples of Dopamine Receptor Blockers commonly prescribed

Older

Haloperidol, thorazine

"Atypicals"

- Risperidone, Aripiprazole
- Olanzapine, Quetiapine

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171 Dopamine Blocker Indications

- Psychosis (uncommon)
- Aggression, irritability, mania
- Tics

172 Dopamine Blockers - Used with caution

- Neurologic side effects restlessness, stiffness
- Sedation
- Weight gain

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- Mood changes, panic/anxiety/depression
 - Separation anxiety
 - Stomach aches before school/ School avoidance

173 Examples of SSRIs commonly prescribed

Older

• Clomipramine

Mainstays

- Fluoxetine, Fluvoxamine, Sertraline
- Citalopram, Escitalopram
- Paroxetine

174 SSRI Indications

- Depression
- OCD
- Anxiety
- Panic attacks

175 SSRI Side effects

- Dizziness
- Sleep disturbance
- Tremor
- Apathy
- Irritability, agitation, aggressive or suicidal ideation

176 Medication Induced Side Effects – What Docs Do

- Primary doctors often prescribe
- Adjunctive use of therapy variable
- Educate families about possible benefits, side effects, risks

177 Medication Induced Side Effects – What You Can Do

- Be aware of use of medication in kids in classroom
- Advise parents of changes in personality, attention, mood, alertness/somnolence
- Tolerate more side effects at onset of medication or after change/dose increases these may be transient!

178 Motivation for seeking CAM in pediatric neurological and developmental disorders

- Lack of scientific answers for causes
- Lack of effective medical treatments
- These conditions have pervasive effects on the child's life and the family's life

179 Attraction of complementary, non-scientific therapies

- Lack of scientific knowledge in general population
- Fear of the science, fears about medication
- Doctors who spend too little time talking with parents
- Alternative practitioners may be more compassionate and spend more time with families

180 Complementary treatments - issues

- Conflict of interest
 - Recommending treatments and charging for those same treatments, e.g. selling supplements from the shop (like auto-mechanics)

Donald L. Gilbert MD MS Cincinnati Children's OSPA 2014 Business increases if kids come during school hours – beware of providers who
recommend regular therapies, e.g. vision therapy, and offer discounts for appts during
school hours

181 Complementary treatments - quackery

- Vision Therapy
- Cranial Sacral Therapy
- Homeopathy
- Website: Quackwatch, NIH Institute Complementary and Alternative Medicine

182 CAM – what doctors and you can do

- Be understanding that reasonable science is complex and reasonable people can be "taken in"
- Acknowledge our own shortcomings
- · Be firm about the science
- Discourage families from using time-consuming or expensive non-validated therapies

183 Tools

- 1. The spectrum of evidence
- 2. Study design randomized controlled trials are the gold standard

184 Sources - PRETTY STRONG

- Major Medical Journals: NEJM, JAMA, LANCET,
 - Major Specialty Journals: NEUROLOGY
 - Medical Society Published, Evidence-Based Practice Guidelines and Patient Pages (examples in this talk)
- Advocacy organization websites with reputable medical advisory boards
 - Hospitals with patient health topic pages, e.g. Cincinnati Children's, Mayo Clinic
 - National Institutes of Health / National Library of Medicine
 - Accredited Continuing Education

185 Sources - NOT SCOTTISH

- Internet hits
 - Some Psychiatry Journals
 - Journals your doctor has never heard of
 - Network TV shows
 - "Other" Advocacy organization websites without reputable medical advisory boards
- Lectures online by sincere but wacky doctors who tell lots of anecdotes
 - Websites by unregulated purveyors of nutritional products and non-validated therapies that you have to pay directly for

186 How do we know if a treatment really works?

• Our best estimate of whether something really works is from Randomized Controlled Clinical Trials – look for this when you are on the internet seeking new treatments

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188 Thank you for your attention!

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